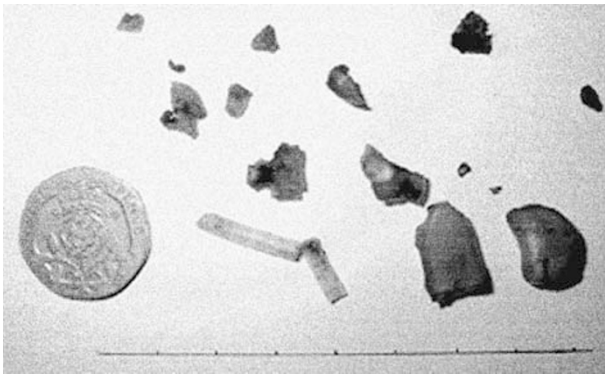

Figure 2 Fragmentation of Miragel explant.

swelling and disintegration.[1] This process must be a very slow one, as typically many of these complications only appear 7–10 years after the initial scleral buckle operation.[1–4]

Comment

Using scissors and forceps in the conventional manner, the removal of Miragel may be difficult, as the explants disintegrate when grasped by instruments. In our patient, the explant was associated with massive periocular fibrosis and extensive dissection was required to free the globe from its mechanical restriction. Our case illustrated that the symptoms can be slowly progressive and that some complications tended to be delayed for many years. We may only be starting to see a number of these cases in this country, even though the explants were withdrawn some years ago. Where Miragel explants had been used extensively in the past, eye units should be aware of the possibility of late complications.

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Sir,

Acute visual loss as an early manifestation of metastatic neuroblastoma

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Neuroblastoma is the most common extracranial solid tumour among paediatric patients. Visual loss is uncommon as an early clinical feature of metastatic neuroblastoma. We report a case of metastatic neuroblastoma and discuss the possible aetiology of visual loss.

Case report

A $2\frac{1}{2}$-year-old boy with a 3-week history of general malaise, irritability, and anorexia was suspected to have encephalopathy. When he failed to improve on medical therapy, he was referred to the regional paediatric neurology unit. Initial investigations carried out at his local hospital, including CT scan and EEG, were reported to be normal. On admission, he was lethargic, irritable but apyrexial, and there were no focal neurological signs. After 2 days, concerns were raised regarding the child's vision and he was referred for an ophthalmic opinion. Ophthalmic examination was difficult due to the child's irritability. He appeared to have profound visual loss to a level of hand movements or worse in both eyes. The left pupil was dilated and poorly reactive, but the right pupil was normal. The discs appeared normal and the ocular movements were full. Visual evoked potentials showed absent responses with normal electroretinograms. Repeat neuroimaging studies demonstrated a large mass in the sphenoid region compressing both optic nerves at the orbital apices (Figure 1). The first clue of neuroblastoma emerged on urinary screening for organic and amino acids. Elevated urinary catecholamine metabolites were found (vanillyl mandelic acid), which was confirmed on repeat samples. After 2 days the right pupil had become involved. He was treated with high-dose intravenous

methyl prednisolone. CT scan of the neck, chest, abdomen, and pelvis demonstrated enlarged cervical and paratracheal lymph nodes and a left paravertebral lesion in the thoracic area. The bone marrow aspirates were found to have between 5 and 8% neuroblastoma cells that were CD56 positive, CD90 positive, and CD45 negative. The CT-guided pernasal biopsy also confirmed the diagnosis of neuroblastoma (Figure 2). Bone scintography revealed multiple areas of skeletal abnormality. A diagnosis of Stage 4 neuroblastoma without an identifiable primary tumour was made.

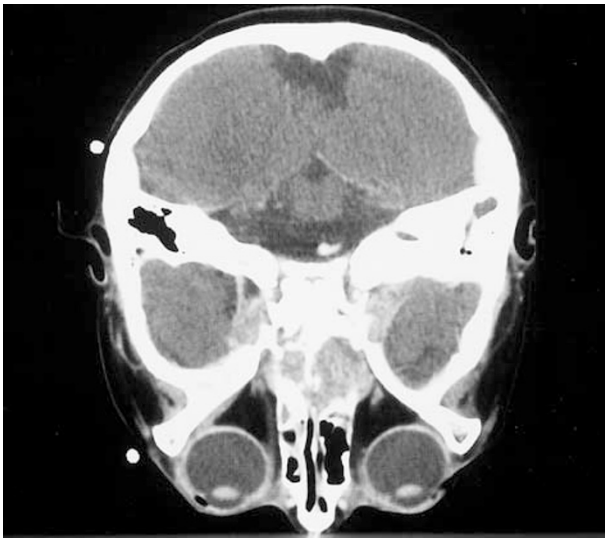

Figure 1 CT scan of the head showing a large mass in the sphenoid region compressing both optic nerves at the orbital apices.

Multiagent chemotherapy was commenced and he was referred to Birmingham Children's Hospital, where he received a course of high-dose retinoic acid. Following treatment, his general condition improved and his vision was stable at 6/60 in the right eye on Logmar charts, but his left eye remained at no perception of light. There was bilateral disc pallor, left more than right. At 2 years follow-up, the child is making good progress and is currently awaiting a repeat MRI scan.

Comment

Neuroblastoma most frequently affects children below the age of 2 years, with 72% of tumours arising in the abdomen, 13% in the mediastinum, and in some cases of metastatic neuroblastoma the primary site is unknown.[4] Three major eye signs of neuroblastoma are proptosis, Horner's syndrome, and opsoclonus.

Olfactory neuroblastoma, also known as esthesioneuroblastoma, resembles neuroblastoma histologically, but originates in the nasal or sinus region. It is uncommon in children, slow-growing, and rarely develops multiple distant metastasis. Catecholamines in the urine and serum are rarely elevated in esthesioneuroblastoma, as they are often in classical neuroblastoma.[1–3] Esthesioneuroblastoma as a diagnosis is unlikely in our case due to age of onset, lack of chronic progressive nasal symptoms or signs, widespread tumour dissemination, and elevated urinary catecholamines.

Worner[3] reported a 2-year-old boy with olfactory neuroblastoma who presented with sudden blindness. Unlike our patient, 3 weeks prior to visual loss he

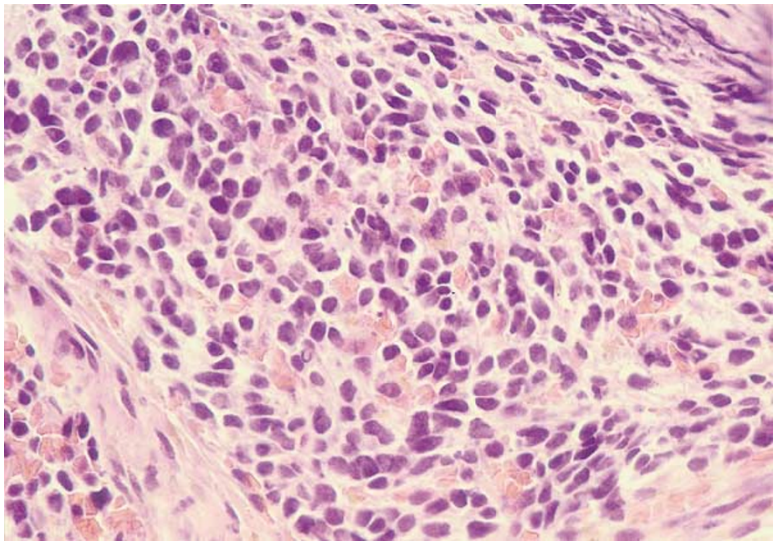

Figure 2 Microscopic view of the sphenoidal neuroblastoma showing discohesive sheets of intermediate-sized cells with minimal cytoplasm and irregular nuclei (haematoxylin and eosin × 400).

suffered from epistaxis, intermittent vomiting, and bilateral proptosis. In a large series by Belgaumi *et al*,[4] visual loss was present in 15 out of 450 patients treated for neuroblastoma, and occurred during the first week in only five of these patients with the rest occurring either during therapy or at the time of recurrence of the tumour. The cause of visual loss in some of these patients was considered to be a paraneoplastic manifestation.[5] Visual loss in our patient was caused by a large sphenoidal lesion compressing both optic nerves.

This case underlines the varied clinical manifestations of undiagnosed neuroblastomas. At presentation, the child was generally unwell, irritable but apyrexial, and visual failure occurred rapidly over 5 days. Neuroblastoma should be considered in this setting. Prompt diagnosis in a substantial number of these patients can be made by a simple and noninvasive urinary screening test for catecholamine metabolites.

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Sir,

Primary nonfamilial ocular amyloidosis

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Ocular amyloidosis is a rare cause of vitreous haze[1,2] and should be included in the differential diagnosis of vitreous opacification. The ophthalmologist should be aware that ocular amyloidosis may occur sporadically and that vitreous biopsy with specific staining with Congo red dye may be indicated.

Case history

A 62-year-old female presented with a 2-year history of floaters in her left eye. Visual acuity was 'counting fingers' in the left eye and 6/6 in the right. Anterior segment examination and pupil reactions were both normal. In the left vitreous there were dense opacities obscuring the retina (Figure 1a), while in the right eye the only abnormality noted was a patch of vitreous condensation overlying the inferior arcuate vessels (Figure 1b). Medical history and examination was unremarkable and there was no significant family history. Serum angiotensin converting enzyme levels, syphilis serology and immunological investigations for HLA B27 markers and autoantibodies were normal. An ultrasound scan of the left eye showed that the retina was flat and there were no intraocular masses.
A vitrectomy with vitreous biopsy was performed. The specimen was stained with haematoxylin and eosin and reported to show no signs of malignancy or infection. Postoperatively the left retina appeared to be normal and the visual acuity improved to 6/6.

A year after surgery the visual acuity had deteriorated to 6/18 in both eyes. Vitreous opacities had recurred in the left eye and similar opacities were apparent in the right. A right vitrectomy and vitreous biopsy were performed, with the cytology specimen being specifically stained with Congo red dye for amyloid (Figure 1c). A diagnosis of amyloidosis of the vitreous was confirmed. A full history and examination failed to identify any