Case Reports of Ocular Adnexal Anaplastic Large Cell Lymphoma (ALCL)

Features	Source				
	Segal et al,[5] 1997	Hu et al,[4] 1998	Coupland et al,[3] 1999	Coupland et al,[3] 1999	Current Study, 2002
Clinical					
Age, y	10	63	48	30	16
Sex	Male	Female	Male	Female	Female
Site	Lateral aspect eyelid	Corneoscleral limbus, maxillary sinus	Eyelid	Multiple	Fornix
Immune profile	UCHL-1, CD3, and Ber-H2	CD45RO	CD45, CD3, CD30, and ALK-1	CD45, CD3, and CD30	UCHL-1, CD30, and ALK-1
Diagnosis	ALCL	T-cell lymphoma of diffuse large cell	ALCL	ALCL	ALCL
Systemic or local/stage	Local/stage II	Systemic/stage IV	Local	Systemic	Systemic
Therapy	Excision; CHOP, methotrexate, and bleomycin	Cyclophosphamide, mitoxantrone hydrochloride, vincristine, methylprednisone, and 13-*cis* retinoic acid	CHOP; local irradiation	CHOP and VIP	CHOP and allopurinol
Follow-up	Disease free at 9 mo	NA	Disease free at 4 y	Dead	Dead

Abbreviations: ALK-1, anaplastic lymphoma kinase; CHOP, cyclophosphamide, doxorubicin hydrochloride, vincristine sulfate, and prednisone; NA, not available; VIP, vincristine, ifosfamide, and prednisone.

tion is made of CD30 staining. The microscopy illustrated in the text is consistent with anaplastic large cell lymphoma. The case we report is also of the T-cell type. Of the 2 cases reported by Coupland et al,[3] only 1 was ALK positive.

Initial symptoms of conjunctival lymphoma may include a mass, irritation, ptosis, epiphora, blurred vision, proptosis, or diplopia; thus, the diagnosis requires a high index of clinical suspicion.[8] It is important to consider lymphoma in patients with unresponsive chronic conjunctivitis. Of the ocular adnexal lymphomas, conjunctival involvement is purported to have the best prognosis, with stage and category being determinants of outcome. In the series of 117 cases reported by Shields et al,[8] the midbulbar and fornix conjunctiva were noted to be the most common sites. Factors predictive of the presence or development of systemic lymphoma are location of the tumor at an extralimbal site (fornix midbulbar conjunctiva) and multiplicity of tumors. Immunohistochemical detection of *p53* and high Ki-67 (MIB1) expression also correlate with disease stage at the initial visit and aggressive disease course and mortality. In our case, the MIB1 proliferation index was high, 70%, and scattered nuclei were positive for *p53*.

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Optic Neuropathy and Macular Chorioretinal Folds Caused by Orbital Cherubism

Cherubism is a rare fibro-osseous disease of the maxilla and mandible usually seen in childhood. Orbital manifestations are proptosis, lower eyelid retraction, and upward displacement of the globes. Previous cases of orbital cherubism with unspecified visual change have been documented.[1,2] We present a case of orbital cherubism with visual loss directly attributable to optic neuropathy and macular striae/scarring that resulted from the effect of the mass or tumor pushing on the eye of the orbital lesion.

Clinicopathologic Report. A 31-year-old hispanic woman with a known history of cherubism was referred by

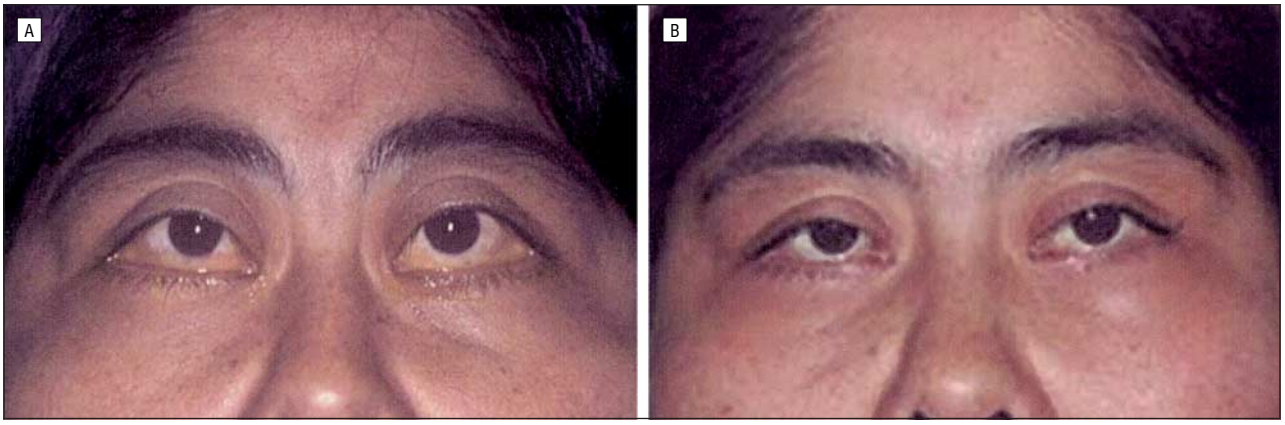

Figure 1. A, Preoperative photograph. B, Photograph taken 4 months postoperatively. Note the improvement in scleral show and decreased upward and inward displacement of the globes.

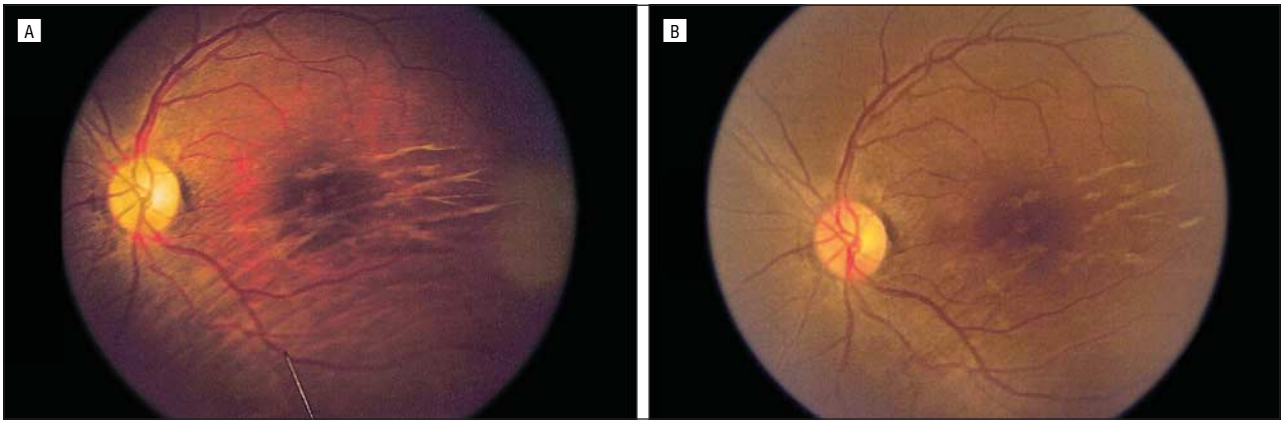

Figure 2. A, Preoperative fundus photograph. B, Postoperative photograph. Note the improvement in macular striae/scarring. Also, there is temporal pallor of the optic nerve.

the Oral Maxillofacial Surgery Service to the Department of Ophthalmology for evaluation of decreased vision in her left eye. She had undergone mandibular osteoplasty 3 months earlier for treatment of facial deformity secondary to cherubism.

When we saw her (**Figure 1**, A), the patient complained of mildly decreased vision in her left eye, intermittent diplopia on far lateral gaze, and tearing. She also admitted to intermittent photopsias and floaters of 4 months' duration. On examination, her corrected visual acuity was 20/20 OD and 20/30 OS with a correction of –1.00+1.50×90 OD and –1.00+1.75×120 OS, respectively. She had a relative afferent pupillary defect in her left eye and scored 15/15 OU on Ishihara color plate testing. Hertel exophthalmometry readings were symmetrical at 18 mm OU at a base of 87 mm. Extraocular motility was diminished on upgaze and lateral gaze in both eyes. Examination of the eyelids revealed a margin-reflex distance of 1.5 mm OD and 2 mm OS with good levator function. She displayed 3 mm of inferior scleral show with 1 to 2 mm of lagophthalmos in both eyes. Results of slitlamp examination revealed inferior punctate keratopathy with mild conjunctival hyperemia in both eyes. The remaining anterior segment examination findings were within normal limits. Funduscopic examination results were unremarkable in the right eye and showed temporal disc pallor and macular chorioretinal folds and scarring in the left eye (**Figure 2**, A). Cranial nerves II through VII were tested and intact. Humphrey visual fields were performed and revealed a generalized depression in the left eye with a relative paracentral scotoma (mean deviation, –14.34 dB, $P<0.5\%$). The visual field in her right eye was within normal limits (mean deviation, –6.01 dB, $P<0.1\%$).

Computed tomographic scans of the sinuses/orbits were performed and showed an expansile, at least partially osseous, mass invading the maxillary sinuses and the orbits bilaterally, with resultant remodeling of the orbits (**Figure 3**, A and B). The orbital extension of the mass was greater on the left than the right, with resultant medial and superior shift of orbital contents. The optic nerves were displaced medially and were draped across the mass. The mass abutted on the globe posteriorly. These findings were unchanged from the scans performed 3 months earlier by the Oral Maxillofacial Surgery Service.

It was decided to perform staged orbital decompression surgery, left eye followed by the right eye, with curettage of the lesion. After obtaining informed consent from the patient, preoperative embolization of the internal maxillary arteries and the transfacial branch from the superficial temporal artery was

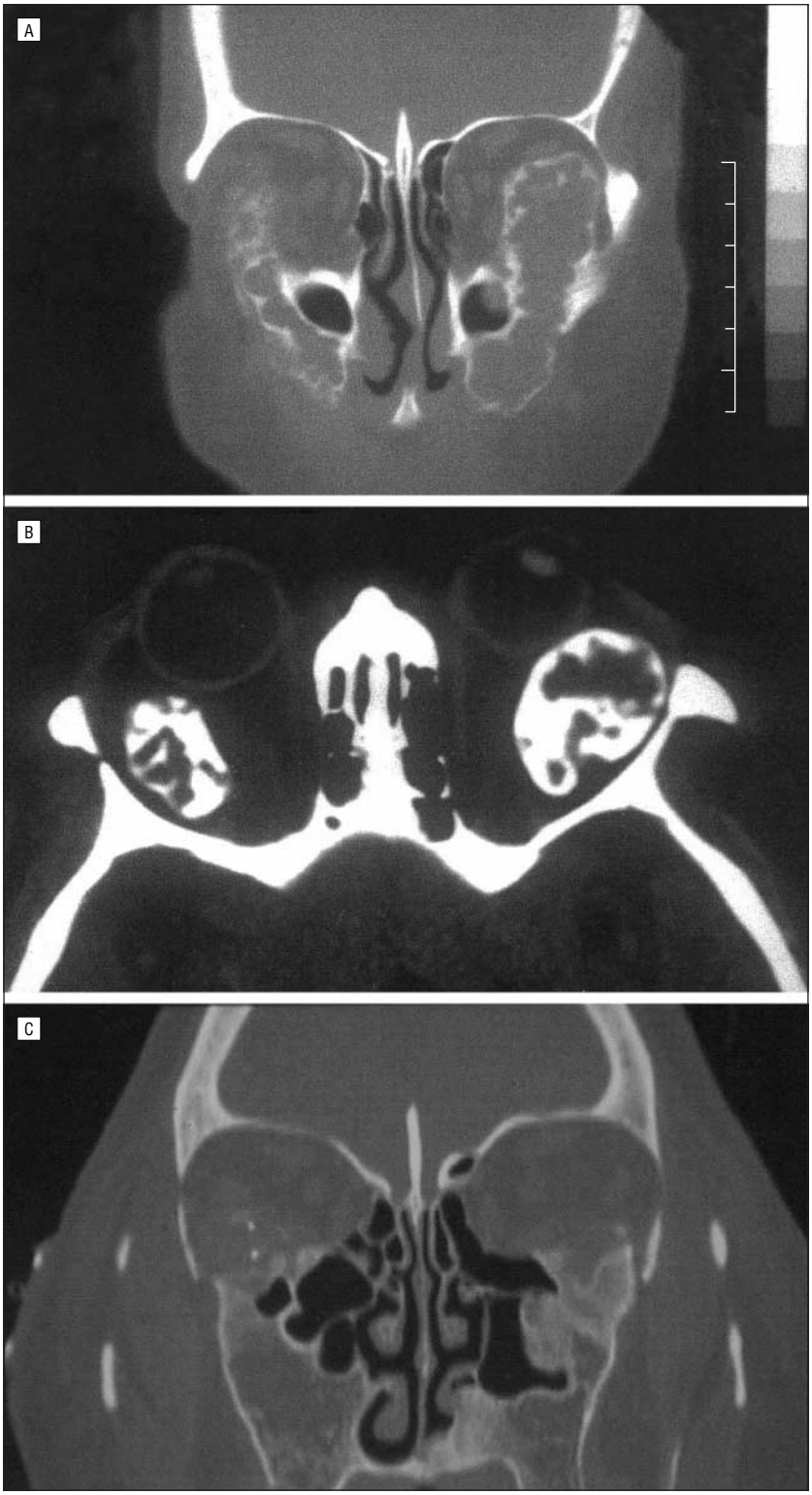

Figure 3. A, Preoperative coronal computed tomographic (CT) scan. B, Preoperative axial CT scan. C, Coronal CT scan 2 days postoperatively. Note the decreased height of the maxillary fibro-osseous columns.

performed to optimize intraoperative hemostasis.

A transconjunctival approach was used to reach the inferior orbital rim periosteum. The periosteum of the floor was reflected off the bone using Freer elevators, and a 2×3-cm bony mass was visualized. Using a reciprocating saw blade and osteotomes, an osteotomy was performed and the bony mass was debulked and sent for histopathological diagnosis. The floor was then smoothed down using a reciprocating rasp. The wound was then irrigated with isotonic sodium chloride and suctioned dry. Hemostasis was achieved with the help of bone wax. The conjunctiva was closed with multiple interrupted 6-0, fast-absorbing plain gut sutures. Intermittent pupillary checks performed during surgery encountered no abnormalities.

On histopathology, sections showed lamellar and woven bone, osteoblast proliferation, and cellular zones composed of spindled, fibrous stroma with numerous, scattered multinucleated giant cells (**Figure 4**). The findings were consistent with the clinical diagnosis of cherubism.

Postoperative CT scans were performed and revealed debulked orbital masses with decreased proptosis (Figure 3, C). On ophthalmic examination 4 months after surgery, her visual acuity had remained unchanged. Hertel exophthalmometry readings were unchanged, but her dystopia had resolved. Although the macular folds had improved, some residual scarring remained (Figure 2, B). Her facial appearance improved with her globes and eyelids returning to a normal anatomic position (Figure 1, right).

Comment. Cherubism is a fibro-osseous disease of childhood characterized by bony expansile lesions of the maxilla and mandible. Jones[3] coined the term "cherubism" after the angelic features seen in Baroque art of the Renaissance period. These figures had round, full-cheeked faces whose eyes "turned up to the heavens."[4] The mode of transmission is autosomal dominant with 80% to 100% penetrance and variable expressivity.[5] The gene was localized recently to chromosome 4p16.3 in a study of 2 affected families.[6] Patients affected with this disease are generally without these distinguishing characteristics at birth. Onset of the disease process occurs between 2 and 4 years of age with painless expansion of the mandible and occasionally the maxilla. This

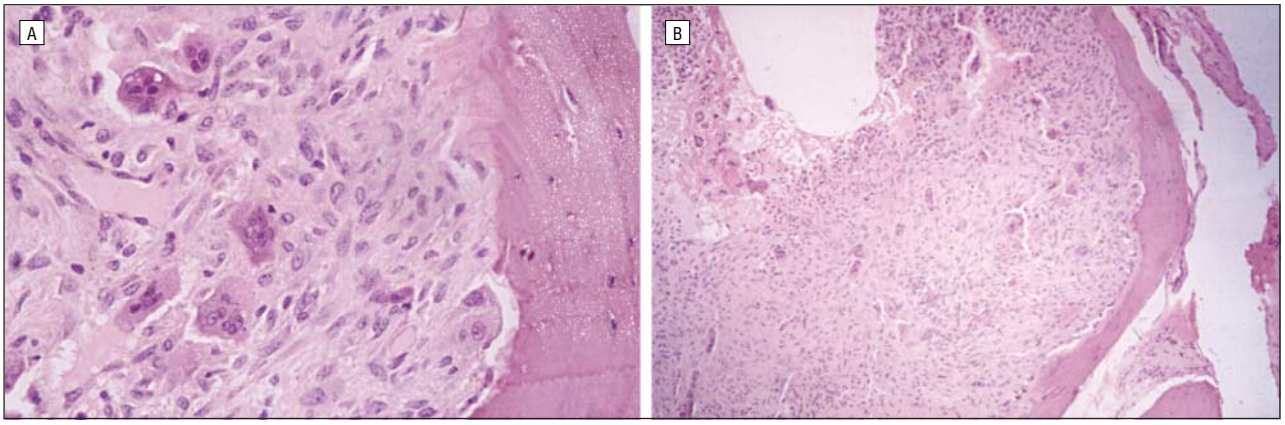

Figure 4. Fibrous stroma with scattered multinucleated giant cell adjacent to bone seen on histopathological examination (hematoxylin-eosin, original magnification: A, ×40; B, ×10).

causes facial deformities and abnormalities of dentition.[3] The disease generally progresses until puberty and may regress in adulthood.[4] Radiologically, the radiolucent bony lesions are cystic and multiloculated in nature and are bilateral.[4] These findings in conjunction with the clinical presentation are usually enough to make a diagnosis. Unless surgical intervention is needed for genetic or reconstructive purposes, a confirmatory biopsy is not obligatory. If a biopsy is performed, the histopathological differential diagnosis of lamellar bone formation with fibrous stroma and giant cell formation includes fibrous dysplasia, aneurysmal bone cyst, central giant cell granuloma of bone, and brown tumor of hyperparathyroidism.[4] However, on the basis of clinical presentation and laboratory data, alternative diagnoses can be ruled out. Indications for treatment include functional problems, such as dental abnormalities and visual compromise, and the need for cosmesis. Treatment of choice is surgical curettage and contouring.[7] Radiation therapy is not recommended because of risks of osteonecrosis and sarcoma transformation.[5]

We present a case of cherubism with documented visual loss secondary to optic neuropathy and macular chorioretinal folds/scarring directly attributable to compression from the fibro-osseous growth within the orbit. To our knowledge, only 3 other cases of cherubism have been described in the ophthalmic literature.[2,4] None of these cases presented with a relative afferent pupillary defect indicative of optic neuropathy. Although Hawes[2] presented a patient with visual loss secondary to macular scarring, he did not attribute this to globe compression from the fibro-osseous mass. Since most reports on this disease are presented in the oral maxillofacial and otorhinolaryngology literature (MEDLINE search of the past decade reveals 2 articles in ophthalmology journals and 37 in the two aforementioned fields), the focus is not on ophthalmic manifestations of disease and etiology of visual loss. It is therefore quite possible that visual loss from optic neuropathy and/or maculopathy secondary to cherubism is underestimated. As such, we feel that routine examination by an ophthalmologist be recommended in the management of a patient with cherubism.

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Endophthalmitis Caused by *Mycobacterium chelonae*: Selection of Antibiotics and Outcomes of Treatment

Mycobacterium chelonae is a rapidly growing acid-fast bacterium that has been reported to cause keratitis,[1] interface infection after laser in situ keratomileusis,[2,3] scleral buckle infection,[4] keratitis in a corneal graft,[5] and periocular infection after dacryocystorhinostomy and ptosis repair.[6] In a review of the literature, a total of 4 cases of endophthalmitis caused by *M chelonae* have been reported.[7-10] The current study reports 5 cases of culture-proved endophthalmitis caused by *M chelonae* at Bascom Palmer Eye Institute, Miami, Fla, between January 1, 1980, and December 31, 2001.

Report of Cases. *Case 1.* A 62-year-old man received an intravitreal injection of triamcinolone acetonide (4 mg/0.1 mL) in October 2001 for clinically significant macular edema reducing visual acuity to 20/70 OD.