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Hemangioblastoma Originated from the Lumbar Spinal Nerve Root
- A Case Report -

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Hemangioblastoma of the spine is a disease that mostly occurs intradurally and intramedullary. We report a case of 62 years old woman with hemangioblastoma originated from the lumbar spinal nerve root not associated with von Hippel-Lindau disease and mimicking schwannoma. Diagnostic procedures, surgical findings, histological and clinical outcomes are discussed.

Key Words: Hemangioblastoma · Lumbar spine · Extradural · von Hippel-Lindau disease

INTRODUCTION

Hemangioblastoma of the spine is relatively rare, accounting for only 7% of all central nervous system cases[4] and mostly occurring in Von Hippel-Lindau (VHL) syndrome. It represents only 1.6~6.4% of all spinal tumors and is almost exclusively occurring intramedullary, commonly occurring at thoracic and cervical cord[2]. Occasionally(<1%), they are associated with spinal nerve roots but mostly they are situated intrathecally[6]. We report a rare case of patient with extradural hemangioblastoma originated from the lumbar nerve root, its radiological findings, surgical treatment and clinical outcome.

CASE REPORT

A 62 years old woman presented with pain in the left leg for 3 months. She had taken physical therapy and acupuncture treatment at regional hospital but her symptoms persisted and progressive lower extremity weakness developed. At the time of admission, she showed motor weakness of left knee extension (Grade 3) with sensory change on L3 and L4 dermatomes. She did not complain any bladder and/or bowel symptom.

Magnetic resonance (MR) imaging of lumbar vertebrae showed that an epidural mass occupied left side of spinal canal on L2-3 area and extended out of left L2-3 neural foramen like dumbbell-shape(Fig. 1 & 2). The epidural mass showed very high signal intensity on T2-weighted images and almost iso-signal intensity with mottled signal void inside the mass on T1-weighted images. On gadolinium (Gd-DTPA)-enhanced images, the mass was enhanced very well(Fig. 1 & 2). The results of laboratory studies did not reveal any abnormality.

Surgery was performed by hemi-laminectomy of left L2 and partial hemi-lamiectomy of left L1 lower and L3 upper for exposure of tumor. Upon laminectomy, gray white epidural mass was found and it was highly vascular. It was compressing the dura from the midline to left foraminal space and total facetectomy of left L2-3 facet joint was additionally done for complete exposure of tumor. Tumor mass was firmly attached to dura and L2 nerve root at L2/3 foramen. Entire tumor was carefully dissected and tumor was originating from L2 nerve root, so resection of left L2 nerve root was done together with the tumor mass at the foraminal area(Fig. 3). Patient recovered uneventfully with improved pain to her left leg. Her muscle strength recovered and became ambulatory on postoperative day 7.

Histology of the tumor revealed it to be capillary hemangioblastoma. Mass was composed of capillary sized vascular structures with foamy interstitial cell infiltration which was characteristic of hemangioblastoma in high power field(Fig. 4). She

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had no family history of any genetic disease. Further examination was done to rule out patient being Von Hippel-Lindau syndrome. Abdomen and pelvic CT revealed left renal cyst but it did not show evidence of renal cell carcinoma(Fig. 5). Brain computed tomography (CT) and retinal examination were negative for tumor involvement. Subsequent genetic testing for VHL gene was done and the result was negative.

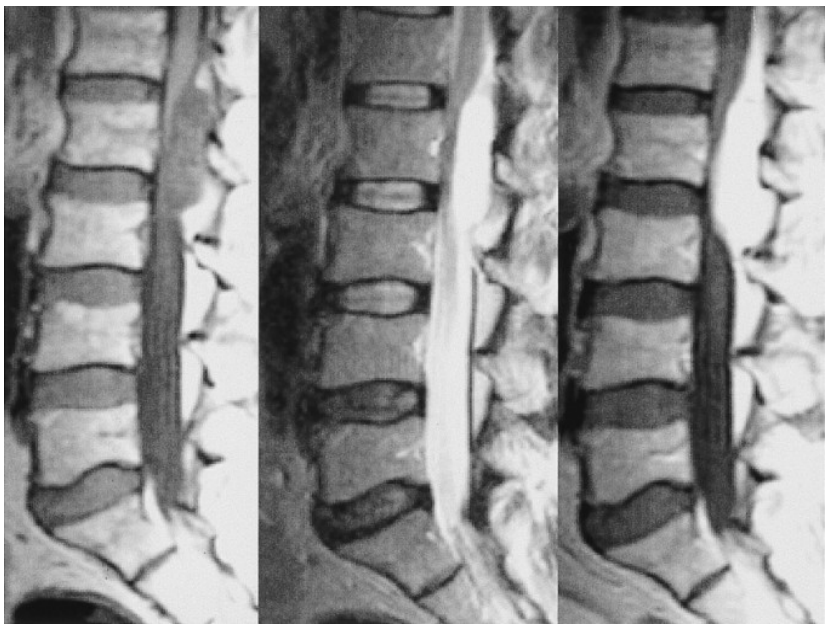

Fig. 1. Sagittal MR images, which show epidural mass on L2-3 area. Left: T1-weighted MR image demonstrating iso-signal mass with mottled signal void inside the mass. Center: T2-weighted image shows high signal intensity of the mass. Right: Gd-DTPA enhanced image revealing well enhanced epidural mass.

DISCUSSION

Hemagioblastoma is World Health Organization (WHO) grade 1 tumor that occurs exclusively in the nervous system. In this case report, we have described an unusual case of hemagioblastoma originated from the lumbar nerve root and situated extradurally. As nerve root hemangioblastomas frequently originate in the dorsal root entry zone or sensory roots[3], they tend to be intradural[2]. Tronnier, et al reported two cases of extradural hemagioblastomas in which one patient was with von Hippel-Lindau disease and one without[7]. Rohde, at al reported a case of intradural and extradural hemagioblastoma of the cauda equina[5]. Although mechanism behind the extradural location of tumor still needs to be revealed, the preoperative differential diagnosis and surgical strategy are key to optimal surgical outcome. Nerve root hemangioblastoma can mimic schwannomas at MR imaging as they share the characteristic of being nerve root in origin. In our case, MR imaging revealed high

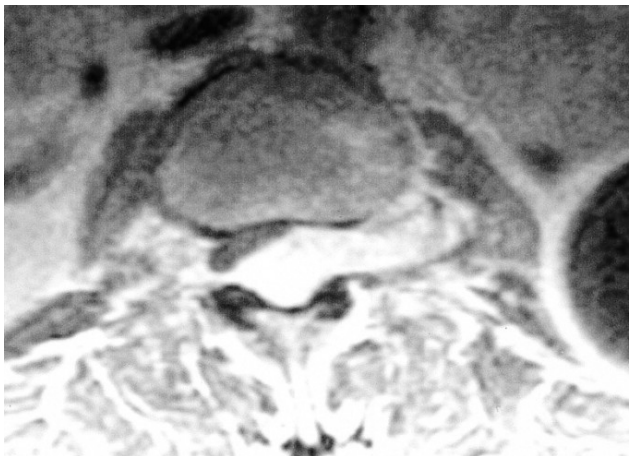

Fig. 2. Gd-DTPA enhanced axial MR image that shows left epidural well enhanced mass extending out of left neural foramen like dumbbell-shape.

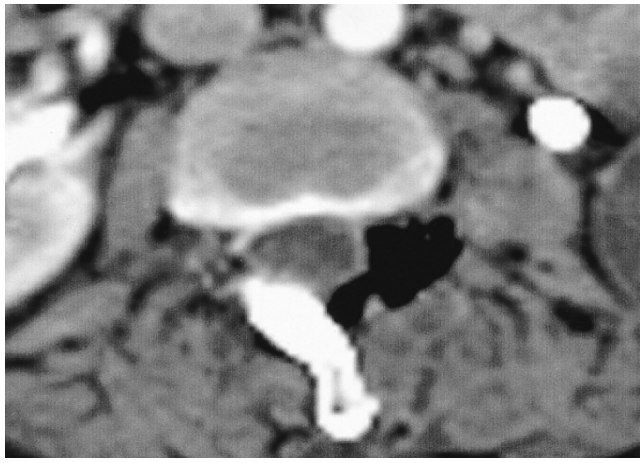

Fig. 3. Postoperative CT image. The tumor mass was completely removed after total facetectomy of left L2-3 facet joint.

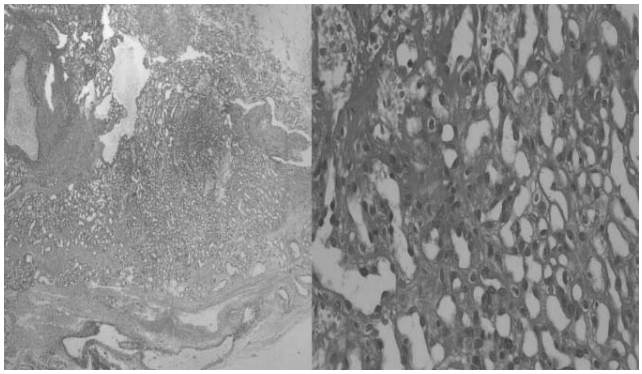

Fig. 4. Microphotographs of specimen shows fragmented spinal cord mass and sponge like structure with central hemorrhage(Left: Hematoxylin and eosin stain x40). In the high power view, it was composed of capillary sized vascular structures and foamy interstitial cell infiltration(Right: Hematoxylin and eosin stain x400).

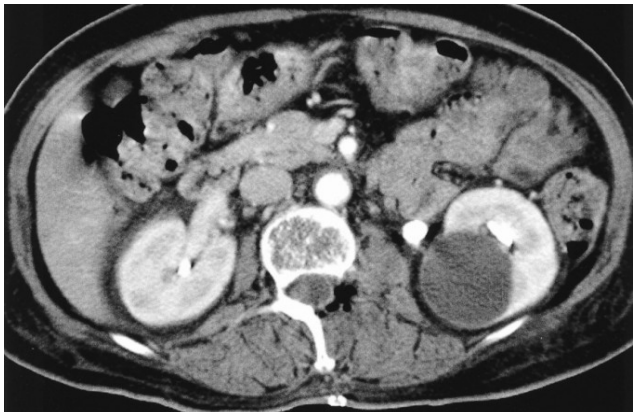

Fig. 5. Abdominal CT image that shows left renal cyst without tumor mass.

signal intensity on T2 weighted image and intense, mottled enhancement on T1 Gd-DTPA enhancement. Lonser, et al, on their surgical manage- ment of 6 lumbosacral nerve root hemagioblastomas, reported on frequent adherence of tumor to the nerve root and surrounding structures (5 of 6 cases)[3]. Once hemangioblastoma is suspected, wide surgical exposure, identification and dissection of tumor capsule, and coagulation of large-caliber feeding vessel must be in mind of surgeon for feasible and successful resection. In our case, the feeding artery was radicular artery joining with L2 nerve root. When nerve root involved with the tumor can be sacrificed, it should be resected to achieve complete removal. After resection of L2 nerve root in our case, there was not neurological deficit. We think that L2 nerve root already lost its function before surgery by long-time compression at the neural foramen.

It can occur sporadically or in association with other visceral tumors and cysts that have been grouped under the familial tumor syndrome of Von Hippel-Lindau (VHL) disease. In a patient with a positive family history of VHL, the finding of a single retinal or cerebellar hemangioblastoma, pheochromocytoma, or renal cell cancer is sufficient to make the diagnosis. Some have argued that the presence of multiple pancreatic cysts is also sufficient. Renal or epididymal cysts alone are not sufficient because they occur frequently in the general population. If no known family history of VHL exists, two or more retinal or cerebellar hemangioblastomas or one hemangioblastoma plus one visceral tumor must be present to justify the diagnosis[1]. In our patient, only combined abnormality was bilateral renal cysts which is not sufficient to diagnose VHL syndrome. Genetic analysis for VHL disease can aid in its diagnosis. VHL gene is a tumor suppression gene located on chromosome 3p25-26, and our patient's gene revealed no such deletion.

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