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Case report

Giant presacral-pelvic cystic schwannoma presenting as a right iliac fossa mass - megacolon in a poliomyelitis patient : a rare case report & review of literature

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ABSTRACT

Sacral & presacral tumours are uncommon & occur in approximately 1 in 40,000 hospital admissions.Schwannoma is one of the tumours that occur in these areas and occasionally presents with enormous dimensions known as Giant schwannoma.It usually grows slowly and is often found incidentally because it presents with vague and non-specific symptoms. Tumour removal is a surgical challenge due to the difficult approach and vascularity.We report a rare case of Giant presacral schwannoma presenting as a right iliac fossa mass with subacute intestinal obstruction- megacolon in a childhood poliomyelitis patient,who underwent total excision of the tumour and HPE report was benign schwannoma with cystic degeneration and confirmed by Immunohistochemistry. The patient was relieved of his bowel symptoms (obstruction). Recurrence was not noted even after 2 yrs of follow up.

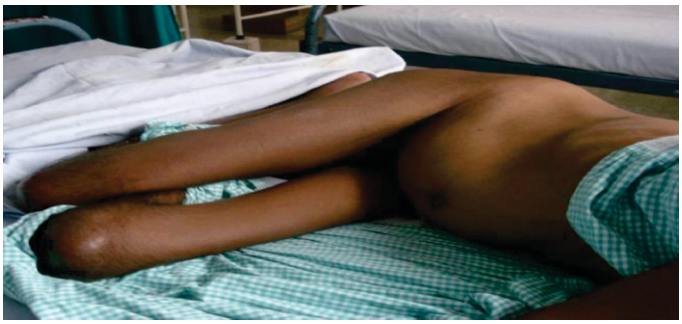
1. Introduction

Giant presacral schwannomas are uncommon and occur 1 in 40000 hospital admissions [1,2]. Benign schwannomas are generally slow growing and painless tumours originating from Schwann cells of peripheral nerve sheath of the neural crest [1,2]. Symptoms occur late unless they are large enough to produce pressure effect. These are slow growing and often found incidentally [3-5]. Due to its non specific clinical and imaging findings preoperative diagnosis is very difficult [3]. FNAC is diagnostic in differentiating benign from malignant schwannoma [6-7]. We report a rare case of giant presacral schwannoma in a patient with poliomyelitis who presented with subacute intestinal obstruction.

2.Case Illustration

A 26 yrs old male patient with childhood poliomyelitis {FIG-1} presented with low back ache since 2 months, vomiting,distension of abdomen and constipation from 2 weeks.On clinical examination two firm masses were papable per abdomen,one in the right iliac region 10 x 7cms non tender,non mobile and another in the suprapubic region measuring 6 x 3cms The upper pole was reachable and lower pole was diffuse. Impaired note was heard over both the masses. On manupulation of the masses,the patient complained of nausea and sensation to defecate. A clinical diagnosis of lobulated retroperitoneal soft tissue tumour with subacute intestinal obstruction was made.

FIG – 1; Photograph of 26 year old male patient with poliomyelitis: with Giant presacral schwannoma with subacute intestinal obstruction.



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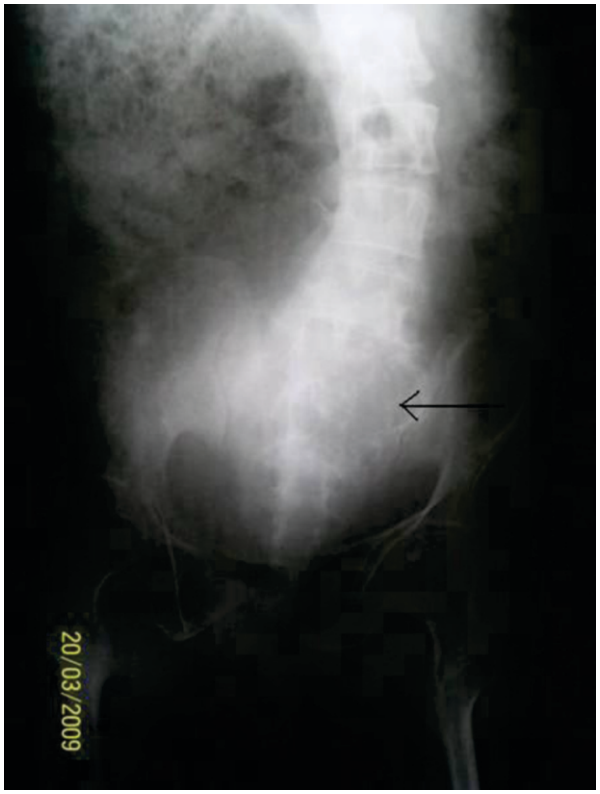
All clinical and lab investigations were within normal limits. USG abdomen and pelvis revealed a cystic swelling in the pelvis[Fig-2]. A large dilated fluid filled colon imaged in the right iliac region on supine abdomen x-ray. Origin of mass could not be commented . CT of the pelvis and MRI scans revealed, a large dilated air fluid filled sigmoid megacolon of 13 cms diameter [Fig-3,CT,MRI].

FIG-2;A supine abdomen x-ray shows scoliosis to left side,sacral bone destruction,B/L femoral bone deformity ,dilated colon on right side.

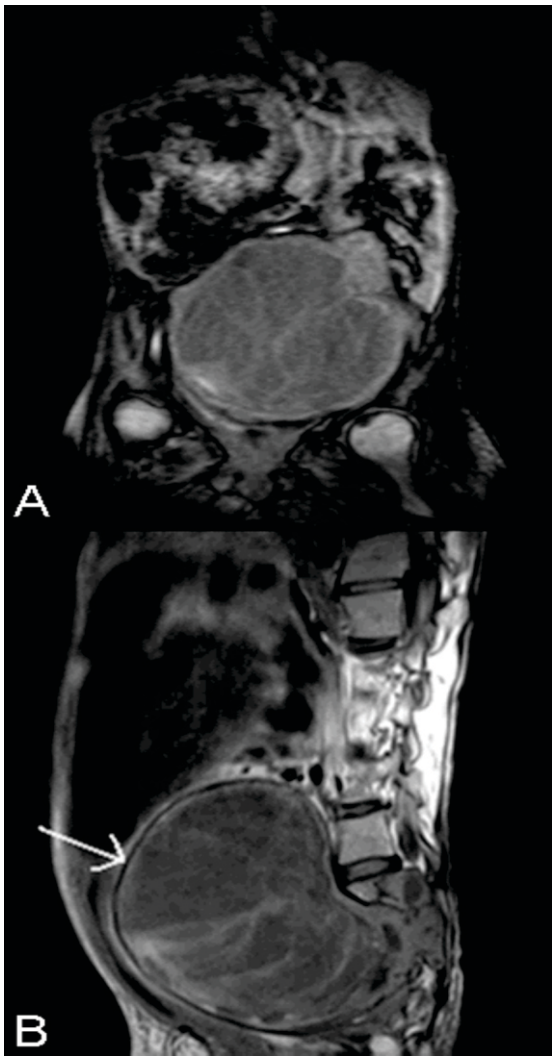


A pelvic tumour in the presacral region measuring 13 x14 x14 cms,arising to the left of midline cystic, encapsulated mass with septations. The tumour was attached to the anterior surface of the sacrum with pressure erosions. Tumour had fine peripheral calcification and small foci of the same within the tumour. A diagnosis of benign retroperitoneal cystic tumour with erosion of sacral cortex with sigmoid megacolon was made.

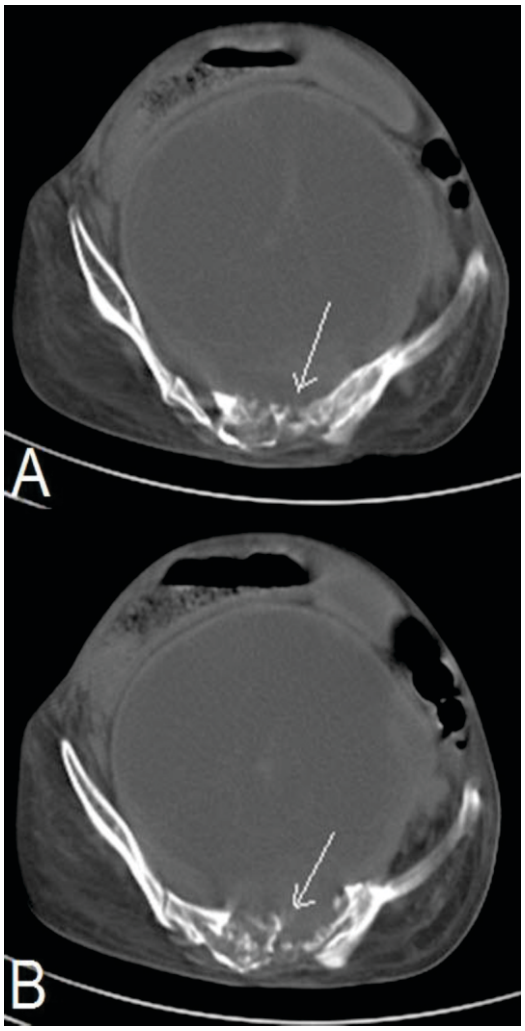
The patient was taken up for surgery under general anaesthesia and by transabdominal/transperitoneal approach. The presacral cystic tumour was totally excised. Peroperative findings revealed hugely distended megacolon in the right lumbar and iliac region.[perop Fig].A large well encapsulated 15 x13 cms tumour was noted adherent to the anterior surface of sacrum compressing over the sigmoid colon,which was the cause for bowel obstruction [perop Fig]. Due to its large size to make space for dissection,the tumour was aspirated and the content was 600 ml of haemorragic fluid[perop-fig]. By blunt dissection the cystic tumour was freed all around though the capsule was densely adherent to the anterior surface of sacrum. The cystic tumour was opened up and contents apirated. The inner cyst wall had multiple septations,calcified, necrotic and haemorrhagic areas. The cystic presacral tumour was totally excised and haemostasis was well maintained as it was very vascular and close to sacral veins.

On histopathological examination [HPR NO:B/1165/08 dated 8-9-08] multiple sections studied from the cyst shows fibrohyalinised collagenous tissue with overlying schwanian cells in hyper cellular areas. Areas of haemorrhage, hyalinization, calcification and cyst change seen. The HPE report was suggestive of benign schwannoma with cystic degeneration.[HPE-Figs]. Postoperative recovery was good . Patient was relieved of bowel obstruction completely and was discharged after a week. Patient is disease free and no recurrence or sensory neurological deficits noted even after 2 yrs of follow up.

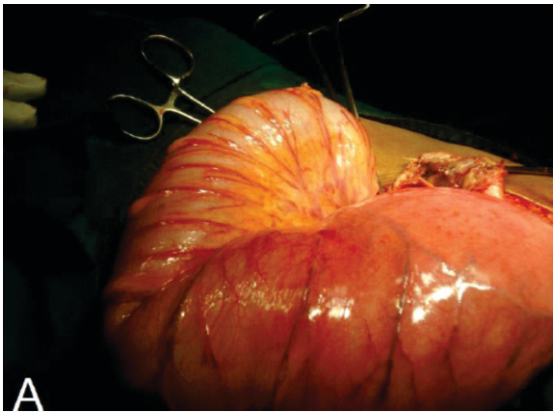
FIG-A&B Coronal and Saggital images showing large mass in the pelvis with a hypointense peripheral pseudocapsule[arrow] and internal septations.



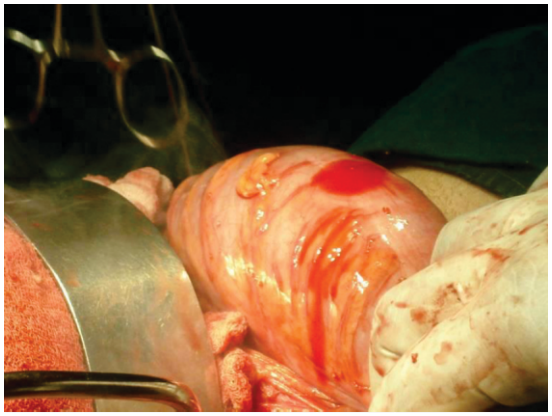
Axial CT sections of pelvis showing large mass with erosions of sacral cortex on left side[arrow] and mass effect over sigmoid colon.



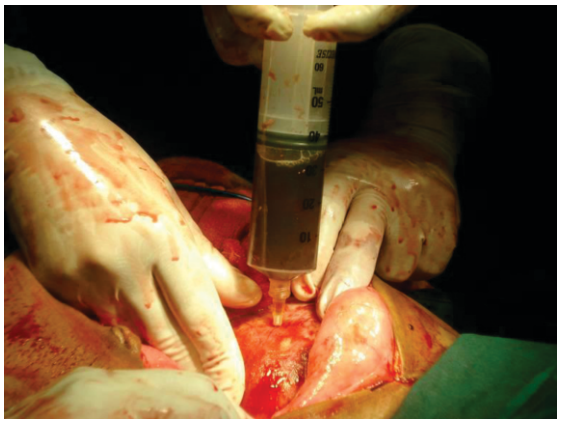
Op Fig 1: Showing hugely distended sigmoid colon (mega colon).



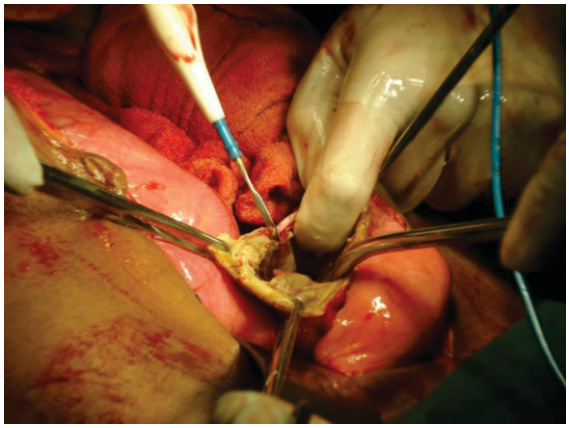
Op Fig 2: Showing the presacral cystic tumour.



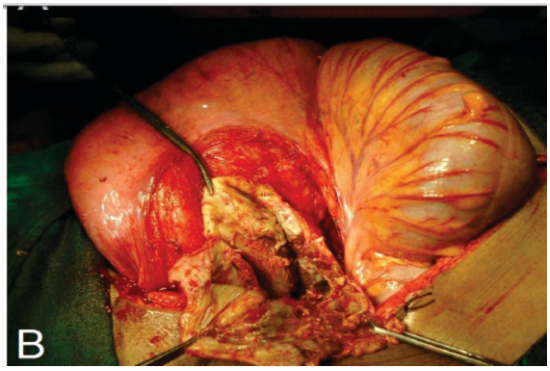
Op Fig 3: Showing aspiration of haemorrhagic fluid from the presacral cystic tumour to decompress



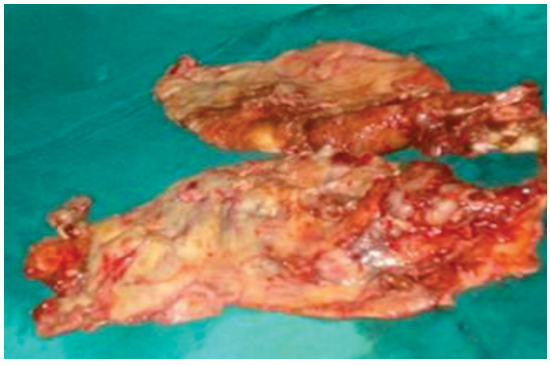
Op Fig 4: presacral cystic tumour being opened up and contents aspirated.



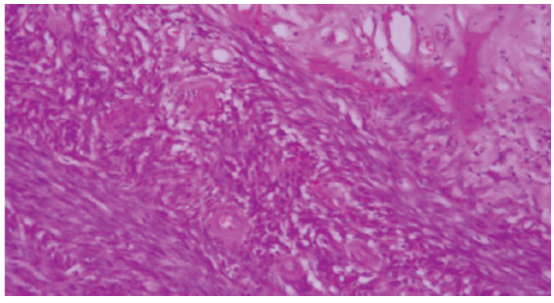
Op Fig 5 : Showing megacolon & the cut open presacral cystic tumour.



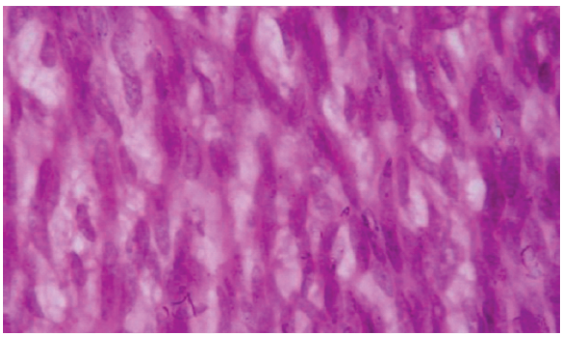
Op Fig 6: Showing totally excised specimen of presacral cystic tumour.



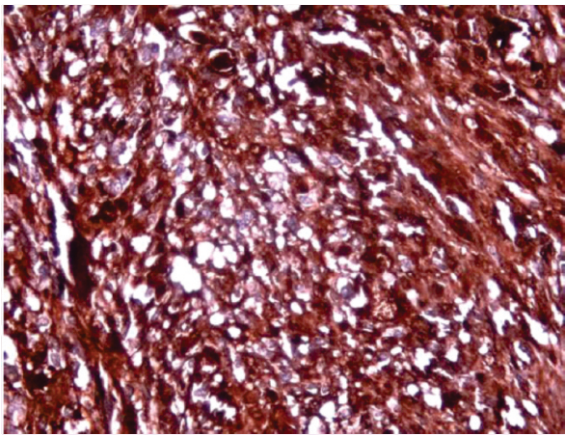
HP Report 1: Microphotograph (200X, H & E) showing palisading of the nuclei with hyaline thickening of the blood vessel.



HP – Report 2: Microphotograph (400X, H & E) showing tumour composed of fibrohyalinized collagenous tissue with overlying Schwann cells in hyper cellular areas – benign cystic schwannoma.



Immuno – histochemistry Microphotograph (200X) showing S-100 immunoreactivity in the tumour cells.



3. Discussion

Giant presacral Schwannomas are benign neoplasm arising from the myelinated nerve sheaths. Malignant schwannomas occur denovo or from transformation of plexiform neurofibromatosis [7-12]. However our patient did not have the disease.

Pelvic schwannoma is rare and accounts for 1% of all Benign schwannoma.11 As these are slow growing and painless tumours, symptoms occur late ,unless they are large enough for pressure effect .They are low backache ,pain in the abdomen and pelvis, urinary and digestive symptoms from bladder and rectal compression.6 However clinical manifestation of constipation and intestinal obstruction with megacolon as noted in our case is very rare.

Giant presacral schwannomas arise from peripheral nerve sheaths within the sacrum or adjacent to sacral foramen and grow out side the bony frame work , to a large size. Erosion of the bone and compressive symptoms occur rather late.4,5,12 Ancient variant of schwannoma is rare and two cases have been reported in literature so far. These undergo degenerative changes such as cyst, haemorrhagic, calcification, and fibrosis [11,13]. The mechanism of degeneration is attributed to vascular insufficiency [4].

Preoperative diagnosis of presacral schwannoma is difficult. CT and MRI are widely used as imaging modalities in evaluation of retroperitoneal soft tissue tumours [4,13]. The MRI characteristics of presacral schwannoma are homogenous or heterogenous / cystic appearance with well defined margin and ISO/ISO intensity ,ISO/HIGH intensity on T1/T2W1 [4]. CT can accurately detect bony destruction and sacro iliac joint involvement4. Definitive diagnosis is based on histo pathology of the biopsied specimen, that can differentiate benign from malignant schwannomas. Immunohistochemistry is confirmatory of benign schwannomas as they show diffuse immune reactivity to S-100 protein [7,9].

Total tumour excision is considered the treatment of choice for presacral schwannomas [5]. Since tumour recurrence and malignant transformation almost never occurs in benign schwannomas local tumour excision can be considered [5,9].
Utmost care is required in surgical removal of retroperitoneal and intrapelvic schwannomas [5]. Generally anterior transabdominal or retroperitoneal approach should be performed for presacral schwannomas to gain control of vascular plexus and protection of intrapelvic organs [14].
If the tumour or tumour capsule is densely adherent to the presacral venous plexus , bleeding can be hazardous and haemostasis can pose a problem . Hence piecemeal or subtotal excision of the tumour can be considered [5]. Though, the benign nature of the the tumour has to be confirmed by histopathology. If the HPE confirms malignancy ,local recurrence after marginal excision has to be expected in upto 72%, while wide marginal excision has been reported in 11.7% 9. In a case report by Foote et al, the attempt to excise a large retroperitoneal schwannoma was abandoned because of danger of uncontrollable bleeding [5,8].

In our case the tumour capsule was densely adherent to sacrum and by blunt dissection total excision of the cystic tumour was done. We had blood loss of 500ml. However there was no extra/intradural extention of the tumour noted. The nerve of origin could not be made out due to the huge size of the tumour.

Regular follow up of the patient for the last 2 years has shown marked improvement of bowel symptoms and he has no recurrence\ sensory neurological deficits and the patient is disease free till date.

4. Conclusion

The duration of symptoms in a patient with giant presacral/pelvic schwannoma is very long and patient presents late when pressure symptoms predominate. Pain abdomen, urinary symptoms, constipation, may be the presenting features. CT /MRI is the preoperative imaging of choice in demonstrating tissue heterogeneity and anatomic location of tumour. Surgery is the treatment of choice for patients with presacral/pelvic schwannomas as it is associated with functional improvement.

Total excision is the standard surgical modality. However piecemeal or subtotal excision can be considered in benign tumours. Regular follow up is mandatory.

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