

HEMANGIOBLASTOMA OF THE SPINAL NERVE ROOT

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ABSTRACT

Spinal root haemangioblastomas are very rare tumors. A 70-year-old male patient admitted to our clinic with low back and both legs pain of one year's duration. Magnetic resonance imaging revealed a solid mass lesion at T-12 level. The contrast enhancing mass was intradural in location. T-12 laminectomy was performed and a highly vascular mass originated at the T-12 nerve root was totally removed. Histopathological examination revealed diagnosis of haemangioblastoma. He was then screened for von Hippel Lindau disease. No stigmata of this disease were found. The postoperative period was uneventful. He was discharged with no complaints.

Spinal root haemangioblastoma should be kept in mind in a patient with highly enhancing spinal intradural mass.

Key Words: Hemangioblastoma, Spinal nerve root, von Hippel Lindau disease

INTRODUCTION

Hemangioblastomas (HBLs) of the spinal region are rare and represent about 5.8 % of all spinal

cord tumors (1). These tumors are more specific to von Hippel Lindau (VHL) than CNS HBLs and about 80% are caused by VHL disease. (2) Sporadic and VHL-associated HBLs are histologically identical and can be cured by complete surgical resection. The tumor is usually intramedullary in location. We present a sporadic case of HBL diagnosed without the clinical features of VHL disease. To our knowledge there are only 3 case reports of sporadic HBL of the spinal nerve root in the literature (3-5).

CASE REPORT

A 70-year-old man presented with a history of low back pain of one year's duration. He had pain in both legs for 6 months. Neurological examination revealed normal signs. Magnetic resonance imaging of the lumbar spinal area revealed an isointense 1x1 cm mass lesion at T-12 level (Figs. 1a-b). The mass was intradural and extramedullary in location. MR with contrast material demonstrated that the mass was heavily enhanced.

Preoperative diagnosis was spinal schwannoma. A T-12 laminectomy in prone position was performed. When the dura opened, a highly vascular mass originating from the T-12 nerve



A



B

Fig. 1: The contrast enhanced T1-weighted axial (a) and sagittal (b) images demonstrate the round enhancing tumor.

root was found. The tumor was totally removed. Postoperative period was uneventful. Pathological examination of the tumor revealed a rich capillary network accompanying stromal or interstitial cells (Fig. 2). The stromal cells were large, polygonal, and showed clear cytoplasmic vacuoles. In some regions of the tumor, the cell nuclei were large, pleomorphic and hyperchromatic. No mitotic activity was observed. The tumor cell cytoplasm was clear and showed weak periodic acid-Schiff positivity in some areas. There was a rich reticulin network, and many of the tumor cells were surrounded by collars of reticulin. Mast cells were distributed throughout mass, and these showed metachromasia with toluidine blue and May-Grünwald Giemsa staining. These findings led us to diagnose hemangioblastoma. There was no familial history of VHL. The patient was evaluated for VHL disease and no clinical feature of VHL was found.

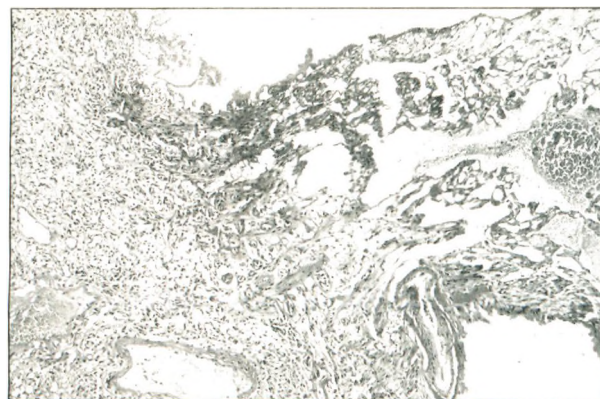


Fig. 2: Hematoxylin and eosin staining demonstrates the classical features of a hemangioblastoma. These include a highly vascular capillary plexus surrounded by vacuolated stromal cells. The relationship to the nerve root is clearly seen at left.

He was discharged with no complaints. He is without any problems at postoperative 6th year follow-up.

DISCUSSION

Hemangioblastomas are low-grade, highly vascular tumors commonly associated with the von Hippel-Lindau Syndrome and most commonly occurring in the cerebellum (6). The most common locations for central nervous system HBLs are the cerebellum, spinal cord and brain stem. They can very rarely be seen in spinal nerve roots (7). Extramedullary HBLs arising from the filum terminale or proximal nerve roots have been reported but are considerably less common. Extramedullary-intradural HBLs represent only about one fifth of all spinal HBLs (8). Only 20 cases have been reported up to date.

von Hippel-Lindau disease demonstrates an autosomal dominant pattern of inheritance and the gene is located on 3p25-26 (9). Some families inherit the gene but it is not expressed. However, it appears that many of these cases represent asymptomatic carriers and if they are carefully screened will be found to have VHL. Nevertheless, VHL rarely can arise without a family history. Such new mutations occur in only 1-3% of cases (10). Our patient had no manifestations of VHL.

Most of the patients in the literature come to attention with spinal cord and root compression signs. There was no neurological deficit in our patient. Since these tumors are very vascular, they can be presented with intratumoral hemorrhage or subarachnoid hemorrhage. But this is a very rare occasion (11).

The intramedullary spinal cord HBLs are mostly situated at cervical and thoracic levels. However, the majority of the extramedullary spinal HBLs were within the lumbar region. The tumor in our patient was at the thoraco-lumbar junction.

Some authors recommend a superselective spinal angiography in order to distinguish between intradural HBL and spinal arteriovenous fistulae (12). MR imaging reveals isointense mass lesion in T1-weighted images. These tumors are heavily enhanced with contrast material. Intratumoral signal void areas may demonstrate the vascular supply of the tumor. The MR imaging in our patient demonstrated these classical features. However, no history of VHL disease led us to make a preoperative diagnosis of spinal schwannoma.

The term nerve root HBL includes three different types; the subdural type partially adherent to the nerve root, the subdural type encasing the nerve root wholly, and the extradural type (11). The tumor in our patient encased the posterior nerve root totally, resembling a spinal schwannoma, and belongs to the second type. In the second and third groups it is not possible to remove the tumor without sacrificing the nerve root. The nerve root-tumor relation in our patient's tumor is very clearly seen in Fig. 2.

Spinal extramedullary HBLs are very rare tumors. A spinal round mass with strong contrast enhancement should alert the surgeon about the presence of a root HBL. These tumors should be totally removed. Total removal of a sporadic tumor will result in the cure of the disease. However, the patient should be screened for any stigmata of VHL disease.

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