chest and lower limb radiographs, abdominal ultrasound and barium meal follow up were normal.

A left parietal craniotomy was carried out and a circumscribed firm mass 2 cm in diameter was palpated under a widened sulcus. When the operated the mass was purulent and contained a 2.5 cm long motile thread-like worm. The mass and worm were excised. Postoperatively the patient gradually recovered from the hemiparesis and is free from seizures and dystonia at two years follow up.

Sections studied showed microabscesses with necrotic material, neutrophils, eosinophils and lymphomononuclears (fig 2). The parasite was identified as that of A cantonensis (fig 3).

Angiostrongylus cantonensis is a metstrongylid nematode with the rat as a definitive host, and slugs and snails as intermediate hosts. Human infection is an example of aberrant parasitism and is caused by ingesting raw or insufficiently cooked food containing the third stage larvae.

Cerebral angiostrongyliasis usually has an incubation period of two weeks with headache, low grade fever and meningeal signs as the common presenting features. Watts reported five cases of this infection with epileptic seizures in one case. Schmutzhard et al reported five patients with A cantonensis meningoencephalitis with one case having bilateral abducens nerve palsy and unilateral papilloedema. She had been known to have Klippel-Feil syndrome since the age of four when she presented with a Sprengel's deformity of the right shoulder.

She had a short webbed neck with a low hairline which exfoliated as far as the C7 vertebral prominence. No occipital swelling was present, but a small punctum was present just above theunion. Neurological examination revealed vertical nystagmus, dysdiadochokinesia, and mirror movements of the hands.

Radiographs of the cervical spine (fig 1) showed ossification of the posterior arch of C1, with the anterior element apparently fused to C2, posterior fusion of C2 and C3, and anterior and posterior fusion of C4-6. A chest radiograph showed fusion of the posterior ends of the first and second ribs on the right, a left hemivertebra at D3 and a scoliosis concave to the right.

CT scan of the skull (fig 2) showed a large midline hypodense mass lesion in the posterior cranial fossa with peripheral flecks of calcification, and likely compression of the inferior pons and medulla and anterior displacement of the fourth ventricle. There was moderate dilatation of the upper part of the fourth, third and both lateral ventricles, and a small midline defect in the posterior falx extending into the occipital bone with adjacent thickening. Appearances following the injection of intravenous contrast were unchanged.

At operation she was seen to have a tract with thick walls which ran inferiorly through the occipital bone into a large dermoid cyst which filled the cisterna magna, causing upward displacement of the cerebellar hemispheres. The posterior arch of C1 was absent.

The cyst was opened and decompressed. A portion of the cyst capsule was adherent to the upper spinal cord and medulla and was not removed. Histologically the cyst capsule was lined with stratified squamous epithelium. The cyst contained hair and scattered foci of calcification. The patient had no complications from surgery and remains well.

Congenital fusion of the cervical vertebrae is due to failure of normal segmentation of the cervical somites during the third to eighth week of gestation. Similarly, dermoid cysts originate during the third to fifth week, when cleavage of the neuroectoderm from the epithelial ectoderm along the mid-dorsal aspect of the embryo may be incomplete. The resultant persistent cutaneous defect may extend from the skin into the substance or central canal of the cervical spine and expand into a cyst containing sebaceous material, hair and epithelial debris. Posterior fossa dermoids may present with symptoms and signs of a mass lesion or with staphylococcal menigitis. The path of infection is via the dermal sinus which may be seen as a small punctum or diploe over the occiput.

We are aware of four previously reported cases of posterior cranial fossa dermoid cyst associated with cerebral fusion anomalies.** Given the rarity of both Klippel-Feil syndrome and intracranial dermoid cyst, the association is probably significant, but is not

Figure 1 CT scan of head showing a left parietal enhancing disc shaped lesion.

Figure 2 Abscess with neutrophils and eosinophils around the cut section of parasite. H & E × 40.

Figure 3 Angiostrongylus cantonensis. H & E × 10.

Posterior fossa dermoid cysts and the Klippel-Feil syndrome

Although the Klippel-Feil syndrome as originally described comprises a triad of short neck, painless restriction of neck movements and low posterior hairline due to the congenital fusion of two or more cervical vertebrae, more serious anomalies of the cardiovascular, renal and central nervous systems may coexist. The following case report illustrates another important association which we feel has not received sufficient emphasis.

A 14 year old girl presented with a 12 month history of bitemporal headache and intermittent vertigo with nausea for two months. From the age of three months she had developed intermittently a soft discharging swelling over her occiput, which had been incised on three occasions in her local casualty department. She had been known to have Klippel-Feil syndrome since the age of four when she presented with a Sprengel's deformity of the right shoulder.

She had a short webbed neck with a low hairline which exfoliated as far as the C7 vertebral prominence. No occipital swelling was present, but a small punctum was present just above the union. Neurological examination revealed vertical nystagmus, dysdiadochokinesia, and mirror movements of the hands.

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We thank Professor CRRM Reddy for identifying the morphology of the parasite and Professor K Subba Rao, NIMS for permitting us to publish the case.

References


Notes

Figure a A lateral view of the cervical spine showing occipitalisation of the posterior arch of C1 with the anterior element apparently fused to C2. There is fusion of the posterior elements of C2 and C3 and of the anterior and posterior elements of C4, C5 and C6. Figure b CT scan showing a large midline hypodense mass lesion in the posterior fossa with peripheral flecks of calcification (solid arrow) and an associated midline defect in the occipital bone (open arrow).

commented upon in reviews of either condition and may well be under recognised. In view of the complications of these cysts, early recognition is important. Posterior cranial fossa dermoids should be added to the list of congenital abnormalities which must be sought in patients with Klippel-Feil syndrome.

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Visual failure following subarachnoid haemorrhage from rupture of an anterior communicating artery aneurysm

Subarachnoid haemorrhage secondary to rupture of an intracranial aneurysm may lead to a wide spectrum of neurological disturbances. Although visual loss may occur, complete and permanent amnesia is unusual unless associated with a large anterior communicating artery aneurysm. We report a case of total blindness with only minimal unilateral recovery following rupture of a small anterior communicating artery aneurysm which was associated with a documented period of reduction in global cerebral blood flow.

A previously healthy 56 year old woman was admitted to hospital following the sudden onset of severe occipital headache associated with dizziness, vomiting and neck stiffness. She was alert and orientated with no focal neurological signs, and both visual fields and acuities were normal. Lumbar puncture revealed uniformly bloodstained cerebrospinal fluid at a pressure of 18-5 cm. She was subsequently referred for neurosurgical assessment.

The following day she became drowsy although remaining orientated and otherwise neurologically intact. Computerised tomography showed diffuse blood interhemispherically and in the sylvian fissures with no aneurysmal mass or clot visible (fig 1a). On the fifth day, four vessel angiography revealed an anterior communicating artery aneurysm filling from the right and measuring 12 x 8 mm projecting down and forwards (fig 1b).

The cerebral blood flow was measured intermittently throughout the admission using radiouclide and blood flow determination with deconvolutional analysis. On the fifth day after the presenting haemorrhage this showed no regional derangement but a global reduction to 615 mls/minute (normal > 875 mls/minute).

On the sixth day she became increasingly drowsy and disorientated with the right pupil becoming dilated and transiently unreactive to light. A repeat CT scan showed absorption of the diffuse blood and an associated hydrocephalus (fig 1c).

The situation remained static over the subsequent days as did the cerebral blood flow and the CT scan appearances, and on the thirteenth day she had aneurysm surgery. A periferal approach was used and the aneurysm, parent vessels and optic apparatus were clearly displayed. The size of the aneurysm was in accordance with angiographic findings. No blood clot was seen and there was no direct compression of the anterior optic pathways. The aneurysm was controlled with a single Sugita clip which was left lying clear of the optic chiasm. She made a prompt recovery although remained mildly disorientated and with no change in her visual disturbance. Subsequent follow up at one year showed an improvement in the left visual acuity to 6/18 although the field remained restricted to a small central patch and there was complete blindness of the right eye.

Anterior communicating artery aneurysms, although close to the anterior visual pathways, rarely produce visual

Figure 1a CT scan on second day showing diffuse blood interhemispherically. 1b Angiogram showing anterior communicating artery aneurysm (12 x 8 mm).
Posterior fossa dermoid cysts and the Klippel-Feil syndrome.

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