



ORBITAL TUMORS

T-cell sinonasal lymphoma presenting as acute orbit with extraocular muscle infiltration

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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 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.

Malignant solitary fibrous tumor metastatic to the orbit

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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.

Orbital, middle cranial fossa, and pterygopalatine fossa yolk sac tumor in an infant

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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.

Orbital dermoid cyst and sinus tract presenting with acute infection

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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.

Orbital decompression for gross proptosis associated with orbital lymphangioma

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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.

Spontaneous regression of a large-cell lymphoma in the conjunctiva and orbit

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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.

Cavernous hemangioma extending to extracranial, intracranial, and orbital regions. Case report

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Extraaxial cavernous hemangiomas are rare intracranial lesions that can be located in different cranial compartments. Extension across different tissue planes such as the subcutaneous tissue, skull, orbital cavity, intracranial dura mater, and extracranial trigeminal divisions within the same patient has not been previously reported. This 32-year-old woman suffered left exophthalmos, left sixth nerve palsy, and trigeminal neuropathy. Magnetic resonance imaging studies revealed an extensive multicompartamental lesion, with enhancement following Gd administration. A left orbitopterional approach allowed removal of several cavernomatous lesions located in the orbit, frontotemporal dura, and lateral wall of the cavernous sinus. A histologically based diagnosis of extraaxial cavernous hemangioma was made. In the postoperative period the patient experienced a regression of her symptoms. The authors report on a case of cavernous hemangioma with a unique extension to different intracranial/extracranial compartments. Although radical removal of the lesion was not feasible, partial excision allowed for satisfactory clinical control of the patient's symptoms.

Histopathological study of orbital lymphangioma in an infant

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BACKGROUND: There has been controversy over whether orbital lymphangioma is a true entity in the orbit or various venous malformations.

CASE: A 1-year-old girl was referred to us with subcutaneous induration of the right upper eyelid. Rapid enlargement of the eyelid mass with blepharoptosis and dislocation of the eyeball were noted within 1 week. A diagnosis of an orbital lymphangioma was made from the clinical course and findings of magnetic resonance imaging. Surgical resection of the tumor was performed to prevent amblyopia.

OBSERVATIONS: Histopathologically, the lumen of the expanded cystic tumor consisted of a single layer of endothelium and was filled with erythrocytes as well as lymph. Immunohistochemical study revealed positive staining for factor VIII-related protein and CD 31, weak positive staining for thrombomodulin, and negative staining for CD 34 at the endothelium of the lymphangioma. The endothelium of the capillary vessels around the lymphangioma was positive for factor VIII-related protein, CD 31, and CD 34. Although a part of the tumor remained in the orbit postoperatively, orthophoria was achieved and no recurrence was observed for 2 years.

CONCLUSION: Immunohistochemical study suggested that the cellular composition of the endothelium of the orbital lymphangioma was different from that of the surrounding capillary vessels.

Growth factor receptor expression in orbital lymphangioma: possible therapeutic implications

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OBJECTIVE: To examine the expression of molecules targeted by imatinib mesylate (STI571;Gleevec) and epidermal growth factor receptor (EGFR) inhibitors in orbital lymphangiomas.

DESIGN: Retrospective observational case series.

PARTICIPANTS: Six patients with orbital lymphangioma treated at four institutions between March 2000 and December 2002.

METHODS: Tissue specimens and medical records from six patients were collected.

Immunohistochemical analysis was performed using antibodies against c-kit and platelet-derived growth factor receptor (PDGFR) alpha and beta and EGFR tyrosine kinase.

The differentiation of idiopathic inflammatory pseudotumor from lymphoid tumors of orbit: analysis of 319 cases

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PURPOSE: To analyze the clinical, morphologic, immunophenotypic, and molecular genetic differences between idiopathic inflammatory pseudotumor and lymphoid tumors of the orbit.

METHODS: 209 patients with IOIP and 110 patients with lymphoid tumors seen between January 1, 1978 and December 31, 1999 in Zhongshan Ophthalmic Center, Sun Yat-sen University were evaluated retrospectively.

RESULTS: More patients with lymphoid tumors had palpable mass than patients with idiopathic orbital inflammatory pseudotumor (IOIP) ($P < 0.0001$), with the percentage of 90% and 65%, respectively, whereas more patients with IOIP had swollen eyelid, eyelid or conjunctival congestion, pain, retinal folds or hemorrhage, and optic nerve atrophy than patients with lymphoid tumors, with the percentage of 55% and 40% ($P = 0.014$), 42% and 24% ($P = 0.001$), 24% and 1% ($P < 0.0001$), 14% and 4% ($P = 0.004$), 7% and 2% ($P = 0.043$), respectively. Ultrasound and computed tomography/magnetic resonance image (CT/MRI) scan usually could not differentiate IOIP from lymphoma. Nearly one third of patients with IOIP could not be easily differentiated pathologically from lymphoid tumor. Immunophenotypic, and molecular genetic analyses can differentiate IOIP from lymphoid tumor based on polyclonal or monoclonal proliferation of lymphocytes with the IOIP being polyclonal and lymphoma monoclonal.

CONCLUSIONS: Idiopathic orbital inflammatory pseudotumor and lymphoid tumor showed distinguishing clinical, morphologic, immunophenotypic, and molecular genetic characteristics.

Orbital lymphoid tumour located within an extraocular muscle

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A rare case of an orbital lymphoid tumour within an extraocular muscle is presented. The tumour displayed features of a progressive indolent systemic lymphoma. There was no response to a trial of chemotherapy and so local radiotherapy was instituted. At follow up 2 months later there was no evidence of disease.

Isolated, benign, intraorbital schwannoma arising from the supraorbital nerve.**Case report and review of the literature**

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The authors describe the rare case of a 65-year-old man, with absent clinical evidence of von Recklinghausen's disease, harbouring an isolated, benign schwannoma of the right supraorbital nerve. The patient presented a progressive, painless proptosis of the eye for the past 2 years, did not complain of any intra- or periorbital pain, and did not experience any form of visual disturbance or field defect. The lesion was completely resected using a fronto-orbitozygomatic approach. The clinicopathological, radiological and surgical features are discussed, and the literature on supraorbital nerve schwannomas reviewed. To the authors' knowledge the fronto-orbitozygomatic approach has not been previously reported for the removal of supraorbital nerve tumors.

Update on hemangiomas and vascular malformations

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PURPOSE OF REVIEW: Our knowledge base in the area of vascular anomalies is growing rapidly. With greater understanding of classification and diagnosis, as well as with the numerous areas of research bringing further awareness on the complexity of these lesions, we are improving our ability to treat them. We will attempt in this article to summarize the developments in the field of vascular anomalies over the last year.

RECENT FINDINGS: Emphasis on correct classification is still a high priority in the literature and yet there remains a great deal of misinformation. Many new developments in the basic science of these lesions are allowing better understanding of why these lesions occur while improving our management in these patients. Advances in laser surgery as well as sclerotherapy techniques have improved our ability to treat extensive lesions and also improve patients' quality of life.

SUMMARY: Many new and exciting areas of discovery occur almost daily in the field of vascular anomalies. Due to the breadth of this topic, it is certain that not all articles can be reviewed however the author has tried to present the most recent and clinically relevant breakthroughs in the field.

High incidence of orbital malignant lymphoma in Japanese patients

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Am J Ophthalmol. 2004 Nov;138(5):881-2

PURPOSE: To review 244 cases of orbital tumors to determine pathologic findings in Japanese patients.

DESIGN: Observational case series.

METHODS: We studied the pathology and origin of tumors in the orbit in 244 consecutive Japanese patients with orbital tumors at our institution from 1981 through 2002 (age 0 to 90 years, mean, 48.7

years; 114 men, 130 women).

RESULTS: The most common tumors were lymphoproliferative diseases (n = 114, 42.5%), including malignant lymphoma (n = 59, 24.1%) and reactive lymphoid hyperplasia (n = 45, 18.4%), pleomorphic adenoma (n = 21, 8.6%), and cavernous hemangioma (n = 18, 7.4%).

CONCLUSION: The incidence of lymphoproliferative diseases, especially malignant lymphoma, was very high in Japanese patients.

Orbital lymphoma: is it necessary to treat the entire orbit?

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PURPOSE: Conformal radiotherapy (RT) has been used for all patients with orbital lymphoma treated at our institution since 1997. We retrospectively reviewed the charts of 23 consecutive patients to test the hypothesis that partial orbit RT is effective and less toxic than whole orbit RT.

METHODS AND MATERIALS: Twelve patients with limited lesions were treated to partial orbital volumes and 11 patients (1 with bilateral disease) with more extensive lesions received whole orbit RT. The dose was 20-30 Gy (median, 25.2 Gy) for 19 patients with low-grade lymphoma and 24-40 Gy (median, 39.6 Gy) for 5 patients with intermediate- to high-grade lymphoma. The follow-up was 12-68 months (median, 34 months).

RESULTS: All patients had a complete response to RT. Intraorbital recurrence developed in previously uninvolved areas not included in the initial target volume in 4 patients (33%) treated with partial orbit RT. All were salvaged by repeat RT or surgery. No patient treated with whole orbit RT developed intraorbital recurrence. The acute and long-term toxicity was similar in both groups. All but 1 patient retained good vision.

CONCLUSION: Patients with orbital lymphoma should be treated to the entire orbit. An effective dose of RT for low-grade lesions is 25 Gy, which results in minimal morbidity even when delivered to the entire orbit.

Esthesioneuroblastoma in Maffucci's syndrome

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Maffucci's syndrome consists of multiple cutaneous hemangiomas, dyschondroplasia, and enchondromas with potential for malignant change. We report a case of a 33 year-old man with Maffucci's syndrome who presented with a several month history of nasal congestion, facial pain, and diminished vision in his left eye. Radiological studies showed a large soft tissue mass centered in the sinonasal area, extending bilaterally into maxillary sinuses and orbits with compression of left optic nerve. Biopsy of the mass showed esthesioneuroblastoma (olfactory neuroblastoma). Chemotherapy resulted in initial improvement, but the tumor recurred and did not respond to further treatment, resulting in his death. Sarcomatous tumors are reported in Maffucci's syndrome, but this is a rare case of a neuroendocrine tumor in a patient with Maffucci's syndrome.