absent b-wave. Type II CSNB is further divided into complete and incomplete types. Complete type CSNB shows refractive error ranging from moderate to high myopia and corrected visual acuity ranging from 20/200 to 20/25. Dark adaptation shows absent rod-dark adaptation. On single flash ERG, oscillatory potentials are absent. The scotopic ERG is extinguished whereas the photopic flicker responses are normal or subnormal. Accordingly, this patient was diagnosed as complete type CSNB.

Patients having osteopetrosis with visual disturbance caused by compression of the optic canal have been recommended for optic nerve decompression. However, primary retinal degeneration has been reported as a cause of visual disturbance; three cases of reduced or extinguished ERGs, two cases of macular degeneration, and a case of marked atrophy of the rod and cone layer with degenerative outer nuclear layer. In our case, CSNB was suggested to be the cause of visual dysfunction. Electrophysiological examination is important in detecting retinal dysfunction in patients with osteopetrosis.

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An unusual impediment to spectacle wear: pilomatrixoma

EDITOR—Pilomatrixoma or calcifying epidermoid of Malherbe is a relatively uncommon benign skin tumour arising from cells of the hair follicle. Sixty per cent present during the first two decades of life although pilomatrixoma can occur at any age. It is more common in females by a ratio of 3:2. Although pilomatrixoma has a predilection for the upper lid and eyebrow, each ophthalmologist will see a relatively small number. A case of pilomatrixoma is presented with discussion of its management.

CASE REPORT
A 48-year-old woman developed a 'thumb sized' lump on her left upper eyelid over the course of 1 year. Because of a pathological fear of doctors and hospitals she had delayed her presentation until the size of the lump interfered with her wearing of spectacles.

She attended the clinic with a gauze dressing attached to her spectacle frame with which she had been attempting to conceal the mass's increasing bulk. On examination she was found to have a very large tumour, approximately 6 cm long and 3 cm in diameter, attached by a broad base to the left upper eyelid and anterior aspect of the temporal region. Granulation tissue was evident on the surface with a purulent discharge (Fig 1A). There was no evidence of local or systemic metastasis.

She underwent primary diagnostic excision of the tumour, and since the patient insisted upon local anaesthesia and day-case surgery, the tissue defect was left to heal by secondary intention (Fig 1B).
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 stromal components contained blood vessels and a scattering of mixed inflammatory cells. Overall, the features were those of benign pilomatrixoma, 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 pilomatrixoma 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. Had the lesion proved malignant, secondary wider excision of the wound margins would have been required, probably with skin grafting to repair the larger defect once the margins were clear. Mohs' surgery would have been an alternative but lengthy procedure.

We endorse the advice of Simpson et al, 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 commoner basal cell carcinoma they should 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 pilomatrixoma.

Figure 2 A gross photograph of the pathological specimen demonstrating the cut surface of the excised lid tumour which highlights the chalky white appearance characteristic of pilomatrixoma.

This, commonly, giant pilomatrixoma has been described which behaves more aggressively and shows rapid growth rate and larger size, the latter features shared with this case. Pilomatrix 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.

The case reported here serves as a reminder that pilomatrixoma has a predilection for the upper eyelid or eyebrow and may occur at any age, including 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. Two infants with an unusual 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 which he was treated in hospital. During his stay some proptosis of the right eye was noted. No pupillary or motility abnormalities were present. Ultrasonography of the orbit revealed the presence of a cystic superotemporal lesion. 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 allow proper access for intact removal, a lateral orbitotomy with the presumptive diagnosis of dermoid cyst was performed. The calcific lesions were seen at the border but could not be safely 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 basa- loid 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 proptosis 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,¹ and cited by Kivelä and Tarkkanen² 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 lacrimal glands and corneal ulceration. ³ Ocular motility is moderately to markedly reduced, whereas visual function is very poor. One case has been described in which useful vision was preserved. ⁴ 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