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Case Report

GIANT LUMBO-SACRAL EXTRADURAL INTRAOSSEOUS SCHWANOMMA IN A ADOLOCENT MALE- RARE LESION

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INTRODUCTION:

Schwannoma are benign slow growing tumor arising from Schwann cells of the nerve sheaths of peripheral nerves and believed to originate from embryonic neural crest cells [1]. Generally, spinal schwannoma presents with pain and/or neurologic deficits when it grows and /or as a large mass extending into the vertebra body and extraspinal space. Giant spinal schwannomas are defined as lesions that extend over more than two vertebral levels or have extraspinal extension of more than 2.5 cm. Small no of cases of spinal giant schwannoma arising from the lumbosacral area have been reported in the literature with a rare incidence [2–4] and limited data. Extradural giant spinal schwannomas in the adolescent age group is of rare occurrence with a strong propensity for schwannomas to occur in neurofibromatosis (NF) type I as multiple lesions in the cervical and lumbar spine [1]. These commonly occur in the intramedullary location. Extradural spinal lesions in young age group are generally malignant with sarcoma, osteoblastoma, lymphoma, or metastasis being the common causes [2]. To date, there have only been five cases of pediatric giant spinal schwannomas, purely extradural in location, without features of NF, reported in the literature [3–7]. A similar case of giant schwannoma in a 20-year-old boy is presented which was treated successfully with an uneventful recovery.

CASE REPORT:

A 20-year-old boy presented to us with a history of gradually progressive back pain for 6 months. He also developed imbalance in walking and urinary incontinence for past 1 month. On neurological examination, there were no neurocutaneous marker; power was grade 4/5 in bilateral ankle dorsiflexors and 3/5 in bilateral ankle plantar flexors. The power in rest of the lower limb muscle groups was 5/5. Ankle reflex was exaggerated in both the lower limbs (4+). The sensory examination was within normal limits and patient had to be catheterized due to urinary incontinence. Magnetic resonance imaging (MRI) was done which showed a ill-defined enhancing lesion noted in the spinal canal extending D12 (Dorsal) to S2 (sacral) level. It was extending in the left para vertebral region via left neural formina of L5 and S1 vertebrae. The extension of lesion in the left para vertebral region was measuring 7.3*7.1*11 cm. It was solid and shows heterogenous post contrast enhancement. Similar extension measuring 1.3 cm*1.1 note in the right paravertebral region at the level l5 vertebrae via right neural foramina. (Fig 1 to 5) Patient underwent complete excision of lesion. Intraoperatively lesion was extradural exiting from the neural left L1 foramina into the retroperitoneum space. Tumour was vascular, and suckable . Skin inscion was extended lateral and retoperitonium space was entered after cutting the underlying muscle. Tumor was seen entering into the vertebral body which was excised and retorpeitoneal tumor was removed and endoscope was used to confirm complete removal of tumor from deeper areas. (Fig 6) On histopathology, tissue revealed hypercellualr areas (Antoni A) and hypocellular areas (Antoni B). The tumor cells were arranged in fascicles and bundles. Verocay body palisading around fibrillary process are seen with hyalinated blood vessels. These findings were suggestive of a schwannoma without evidence of malignant potential. Postopeatively his power improved to 4/5 in bilateral ankle plantar flexors and 5/5 in bilateral ankle dorsiflexors and he was able to walk without support. At 1 month follow up his

preoperative motor weakness recovered completely and his bladder sensation and voiding improved significantly. The follow-up MRI after 1 year showed no evidence of recurrence of the lesion (Fig. 4). Patient has been completely asymptomatic over the last 4 months with normal motor strength and bowel-bladder function

DISSCUSION:

The goal of retroperitoneal schwannoma surgery is to achieve a complete removal of the tumour while preserving neurological function. Intraosseous schwannomas are rare benign tumors, accounting for < 0.2% of primary bone tumors. The most common intraosseous lesions include the mandible and sacrum, while spinal lesions are less common. The first case of spinal schwannoma of a lumbar vertebral body was reported by Dickson *et al*[8] in 1971, where the lesion was located at the level of L3, but with extension to the spinal canal resulting in compression of the thecal sac. The tumors originally arose from the abutting nerve root, extending into the spinal canal and invading the vertebrae, suggesting that lesions likely involved intraosseous invasion of the extraosseous nerve sheath tumor originating from the spinal nerve root. Wang *et al*[9] described the largest series of cases of intraspinal schwannoma to date, with a retrospective analysis of the clinical features, surgical strategies, and outcomes of 20 cases . In that study, all of the lesions extended from layers A to D of the Weinstein-Boriani- Biagini classification, and all lesions exhibited extraosseous components extending into the spinal canal. In 2001, Sridhar *et al*[10] classified benign nerve sheath tumors, in which V-type tumors caused vertebral erosion. Park *et al*[11] modified this classification by adding type VI and type VII lesions; type VI is a complete intraspinal tumor, while type VII is an intraspinal tumor with vertebral erosion and expansion into the nerve foramen. Mohanty *et al*[12] proposed a new subtype (type VIII) involving intraosseous tumors with an exophytic component, but without any intraspinal or neural foraminal extension. Type VIII, which has an exophytic component, may present with symptoms related to compression of the surrounding structures such as dysphagia or pain, while no radicular pain or myelopathic features are present. Because the tumor does not involve the spinal canal or neuroforamina, it is not necessary to separate the tumor from the nerve root, making the operation safer. Postoperative nerve root numbness was reported in some patient. MRI has better resolution and more specificity than CT as it delineate the tumour from the surrounding structures. CT images failed to adequately reproduce a stroma heterogeneity, the main characteristic sign for the ancient Schwannomas. Schwannomas posses low signal intensity in T1- weighted images similar to muscle and a high signal on T2 weighted images similar to fat. There are recommendations beside standard T1 and T2 weighted images to make a fat suppression sequence (STIR) on which the schwannoma will maintain its high signal allowing a delineation from pure lipomatous tumours[5]. Schwannoma typically enhances dramatically with gadolinium contrast on MRI imaging. The malignancy sign can be asymmetry, irregular infiltrative borders, big tumour size, infiltration, heterogenic pattern and mixed intensity [4,5,3]. Giant invasive retroperitoneal schwannomas are defined as lesions that erode spine vertebrae and extend posterior and laterally into myofascial planes. [11-14]. Giant invasive schwannomas growing in all directions is a challenge to the surgeon. The usual objects of discussion are the surgical approaches and techniques. Even a minimal invasive surgery such as endoscope-assisted minilaparotomy or a laparoscopicsurgery of retroperitoneal tumours has been mentioned in the literature as the choice of the operative technique [11-14]. Schwannomas can cause compressions to the surrounding organs and mimic spine disc herniation, pancreatic cyst, adrenal lesion, psoas muscle abscess, hepatic tumor, bowel or urinary bladder dysfunction like pathology [5]. Different surgical approaches were mentioned: hand-assisted transperitoneal laparoscopic excision, laparoscopic resection, mini-invasive anterior retroperitoneal approach, parasagital incision, anteriorn transabdominal approach, laparotomy through a median incision, suprapubic incision, transverse suprapubic incision, external incision over the swelling, posterior approach (sacral laminectomy), oblique skin incision. In our case the tumor was approached posteriorly and endoscope was used which helped to excise complete tumor where visulisation using microscope was not possible.

CONCLUSION:

Complete surgical resection is the treatment of choice for giant schwannomas with the goal of symptom relief and prevention of recurrence. In our case, resection led to improvement of preoperative symptoms. Complete resection is highly effective for the prevention of recurrence. Giant lumbo sacral schwannomas are benign and slow growing, with an excellent response to complete resection and a good prognosis.

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FIGURE LEGEND

FIG 1- MRI Dorso Lumbar spine, saggital images, T1 contrast

Fig 2- MRI T2 weighted images

Fig 3 & 4 - MRI T1 contrast axial images

Fig 5- MRI DL spine Coronal images

Fig 6 – Tumor specimen