

HAEMANGIOBLASTOMA

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The haemangioblastoma of the central nervous system is a vascular neoplasm usually found in adults occurring most commonly in the cerebellum, either in the midline or in one hemisphere. Occasionally the lesion is encountered in the medulla, the spinal cord or the supratentorial compartment.

Haemangioblastomas are relatively uncommon; their incidence ranges from 1 to 2.5 percent of all intracranial tumours¹. Although usually single, haemangioblastoma may be multiple and may be associated with the von HippelLindau syndrome².

CASE REPORT

A 45 year old Sudanese patient was admitted to the Neurosurgical Division of the King Khalid University Hospital because of headache and repeated vomiting associated with unsteadiness on walking which started two months ago. The patient gave a history of a brain operation 14 years ago in Egypt after which he remained free of symptoms until his present complaints developed. On examination the patient had gross bilateral papilloedema with retinal haemorrhages. There was no other cranial nerve affection. Tendon reflexes were symmetrical but hyperactive. Motor power and sensation were intact. The gait was ataxic. On admission the haemoglobin was 18.4 g/dl, the red blood cell count (RBC) $5.9 \times 10^{12}/L$ and the packed cell volume (PCV) 56.2% with a normal white and differential blood cell count. The computerized tomography (CT) scan of the brain revealed a partially solid, partially cystic tumour in the right cerebellar hemisphere with irregular ring like enhancement following the injection of contrast medium. The lesion measured approximately 5.5 cm in diameter and resulted in marked occlusive hydrocephalus. Because of the increased intracranial pressure a right frontal ventriculoperitoneal shunt was performed on the second day of admission. Postoperatively the patient became free of symptoms except for a moderately ataxic gait. Two weeks following operation the haemoglobin was 20.4 g/dl, RBC $6.47 \times 10^{12}/L$ and PC 62.7%. No blood transfusion had been carried out upto this stage. On 7.11. 1986 the old posterior fossa craniotomy was extended and total excision of the cerebellar tumour was achieved. The post-operative course was uneventful, and on discharge the patient was free of symptoms. Three weeks after this operation the haemoglobin was 124 g/dL, RBC $3.83 \times 10^{12}/L$ and PCV 36.1%. On gross examination the tumours removed was as dark brown soft mass measuring 4x3. 5x2.5cms (Figure I).

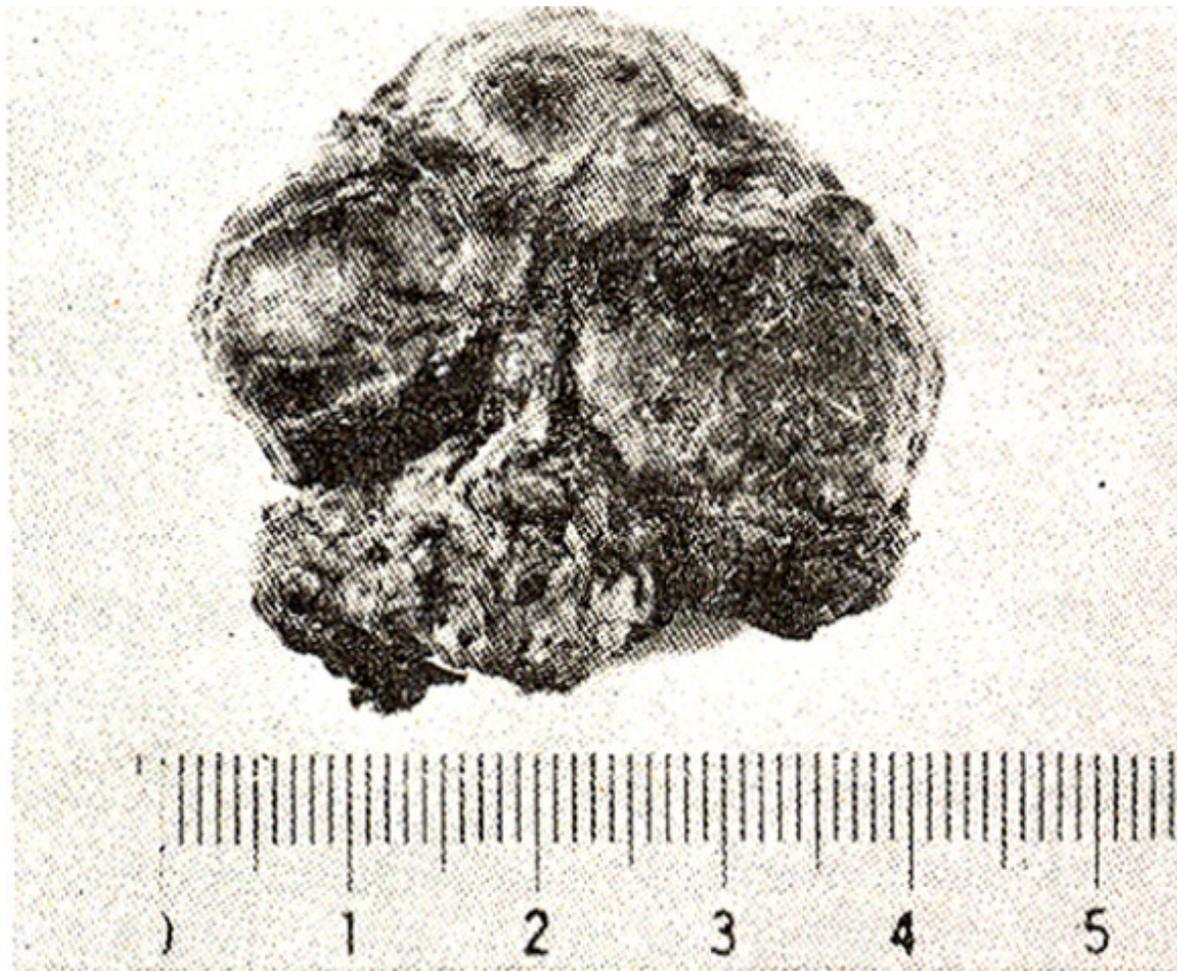


Figure 1. Gross photograph of the dark brown cerebellar tumour measuring 4x3.5x2.5cms.

Sectioning of the tumour revealed brownish solid substance with several cystic spaces of varying sizes, some of which contained yellowish fluid. The histological examination showed a tumour composed of endothelial lined vascular channels of variable sizes containing blood. The vascular spaces were separated by large pale stromal cells of generally uniform cytological appearance (Figure 2).

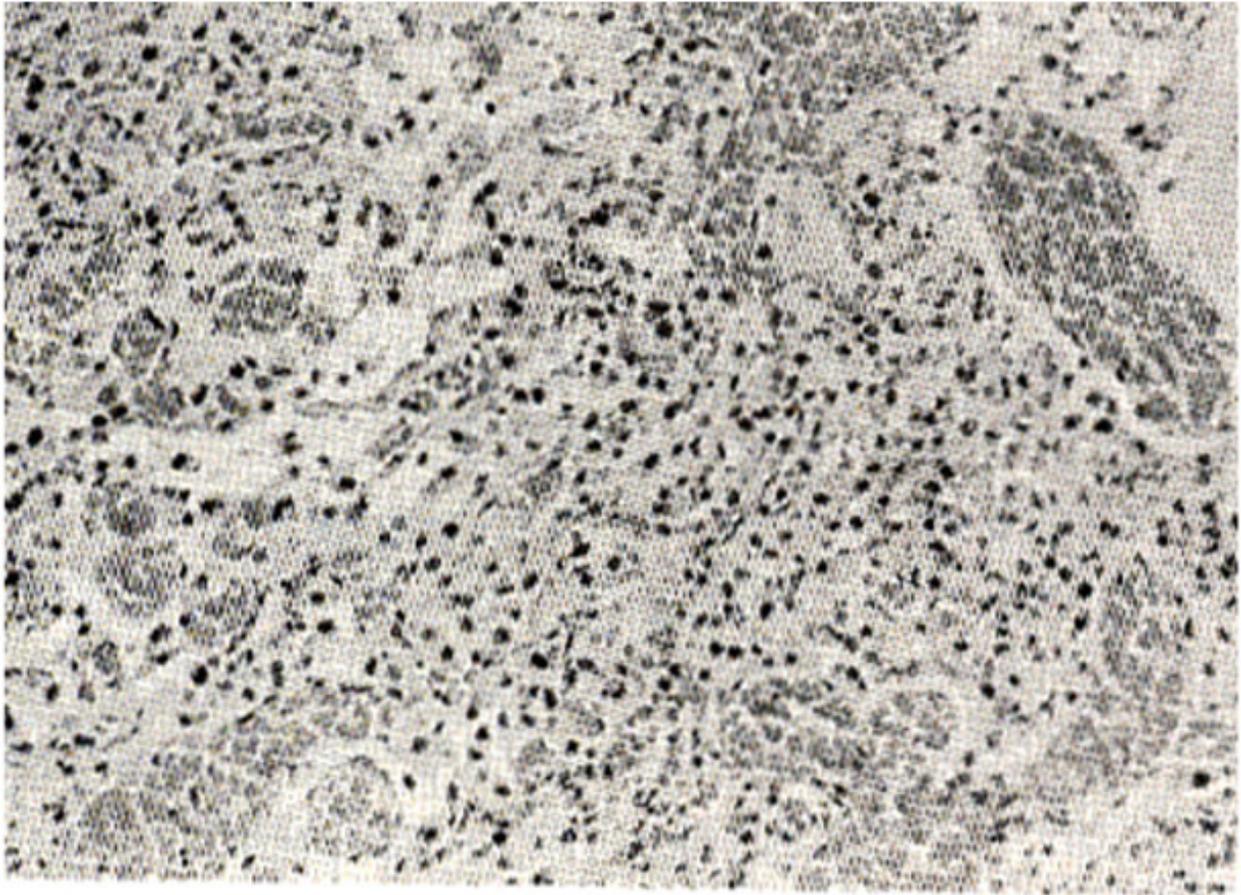


Figure 2. Histological section of the haemangioblastoma showing endothelial lined vascular channels with large pale Haematoxylin and Eosin x 250.

In a few areas the nuclei of the stromal cells were large, hyperchromatic and showed some pleomorphism. No mitotic figure was seen. Scattered mast cells were seen. Many of the stromal cells gave a positive lipid staining with Oil Red O stain on frozen sections. However the stromal cells gave a negative staining for both G.F.A.P. and Factor VIII R: Ag by the immunoperoxidase technique. Electron microscopic examination of the tumour revealed vascular channels and three major types of cellular elements: endothelial cells, pericytes and stromal cells (Figure 3).

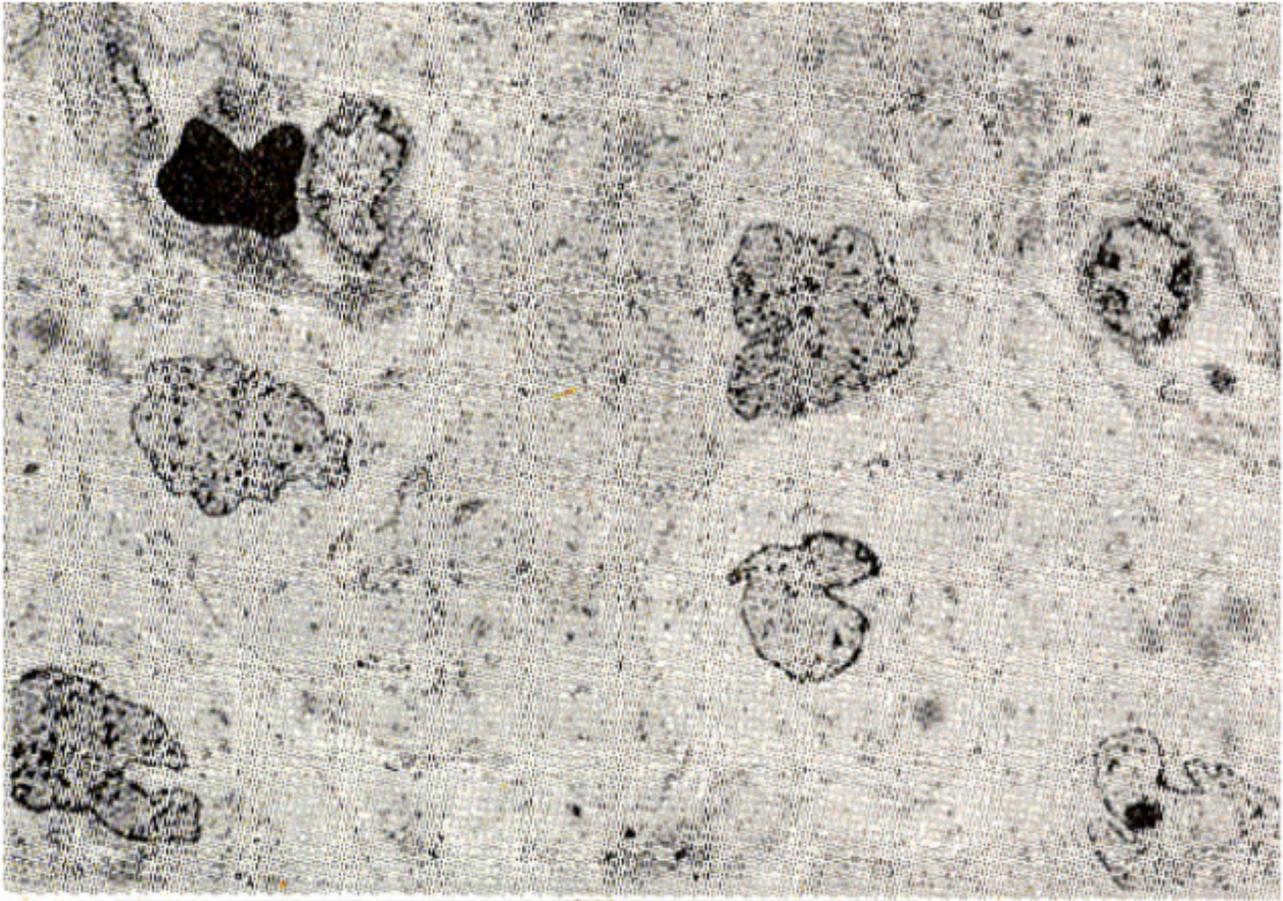


Figure 3. Electronmicrograph of the tumour showing endothelial cells, and dark and light stromal cells.x 3,450.

The vascular channels were lined by endothelial cells of varying shapes and sizes. The endothelial cell cytoplasm showed many mitochondria, rough endoplasmic reticulum, and Golgi apparatus. Pinocytotic vesicles were seen in the plasma membrane. The endothelial cells along with the pericytes were surrounded by basal lamina. The stromal cells revealed abundant cytoplasm and irregular cell contour. The cytoplasm was electron dense in some and electron lucent in others. The size and distribution of cytoplasmic organelles varied. They showed microfilaments, coarse granules, mitochondria and lipid droplets. In some cells a whorl-like arrangement of smooth endoplasmic reticulum was present. Occasional desmosomal contacts were seen between stromal cells apposed to each other.

COMMENTS

The haemangioblastoma of the central nervous system is a distinct clinical and pathological entity. It is appealing to the surgeon because of its curability, to the pathologist because of its distinctive morphology, to the geneticist because of its potential association with a myriad of other genetically patterned abnormalities and to all because of the apparent functional capacity of some lesions to stimulate erythropoiesis³. Histologically the neoplasm is composed of three distinct cell types: endothelial cells, pericytes and stromal cells. The nature and derivation of the stromal cells remains

uncertain. They have been suggested in previous reports to originate from astrocytes⁴, pericytic cells⁵, histiocytic cells⁶ and endothelial cells⁷. It has also been suggested that cells identified as “stromal cells” of haemangioblastomas on light microscopy are a heterogenous group of cells including astrocytic as well as other elements and that they resemble each other on ordinary stains because of the “equalizing effect” of cell lipidization⁸.

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