

Case Report

Management of presacral retroperitoneal schwannoma: highlighting the various features

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ABSTRACT

Schwannomas are relatively rare tumours. They occur in approximately 1 of 40,000 hospital admissions. Usually asymptomatic unless very big in size. They may present with symptoms of visceral compression viz. constipation, urinary frequency and urgency, or the nerve compression like sciatica, lower extremity weakness, tingling and numbness. We here highlight the various clinical, radiological and pathologic features the tumour and importance of proper pre-operative imaging, planning regarding surgery and the type of consent required before the surgery in a 30 years old male with a large pelvic mass, diagnosed and supposed to be Schwannoma arising from the S2 sacral foramina. We were able to enucleate the mass intact without any visceral or neuro-vascular injury with minimal blood loss.

Keywords: Schwannoma, Sacral tumour, Neurilemmoma, Retroperitoneal pelvic tumour

INTRODUCTION

Sacral and presacral tumours are uncommon. They occur in approximately 1 of 40,000 hospital admissions.¹ Schwannomas are benign tumours arising from Schwann cells of the peripheral nerve sheath. Spinal schwannomas account for 25% of all primary spinal tumours. Sacral schwannomas are 1% to 5% of all spinal schwannomas.²

It usually grows slowly. It is found incidentally, because it presents with vague and non-specific symptoms. They most commonly present in the fourth decade of life. They are of three types - intradural-extramedullary in 70-75%, extradural in 15% and with both components (hourglass) in another 15%. The intramedullary variety is rare (1%).³

The management of these large sacral tumours is complex. These tumours are mostly monofocal, and usually arise from the exiting sacral nerve roots. Lesions are mostly asymptomatic, but may present with long history with symptoms of compression viz. constipation, urinary frequency, urgency, and nerve compression symptoms like

sciatica, lower extremity weakness, numbness and tingling. The treatment of these tumours is complete resection, with an approach dictated by the size and location of the tumour.⁴ Unexpected neurologic deficits and tumor recurrence after surgery should be considered when surgery is performed. This is due to the difficult access to the lesion, the high chances of local recurrence, and its extreme vascularization that causes a significant intra-operative blood loss. We here report a case of giant retroperitoneal sacral Schwannoma that arise from S-2 sacral foramina without any erosive features and present with a long 12 months history of constipation, urinary frequency and urgency and mild dull chronic pelvic pain.

CASE REPORT

A 30 years young male presented to our urology outpatient department with complaints of chronic constipation, followed by urinary frequency and urgency for last 12 months. It looked to us as lower urinary tract symptoms due to constipation and a routine work up was advised alongwith a sonography of the abdomen and pelvic region.

All routine investigation came normal and sonography suggested a large pelvic mass. This mass was further evaluated by contrast enhanced computed tomography (CT) and magnetic resonance imaging (MRI) of the abdomen and pelvic region. CT suggested a large pelvic heterogenous mass with mild post contrast enhancement (Figure 1 and 2). Origin of the mass could not be reported. MRI suggested a well-defined solid cystic mass of 15.4 cm in the pre-sacral region arising from the right S2 sacral foramina (Figure 3-5). Isointense to hypointense on T1W1 images (Figure 3), heterogeneously hyperintense on T2W1 images (Figure 4) and there was mild heterogeneous enhancement on post contrast images (Figure 5). It was compressing the rectum as well as the bladder and right ureter. But the planes between them and the prostate were maintained. We planned this for surgery.

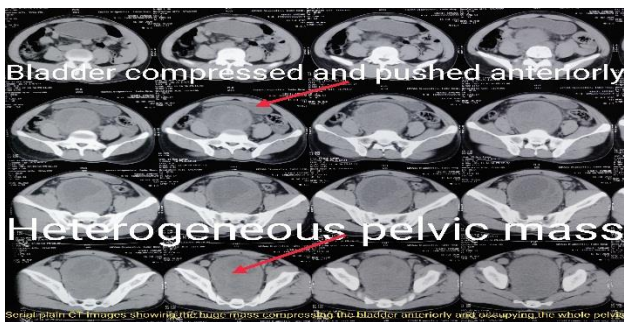


Figure 1: Heterogeneous pelvic mass.

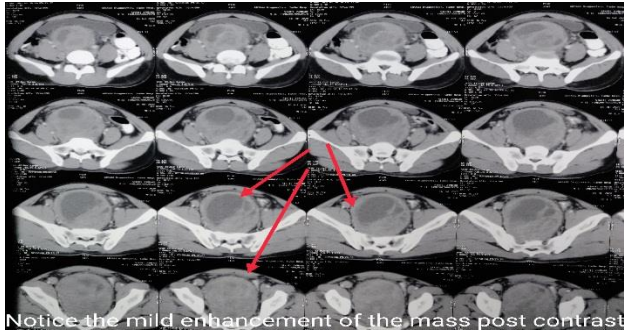


Figure 2: Notice the mild enhancement of the mass post contrast.

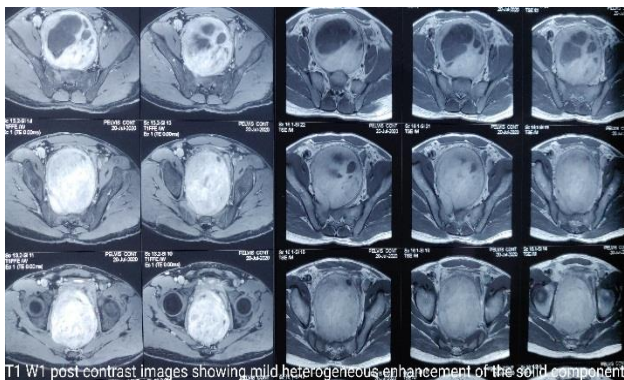


Figure 3: T1W1 post contrast images showing mild heterogeneous enhancement of the solid component.

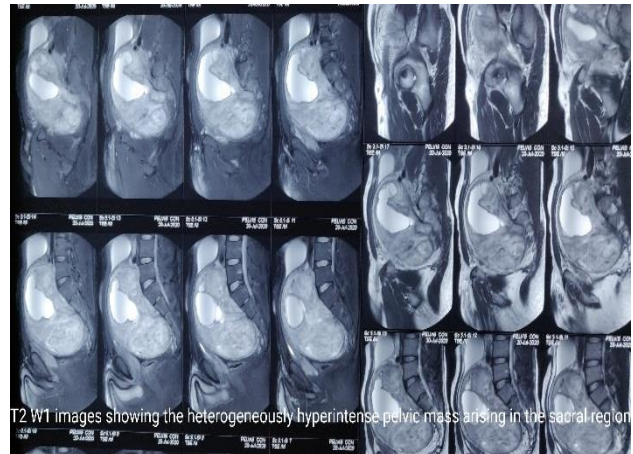


Figure 4: T2W1 images showing heterogeneously pelvic mass arising in the sacral region.

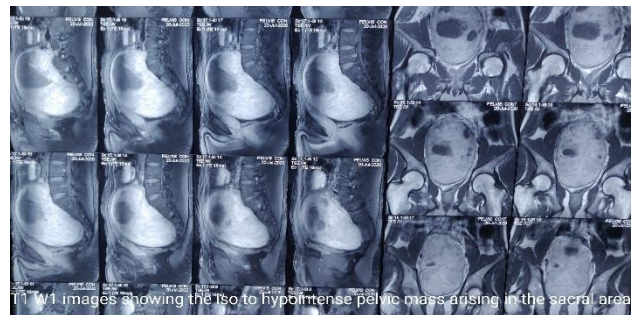


Figure 5: T1W1 images showing the iso to hypointense pelvic mass arising in the sacral region.

Pre-operative discussion with the patient involved discussion about the type of surgery, approach to the tumour, resectability, bleeding, injury to the sacral nerve roots, erectile dysfunction, ejaculation related problems and rectal and urinary incontinence due to nerve injury. We planned to do diagnostic cystoscopy and ureteric catheterisation before the main procedure. But to our surprise we were not able to do cystoscopy properly as the tumour compressed the prostate and the bladder too much anteriorly. Ureteric stenting became out of question with such a difficult cystoscopy. We proceeded with exploratory laparotomy by anterior transabdominal approach. Intra-operatively we were able to find a good intracapsular plane of dissection to enucleate the tumour and were able to isolate both the ureters. Though there was blood loss of around 800 ml, we were able to remove the tumour completely and intact. There was no intra-operative organ or nerve injury. Post-operatively patient recovered well. Till the last follow up of one month his bowel and bladder movements were normal and there was no complaint related to erection or ejaculation. Histopathology report suggested neurilemmoma with the typical features (Figure 6 and 8). Schwannomas are composed of cellular Antoni A areas alternating with hypocellular Antoni B areas. Antoni A areas are composed of interlacing bundles of spindle cells (Schwann cells) as shown here with wavy or oval nuclei, eosinophilic

cytoplasm (Figure 6 marked with arrow). Antoni A areas with consist of interlacing bundles of spindle cells (Schwann cells) and Antoni B areas are hypocellular and less compact than Antoni A areas. They consist of haphazardly arranged spindle cells in a loose myxoid or hyalinized stroma containing a few inflammatory cells and delicate collagen fibres. They also contain variably-sized, ectatic deformed blood vessels which frequently have hyalinized walls and contain thrombi (Figure 7).



Figure 6: Gross pathologic appearance of the Schwannoma.

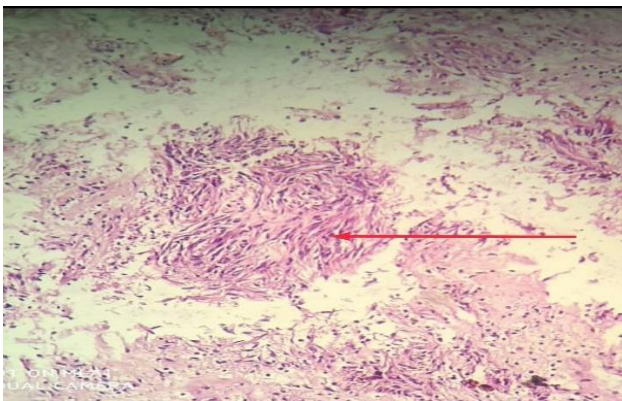


Figure 7: Histopathology of Antoni A.

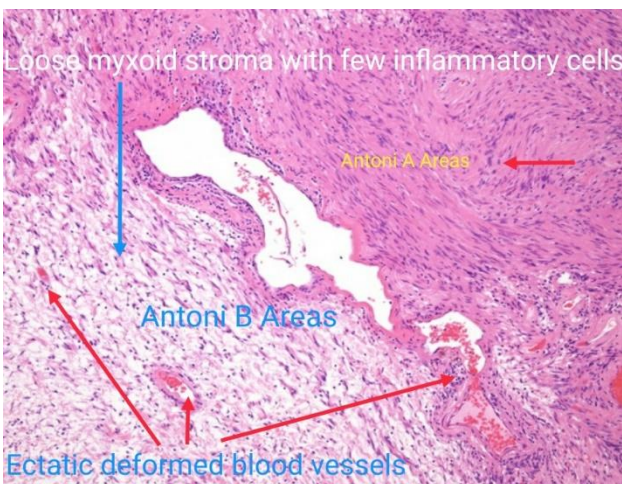


Figure 8: Histopathology of Antoni B.

DISCUSSION

Schwannoma is a benign tumour, arising along sensory nerve roots. Mandible and sacrum are the most common sites of involvement.³

A sacral schwannoma is usually asymptomatic when it is small. Sacral schwannomas are usually found incidentally or when patients present with vague pelvic or lower extremity complaints related to the compression due to the mass effect of the tumour. Our patient came with vague complaints of constipation and related lower urinary tract symptoms and the tumour was diagnosed incidentally on sonography. Lee et al also reported similar complaints in their patient.⁵

Patients usually present in their 2nd to 5th decade of life with female preponderance. Our patient was a male and presented at the age of 30 years.⁶

CT and MRI are the most useful imaging modality for preoperative diagnosis of this tumor. CT gives the details of bony involvement and presacral extension. MRI tells about the heterogeneity of the tumour, enhancement after the gadolinium contrast and tell the extent of the tumor. In our case, we diagnosed the tumour by MRI. The mass was iso to hypointense in T1W1 (Figure 3) and heterogeneously hyperintense on T2W1 images (Figure 4). Post contrast the solid component showed mild heterogenous enhancement (Figure 5). There was no bony invasion/erosion except the compression of the pelvic structures by the tumour with right sided hydroureteronephrosis. Hughes et al also suggested similar characteristics of the tumour on MRI.⁷

The surgical approach for this tumour depends upon the degree of bony sacral involvement and retroperitoneal extension. The posterior approach is suited for the tumor involving the spinal canal and intra-osseous sacrum involvement. An anterior transabdominal or retroperitoneal approach should be performed for a presacral retroperitoneal tumor to gain control of the vascular plexus that encompasses the tumor and to ensure identification and protection of the intrapelvic organs.¹

Different approaches for the excision of the tumour are described viz. a piecemeal subtotal excision or simple enucleation or sometimes an aggressive approach with the aim of achieving a complete resection.¹

We used the anterior transabdominal approach. We also tried stenting the ureters first due to compression on right side, but failed due to anterior displacement of the prostate and the bladder by the tumour. We were able to enucleate the tumour completely without damaging the adjacent neuro-vascular structures and other visceral organs. Till last follow up there was no evidence of recurrence of the tumour in our case. Dominguez et al also used a conservative approach with intracapsular enucleation

alone and gave a favourable result with only 16 percent recurrence rate.⁶

Schwannomas do not exhibit rich blood supply, still there is relatively more blood loss during excision of the tumour. This is due to intraoperative haemorrhage from venous plexus of sacral canal. Managing the venous plexus haemorrhage can effectively decrease the intraoperative blood loss. Pongsthorn et al reported the intraoperative blood loss was 2572 g (range 483-5301 g).¹ We had intraoperative blood loss of around 800 ml.

Usually a long-term follow-up is required to identify the recurrence of the tumour. In study by the recurrence rate was zero percent with a follow-up period of 7 months to 27 months (mean 18 months).⁸ We are also in follow-up of our patient for last one month only and there is no recurrence.

CONCLUSION

The typical vague clinical features, MRI findings of our case of a well-circumscribed lesion with a heterogenous signal intensity on T2-weighted image, without any bony erosion was very helpful for preoperative decision-making and giving an idea of the Schwannoma. With the transabdominal anterior approach, we were successfully able to enucleate the tumour with minimal blood loss. We should always try to safeguard the nerve roots, even if they look involved as the preservation of the function is more important than the presence of residual tumour.

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Ethical approval: Not required

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