



ORBITAL TUMORS

Orbital invasion by periocular basal cell carcinoma

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OBJECTIVES: To present a large series of patients with orbital invasion by periocular basal cell carcinoma (BCC).

DESIGN: Retrospective, noncomparative, interventional case series.

PATIENTS: All cases diagnosed with orbital invasion by periocular BCC between January 1985 and July 2004 in 3 Orbital Units in Australia. **METHODS:** The clinical records of all patients were reviewed.

MAIN OUTCOME MEASURES: Patients' demographics, clinical presentation, histologic subtypes, treatment modalities, recurrence rate, and tumor-related death.

RESULTS: There were 64 patients (49 males) with a mean age of 70.13 years. Most tumors (84.4%) were recurrent or previously incompletely excised, and the medial canthus was most frequently involved (56.2%). Signs suggestive of orbital involvement included a mass with bone fixation (35.7%), limitation of ocular motility (30.4%), and globe displacement (17.6%). There were no signs suggestive of orbital invasion in 35.7%. Most patients (51.6%) had infiltrative histologic findings, and perineural invasion was present in 19.3%. Treatment modalities were mainly exenteration alone or combined with radiotherapy. During a mean follow-up period of 3.6 years, 3 cases of recurrence (4.7%) were diagnosed. Only 1 patient (1.6%) died from tumor-related causes.

CONCLUSIONS: Orbital invasion by periocular BCC is an uncommon event that may be associated with significant ocular morbidity and, rarely, death. Because orbital invasion may often be clinically silent, clinicians need to be alert to the possibility in high-risk tumors and consider appropriate imaging. Surgical treatment with exenteration or excision, with or without radiotherapy, results in a low recurrence and mortality rate.

Primary embryonal carcinoma of the orbit in a 10-month-old female: a seven-year follow-up

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PURPOSE: Extra-gonadal germ cell tumors (GCTs) are rare and can be highly aggressive. If correctly identified and treated with multimodality chemotherapy, their prognosis can be significantly improved. We

examined a 10 month-old female with primary embryonal carcinoma of the orbit.

DESIGN: Case report and literature review.

METHODS: Case study with 7-year follow-up and literature review of intracranial and intraorbital GCT cases. RESULTS: The patient presented with progressive proptosis and ophthalmoplegia. CT scan revealed an orbital apex mass and biopsy demonstrated a nongerminomatous GCT--an embryonal carcinoma. The patient is tumor-free 7 years after multimodality chemotherapy. She has mild amblyopia and a right micro esotropia.

CONCLUSIONS: Nongonadal GCTs of the orbit can occur and should be considered in the differential diagnosis of a young child with proptosis and ophthalmoplegia. Five-year survival rates improve significantly with accurate identification and treatment.

New ophthalmic manifestations of branchio-oculo-facial syndrome

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Am J Ophthalmol. 2005 Feb;139(2):362-4.

PURPOSE: To report new ocular manifestations of branchio-oculo-facial (BOF) syndrome.

DESIGN: Case report.

METHODS: A 10-year-old girl with known BOF syndrome was referred because of a fundus lesion in her left eye.

RESULTS: She had undergone excision of a left orbital dermoid cyst at age 18 months and a branchial cleft fistula from the right side of neck at age 4 years. Examination disclosed openings of sinus tracts on each side of the nose connecting the lacrimal sac to skin. In the right eye, an iris pigment epithelial cyst was confirmed with ultrasound biomicroscopy. In the left eye, there was a combined hamartoma of the retina and retina pigment epithelium.

CONCLUSION: BOF syndrome can display mild to severe craniofacial, auricular, oral, and ophthalmic anomalies. In this case, the ophthalmic manifestations included lacrimal sac fistula, orbital dermoid cyst, iris pigment epithelial cyst, and combined hamartoma of the retina and retinal pigment epithelium.

Carcinoid tumour metastatic to the orbit with infiltration to the extraocular orbital muscle

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APMIS. 2005 Feb;113(2):135-9.

Ninety-three percent of symptomatic patients with small intestinal carcinoid tumours have metastases. The most common sites of metastases are lymph nodes and liver. Orbital metastases have rarely been described and the majority of them involve the choroid rather than extraocular orbital structures. We report a patient who developed proptosis, impairment of vision and reduced ocular motility on the left side, eighteen months after operation for primary intestinal carcinoid tumour with hepatic metastases. CT and MR studies revealed the tumour mass infiltrating the inferior rectus muscle. Biopsy examined by imprint and frozen section showed tumour consistent with metastatic carcinoid. The tumour was removed. HE and staining for cytokeratin, chromogranin, NSE, serotonin, somatostatin and gastrin

showed that the tumour tissue corresponded to that of the primary intestinal carcinoid tumour. Intramuscular orbital metastasis from a carcinoid tumour is a rare occurrence. Diagnosis may be difficult, especially where no evidence of primary carcinoid tumour is present. Metastatic orbital carcinoid should be suspected in patients with a clinical history of carcinoid tumour and who develop ocular complaints and mass lesion in the orbit. Complete surgical removal of the tumour is important for optimal restitution of vision and eye movements.

Upper eyelid oedema from a dumbbell-shaped dermoid cyst

Eibl KH, Kampik A, Hintschich C.

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No abstract available

Prenatal detection of orbital rhabdomyosarcoma

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No abstract available

Congenital orbital and disseminated extrarenal malignant rhabdoid tumor

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A 5-week premature infant boy with tumorous malformations underwent biopsy of two truncal masses and exenteration of the left orbit. Specimens were examined histologically. Histologic reports, slides, and clinical photographs were reviewed. A diagnosis of malignant rhabdoid tumor was made. Malignant rhabdoid tumors can present as local or disseminated neoplastic disease involving the orbit and should be considered in the differential diagnosis of rapidly progressing orbital lesions presenting in early infancy. We review the current classification of rhabdoid tumors and the previous literature on orbital rhabdoid tumors.

Orbital metastasis as the initial finding of breast carcinoma: a ten-year survival

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A 50-year-old woman with a clinically negative breast examination was diagnosed with metastatic breast cancer only after pathologic evidence of an orbital metastasis. She presented with pressure and pain above the right eye associated with enophthalmos and pseudo-Brown syndrome. The diagnosis of breast cancer based on initial ophthalmic findings requires a strong level of suspicion. Multiple reports show that survival in these cases is limited, usually only 1 year after diagnosis. We present a case of 10-year survival after the diagnosis of breast cancer presenting as an orbital mass. These new findings provoke us to continue aggressive systemic and ophthalmic treatment for such patients.

Expanding miragel scleral buckle simulating an orbital tumor in four cases

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Ophthal Plast Reconstr Surg. 2005 Jan;21(1):32-8.

PURPOSE: To describe four patients with an enlarging orbital mass from a swollen MIRAgel scleral buckle that simulated an orbital neoplasm.

METHODS: In a retrospective, single-center case series at the Ocular Oncology Service at Wills Eye Hospital of Thomas Jefferson University, 4 eyes of 4 patients were referred for evaluation and treatment of a suspected orbital tumor.

RESULTS: The initial presenting features were orbital mass (case 1), strabismus (case 2), and conjunctival mass with orbital extension (cases 3 and 4). Each patient vaguely recalled previous uncomplicated retinal detachment surgery 12 to 20 years earlier. Confirmation of the buckling implant material was made with the retina surgeon in 3 cases. A nontender, forniceal conjunctival mass, deep to the Tenon fascia and appearing as a translucent firm elevation was seen in all 4 cases. Axial CT (case 1) revealed a circumscribed anterior temporal orbital mass, believed to be a large inclusion cyst, 4 times thicker than the nasal scleral buckle. Ocular ultrasonography depicted an echolucent mass in the episcleral region (cases 3 and 4) that was 2 times thicker than the nasal scleral buckle (case 3). Excision was attempted in case 1, but only piecemeal removal was achieved, leading to extensive postoperative inflammation and decreased vision. The other 3 cases were followed conservatively without excision because they were each recognized to be a swollen MIRAgel implant and not an orbital tumor.

CONCLUSIONS: MIRAgel scleral buckle material can greatly enlarge over a period of 10 years and simulate an orbital tumor or orbital cyst. Patients often do not recall details of the retinal surgery. Caution is advised regarding excision of this material because it is friable and can lead to extensive postoperative inflammation.

Leiomyoma of the orbit and periocular region: a clinicopathologic study of four cases

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Ophthal Plast Reconstr Surg. 2005 Jan;21(1):16-22.

PURPOSE: Leiomyomas are exceedingly rare tumors of the orbit and periorbital region. Our objective is to describe the clinical, histopathologic, and radiologic features and the management of 4 cases of orbital leiomyomas and to summarize the cases from the literature.

METHODS: This retrospective review describes 4 patients with orbital leiomyoma diagnosed by clinical and histopathologic studies. Electron microscopy was performed in 2 cases. Immunohistochemical techniques were performed with 5 monoclonal antibodies. Selected papers describing well-documented cases of orbital leiomyoma in the English literature published since 1960 were reviewed.

RESULTS: CT and MRI showed well-circumscribed contrast-enhancing mass lesions. Three tumors were completely excised and 1 had subtotal excision. All 4 tumors showed immunoreactivity for actin, desmin, and vimentin. Follow-up examination showed no evidence of recurrence in 3 patients. One patient was lost to follow-up.

CONCLUSIONS: Leiomyoma should be considered in the differential diagnosis of a well circumscribed mass lesion involving the orbit and periorbital region. Immunohistochemistry provides conclusive evidence to confirm the diagnosis.

Periocular hemangiomas in childhood--functional and esthetic results

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Strabismus. 2004 Jun;12(2):103-10.

INTRODUCTION: Hemangiomas are the most common tumors of the eyelids and orbit in childhood. These tumors can produce ptosis, strabismus and anisometropia, resulting in amblyopia. The treatment of hemangiomas is a challenge with respect to the functional results (prevention of amblyopia) and cosmetic outcome.

MATERIAL AND METHODS: The history, clinical findings, magnetic resonance imaging (MRI), management and outcome of 15 children (3-9 months old) with hemangiomas of the eyelids and/or orbital involvement are reviewed.

RESULTS: Seven patients with small superficial hemangiomas were merely kept under clinical observation. In seven other patients with threatened or existing occlusion of the visual axis or refractive errors, treatment was indicated. Four children were given local Neodymium: YAG-laser therapy. In patients with large subcutaneous eyelid hemangiomas and involvement of the orbita we decided to treat with interstitial Neodymium: YAG-laser therapy in combination with systemic corticosteroids. One patient with an unknown tumor in the medial canthus was diagnosed by biopsy. During the follow-up period of 12-24 months, all untreated patients and 6 of 7 treated children showed involution of their tumors. One patient with a large eyelid and orbital hemangioma with occlusion of the visual axis did not respond to laser therapy and systemic corticosteroids; excision of the tumor was necessary. All children with eyelid hemangiomas with orbital involvement (n = 3) suffered from anisometropia with astigmatism and were treated for amblyopia. During 24 months of amblyopia treatment, the visual acuity improved in two of three cases to 0.4 and 1.0.

CONCLUSION: Periorbital hemangiomas must be managed by individual and interdisciplinary diagnostic and therapeutic approaches. Therapy of amblyopia remains mandatory in all cases of large eyelid hemangiomas and/or orbital involvement.

Mucosa-associated lymphoid tissue lymphoma with intraocular involvement

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PURPOSE: To report the clinicopathologic features of a patient with mucosa-associated lymphoid tissue (MALT) lymphoma of the conjunctiva and associated intraocular involvement.

METHODS: This study is a retrospective clinicopathologic correlative case report summarizing the clinical, radiologic, and histopathologic findings of a patient with conjunctival MALT lymphoma and associated intraocular involvement.

RESULTS: Ophthalmic examination and fluorescein angiography demonstrated progressive conjunctival infiltration bilaterally, marked uveal effusions in the left eye, and cellular white infiltrates of the choroid in the right eye. MRI of the orbit revealed a diffusely infiltrating intra- and extraocular lesion extending around the globe and optic nerve in the left eye without evidence of intracranial extension. Conjunctival biopsy showed low-grade tumor cells, consistent with the diagnosis of MALT lymphoma. The patient was successfully treated with external beam radiation with marked clinical improvement.

CONCLUSION: Conjunctival MALT lymphomas, typically indolent and localized tumors, may extend into the orbit and invade local tissues such as the choroid with devastating consequences. A conjunctival biopsy can provide an easy diagnosis of this treatable entity.