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The excised skin tumour, measuring 55×25×25 mm, was ulcerated, nodular, and crusted. On section, the tumour was well circumscribed and had a chalky white appearance with small foci of calcification (Fig 2). Histological examination showed two main cell types: basophilic cells located peripherally in cell islands and showing occasional mitotic figures and eosinophilic shadow cells, found towards the central areas of the cell masses, with no nuclear staining. Intervening stroma composed of small blood vessels and a scattering of mixed inflammatory cells. Overall, the features were those of benign pilomatricoma, which was well circumscribed and appeared to have been completely excised. The wound healed very satisfactorily without further surgery, and she was pleased with the cosmetic result.

COMMENT

The tumour in the case presented was of uncertain diagnosis clinically, partly as a result of the patient’s age. Differential diagnosis included basal cell carcinoma, squamous cell carcinoma, keratoacanthoma, and neurofibroma. Other less typical aspects of this case were the rapid growth rate and large size of the tumour. In view of the possibility of malignancy, it was felt to be important to establish a histological diagnosis. Incisional biopsy, if small, could have led to a misdiagnosis of basal cell carcinoma, owing to the pleomorphism of pilomatricoma with basophilic cells present peripherally, and would have required subsequent definitive surgery. We opted for excision biopsy with a narrow margin of clearance as the primary procedure.

Once the lesion was established as benign, no further surgery was necessary and the wound healed by secondary intention. Mohs’ surgery would have been an alternative but lengthier procedure.

We endorse the advice of Simpson et al.2 regarding the importance of making a tissue diagnosis before undertaking definitive surgery in cases such as this, where the clinical diagnosis and nature of the lesion are uncertain. Thus, for benign hair follicle tumours, wide margins of excision are avoided although for the common basal cell carcinoma they would be required. Notably, over a 30 year period at the Institute of Ophthalmology, there were 2447 cases of basal cell carcinoma compared with 94 cases of pilomatricoma.2

Less commonly, giant pilomatricoma has been described which behaves more aggressively and shows rapid growth rate and larger size, the latter features shared with this case.4 Pilomatricoma carcinoma occurs rarely and is diagnosed on the basis of frequent mitoses in the basaloid cells and invasion of adjacent fat, muscle, and blood vessels.6

The case reported here serves as a reminder that pilomatricoma has a predilection for the upper eyelid or eyebrow and may occur at any age, although it is commoner in children. If these cases are to be spared the greater cosmetic disfigurement of wide margin excision the importance of first establishing a diagnostic biopsy is re-emphasised.

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ECtopic teeth in the orbit of a neonate

EDITOR,—Congenital orbital teratomas are rare tumours derived from more than one germinal layer.1 The inclusion of dental elements in such an orbital tumour has rarely been reported as a bizarre finding.1 2 We encountered an intracranial primary orbital teratoma with presence of mature dental elements in the orbit and preservation of good visual function after surgery.

CASE REPORT

A full term baby boy was born following an uneventful pregnancy and vacuum delivery. He was small for dates and developed jaundice for which he was treated in hospital. During his stay some prognosis of the right eye was noted. No pupillary or motility abnormalities were present. Ultrasonography of the orbit revealed the presence of a cystic lesion superotemporally. Computed tomography (CT) scan confirmed the presence of the cyst and showed calcifications in the orbital apex as well (Fig 1). Since the lesion was retrobulbar and the anterior route was unlikely to provide proper access for intact removal, a lateral orbitotomy with the presumptive diagnosis of dermoid cyst was performed. The calcific lesions were seen at the apex but could not safely be removed. Histology showed the presence of surface ectoderm (squamous epithelium), with sweat and sebaceous glands. Striated muscle was present as well as mucous glands.

The postoperative period was uneventful until 2 months after the operation. A recurrence of the cyst was clinically suspected and confirmed with a CT scan. A transcranial superior orbitotomy was performed and the abnormal tissues were removed, including two mature teeth (Fig 2). Light microscopy showed the presence of ectodermal tissue (squamous epithelium, epithelium with basaloid features, teeth incisors which show differentiation corresponding to the age of the patient), and mesoderm (prominent connective tissue, bone fragments).

Twelve months postoperatively the child exhibits only slight ptosis and slightly reduced elevation of the right eye. There was no afferent pupillary defect and visual acuity was equal to the contralateral eye as measured with the Bock candy bead test.

COMMENT

The orbit is a rare, but well known location of congenital teratoma. These choristomas are composed of tissues of at least two germinal layers. Tissue types present in the lesion may be differentiated to a varying degree. In a bizarre case, described by Mizuo,3 and cited by Kivela and Tarikanen4 a teratoid fetus was found. Tissue differentiation to a mature degree is unusual.

Orbital teratomas are usually present at birth, although adult cases have been described. The neonate typically presents with moderate (but obvious) to massive proptosis and its sequelae, such as lagophthalmos and corneal ulceration.5 Ocular motility is moderately to markedly reduced, whereas visual function is very poor. One case has been described in which useful vision was preserved.5 The salvage of vision, and indeed the eye, is related to the age of the tumour. Management used to comprise exenteration, although the introduction of microscopic
operative techniques has resulted in less aggressive treatment with preservation of the eye.

Our case demonstrates some unusual features of orbital teratoma. The presence of globe displacement was not very obvious with the eye closed. In the presence of tumour mass in the orbital apex, ocular movements were still full, whereas visual function did not appear to be severely reduced. Following complete excision of the tumour, vision had not changed clinically. A high index of suspicion in the presence of a cystic lesion in the neonate, even in the absence of obvious proptosis, particularly in the presence of calcifications, may lead to the diagnosis of orbital teratoma. Surgery by a route allowing complete excision should be planned as soon as possible.

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5 Chang DF, Dallow RL, Walton DS. Congenital and acquired lacrimal gland fossa mass (arrow). This mass, located anteriorly, displaced the globe downward and appeared oblong.

Orbital coccidiomycosis presenting as a lacrimal gland fossa mass

EDITOR—A wide spectrum of pathological processes such as tumour, infections, and inflammatory diseases can present as a mass in the superotemporal orbital region. The clinical history and radiographic findings often provide the clues necessary for diagnosis and planning of treatment. Here, we report an unusual case with a lacrimal gland fossa mass caused by Coccidioides immitis, a fungus endemic to the southwestern United States, Mexico, and Central and South America.

CASE REPORT

A 36-year-old Filipino man with no history of head trauma presented with a 2 week history of pain and proptosis of the right eye, accompanied by intermittent fevers and chills. Medical history was significant for a mild leucocytosis phy of the orbits showed an increase in the right eye and 14 mm in the left eye. His grew.

Visual acuity was 20/20 in each eye, and anterior and posterior segments appeared normal. A trial dose of intravenous amphotericin B (50 mg intravenously each day for 3 weeks), oral fluconazole (800 mg/day) was used as a maintenance agent upon discharge. One year after the initial presentation, the patient remained asymptomatic and a repeat CT scan of the orbit was performed which revealed resolution of the mass.

COMMENT

Coccidiomycosis is a pathogenic, endemic fungal disease caused by the organism Coccidioides immitis. The ocular manifestations of coccidiomycosis are many and can involve certain extracapsular or intracapsular tissues. However, to our knowledge lacrimal gland invasion by the organism has not been reported previously. Our patient presented with a painful proptosis suggestive of a bacterial orbital cellulitis, for which he was treated with intravenous antibiotics. The tentative diagnosis of dacrocyoadenitis was based on the moderate improvement with intravenous antibiotics and the CT scan findings which were consistent with an inflammation of the lacrimal gland. The usual incidences of lacrimal gland masses are benign mixed tumour (12%), dacryocystitis (6%), malignant epithelial tumours (4%), inflammatory pseudotumour (idiopathic orbital inflammation) (50%), sarcoid (13%), lymphoid masses (13%), and acute dacryo­adenitis (1%).

Orbital infections that involve the lacrimal gland initially produce only non-specific inflammatory changes on CT scan, such as thickening of the scleral-uveal rim, enhancement of the lacrimal gland, or thickening of an extracapsular muscle, changes that often mimic the CT findings in idiopathic orbital inflammation in the lacrimal gland region. An abscess can be clearly delineated by CT scan only when necrotic tissue is present within encapsulated fibrous tissue. Since a CT scan may not reveal an abscess during the initial presentation, orbital ultrasound has been recommended when an abscess is suspected. In a review of 15 cases with orbital abscess, ultrasound consistently revealed well defined lesions of low internal reflectivity with high reflective borders. Balchunas et al examined 32 patients with lacrimal gland/fossa masses who had CT scans and/or standardised A-mode echography (SAE), and concluded that the initial examination of a lacrimal gland/fossa mass should begin with SAE since its specificity exceeds that of CT scans in this region. However, many orbital surgeons prefer to rely on CT scanning.

The current treatment of choice for ocular coccidiomycosis is intravenous amphotericin B, which has been proved to be effective in the treatment of extrapulmonary dissemination of Coccidioides immitis. Drainage is indicated if the abscess is large or if it is