A case report of a giant presacral cystic schwannoma with sigmoid megacolon

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ABSTRACT

Schwannomas are peripheral nerve sheath tumours with a slow growth rate. Giant sacral schwannoma with anterior cortex erosion and associated intrapelvic extension are uncommon. Though they tend to be large when initially found, most Giant schwannomas are clinically asymptomatic. The tumour appears heterogenous due to long standing degeneration. Herein, we present a case of a large purely cystic presacral schwannoma in a patient with poliomyelitis, which has displaced adjacent organs including urinary bladder and sigmoid colon, with an initial presentation of constipation. The tumour was partially excised and diagnosis was confirmed by histo-pathology and immunohistochemistry.

CASE REPORT

CASE REPORT

A 35years old male with poliomyelitis (Fig.1) involving both lower limbs, presented with low back ache since two months, severe abdominal pain, abdominal distention and constipation since 2 weeks. On physical examination, two soft tissue masses were palpable in the suprapubic and right lumbar regions. All other clinical and lab parameters were within normal limits. Initial supine abdominal radiograph (Fig.2) showed scoliosis to the left side, bone destructions in the left side of the sacrum, bilateral femoral bone deformities and dilated colon on the right side. The patient was subjected to further radiological examinations for origin and characterization of the mass lesion.

Abdominal sonography revealed a cystic lesion involving the pelvis and a large dilated fluid filled colon in the right lumbar region. Further evaluation by computed tomography and MRI scans revealed a large dilated air and fluid filled sigmoid megacolon with a maximum diameter of 13 cms (Fig.3) and a cystic encapsulated mass in the presacral region measuring 13 x 14 x 14cms with thick septations, arising to the left side of the midline (Fig. 4,5 & 6).

On MR imaging, the tumour showed a thin pseudo-capsule, which was hypointense in all sequences (Fig.5). The tumour was attached to the anterior surface of the sacrum with minor involvement of the underlying bone. This latter feature was more clearly appreciated on the CT scan (Fig.7). Fine peripheral calcification and small foci of calcifications within the tumour, readily identified on CT could not be identified on the MR images (Fig.7). Initial diagnosis was a benign retroperitoneal cystic tumour with pressure erosions over the anterior cortex of the left sacrum and associated sigmoid megacolon. Patient was taken up for surgery. Surgery was done through an anterior approach. Sigmoid megacolon (Fig.8) was indentified in the right lumbar region and a large well encapsulated cystic tumour was found adherent to the anterior aspect of the sacrum (Fig. 9). As it was adherent complete excision of the tumour was unsuccessful. The cystic component was aspirated using a syringe (Fig.10) and tumour was partially resected (Fig.11). Haemorrhage was noted inside the tumour. The post operative course was uneventful. Patient recovered completely after 10 days.

Pathological examination revealed irregular cut open cyst wall measuring 8 x 6 cms with smooth grey brown surface and maximum wall thickness of 6mm with areas of haemorrhage (Fig.12). Microscopically, multiple sections studied from the cyst wall shows palisading of the nuclei with hyaline thickening of the blood vessel, fibrohyalinized collagenous tissue with overlying schwannian cells in hypercellular areas, areas of haemorrhage, calcification and cystic changes, features suggestive of benign cystic schwannoma (Fig.13). Immunohistochemistry demonstrated cystoplasmic positivity of S-100 protein in >90% of cells confirming the diagnosis (Fig.14).

DISCUSSION

Sacral tumours with presacral extension are uncommon, cited historically as being responsible for 1 in 40,000 hospital admissions (1). Schwannomas are peripheral nerve sheath tumours arising from neural crest-derived Schwann cells (2). Although the tumour arises from the peripheral nerve sheath, it rarely elicits any clinically detectable neuro-logical deficits. Most Schwannomas are benign. Malignant tumours have been reported but are usually associated with von Recklinghausen's disease (3). Giant sacral schwannomas (GSS) arise from a peripheral nerve within the sacrum and produce moderate to massive bone destruction before extending into the soft tissues. In contrast, giant presacral Schwannomas (GPSS), as illustrated in this case report and in another case report (4), presumably arise from a peripheral nerve in or adjacent to an anterior sacral foramen. Therefore, they are able to grow outside of the confines of the bone with a limited degree of secondary bone involvement. Minimal bone destruction and relative absence of compression of neurovascular structures can explain why tumours are able to grow to such a large size before presentation either as incidental findings or with only minor complaints (5).

Sacrococcygeal and presacral masses have a wide range of differential diagnosis (6). MR and CT are ideally suited to detect sacral pathology and delineate the soft tissue and bony components. Bone destruction was best visualized on CT. Multiplanar imaging in both MR and CT helps in exact identification of the level of involvement and level of nerve root involvement as this is important for pre-operative surgical staging. Morphological features in cross-sectional imaging exclude differential diagnoses such as aneurysmal bone cyst in children and adolescents, abscess, GCT and chordoma. Giant sacral schwannoma may therefore be easily mistaken for one of these conditions, whereas GPSS appears well defined with only minor bone involvement, just lateral to the midline, corresponding to one of the anterior sacral foramina, unlike chordoma which is almost invariably a midline lesion (7).

MR imaging features which suggest the diagnosis of a neurogenic tumour include the presence of a fusiform shaped mass with an entering and exiting tail representing the host nerve (8). Typical features like eccentrically placed schwannoma (9), the "split fat sign" (10), the " target sign" (8) and the "fascicular sign"(11), are not seen in GPSS by virtue of the enormous size of the lesion as compared with the underlying nerve (5). Heterogeneity can be seen on MR imaging, which reflects degenerative change of long -standing schwannomas. The so- called "ancient schwannoma" refers to a degenerative schwannoma typified by cyst formation, hemorrhage, calcification and fibrosis (11). The diagnosis of GPSS can be suggested when identifying a large, well defined, heterogenous presacral soft tissue mass with a low signal intensity rim (fibrous pseudocapsule), arising just to one side of the midline, with minor bone erosion. Features claimed to be more suggestive of a malignant peripheral nerve sheath tumour (MPNST) formerly known as malignant schwannoma than the benign counterpart include large size (>5 cm) and heterogeneity (are also seen in GPSS). MR imaging can also differentiate malignant schwannoma from benign types by a lack of a pseudo-capsule, indistinct margins and satellite lesion (5). Biopsy is still required for final diagnosis. Preoperative diagnosis of a presacral schwannoma is difficult. Radiological examinations such as ultrasonography, CT or MRI usually cannot discriminate retroperitoneal tumours from retroperitoneal schwannomas. However, cystic changes may be one of the characteristic radiological features (12).

Grossly, benign Schwannomas are well-circumscribed and encapsulated masses; malignant schwannomas are poorly defined masses with frequent invasion of adjacent soft tissues in contrast to the benign types. Microscopically, benign Schwannomas show a mixture of 2 growth patterns: Antoni A and Antoni B. The Antoni A growth pattern presents elongated cells with cytoplasmic processes arranged in fascicles in areas of moderate to high cellularity with little stromal matrix. The Antoni B growth pattern shows a lower density of cells with a loose meshwork of cells along with micro cysts and myxoid changes. Degenerative changes like nuclear pleomorphism, xanthomatous change and vascular hyalinization may be found in benign schwannomas. A wide range of histological findings can be observed in malignant schwannomas such as patterns of reminiscent of fibro sarcomas or malignant fibrous histiocytomas. A possibility of malignant schwannoma is high, in association with von Recklinghausen's disease. Biopsy is still required to rule out the malignant schwannoma. It is thus suggested to perform more radical surgery, including excision of adjacent organs (13).

We report a rare giant presacral schwannoma, the interesting findings are that its clinical manifestation of constipation, severe abdominal pain and distention associated sigmoid megacolon in a patient with childhood history of poliomyelitis. The surgical outcome was fair at 6 months of postoperative follow up. Recurrence is rare and patients live a long, symptom free life even after partial resection (14). To our knowledge, presacral schwannoma presenting with such clinical manifestations has not been mentioned in any English literature.

TEACHING POINT

Sacrococcygeal and presacral masses have a wide range of differential diagnosis. A multimodality approach can demonstrate the exact location and extent of the mass and can also differentiate benign tumors with malignant aiding in the planning for treatment.

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FIGURES

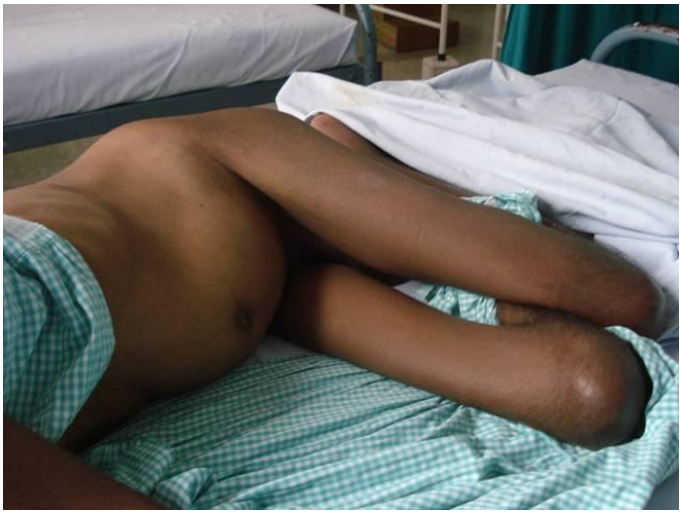

Figure 1. 35 yrs old male with giant presacral schwannoma: Photograph of the patient with poliomyelitis involving both lower limbs.

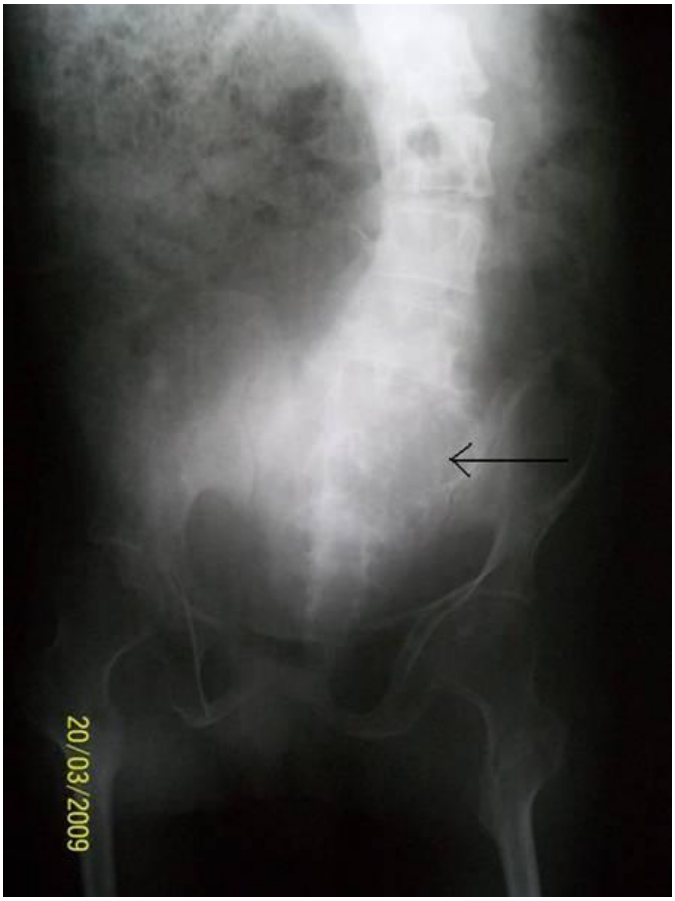

Figure 2. 35 yrs old male with giant presacral schwannoma: Supine abdomen radiograph showed scoliosis to the left side, bone destructions in the left side of the sacrum, bilateral femoral bone deformities and dilated colon on the right side.

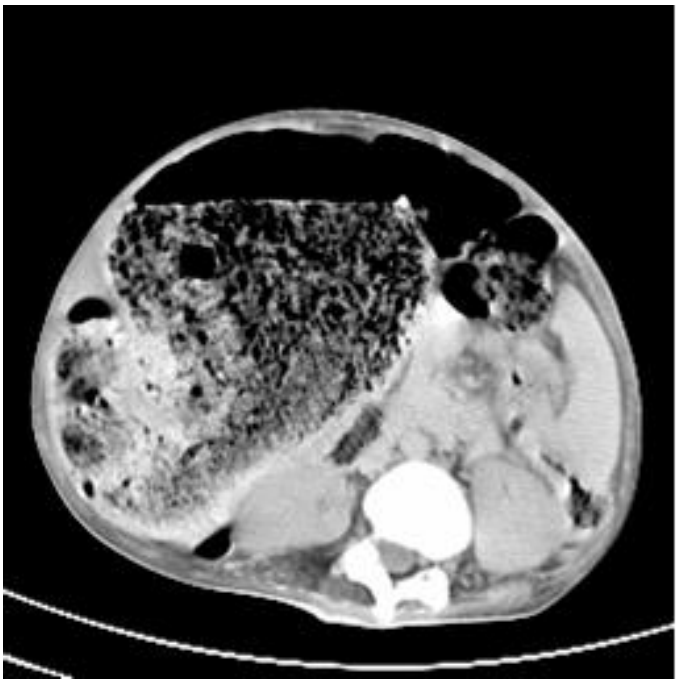

Figure 3. 35 yrs old male with giant presacral schwannoma: Axial non-contrast CT scan (130kv/150mAs) at the level of the kidneys, showing enlarged dilated air and fluid filled sigmoid colon.

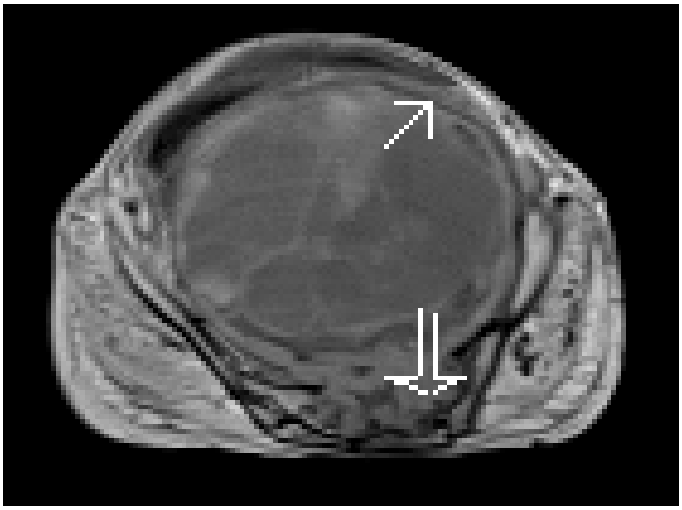

Figure 4. 35 yrs old male with giant presacral schwannoma: Axial T1 weighted image (0.35T, TR/TE, 630/22) showing a large mass lesion with a hypointense peripheral rim (white arrow) and focal bone destruction in the anterior aspect of the sacrum on the left side (white open arrow).

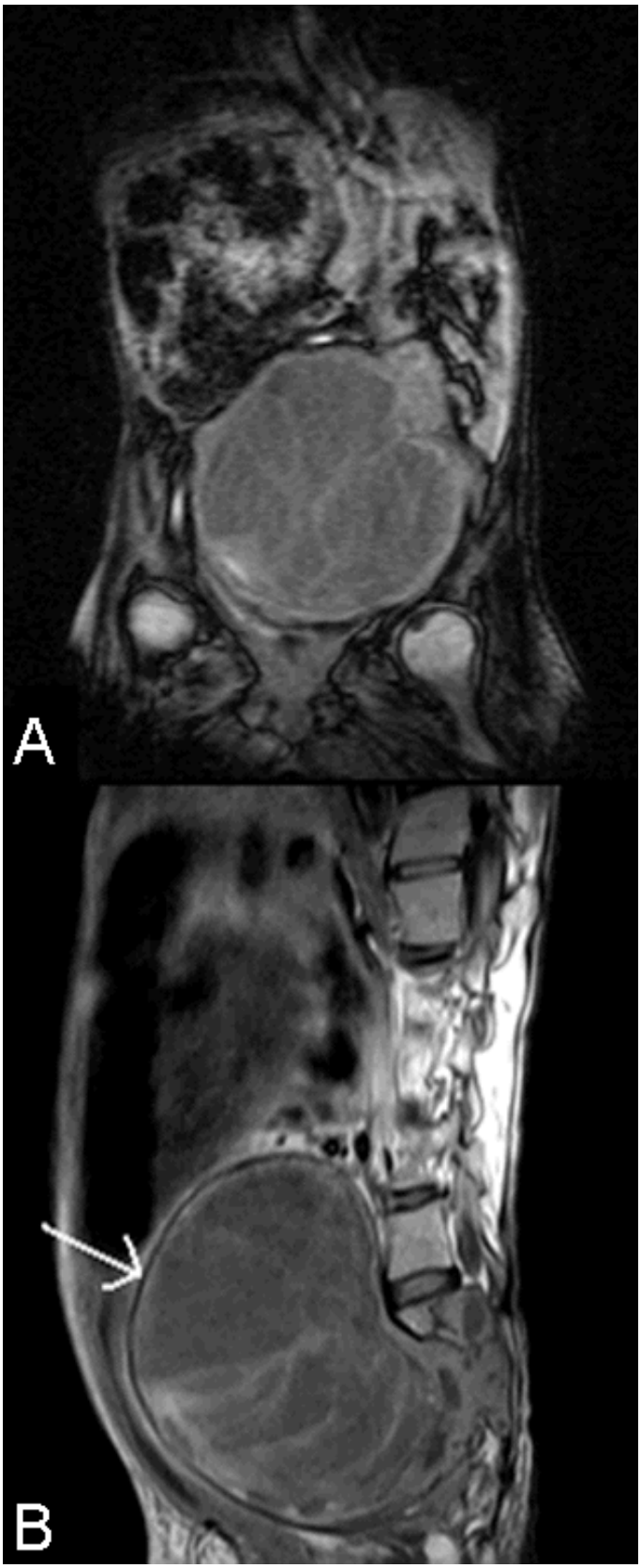

Figure 5. A & B: 35 yrs old male with giant presacral schwannoma: Coronal and sagittal T1 weighted images (0.35T, TR/TE, 500/17) showing large mass in the pelvis with a hypointense peripheral pseudocapsule (arrow) and internal septations.

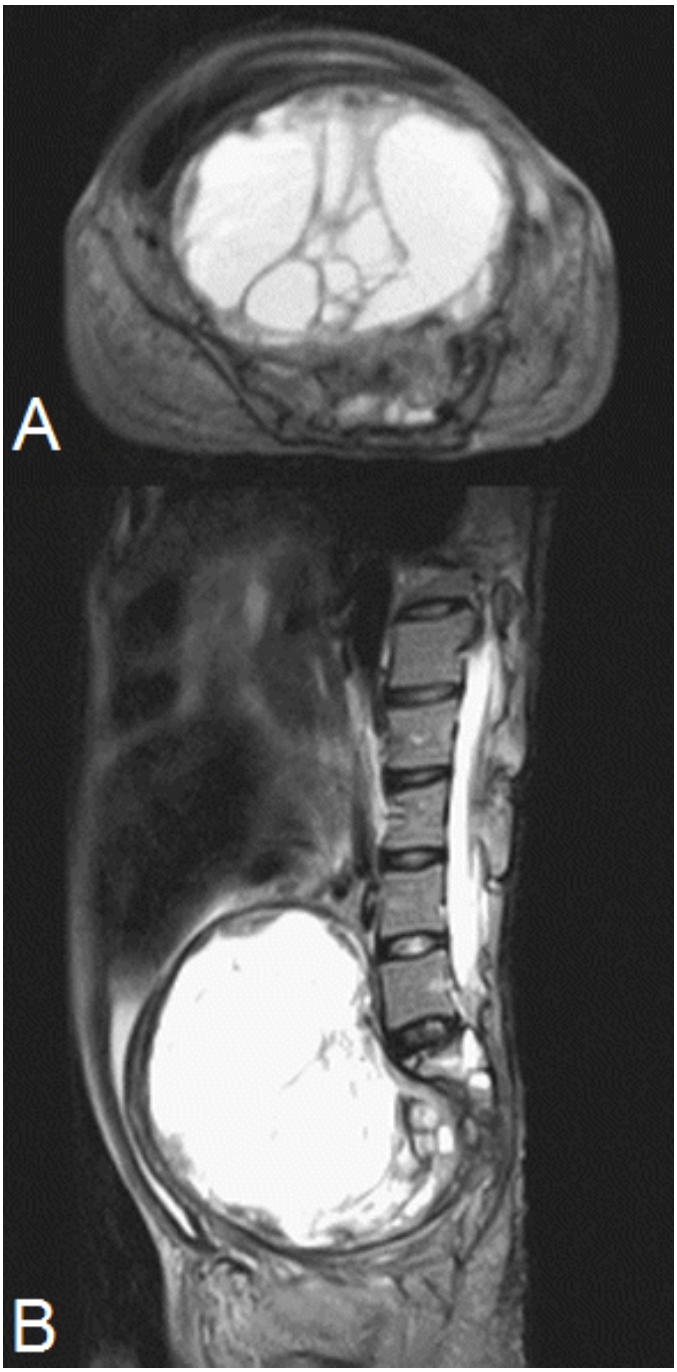

Figure 6. A & B: 35 yrs old male with giant presacral schwannoma. T2 weighted coronal and sagittal images (0.35T, TR/TE, 4600/139) showing large hyperintense mass in the pelvis with septations and a peripheral hypo intense rim (pseudocapsule).

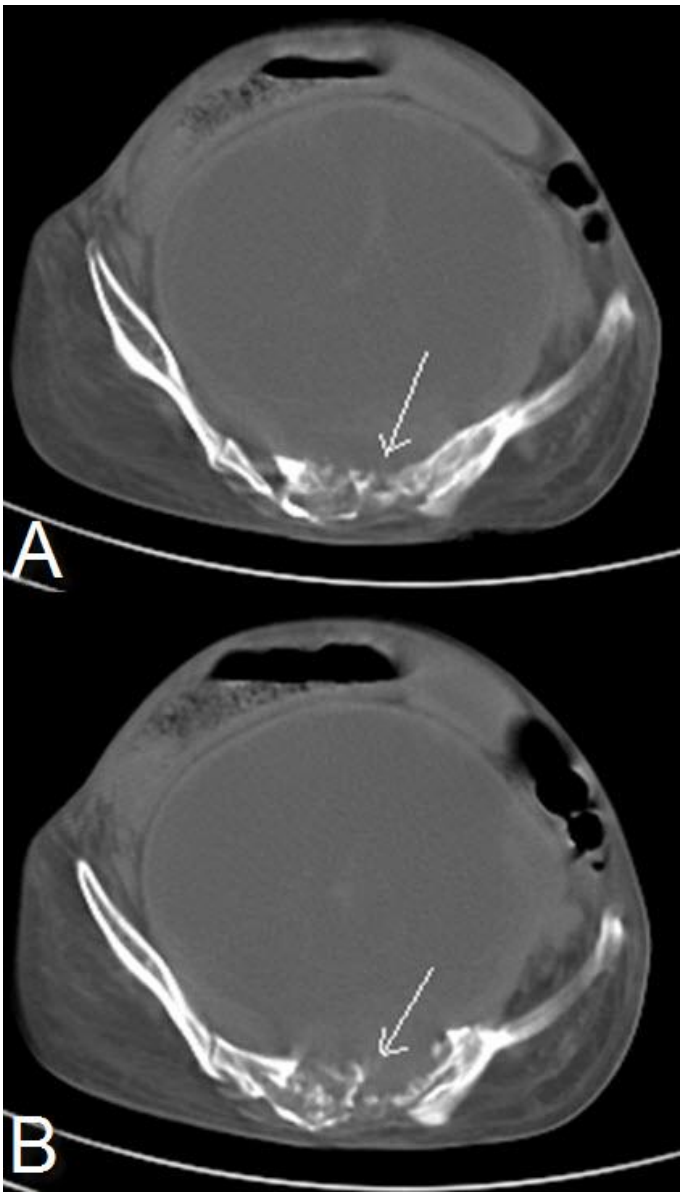

Figure 7. A & B: 35 yrs old male with giant presacral schwannoma: Bone window of axial CT sections (130kv/150mAs) of the pelvis showing large mass with erosions of the anterior cortex of the sacrum on the left side (arrow) and mass effect over the sigmoid colon.

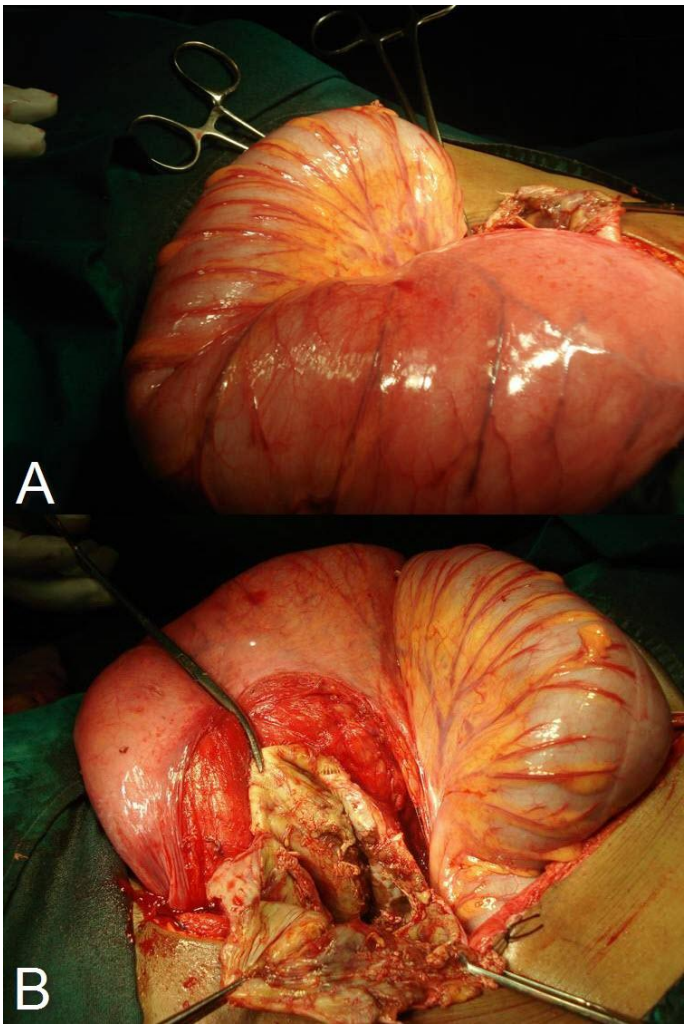

Figure 8. A & B: 35 yrs old male with giant presacral schwannoma: Perioperative photograph showing sigmoid mega colon.

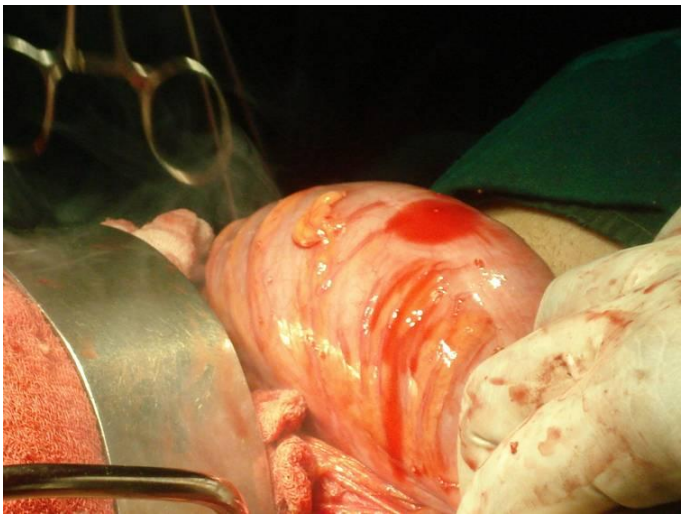

Figure 9. 35 yrs old male with giant presacral schwannoma. Perioperative photograph showing the cystic mass in the pelvis.

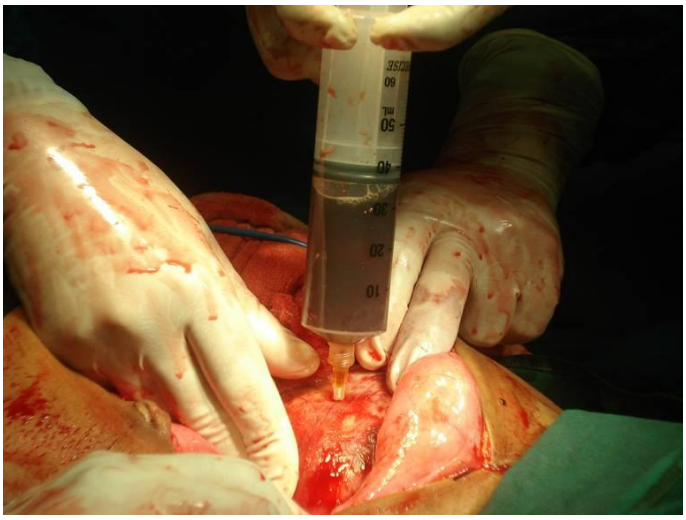

Figure 10. 35 yrs old male with giant presacral schwannoma: Perioperative photograph showing aspiration of fluid from the cystic mass.

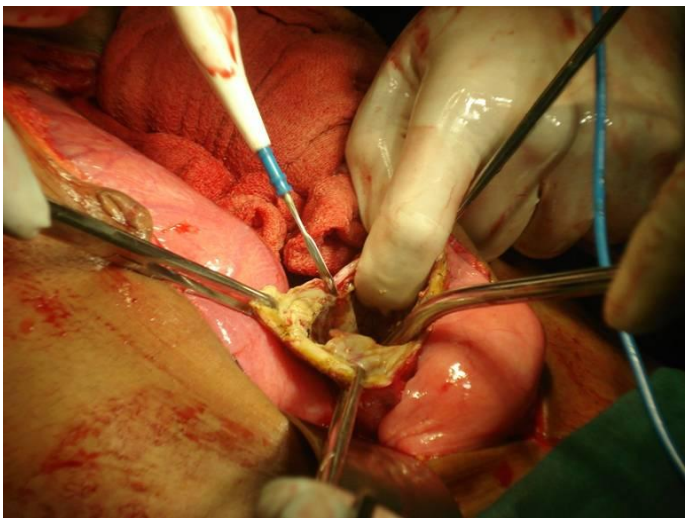

Figure 11. 35 yrs old male with giant presacral schwannoma: Perioperative photograph showing partial resection of the cystic mass.

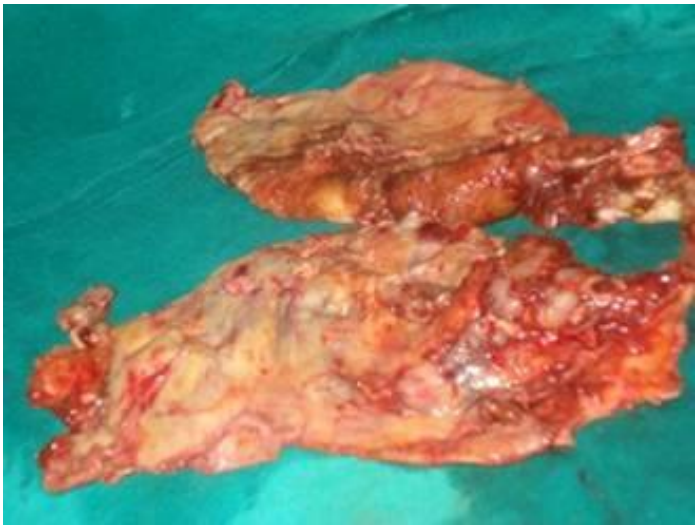

Figure 12: 35 yrs old male with giant presacral schwannoma: Partially resected specimen showing areas of hemorrhage.

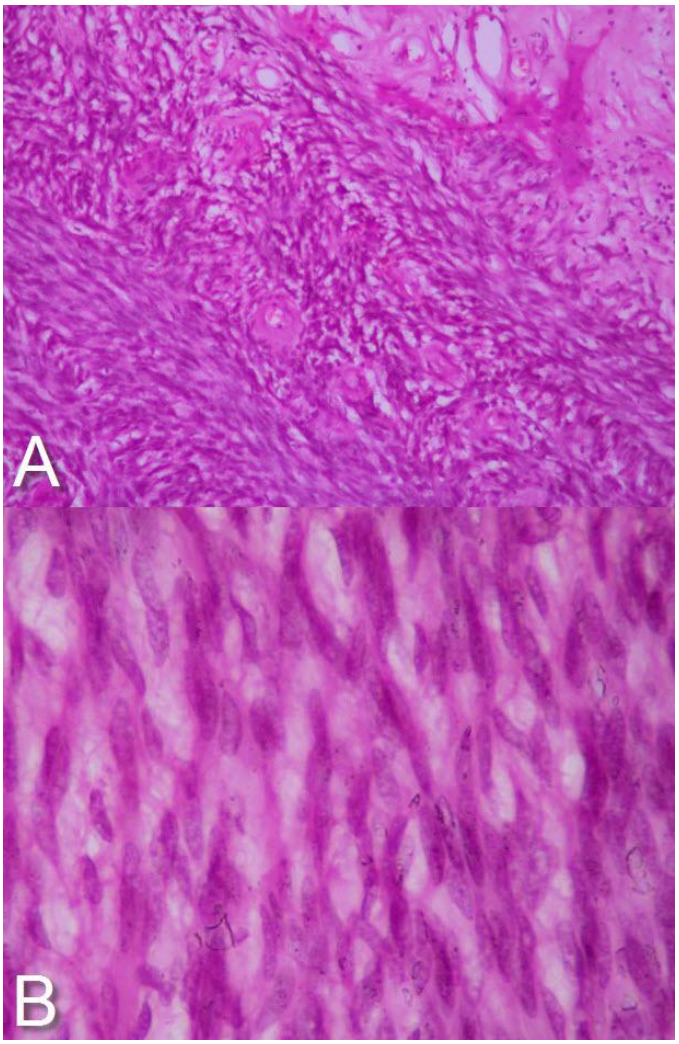

Figure 13. A: 35 yrs old male with giant presacral schwannoma: Microphotograph (200X, H & E) showing palisading of the nuclei with hyaline thickening of the blood vessel. B: 35 yrs old male with giant presacral schwannoma: Microphotograph (400X, H & E) showing tumor composed of fibrohyalinized collagenous tissue with overlying Schwann cells in hyper cellular areas.

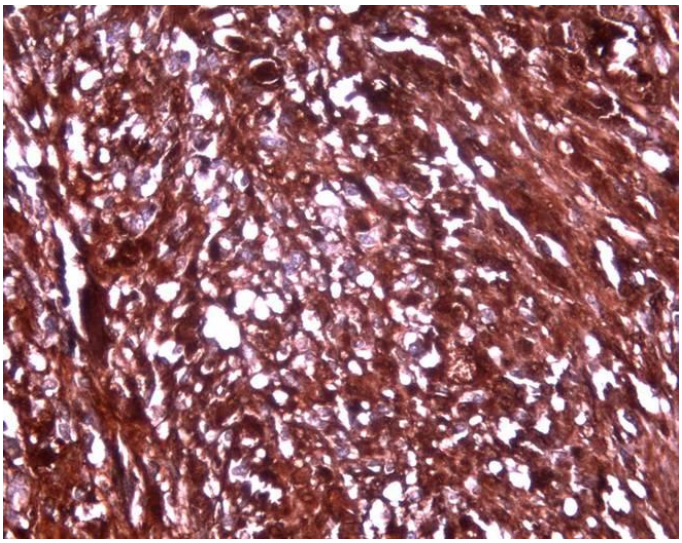

Figure 14. 35 yrs old male with Giant Presacral Schwannoma: Microphotograph (200X) showing S-100 immunoreactivity in the tumor cells.

ABBREVIATIONS

GPSS = Giant presacral sacral schwannoma
CT = Computed Tomography
MRI = Magnetic Resonance Imaging
GCT = Giant cell tumor
GSS = Giant cell sacral schwannoma
MPNST = Malignant peripheral nerve sheath tumour

KEYWORDS

CT, MR imaging, giant schwannoma, presacral schwannoma, cystic schwannoma, megacolon