ISSN No:-2456-2165

A Case Report of Ancient Presacral Schwannoma

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Abstract:-

- ***Introduction:*** **Schwannomas are peripheral nerve sheath tumors arising from Schwann cells. They are usually found in head and neck, mediastinum and extremities but rarely found in pelvis. Pelvic schwannomas have a slow growth rate, remain asymptomatic and get incidentally detected.**

- ***Materials and Methods:*** **A 54 year old male presented with right loin pain and burning micturation since 2 days. Ultrasonography showed right mid ureteric calculus with a thick walled cystic swelling in presacral region. Digital rectal examination revealed a firm, fixed posterior extra rectal mass. The upper limit of the mass could not be reached. Patient was evaluated and on emergency basis Ureterorenoscopy with right sided DJ stenting was done. CE-MRI of pelvis showed a large well defined T1 hypointense, T2 iso-hyperintense solid cystic lesion in presacral region. Mass effect over rectum and urinary bladder anteriorly. Posteriorly, abutting the sacrum and in close relation with right S3 exiting nerve root – F/S/O Schwannoma. Intraoperatively, cystic swelling noted in presacral region measuring 12×10cm. Entire tumor was excised and its origin from nerve root identified and excised without injuring it.**

- ***Results:*** **Histopathological report revealed benign nerve sheath tumor composed of Schwann cells with focal Verocay body formation. Immunohistology confirmed Schwannoma with diffuse positivity of S-100 protein in the cytoplasm of tumor cells.**

- ***Conclusion:*** **Schwannomas with degenerative changes like cyst formation, calcification and hemorrhage are called as 'Ancient Schwannomas'. MRI is the most preferred imaging technique. Complete surgical excision remains the treatment of choice. Recurrence rate is usually low with good prognosis.**

Keywords: *Schwannoma, Nerve Sheath, Schwann Cells.*

I. INTRODUCTION

Schwannomas are peripheral nerve sheath tumours which arise from Schwann cells of nerve fibres. They are usually found in regions of head and neck, mediastinum[1] and extremities but rarely found in pelvis 1-5%[2]. Since pelvic schwannoma have a slow growth rate, they remain asymptomatic[3,4] and get incidentally detected during medical investigations. Because of its rarity here we present a 54 year old male case with presacral schwannoma.

II. CASE REPORT

A 54 year old male patient was admitted to our hospital with sudden onset of pain abdomen with burning micturation since 2 days who was initially treated at local hospital conservatively and was referred for further evaluation and management. On examination patient had tenderness in right lumbar and iliac region. Digital rectal examination revealed a firm, fixed posterior extra rectal mass. The upper limit of the mass could not be reached.

Ultrasonography showed right moderate hydroureteronephrosis due to mid ureteric calculus with a cystic swelling in presacral region. Later patient was subjected to NCCT-KUB which showed a calculus measuring 9.2x6.9x12.8mm (APxTRxCC) (1267HU) noted in right mid ureter at the level of superior endplate of L5 vertebra causing moderate upstream dilatation of pelvicalyceal system and ureter with perinephric and periureteral fat stranding. A well defined large hypodense thick walled cystic lesion measuring 11.4x9.7x13.0cm (APxTRxCC) with few tiny calcifications within noted in presacral region.

ISSN No:-2456-2165

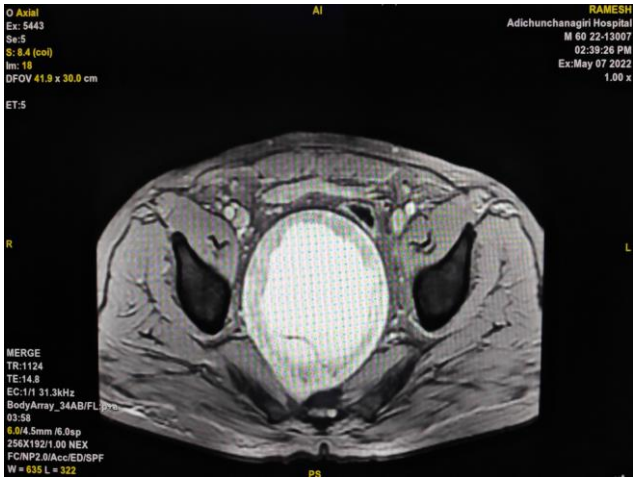

Fig 1 CE-MRI Pelvis Showing Tumor (Axial view).

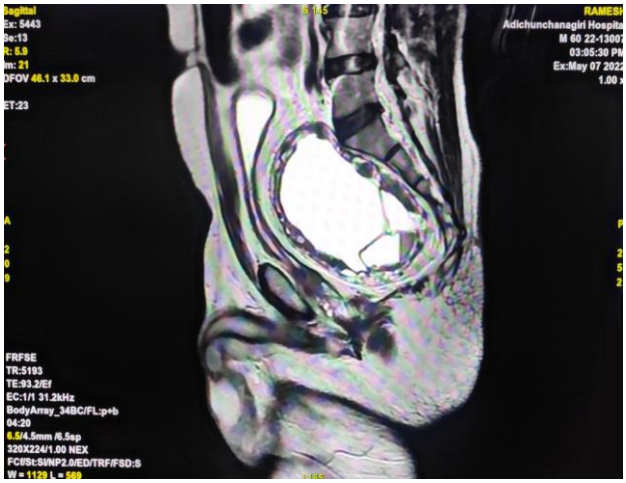

Fig 2 CE-MRI Pelvis Showing Tumor (Sagittal view).

The mass was anteriorly displacing the rectum with maintained fat plane. Posteriorly abutting the sacrum with no erosions/ destructions – likely benign etiology.

After evaluation on emergency basis patient was taken up for Ureterorenoscopy and right sided DJ stenting was done. Later he was subjected to CE-MRI OF PELVIS which showed a large well defined T1 hypointense, T2 iso-hyperintense solid cystic lesion noted in presacral region. Few internal fluid-fluid level areas with corresponding areas of blooming on GRE sequence – S/O Hemorrhage. Multiple tiny GRE blooming foci long the walls – S/O calcifications. Extensions – Anteriorly, significant mass effects over rectum and urinary bladder in the form of compression and displacement. Posteriorly, abutting the sacrum with mild extension into the right S3 neural foramina in close relation with right S3 exiting nerve root and causing mild expansion of neural foramina with no erosions/ destructions. Laterally, abutting the internal iliac vessels with maintained fat plains. Superiorly, extending upto lower border of L5 vertebra. Inferiorly, abutting the pelvic floor muscles – F/S/O Presacral peripheral nerve sheath tumor with cystic degeneration and hemorrhage – likely Schwannoma.

Patient was planned for elective laparotomy and excision of presacral tumor. Thick walled cystic swelling noted in presacral region measuring approximately 12×10cm abutting the rectum anteriorly, abutting the sacrum posteriorly. Blunt dissection done and tumor separated from surrounding structures. Fluid aspirated from cyst and sent for fluid cell cytology. Due to dense adhesions in presacral region, tumor was removed in piece meal with proper care taken to prevent spillage. Entire tumor was removed and its origin from nerve root identified and excised without injuring it. In view of bleed from presacral plexus, packing was done. Pack removed after 48 hours with achievance of hemostasis. Post operative period was uneventful. Pelvic drain was removed on post operative day 5 and discharged on post operative day 6.

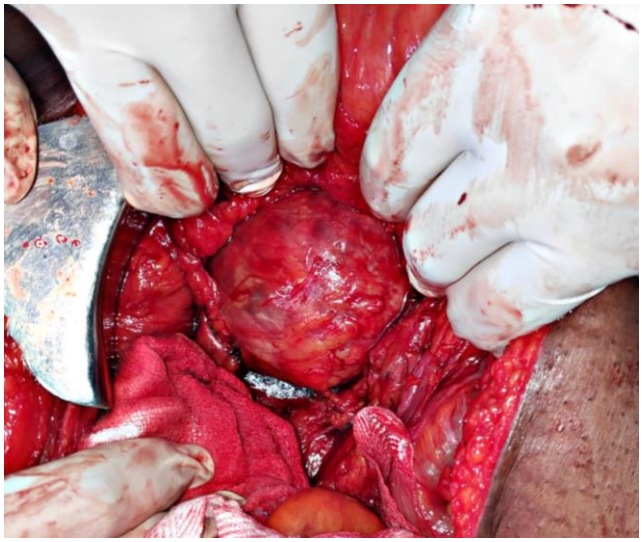
Fig 3 Intraoperative Tumor Image.


Fig 4 Specimen Removed in Piece Meal.

ISSN No:-2456-2165

III. RESULTS

Histopathological report confirmed Schwannoma with sections showing fragments of cystic benign nerve sheath tumor composed of extensive areas of collagenization, hemorrhage and fibrin deposition with aggregates of Schwann cells showing degenerative nuclear atypia and focal Verocay body formation. Immunohistochemistry showed S100 positive. Later patient underwent Ureterorenoscopic lithotripsy for right mid ureteric calculus after 1 month.

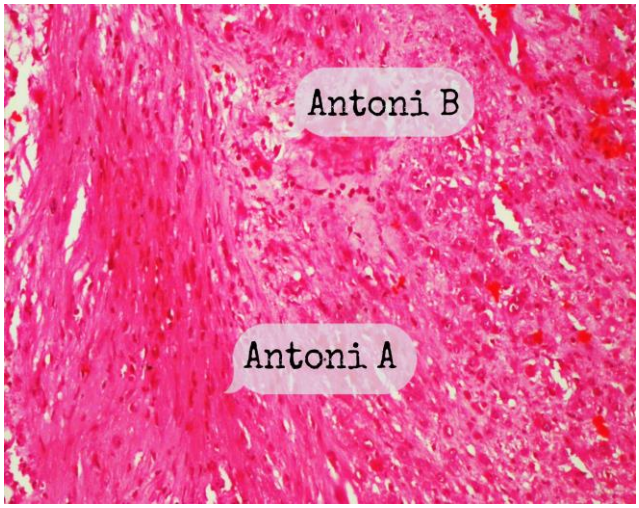

Fig 5 HPE Image Showing Palisading Spindle Shaped Cells.

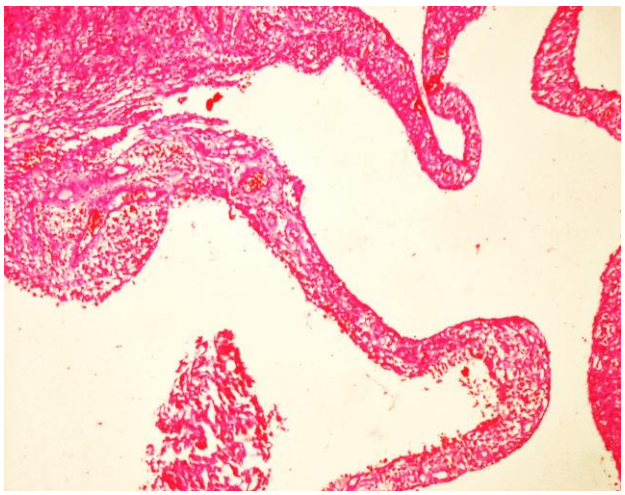
Fig 6 HPE Image Showing Cystic Spaces and Hemorrhage.

IV. DISCUSSION

Schwannoma also called as neurilemmoma or neurinoma are peripheral nerve sheath tumor which are rarely seen in the presacral region[5]. Nerve sheath tumors are classified into 3 types depending on their localization and spread. Type 1 is tumor localized to sacrum. Type 2 is the one which protrudes to adjacent cavities by invading the anterior or posterior sacral wall. Type 3 is tumor localized to presacral region.

Histologically they are classified into two types. Antoni A type, which have highly ordered cellular areas consisting of spindle shaped cells with nuclear palisading called verocay bodies. Antoni B type, which have loose myxoid areas[6].

Immunohistologically, diffuse positivity of S-100 protein in the cytoplasm of tumor cells is necessary for definitive diagnosis[8]. Schwannomas with degenerative changes including cyst formation, calcification and hemorrhage are called as 'Ancient Schwannomas'.

MRI is the most preferred and precise imaging technique to know the extent and relation to surrounding structures. Complete surgical excision, either open or laparoscopic is mandatory for treatment of schwannoma. Recurrence rate is usually low with good prognosis. Choice of open surgical approach depends on type of tumor[9,10], ie; transabdominal or posterior approach.

V. CONCLUSION

Schwannomas with degenerative changes like cyst formation, calcification and hemorrhage are called as 'Ancient Schwannomas'. MRI is the most preferred imaging technique. Complete surgical excision remains the treatment of choice. Recurrence rate is usually low with good prognosis.

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