Spontaneous Regression of a Large-Cell Lymphoma in the Conjunctiva and Orbit
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Abstract: Spontaneous and complete regression of malignant neoplasms is extremely unusual. To our knowledge, this case report is the first description of spontaneous regression of an extranodal malignant lymphoma occurring in the conjunctiva and orbit. A 40-year-old woman noticed a pink conjunctival mass at the medial aspect of her left eye that had been present for 3 weeks. She presented on May 5, 2003. Ophthalmologic examination showed a salmon-colored mass along the lateral side of the caruncle. CT revealed a mass in the medial orbit. Surgical biopsy exhibited a malignant lymphoma, diffuse large B-cell type. After biopsy, the tumor spontaneously decreased in size and completely disappeared in 5 weeks. At 6 months’ follow-up, the tumor had not recurred.

Regressions of malignant neoplasms spontaneously or in response to seemingly nonspecific therapies is a rare but well-recognized phenomenon.1–4 Diseases most frequently associated with spontaneous regression are renal cell carcinoma, neuroblastoma, malignant melanoma, choriocarcinoma, and related gestational trophoblastic neoplasms.5 Spontaneous regression of malignant lymphomas has been reported in both nodal and extranodal sites.1,6–7 In this report, we describe the first case of spontaneous regression of an extranodal malignant lymphoma occurring in the conjunctiva.

CASE REPORT
A 40-year-old woman visited the Department of Ophthalmology at Kaohsiung Medical University Hospital for investigation of a conjunctival mass affecting the medial side of her left eye on May 5, 2003. The patient stated that the conjunctival mass had been present for approximately 3 weeks and was increasing in size. Her medical history was noncontributory except for being a carrier of hepatitis B.

Ophthalmologic examination revealed a relatively flat lesion with a smooth surface and a salmon-pink appearance growing along the lateral side of the caruncle (Fig. 1, A and B). Orbital CT showed an ill-demarcated, soft-tissue density mass located in the medial aspect of the left orbit with a clear retrobulbar fat space (Fig. 2, A and B).

Incisional biopsy was performed on May 13, 2003. Histopathologic findings revealed monotonous diffuse lymphocytic infiltration within the submucosa and tumor cells containing round hyperchromatic nuclei and scanty cytoplasm with an abundance of mitotic figures (Fig. 3A). Immunohistochemical examination showed diffuse positive leukocyte common antigen (Fig. 3B) and L-26 (Fig. 3C) and scattered positive CD45RO (Fig. 3D). In addition, there was diffuse positive stain with CD20, scattered positive CD3, positive for κ-light chain and negative for λ-light chain, and a high Ki-67 proliferation

FIG. 1. A and B, Conjunctival tumor with salmon-pink appearance on the left eye of the patient. C and D. Five weeks later, the tumor completely disappeared after an incisional biopsy.
index (mean, 35% in lymphoma cells) (not shown). Based on the above findings, a diagnosis of malignant lymphoma, diffuse large B-cell type was established.

Systemic investigations, including a complete blood count, chest radiography, abdominal echography, and abdominal CT scan were unremarkable. Ga-67 citrate whole-body scan showed increased radioactivity in the left orbital area. The patient was advised of the diagnosis and referred to a consultant in the hematology oncology clinic for further investigation and treatment. However, after biopsy, the tumor gradually decreased in size without anticancer medication. Finally, the salmon-pink mass completely disappeared after 5 weeks (Fig. 1, C and D), and follow-up CT of the orbit performed on September 26, 2003, showed complete regression of the tumor located at the medial aspect of the left orbit (Fig. 2C, 2D).

**DISCUSSION**

Spontaneous regression of malignant lymphoma is a rare but well-recognized condition. The first description of spontaneous regression of malignant lymphoma was reported by Burkitt and Kyalwazi in 1966. In low-grade non-Hodgkin lymphoma, spontaneous regression may be observed in up to 23% of patients without initial therapy. Gattiker et al. retrospectively reviewed 209 cases of non-Hodgkin lymphoma for the phenomenon of spontaneous regression. They reported that spontaneous regression was found in 18 of 140 cases of nodular lymphoma and in only 2 of 69 cases with diffuse lymphoma. The reason for the difference of frequency in spontaneous regression between the two histologic types remains unclear. The grade might be an important factor in the occurrence of spontaneous regression in non-Hodgkin lymphoma. In recent reports, the rate of spontaneous regression of high-grade non-Hodgkin lymphomas is much lower.

The mechanism of spontaneous regression of cancer remains unclear. Papac proposed several mechanisms for spontaneous regression of a neoplasm including immune modulation, tumor inhibition by cytokines or growth factors, elimination of certain carcinogens, induction of differentiation, tumor necrosis or inhibition of angiogenesis, and apoptosis. For example, spontaneous regression of malignant lymphoma is thought to be caused by modulation of the host immune system against concurrent bacteria or viral infection and by a traumatic effect including reduction of tumor load by biopsy. Krikorian et al. suggested an immunomodulatory effect as a possible mechanism for the regression of gastric lymphomas after cimetidine therapy. Although not ordinarily considered an antineoplastic or cytotoxic drug, cimetidine has been observed to have a number of effects on the immune system, including enhancement of natural killer cell activity of human lymphocytes treated with interferon and response to interleukin-2 in stimulated human lymphocytes. In our patient, the
possible mechanism for the spontaneous regression occurring after an incisional biopsy of the conjunctival neoplasm is an activation of the host immunity by the surgical intervention.

Although our patient is currently in complete remission, regression should be monitored over a prolonged period. It has been reported that a patient with large-cell lymphoma died of recurrence 13 years after spontaneous remission.10 Therefore, our patient will be closely followed.

REFERENCES

Orbital Decompression for Gross Proptosis Associated With Orbital Lymphangioma

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Abstract: A 14-year-old boy with 11 mm of proptosis and exposure keratopathy secondary to an orbital lymphangioma underwent surgical debulking with a carbon dioxide laser through a lateral orbitotomy combined with a 3-wall orbital decompression. The proptosis was reduced by approximately 2 mm as a result of the debulking procedure, but a further 5 mm reduction was achieved with the orbital decompression. No serious adverse effects were encountered. Bony orbital decompression may be a useful alternative treatment in patients with severe proptosis secondary to orbital lymphangioma.

Orbital lymphangiomas are rare vascular malformations. Their precise nature has been the subject of debate, with some authors classifying them as venous-lymphatic malformations and others grouping them together under the umbrella term of venous anomalies.1,2 In 1999, the Orbital Society suggested that all vascular malformations should be classified on the basis of their hemodynamic properties as no flow, venous flow, or arterial flow.3 Lymphangiomas have no significant direct communication with the venous system and belong in the no-flow category. The management has also been controversial. It may be possible to excise a well-circumscribed lesion, but many are diffusely infiltrative, and complete excision is virtually impossible. MRI may reveal cystic parts of the lymphangioma that can be drained and a limited debulking attempted, but even this risks damaging important orbital structures. Systemic steroids, direct injection of sclerosing or immunopotentiating agents, and radiotherapy have been described, but most authors agree that conservative management is recommended where possible.4,5 Surgical intervention is reserved for patients with visually threatening or cosmetically disfiguring disease, and multiple resections may be required.6

CASE REPORT

A 14-year-old boy presented with an 8-year history of gradually increasing right proptosis. Over the previous 6 months, he had development of ocular discomfort as a result of corneal exposure and was distressed by his appearance. There was no history of orbital inflammation or any acute exacerbations of the proptosis. Examination revealed best-corrected visual acuities of 20/20 OU, with Hertel exophthalmometry measuring 27 mm OD and 16 mm OS (Fig. 1). The right globe was displaced 11 mm laterally and 6 mm inferiorly, and there was 3 mm of lagophthalmos associated with moderate exposure keratopathy. Extraocular movements showed mild global restriction OD. Performing the Valsalva maneuver produced no change in the signs.

Orbital CT demonstrated a soft tissue intraconal mass in the right orbit, displacing the rectus muscles.
but with no bony changes. There was minimal enhancement with contrast medium. MRI confirmed a multilobulated heterogeneous lesion encircling the optic nerve with evidence of recent hemorrhage (Fig. 2). Ultrasonography showed a lesion with mixed echogenicity. Doppler studies revealed no vascular flow. A diagnosis of orbital lymphangioma was made and a surgical plan was formulated to retroplace the globe because of the corneal exposure.

The patient underwent a lateral orbitotomy through a swinging eyelid approach. The lymphangioma was debulked through the use of a CO2 laser but was fibrous and infiltrative, making safe intraconal excision difficult. At this stage, there was only a mild reduction in proptosis, estimated during surgery at approximately 2 mm. Therefore, a 3-wall orbital decompression was performed at the same time, which reduced the proptosis by a further 5 mm, judged by the total postoperative reduction measured at 7 mm. The patient made an uncomplicated recovery, and the histopathology confirmed the diagnosis.

One month after surgery, there was 4 mm of residual proptosis with no lagophthalmos and resolution of the exposure keratopathy. At 8 months, there was no worsening of the ocular motility and no complications. The patient was satisfied with his cosmetic appearance, and exophthalmometry measured 19 mm OD and 15 mm OS (Fig. 3). Repeat imaging demonstrated the

FIG. 1. Preoperative orbital lymphangioma. View from below, demonstrating severe proptosis of the right eye.

FIG. 2. T1-weighted MRI showing heterogeneous signal from diffuse lymphangioma filling right orbit.

FIG. 3. Marked reduction in proptosis is evident 8 months after surgery.

FIG. 4. Coronal CT scan demonstrating effect of 3-wall decompression.
effect of the decompression, with no impairment of sinus drainage (Fig. 4).

**DISCUSSION**

To our knowledge, this is the first reported use of bony orbital decompression in the management of orbital lymphangioma. The procedure is well recognized in thyroid eye disease and has been used for relieving optic nerve compression secondary to sphenoid ridge meningiomas, pseudotumors, and other orbital conditions.7,8 The indication for surgery in this case was severe, cosmetically unacceptable proptosis with exposure keratopathy. Debunking of the lesion was attempted with the CO2 laser, but because of its markedly fibrotic nature, there was minimal reduction of proptosis achieved by this method. Three-wall decompression, however, resulted in a much greater degree of retroposition. The potential risks of this procedure are the same as its use in thyroid orbitopathy, restrictive diplopia being the commonest complication. In addition, there is the risk of spread of the lesion in the maxillary and ethmoid sinuses, with subsequent obstruction and sinus dysfunction. However, these risks were deemed acceptable by both patient and surgeon, in the absence of a better alternative.

The patient had a marked reduction in proptosis, resolution of the exposure keratopathy, and he was happy with his appearance. Further studies are recommended to assess the use of orbital decompression for severe proptosis secondary to lymphangioma.

**REFERENCES**


**Orbital Dermoid Cyst and Sinus Tract Presenting With Acute Infection**

**Timothy S. Wells, M.D., and Gerald J. Harris, M.D., F.A.C.S.**

**Abstract:** A 9-month-old infant presented with orbital cellulitis and recent discharge from a hair-bearing pit above the eyebrow. Orbital imaging demonstrated a tubular lesion and sinus tract extending from a hypoplastic sphenoid wing, through the lateral orbit, to the skin surface. Complete excision of the dermoid cyst and sinus tract was performed through an eyelid crease approach.

Although dermoid cysts are the most common orbital lesions in childhood,1 few draining dermoid sinuses of the orbit or frontotemporal region have been reported.2–7 We describe a 9-month-old male infant who presented with orbital cellulitis and was found to have a dermally lined, orbito-cutaneous sinus and tract.

**CASE REPORT**

A 9-month-old male infant presented to his pediatrician with fever, left periorbital edema, and recent discharge from a congenital “mole” above the left eyebrow. Despite a 3-day course of oral amoxicillin trihydrate, edema and erythema increased, and the patient was admitted to Children’s Hospital of Wisconsin for intravenous administration of ampicillin sodium/sulbactam sodium. The authors were consulted on the third hospital day. Examination revealed mild residual left upper eyelid edema and erythema and a hair-bearing pit superior to the tail of the left eyebrow (Figure, A). No material could be expressed by palpation of the surrounding area. Orbital CT showed a subperiosteal, low-density elevation along the lateral orbital wall that extended posterior to a hypoplastic greater sphenoid wing; a small canal perforated the lateral orbital wall (Figure, B). The paranasal sinuses were unremarkable. After additional antibiotic treatment, MRI demonstrated a sinus tract in continuity with the orbital lesion (Figure, C through E).

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Surgical management involved an anterior orbitotomy—with a contingent, consecutive frontal craniotomy planned if the posterior aspect of the lesion could not be cleanly separated from dura. The lesion was approached with an eyelid crease incision and subperiosteal dissection. A narrow isthmus, which extended through a small defect in the lateral orbital rim, was intentionally divided to facilitate dissection and delivery of the anterior and posterior components. The posterior portion was followed in a bony fossa within the greater sphenoid wing and farther posterior to an area of deficient bone and thick membranous tissue overlying the middle cranial fossa. The lesion was separated without dural disruption. The anterior component was dissected in continuity with a sinus tract that extended laterally, superiorly, and superficially to a dermal pit, which was then circumscribed with a small elliptical incision. The lesion was removed in its entirety through the anterior approach (Figure, F). Histopathologic examination revealed a cavity lined by squamous epithelium and containing amorphous eosinophilic keratinous debris. Small skin appendages surrounded the lesion’s wall and hair shafts extended into the cavity’s center. There were no adverse sequelae of the surgery and there was no CT evidence of recurrence 7 months after surgery.

DISCUSSION

During development of the cranial bones, the embryonic ectoderm and periosteum are in apposition at suture lines. It is believed that abnormal sequestration of surface ectoderm at these lines underlies the genesis of orbitofacial dermoid cysts. Continued desquamation of keratinized epithelium and secretion of dermal glands usually cause progressive expansion of the lesions. Orbital dermoid cysts are most commonly superolateral, with attachment to the underlying frontozygomatic suture. Lesions are usually external to the bony rim, connected by a fibrous stalk or broadly adherent to an external fossa. Dermoid cysts can also occur intraorbitally, and they can extend as “dumbbell” lesions through the bones that separate the orbit, frontal sinus, temporal fossa, and intracranial cavity.

Dermoid sinuses presumably develop in a similar fashion but retain a connection to the skin surface. Because the tract allows egress of keratin and dermal glandular products, the mass effect may be limited. However, the tract also permits entry of skin flora and the opportunity for clinical infection. In our case, bacterial cellulitis was the presenting sign. The absence of associated sinusitis prompted additional imaging studies, which revealed the underlying anomaly.

In contrast to nasolabellar dermoid lesions, which have a draining sinus in 10% to 45% of cases, those
in the frontotemporal or orbital regions rarely have this association. In a few previously reported cases,\textsuperscript{2–7} the lesions were continuous from the skin surface to the frontal, temporal, or sphenoid bones, with or without extension in the orbital cavity. To our knowledge, only one patient had a history of orbital cellulitis.\textsuperscript{3}

REFERENCES

Unilateral Microblepharon
Francesco P. Bernardini, M.D.*, Robert C. Kersten, M.D.†, Carlo de Conciliis, M.D.‡, and Martín H. Devoto, M.D.§

Abstract: We report the clinical features and surgical treatment of a 15-year-old girl with unilateral microblepharon. The anomaly was characterized by a vertical shortage of upper and lower eyelid skin, causing nocturnal lagophthalmos, corneal exposure, and cosmetic deformity. Treatment consisted in hard-palate grafting and lateral tarsal strip suspension of the lower eyelid of the affected side. The outcome was considered satisfactory by the surgeon and by the patient. No further surgery was required.

Microblepharon, a rare congenital abnormality first described in 1848 by Cornaz,\textsuperscript{1} is characterized by a vertical shortage of upper and lower eyelid skin resulting in various degrees of lagophthalmos, scleral show, and corneal exposure, depending on the severity of the defect. Microblepharon is described as a bilateral defect, and the suggested treatment for severe forms is bilateral skin grafting to replace the missing anterior lamella.\textsuperscript{2,3} To the best of our knowledge, unilateral microblepharon has not been described. We report one patient affected by unilateral microblepharon and suggest an alternative surgical option that proved to be effective for the correction of the mild form of this disease.

CASE REPORT
A 15-year-old girl was referred to our clinic for symptoms related to nocturnal lagophthalmos and corneal exposure. She was also disturbed by the appearance of her left eye, which appeared to be “too big,” and by the misdirection of the upper eyelashes. The parents stated that the patient’s left eye was wide open during sleep, and it was kept lubricated with antibiotic ointment at night. The eyelid defect was present since birth; there was no family history of eye or eyelid disease and no childbirth trauma, previous eyelid surgery, or dermatologic treatment. Two years before presentation, the patient underwent permanent lateral tarsorrhaphy without substantial benefit. Ophthalmic plastic evaluation revealed cicatricial ectropion with 5 mm of inferior scleral show OS and 1 to 2 mm OD (Fig. 1). A moderate facial bone asymmetry was noted, with left malar hypoplasia and hemifacial microsomia, which may have contributed to the lower eyelid malposition. There was 14 mm of upper eyelid skin between the eyebrow and the

FIG. 1. Marked left scleral show caused by shortage of lower eyelid skin; the previous lateral tarsorrhaphy is visible at the left corner.
eyelid margin OS and 20 mm OD. As a consequence of the shortage of upper eyelid skin, the left upper eyelid margin was everted, with the eyelashes pointing upward, and 5 mm of eyelid lag in downgaze (Fig. 2). Although the voluntary closure of the eyelids showed no lagophthalmos on either side, under general anesthesia, 12 mm of lagophthalmos was evident in the left eye compared with 4 mm in the right eye. Slit-lamp examination showed moderate epithelial keratopathy OS involving the inferior half of the cornea. The remainder of the ophthalmologic evaluation was normal and Hertel exophthalmometry was 19 mm OU. We elected to proceed with hard palate mucosa grafting and lateral canthoplasty to elevate the left lower eyelid as an initial step. The patient was offered simultaneous skin grafting to lengthen the anterior lamella of the left upper eyelid and to correct the upward tilting of upper eyelid lashes, but she declined. Hard-palate grafting and lateral tarsal strip suspension of the lower eyelid were carried out in the standard fashion, making sure to elevate the lateral canthus by approximately 3 mm. Postoperative evaluation revealed the operated eyelid to be in good position, with the scleral show reduced to within 1 mm of the contralateral, nonoperated side. According to the patient and her family members, eyelid apposition during sleep was complete. The symptoms of dryness completely resolved, and slit-lamp examination showed resolution of the epithelial keratopathy. At the 6-month follow-up visit, the lower eyelid was still in a good position and the patient declined further surgery (Fig. 3).

DISCUSSION

Microblepharon is a rare condition characterized by vertical shortage of eyelid skin, resulting in various degrees of cicatricial ectropion and lagophthalmos. In severe forms, corneal exposure may mandate early surgical intervention including upper and lower eyelid skin grafts and a lateral tarsal strip. In mild forms, a lateral tarsal strip of the upper and lower eyelids in association with lateral tarsorrhaphies may correct lagophthalmos when lubricating eye drops do not suffice to keep the corneal surface intact.

As far as we are aware, unilateral forms of microblepharon have never been described before, and the cosmetic deformity resulting from this condition may be more disturbing to the patient than the functional symptoms. It is distinguished from congenital lower eyelid retraction and from euryblepharon by the associated shortage of anterior lamella affecting the ipsilateral upper eyelid. Previous reports have recommended the use of full-thickness skin grafting to the lower eyelid combined with lateral tarsal strip to lengthen the vertical eyelid dimensions or lateral tarsal strip in combination with lateral tarsorrhaphy. Our patient declined skin grafting because of concerns about cosmesis. The patient had failed to notice any benefit from the previous lateral tarsorrhaphy. A hard-palate graft combined with lateral tarsal strip elevated the lower eyelid enough to reduce nocturnal lagophthalmos and restored a symmetrical position to the lower eyelid. In conclusion, we believe that a posterior spacer with a lateral canthopexy may be effective for the treatment of nocturnal lagophthalmos secondary to mild forms of microblepharon and that it is an alternative to correct the lower eyelid deformity in unilateral forms.
A previously healthy 15-month-old Hispanic girl was referred for evaluation of rapidly progressive right-eye proptosis. On examination, fixation was central, steady, and maintained bilaterally. An right afferent pupillary defect was detected. Proptosis of the right globe was noted. Ocular motility was normal, as was the remainder of the ocular examination.

Further evaluation of the patient included imaging of the orbit and brain with CT and MRI (Fig. 1A). The scans revealed a large, soft-tissue mass that involved the right orbit, infratemporal fossa, middle cranial fossa, pterygopalatine fossa, and maxillary sinus. Bony destruction of the right lateral orbital wall and erosion of the walls of the maxillary sinus were present. The mass extended into the right orbit, involved the intraconal space and apex, and displaced the optic nerve superiorly. No definitive primary site was identified because the tumor was discovered simultaneously in several different anatomic compartments.

A lateral orbitotomy was performed to take a biopsy specimen and partially debulk the mass. Histologic evaluation revealed a tumor with a predominant alveolar–glandular pattern with anastomosing tubules and irregular gland-like formation within a myxoid and fibrous stroma. A focus of reticular pattern with a loose meshwork of microcystic spaces was also present. The neoplastic cells had lightly eosinophilic cytoplasm and pale to hyperchromatic atypical nuclei with rare nucleoli (Fig. 2A). No Schiller-Duval bodies were noted, but a rare hyalin globule was noted. Immunoperoxidase stains were performed and were negative for synaptophysin, lymphoid markers including CD30, desmin, myogenin, and epithelial membrane antigen. The neoplastic cells were strongly positive for α-fetoprotein (Fig. 2B) and cytokeratin. The morphologic and immunohistochemical results supported the diagnosis of an endodermal sinus tumor. Chromosomal analysis performed showed 46, XX/47,xx,del(6)(q21q25), + mar[1], further supporting the diagnosis.5

Further systemic evaluation included a CT of the chest, abdomen, and pelvis, bone marrow biopsy, lumbar puncture, and bone scan; the results were all negative. Serum α-fetal protein was elevated at 21,547 ng/L (normal = <6.4 ng/L), whereas β-human chorionic gonadotropin was normal at less than 2 U/L (normal = <2 U/L). The patient underwent chemotherapy with a germ-cell tumor protocol with cisplatin, VP-16, and bleomycin and had dramatic regression of the tumor (Fig. 1B). There was resolution of proptosis and the right afferent pupillary defect at 5 months. The child fixes well, and there is no evidence of strabismus.

REFERENCES

CASE REPORT

Orbital, Middle Cranial Fossa, and Pterygopalatine Fossa Yolk Sac Tumor in an Infant
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Abstract: A yolk sac tumor, also known as an endodermal sinus tumor, was diagnosed in a 15-month-old infant who presented with rapidly progressive right eye proptosis. Imaging of the orbits and brain revealed a mass in the right orbit, middle cranial fossa, and pterygopalatine fossa. A lateral orbitotomy was performed to take a biopsy specimen and to partially debulk the tumor secondary to signs of optic nerve compromise. The biopsy specimen revealed a yolk sac tumor, and the patient underwent systemic chemotherapeutic treatment. Because orbital endodermal sinus tumors have been infrequently reported, there are no firm prognostic or treatment guidelines. Our case demonstrates that early recognition, limited orbital debulking, and chemotherapy can have an excellent short-term outcome.

Germ cell tumors are neoplasms derived of primordial germ cells, which arise in both gonadal and extragonadal sites. The most common extragonadal germ cell tumors (EGGCT) are benign teratomas, which typically involve midline structures such as the mediastinum, intestinal, retroperitoneum, or sacrococcygeal area.1 These tumors infrequently involve the head and neck region.1,2 Endodermal sinus tumors, also known as yolk sac tumors, and embryonal carcinoma are malignant forms of EGGCT. They have rarely been reported to involve the orbit.1–4

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DISCUSSION

This patient’s diagnosis of endodermal sinus tumor was arrived at by morphologic and immunohistochemical results. It was also supported by chromosomal analysis; genetic alterations at chromosome 6 have been frequently observed in many types of malignant tumors and specifically in 72% of yolk sac tumors.5

Primary orbital endodermal sinus tumors have been encountered so infrequently that no firm prognostic and treatment guidelines exist. EGGCTs are generally highly malignant and lethal. However, those originating in the orbit may have a better prognosis compared with those originating on other extragonadal sites simply because they can be detected at a smaller size. Also, some authors have suggested that orbital endodermal sinus tumors of the orbit occur at a younger age and may have a better chance for

FIG. 1. Coronal MRI of orbits and brain using T2 fast spin-echo with fat saturation. (A), At diagnosis, a mass exhibiting T2 hyperintensity involved the right orbit, infratemporal fossa, middle cranial fossa, and maxillary fossa. (B), Regression of the mass at 5 months after orbital debulking and chemotherapy.

FIG. 2. Histopathologic and immunohistochemical features of the orbital endodermal sinus tumor (original magnification, × 100). (A), Hematoxylin and eosin stain shows irregularly anastomosing tubules and glands within a fibromyxoid stroma (no Schiller-Duval bodies are present). (B), Positive cytoplasmic immunoreactivity for α-fetoprotein.
long-term survival if they are treated aggressively with chemotherapy and surgery. Although a case of chemotherapy-resistant endodermal sinus tumor of the orbit has recently been reported, our case demonstrates that limited orbital debulking and chemotherapy treatment of this lesion can have an excellent short-term outcome. Despite an uncertain ultimate prognosis, our patient’s short-term quality of life has vastly improved.

REFERENCES

Malignant Solitary Fibrous Tumor Metastatic to the Orbit
Carolyn Glazer-Hockstein, M.D.*, Nasreen A. Syed, M.D.*, Michael Warhol, M.D.,†, and Roberta E. Gausas, M.D.*

Abstract: A 61-year-old man with a history of malignant solitary fibrous tumor of the chest had development of unusual sites of metastasis involving the sphenoid wing of the orbit and soft tissues of the cheek. He was found to have a solitary fibrous tumor, an uncommon type of spindle cell neoplasm that most often arises in the pleura, which was metastatic to the orbit. This is the first reported case of malignant solitary fibrous tumor metastatic to the orbit. The clinical and histopathologic findings of metastatic malignant solitary fibrous tumor are described.

A 61-year-old man presented with intermittent horizontal diplopia and masses of the right orbit and cheek in January 2001. His medical history included resection of malignant solitary fibrous tumor (SFT) of the chest wall and right upper lung and resection of a right frontal lobe meningioma in 1997. The chest wall and lung tumor was very large, with invasion of the rib and chest wall muscle and extension to the parietal pleura. Recurrence of the chest SFT occurred in 1998, for which he underwent radiation therapy. The patient also developed a right cheek mass in 1998. Pathologic diagnosis after excision was interpreted as fibrous histiocytoma. This cheek mass recurred in 1999, and the patient underwent re-excision. Unfortunately, histopathologic confirmation of the diagnosis of fibrous histiocytoma could not be performed because the slides were unobtainable.

At presentation to the orbital service in 2001, corrected visual acuity was 20/25 OD and 20/20 OS. No afferent pupillary defect was present and ocular motility was full. Hertel exophthalmometry revealed 3 mm of right proptosis associated with swelling of the right temporalis fossa. Dilated fundus examination was normal.

Orbital CT and MRI revealed a well-circumscribed homogenous lesion of the right sphenoid wing measuring 2 × 2 cm that was associated with frank bony destruction. The lesion extended into the lateral aspect of the right orbit and was immediately adjacent to the right lateral rectus muscle. A second lesion was present in the right cheek. The lesion measured 6.0 × 3.0 × 3.0 mm and eroded the anterior face of the maxillary sinus. This mass was distinct from the sphenoid wing lesion and radiologically did not represent direct extension of tumor. Incidentally, another lesion was present in the anterior and lateral wall of the right maxillary sinus. CT imaging characteristics of the maxillary sinus lesion were consistent with mucocele (Figs. 1 and 2).

The patient underwent lateral orbitotomy with complete resection of the right orbital mass and removal of the right cheek mass by means of a Weber-Ferguson approach. During surgery, the orbital mass was noted to extend through the sphenoid wing and involve the dura.

PATHOLOGIC DESCRIPTION
The orbital and cheek tumors were composed of plump spindle and epithelioid cells interspersed with keloid bands of collagen typical of localized fibrous tumor. The presence of significant pleomorphism, necrosis, and mitotic counts higher than 4 per 10 high-power fields favored malignancy. Immunohistochemical analy-
sis was positive for vimentin, CD34, and bcl2; cytokeratin and calretinin staining was negative. The microscopic findings and the immunohistochemical analysis supported the diagnosis of malignant SFT (Fig. 3). The margins of the cheek mass confirmed the radiologic findings, in that it was distinct from the orbital lesion.

**DISCUSSION**

SFT are uncommon tumors of mesenchymal origin that arise from pleural and extrapleural locations including lung, pericardium, mediastinum, nasopharynx, paranasal sinuses, thyroid gland, salivary glands, and orbit.1 Recently, they have been reported to occur in the lacrimal gland and sac.2,3 SFT are usually benign, rarely recur locally, and rarely metastasize. Malignant pleural SFT may metastasize to sites such as bone, liver, and skin.4,5 Only 10% to 15% of intrathoracic SFT are histologically or clinically malignant, and such malignant cases are rarely described as originating in extrathoracic sites.5 Brunnemann et al.1 studied 24 cases of extrapleural SFT and found them clinically and histologically similar to their pleural counterparts.

Primary orbital SFTs have a nonaggressive clinical course and are treated by en bloc resection.6 Patients may present with proptosis, blurred vision, eyelid swelling, dysmotility, blepharoptosis, or a palpable eyelid mass.7 Gigantelli et al.7 reviewed the radiographic presentations of 7 patients with primary orbital SFT and found intrale
tional heterogeneity and a predominantly low T2 signal intensity on MRI to be distinctive of SFT. No bone destruction was found in their series and the low incidence of pressure-induced bone remodeling was attributed to the nonaggressive biologic nature of these tumors. Woo et al.2 reported bony erosion in 2 cases of primary SFT of the lacrimal sac. In contrast, the frank bony destruction seen on CT in our patient reflects the malignant nature of this metastatic lesion not seen in previously described orbital and peri-orbital SFT.

Histologic characteristics that identify SFT included spindled to plump cells that grow in a random or "patternless" pattern.4,6 Cellularity may vary and there may be bands of collagen interspersed between tumor cells. SFT can mimic other spindle cell tumors of the orbit including...
fibrous histiocytoma, hemangiopericytoma, meningoima, schwannoma, and fibromatosis. Histopathologic analysis with CD34 immunoreactivity has helped improve diagnosis of this tumor. CD34 reactivity demonstrates that SFT is of mesenchymal stromal cell origin. Westra et al. found strong and generalized CD34 positivity helpful in differentiating SFT from other histologically similar neoplasms. SFT are not immunoreactive for S100 protein like schwannoma, or for epithelial membrane antigen, like meningoima.

Certain histologic features further characterize malignant SFT; however, malignant SFT exhibits similar CD34 immunoreactivity as benign SFT. Pleomorphism, mitotic activity, tumor size, necrosis, and increased cellularity are demonstrated in malignant SFT. These characteristics suggest that the tumor may behave in a more aggressive manner with invasion of adjacent tissues, recurrence, and distant metastases.

The most common sites of primary malignant tumor metastatic to orbital bones are the breast, prostate, thyroid, and gastrointestinal tract. Metastases to orbital bone typically demonstrate an irregularly enlarged or a well-defined mass associated with bony destruction as seen in our case. This feature of bone destruction has not been described in other cases of orbital SFT. Complete tumor resection in cases of orbital SFT is considered the best form of management, with only two reported cases of local recurrence.9,10

The differentiation of benign and malignant SFT has historically rested on the histopathologic findings of cellular atypia; however, several cases of local recurrence and distant metastasis have been described in patients whose primary tumors lacked atypical features. Further, recurrence and distant metastasis have been described to occur more than 5 years after primary excision. The difficulty in identifying tumors with aggressive behavior requires that all tumors be surgically managed similarly. Extrathoracic SFT are routinely managed by en bloc resection with clear margins. Management of orbital SFT should follow the same principles, with consideration of the proximity of the tumor to the optic nerve and globe. In our case, complete tumor resection including involved bone and dura was performed to provide local control. At 18 months’ follow-up, our patient’s metastatic disease was stable with systemic chemotherapy.

SFT is a rare orbital lesion that most frequently presents with unilateral proptosis. Histopathologic examination reveals a spindle cell neoplasm, but diagnosis relies on immunohistochemical markers, especially CD34 positivity and S100 nonreactivity. To our knowledge, this is the first reported case of metastatic malignant SFT to the orbit. We suggest that malignant SFT be included in the differential diagnosis of orbital metastatic disease.

REFERENCES

T-Cell Sinonasal Lymphoma Presenting as Acute Orbit With Extraocular Muscle Infiltration
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Abstract: We describe a rare case of sinonasal T-cell lymphoma in an 11-year-old boy who presented with a right acute orbit characterized by proptosis, eyelid edema and erythema, limitation of eye movements, and excruciating pain on the right side of his face. Orbital computed tomography showed progressive right extraocular muscle enlargement. One biopsy specimen showed extensive tissue necrosis and an infiltrate of atypical cells with pleomorphic nuclei

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within the walls of blood vessels. Immunohistochemical studies demonstrated that these cells were positive for leucocyte common antigen (CD45), CD3 cytoplasmic, CD45RO, and terminal deoxynucleotidyl transferase and negative for CD20, CD57, CD56, CD99 and Epstein-Barr virus. Chemotherapy for T-cell non-Hodgkin lymphoma was initiated, but the patient’s status deteriorated and the child died of respiratory insufficiency, sepsis, and central nervous system infection.

In childhood, the classic signs of orbital inflammation such as proptosis, eyelid edema and erythema, pain, limitation of eye movements, and loss of vision are usually related to cellulitis caused by sinusitis. Less frequently, this clinical picture may be the manifestation of an idiopathic orbital inflammation, rhabdomyosarcoma, metastatic neuroblastoma, leukemic infiltration, ruptured dermoid cysts, or even abrupt formation of chocolate cysts with an orbital lymphangioma.

We report an extremely rare case of sinonasal T-cell lymphoma in a Brazilian child of Japanese descent who presented with an acute orbit with extraocular muscle enlargement associated with apparent sinus disease. To our knowledge, this is the first report of extraocular muscle infiltration as a presenting sign of sinonasal T-cell lymphoma.

**CASE REPORT**

An 11-year-old boy was admitted to the hospital with a 2-month history of right ethmoid-maxillary sinusitis and cellulitis. He was initially treated elsewhere with intravenous antibiotics and sinus drainage without success. During the previous 2 weeks, the proptosis of the right eye had worsened and he had lost vision in both eyes.

On examination, the patient presented with a right acute orbit characterized by proptosis, eyelid edema and erythema, limitation of eye movement, and excruciating pain on the right side of his face (Fig. 1). There was bilateral mydriasis and no light perception OU. Biomicroscopy of the anterior segment and ocular fundi was unremarkable in both eyes. A general physical examination disclosed that the boy had fever (38°C), areas of alopecia in the scalp, and palatal ulcers. There was no evidence of liver, spleen, or lymph node enlargement. Computed tomography of the chest and abdomen also failed to detect any abnormality. Results of a bone marrow biopsy were normal.

Two orbital computed tomography scans (obtained 1 month apart) from his initial treatment showed opacification of the right maxillary and ethmoid sinuses and progressive right extraocular muscle enlargement. The ethmoidal labyrinth and medial wall of the orbit were thickened (Fig. 2A).

The first computed tomography scan obtained at our hospital 2 months after the initial presentation showed signs of right ethmoidectomy and a massive right extraocular muscle infiltration. There was also an area of edema in the frontal lobe. Magnetic resonance imaging showed a frontal lobe area of edema and diffuse meningeal infiltration (Fig. 2B). Biopsy specimens were taken immediately from the right orbit and the inferior turbinate. Histopathologic examination revealed tissue edema, dilated vessels, and a mild acute infiltrate, consistent with an idiopathic orbital inflammation. Cultures and special stains for bacteria (Brown-Hopps) and fungi (Gomori-Methamine-Silver) were negative. Antinuclear, antineutrophil cytoplasmic, and anti-SM antibodies were all negative. As the clinical picture was not consistent with idiopathic orbital inflammation, another biopsy was performed with the same results. Only in the third biopsy, did tissue samples from the medial rectus muscle, orbital soft tissue, and maxillary sinus showed extensive tissue necrosis and an infiltrate of atypical cells with pleomorphic nuclei within the walls of blood vessels (Fig. 3). The immunohistochemical studies showed that these cells were positive for leucocyte common antigen (CD45), CD3 cytoplasmic, CD45RO, and terminal deoxynucleotidyl transferase and negative for CD20, CD57, CD56, CD99, and Epstein-Barr virus. A bone marrow biopsy specimen was normal, and clonal TCR rearrangement was not observed. Blood cultures and lumbar puncture results were negative.

![FIG. 1. Clinical presentation: Right acute orbit signs including proptosis, chemosis, eyelid erythema, and edema.](https://example.com/fig1.png)
Computed tomography scans showed an area of infiltration in the frontal lobe base and complete obliteration of the soft tissues of the infratemporal fossa on the right side.

Chemotherapy for T-cell non-Hodgkin lymphoma was initiated, consisting of vincristine, daunorubicin, L-asparaginase, prednisone, and intrathecal injections of aracytin, methotrexate, and dexamethasone. Four weeks after treatment began, no objective response was observed and a rescue chemotherapeutic schedule of high-dose methotrexate and cyclophosphamide was tried. The patient’s status continued to deteriorate and the child died of respiratory insufficiency, sepsis, and central nervous system infection.

**DISCUSSION**

In the past, the association of sinus disease, palatal and nasal ulcers, and orbital swelling was described with nonspecific names such as “nonhealing midline granuloma of the Stewart type,” “lethal midline granuloma,” “idiopathic midline destructive disease,” “malignant granuloma,” “rhinitis gangrenosa progressiva,” “midline malignant reticulosis,” and “polymorphous reticulosis.” It became apparent that these terms described a heterogeneous group of diseases and with the advent of immunophenotypic staining, it was demonstrated that the atypical cells described in most of these cases were of T or NK lymphoid origin. Now, T/NK sinonasal lymphoma is a diagnostic entity in the World Health Organization lymphoma classification.

The malignant cells in this type of tumor are typically angiocentric, infiltrating and destroying blood vessel walls. This angiodestructive pattern of growth promotes extensive necrosis mixed with inflammatory cells. In this context, identification of atypical malignant cells can be difficult, especially because these cells tend to be scarce. Also, the diagnosis of sinonasal lymphoma is not usually suspected clinically. Surface crusting and secondary changes caused by local infection explain the need for abundant biopsy material to establish a correct diagnosis. In our case, the first biopsy showed only nonspecific inflammation with necrosis as a prominent finding. Two more biopsies were necessary to demonstrate positivity for T-cell markers.

Sinonasal T/NK lymphomas are common in Asia and in certain groups of South and Central America individuals of Indian origin and are rare in Europe and the United States. This fact strongly suggests a racial predisposition because there is genetic evidence that the native American populations originate from Asia. Our patient was of Japanese descent.

Morphologically, the cytologic atypia of T or NK lymphomas is variable, with cells of different sizes. The...
phenotype of these tumors is characterized by positivity for CD3 cytoplasmic and CD45RO, and negativity for CD57. NK cells usually lack markers of immaturity such as terminal deoxynucleotidyl transferase and are positive for CD56. In almost all cases, Epstein-Barr virus infection can be demonstrated.

Our case is consistent with the T-cell phenotype. terminal deoxynucleotidyl transferase was positive, indicating a lymphoblastic nature of the malignant cells. Positivity for Epstein-Barr virus was not demonstrated and CD56 was negative, ruling out an NK cell origin.

Sinonasal T/NK cell lymphomas have a poor prognosis, and local radiotherapy plus chemotherapy is believed to be superior to chemotherapy alone. Significant orbital involvement is rare, explaining the scarcity of descriptions of this tumor in the ophthalmic literature. We were able to find only 9 cases of primary T-cell orbital lymphoma. Because of the angiodestructive pattern of the tumor, swelling and erythema are common findings. Thus, it is not surprising that these tumors can present as an acute orbit, as in the case described by Meyer. To the best of our knowledge, the present case is the second described in the pediatric population, and the first in which extraocular muscle enlargement was an early and prominent finding. Even though orbital B-cell lymphomas can involve the extraocular muscles, we are not aware of any case of sinonasal T-cell lymphoma with such a clinical presentation.

The clinical presentation of our case is atypical. In childhood, idiopathic orbital inflammation usually does not present with extraocular muscle infiltration and severe visual loss. Specific myositis (systemic lupus erythematosus, scleroderma, Crohn disease, Whipple disease, and sarcoidosis) is rare in children or present with a more chronic course. The combination of an acute orbit and sinusopathy usually is the expression of orbital infection. However, orbital cellulitis usually appears radiologically as a subperiosteal abscess, a true orbital abscess, or a diffuse fat infiltration and not as progressive extraocular muscle enlargement.

The radiologic pattern of our case also ruled out the other neoplasias that can present as an acute orbit in children. Extraocular muscle enlargement with relatively preserved orbital fat is not seen in rhabdomyosarcoma, lymphangioma, neuroblastoma, or leukemic infiltrations.

The most typical findings of sinonasal T-cell lymphoma in the present case were the progressive meningeal infiltration of the anterior cranial fossa, formation of an abcess-like lesion in the cerebral parenchyma, and the presence of edema in the temporal and infratemporal fossae. These radiologic findings plus the presence of palatal and scalp abnormalities are highly suggestive of angiocentric sinonasal lymphoma.

Early recognition is essential for improving the chances of survival. We hope that this report will help other clinicians to identify this rare tumor early in the disease process.

REFERENCES


Blindness From Acute Angle-Closure Glaucoma After Blepharoplasty

Abstract: An elderly man had acute angle-closure glaucoma soon after undergoing bilateral lower eyelid blepharoplasty that resulted in complete loss of vision in one eye. Glaucoma is a rare complication of blepharoplasty surgery; only two previous cases have been reported to our knowledge. Risk factors for the development of glaucoma in the setting of blepharoplasty are discussed.

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Blepharoplasty is a frequently performed operation with few serious complications, which are often ophthalmic in nature and potentially sight-threatening. We present a case of acute angle-closure glaucoma after blepharoplasty that resulted in complete loss of vision in the affected eye. As far as we are aware, this complication has only been reported on two previous occasions in the medical literature, and in both cases, the outcome was favorable. The circumstances surrounding blepharoplasty that can increase the likelihood of precipitating angle-closure in susceptible patients are discussed.

CASE REPORT

A 73-year-old hypermetropic civil engineer presented to the eye department with a 2-week history of increasing ocular pain and decreasing vision OD. The day before the onset of these symptoms he had undergone bilateral lower eyelid blepharoplasty under general anesthesia by the regional plastic surgery team. Bilateral excess lower eyelid skin excision and removal of the medial fat pad on the right side was performed. After surgery, he had uncontrolled bleeding on the right side; further exploration of the right lower eyelid was undertaken later that day. Local infiltration with 10 ml lidocaine 2% with 1 in 80,000 epinephrine was administered. The bleeding vessels within the orbital fat were identified and hemostasis was achieved with cautery. He was discharged home the following day. Before surgery, his visual acuity was reported as 6/6 OD; it was not checked again after surgery. Twenty-four hours later, the patient noted that his right eye was red and sore, with reduced vision, but he did not report this. As his symptoms persisted he saw his family physician later that week and was treated with oral antibiotics for a possible postoperative infection. After 2 weeks of increasing ocular pain and decreasing vision, he was referred to our department.

On examination the visual acuity was 1/hand movements OD and 6/9 OS. There was a right relative afferent pupillary defect, with a fixed, mid-dilated pupil (Figure). The cornea was hazy, the anterior chamber was shallow, and no fundus view was possible. The left eye was white and quiet. The intraocular pressures were 64 mm Hg OD and 20 mm Hg OS. Acute angle-closure glaucoma was diagnosed, and the patient was admitted and given intravenous acetazolamide, mannitol, and frequent topical pilocarpine 4%, timolol 0.5%, and betamethasone 0.1%. Initial response to therapy was good, with an intraocular pressure of 25 mm Hg sustained for a period of 48 hours. However, it rose again to 60 mm Hg on maximal medical therapy, and he underwent a right augmented trabeculectomy with 5-fluorouracil under local anesthesia the following day. Thereafter, the intraocular pressure remained controlled at 20 mm Hg on no medication. An ultrasound scan of the orbit did not demonstrate any significant retrobulbar hemorrhage. The vision after surgery was hand movements with persisting signs of decompensation in the cornea. Two months later, the visual acuity had dropped to no light perception. The vision has remained at 6/9 in the fellow eye, which had an anterior chamber depth of 2.3 mm and open angles. Despite this, he has undergone prophylactic YAG laser peripheral iridotomy.

DISCUSSION

Blepharoplasty is a common operation and ocular complications are rare. The most serious complication is visual loss, which has been estimated to occur in 0.04% of cases. Due to the relative infrequency of this complication and the lack of appropriate experimental models, a wide range of mechanisms have been proposed. Acute angle-closure glaucoma has an approximate incidence in the general population of 0.09%, with only two previously reported cases of acute angle-closure associated with blepharoplasty.

Our patient had axial hypermetropia, which is a risk factor for angle-closure (refractive error was +1.00 DS OD, +0.75 DS OS). In addition, prolonged pupillary dilation with the likelihood of peripheral iridocorneal contact further increases the risk of angle closure. Our patient was only exposed to adrenaline in the affected eye, which we surmise may have precipitated mydriasis. Lidocaine is also thought to cause mydriasis and transient internal ophthalmoplegia by anesthesia of the short ciliary nerves or ciliary gan-
This has previously been observed in conjunction with blepharoplasty, but in the series of three cases reported,\textsuperscript{6} angle closure did not occur. Anticholinergic drugs used as muscle relaxants in general anesthesia have the potential to cause pupillary dilation, although these were avoided in our case. Other causes of pupillary dilation, such as dilation of a dark-adapted eye under the postoperative bandaging and a heightened anxiety state, could also have contributed.\textsuperscript{2}

Unlike the two previously reported cases,\textsuperscript{2,3} our patient has no functional eyesight left in his right eye. We recommend that all patients undergoing blepharoplasty be examined before surgery at the slit-lamp and have their vision assessed. An ocular cause for postoperative pain following blepharoplasty must be included in the differential diagnosis, especially when the patient complains of visual symptoms. Patients must be warned to seek medical help early if a problem develops after surgery. As blepharoplasty is undertaken by a range of specialists, examples of proformas to use as a preoperative assessment before surgery are available\textsuperscript{7} to highlight potential problems and complications, and their use should be considered.

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