Spinal Cord Haemangioblastoma and MR Imaging

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Abstract : Haemangioblastomas are true vascular neoplasms which progress inexorably to a catastrophic clinical situation if left untreated. They are mostly intramedullary tumours rarely seen in the extramedullary space.An unusual case with a solitary spinal cord haemangioblastoma located purely in the intradural extramedullary space is presented. MR imaging accurately localized the mass and showed the precise delineation of the tumour after Gd-DTPA.

Key Words: Haemangioblastoma,Magnetic Resonance

INTRODUCTION

Haemangioblastoma consists of a proliferation of vascular spaces and endothelial cells separated by fat-laden stromal cells(4). It occurs as a solitary vascular lesion or associated with von Hippel-Lindau's Disease which is a heredofamilial disease transmitted by an autosomal dominant gene and characterized by vascular tumours of the retina and central nervous system or cysts or tumours of the kidney, pancreas or lung(2,10,16,20).

It comprises approximately 2 to 3 % of primary spinal cord neoplasms(20).Most of these tumours are located in the intramedullary space (14,15,18,20) and rarely in the extramedullary space(14,19). Myelography, selective angiography and MRI aid in establishing the diagnosis (5,6,11,14,20).

Case Report

This 45-year-old female presented with a history of pain in the thoracic region and difficulty in walking for 6 months.On neurological examination there was significant weakness in both upper extremities,more on the right,and in both legs. Posterior column dysfunction including absent proprioception and a positive Romberg sign was observed. Sensory level to pinprick was at C5-6 on the right. Evaluation of the spinal area with MR demonstrated a mass brilliantly enhanced with the contrast Gd-DTPA displacing the cord to the left side and localizing in the intradural extramedullary space between C5 and T2 levels (Fig 1,Fig.2).

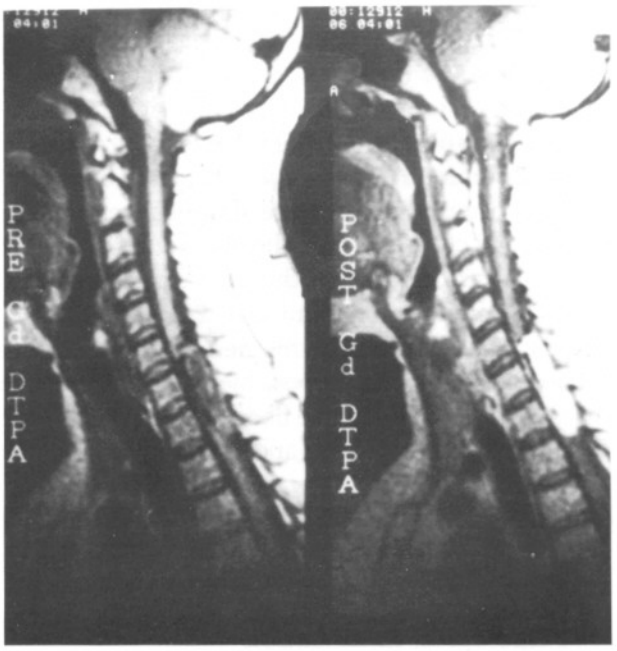

Fig. 1 : Pre-and post-contrast magnetic resonance images (sagittal view) showing the tumoral mass localized between C6 and T2.

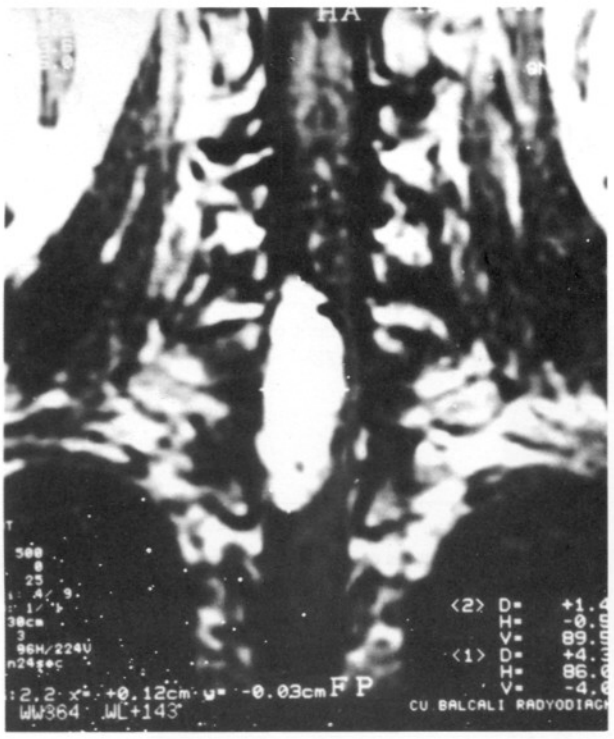

Fig. 2 : Post-contrast magnetic resonance image (coronal view), showing the enhanced mass in the right side of the cord and compressing it to the left side.

Operation and Postoperative Course

The operation was performed in the prone position and total laminectomy was performed to the C6,C7,TI and T2 vertebrae.Under the operating microscope the dura was opened longtudinally. A mass compressing the cord and displacing it to the left side was observed in the subarachnoid space.On the pial surface there was a vessel coil composed of feeding arteries and a large draining vein at the cranial end of the tumour. The distal one third of the tumour was captured by the posterior nerve fibers of the C8 root. The tumour was dark red and very fragile. The dissection of the nerve fibers from the tumor surface was impossible due to massive haemorrhage, so the nerve fibers were sacrificed, the feeding vessels coagulated and the draining vein was interrupted. The tumour was removed in one piece. The postoperative course was uneventful. After a month there was only anaesthesia in the C8 dermatome.

Pathological Examination

Macroscopically the tumor was 4x1x1.5 cm. The sectioned surface was spongy and red colour. The surgical specimen was fixed in 10 percent formalin embedded in paraffin and stained with Hematoxylin-eosin, reticulin factor 8.

Microscopically the tumour was composed of large numbers of closely-packed blood vessels,lined by plump endothelial cells. There were a few hyperchromatic stromal cells between the capillary channels (Fig 3). Histopathological diagnosis was haemangioblastoma.

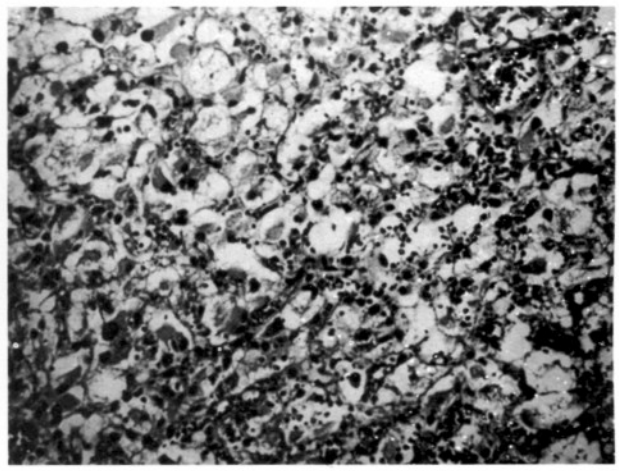

Fig. 3 : The vascular and stromal cells are seen.H–E X 375

Discussion

Haemangioblastomas are rare,benign vascular tumors of the central nervous system. They occur as solitary lesions (12,17,19) or associated with von Hippel-Lindau's Disease(2,10,16). They are frequently in association with cysts(9,13,18) and more than one lesion may be seen in the spinal cord at one time (2,9,18). Syrinx is present in approximately 70 % of cases, pial varices in roughly 40 %, and root involvement in about 15 % (20). Symptoms and signs are due to cord compression and progress to paraparesis, posterior column dysfunction and sensory loss (11,18,17). Haemangioblastomas may be found anywhere in the spinal cord but occur mostly in the cervical and thoracic regions (5,8,9). It is noteworthy that subarachnoid haemorrhage in the unoperated cases is extremely rare(7,12).

Plain X-Rays have no diagnostic value,but on myelographic examination complete or partial block may be observed(1,8) and Myelo-CT may demonstrate the extramedullary isodense mass at the same level.

The method of choice in diagnosis was selective angiography with catheterization of the arteries of the spinal cord with the use of the subtraction technique (12,17,20). At present Magnetic Resonance Imaging with Gd- DTPA enhancement is the most useful tool for precise localization and definition of the tumor margin and differentiation of any accompanying cyst(3,5,11,14). Once a spinal lesion has been demonstrated the association of haemangioblastoma with von Hippel- Lindau's disease should be made to exclude cerebellar, brainstem and visceral lesions (8,20). At the same time the entire spinal cord should be examined to rule out multiple haemangioblastomas.

The treatment of choice is total extirpation utilizing microtechnique. These are highly vascular tumours and if accidently cut into or decompressed, bleeding not only obscures the anatomical planes,but may result in catastrophic problems(13). Removal can be achieved with reasonable morbidity.

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