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Recurrent spontaneous subarachnoid haemorrhage due to spinal haemangioblastoma

Sir: Spontaneous subarachnoid haemorrhage (SAH) of spinal origin is uncommon and accounts for less than 1% of all cases of non-traumatic SAH.^{1,2} When haemorrhage occurs from a lesion in the high cervical region the clinical features may be difficult to distinguish from SAH due to an intracranial lesion. Spinal tumours are a recognised cause of spinal SAH,³ but spinal haemangioblastomas presenting in this manner have only been described twice previously.^{2,4} Recurrent SAH due to cervical haemangioblastoma has not been reported before.

A 37 year old man presented with sudden onset of headache, vomiting, photophobia and neck stiffness. For one month he had been aware of mild numbness and clumsiness of his left arm. On examination he had meningism plus minimal weakness and proprioceptive deficit in the left arm. SAH was confirmed by lumbar puncture, and the clinical signs in the arm were attributed to a right parietal lesion, possibly an arterio-venous malformation. Cranial computed tomography (CT) showed blood in the 4th ventricle, but no structural lesion. Bilateral carotid and left vertebral angiography showed no source of haemorrhage. A sec-

ond SAH occurred 4 weeks later which produced no permanent neurological deficit, and 2 weeks after this repeat angiography was again normal. He was discharged home, but 14 weeks after his initial SAH he collapsed with severe headache and neck stiffness during sexual intercourse. On examination there was again neck stiffness and mild weakness of the left arm. CT showed intraventricular blood, but, as previously, none in the subarachnoid space. On myelography the cervical cord was expanded, and selective angiography of the vertebral and left thyrocervical vessels showed a vascular intramedullary tumour at C.2 (fig). Laminectomy was performed 6 weeks after the third SAH and revealed a haemangioblastoma. At this operation five arterial feeding vessels were obliterated, and at a second procedure the lesion was totally excised. The patient made a good post-operative recovery, but is left with some residual weakness of the left arm.

Haemangioblastomas account for 1.6% to 2.1% of all spinal cord tumours and 3.3% of intramedullary tumours.⁵ Forty percent of the spinal tumours occur in the cervical region of which 60% are intramedullary, and they most commonly present with features of spinal cord compression.⁵ Of two previously described lesions which presented as SAH, one was extramedullary at L.2⁴ and the other was an intramedullary cervical lesion.² When the haemorrhage arises

within the cord in the cervical region, bleeding may extend intracranially, and the clinical differentiation from subarachnoid bleeding from an intracranial source may be extremely difficult.^{6,7} Recurrent haemorrhages at short intervals are characteristic of aneurysmal subarachnoid bleeding, and the natural history of the condition in our patient increased the diagnostic difficulties.

In retrospect, the weakness in the left arm should have been a pointer to the true site of the lesion, but this symptom was mistakenly attributed to an intracerebral lesion, initially a possible arterio-venous malformation, and latterly cerebral ischaemia. Furthermore, the absence of blood in the subarachnoid space following the ictus on two occasions was an indication that it had tracked to the ventricular system via the central spinal canal. If cerebral angiography is negative in cases of recurrent haemorrhage a lateral cervical series should be included to exclude a cervical lesion. This would not prolong the procedure or put the patient at increased risk. Spinal haemangioblastoma is a rare cause of spontaneous SAH and may lead to diagnostic difficulties when its presentation mimics intracerebral SAH. This is the first report of recurrent SAH from such a lesion and it should be considered when other more common sources of haemorrhage have been excluded.

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Fig Vertebral angiogram, anteroposterior projection, showing vascular mass.

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Extramedullary haemopoiesis in the central nervous system: an unusual cause of epilepsy

Sir: Myelofibrosis is characterised by progressive splenomegaly, fibrosis of the bone marrow, a leuco erythroblastic peripheral blood picture and extra medullary haemopoiesis.¹ Extramedullary haemopoiesis predominantly affects the reticulo-endothelial organs such as the spleen, liver and lymph nodes and, less frequently the kidneys, skin, heart, pleura and mesentery.^{2,3} Only very rarely is the central nervous system involved.⁴ We describe a case of idiopathic myelofibrosis, associated with epilepsy, in which necropsy revealed multiple "tumours" of extramedullary haemopoietic tissue in the cranial dura mater, together with microscopic foci in the brain and leptomeninges.

A 65 year old man with idiopathic myelofibrosis had been maintained on busulphan with occasional transfusions of whole blood. Two years following diagnosis, he was admitted to hospital for elective splenectomy, with symptoms attributable to its massive size. The spleen, which weighed 3.8 kg, was removed and subsequent microscopic examination confirmed extensive extramedullary haemopoiesis. The procedure was uneventful and the patient made a good recovery.

Seven months later, however, purplish umbilicated nodules, ranging in size from 0.5 to 2 cm maximum dimension, appeared in the skin of the upper and lower limbs and abdomen. Biopsy of some of these lesions showed extramedullary haemopoiesis in the dermis and subcutaneous fat. Over the next few weeks more skin nodules appeared and, at the same time, the patient's haematological status deteriorated. Anaemia became more severe, the interval between transfusions became shorter and he complained of increasing tiredness and lethargy. Four weeks following the first appearance of the skin lesions, the patient had an epileptiform seizure with loss of consciousness, cyanosis, urinary incontinence and jerking move-

ments of both upper limbs. The patient recovered spontaneously, but two further similar episodes occurred during the next few hours until he was stabilised on anti-convulsant therapy.

The patient had no previous history of epilepsy and all investigations, including CT scan, failed to elucidate the cause of his seizures. Twelve days later, however, his seizures recurred. On this occasion, there was loss of consciousness accompanied by twitching of the facial muscles and all four limbs. The seizures were continuous with increasing periods of apnoea and finally, respiratory arrest and death.

At necropsy, the sclerae were mildly icteric and multiple, well-circumscribed, purplish nodules, up to 3 cm maximum dimension were present in the skin. The bone marrow appeared pale. The liver was enlarged, weighing 1900 g, and there was marked para-aortic lymph node enlargement. Small discrete pale nodules were present in the kidneys, mesentery, pleura, heart and oropharynx. Microscopy of the liver, lymph nodes and the nodular lesions confirmed extramedullary haemopoiesis. The inner aspect of the cranial dura mater contained several nodules of fleshy reddish-brown tissue, the largest 3 cm maximum dimension (fig 1). The larger nodules had indented the underlying cortex. Microscopic examination showed these nodules to consist of extramedullary haemopoietic tissue (fig 2). Apart from the cortical indentations,

the brain and leptomeninges appeared normal to the naked eye. Microscopic deposits at extramedullary haemopoiesis however, were identified in the leptomeninges of the mid-brain and medulla, as well as the choroid plexus of the fourth ventricle. Tiny focal deposits were also noted in the occipital cortex. There was no evidence of blast transformation, a recognised late complication of myelofibrosis.^{5,6}

Extramedullary haemopoietic tumours of the spinal and cranial dura mater are rare. Those in the spine may result in symptoms attributable to cord compression and, if recognised, may be treated successfully by radiation therapy.⁷ Deposits in the cranial

Fig 1 Inner surface of cranial dura mater showing tumour-like nodules of extramedullary haemopoiesis.

Fig 2 Microscopy of dural nodule showing extramedullary haemopoietic tissue ($\times 200$).