Extradural 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. Extramedullary haemopoiesis predominantly affects the reticuloendothelial organs such as the spleen, liver and lymph nodes and, less frequently, the kidneys, skin, heart, pleura and mesentery. Only very rarely is the central nervous system involved. We describe a case of idiopathic myelofibrosis, associated with epilepsy, in which necropsy revealed multiple “tumours” of extradural 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 extradural 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 extradural 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 movements 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.

Extradural 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. Deposits in the cranial...
Nervous system involvement is common in parovirus infection. The exact mechanism by which parovirus causes neuralgic amyotrophy is unknown. It is believed that the virus infects and destroys the nerve cells, leading to inflammation and pain.


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