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RESEARCH ARTICLE

Presacral nerve sheath tumor – An interesting case report

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Abstract

A new case of presacral nerve sheath tumor was reported. An extensive electronic search of the relevant literature since 1990 was carried out using medline. Present study retained only the articles reporting one or several cases. When the article was unavailable, we considered the relevant abstracts which should report clinical patterns and therapeutic modalities. Reviews of the literature, systemic reviews, letters to editors and incomplete abstracts were excluded. A descriptive analysis of collected sample including our case was performed. Morphological, therapeutic and outcome variables were reported.

INTRODUCTION

Schwannomas are benign encapsulated tumors of Schwann cells that grow slowly along the peripheral myelin nerve fibres. Sacral spinal schwannomas are very rare and the incidence of sacral schwannomas range from 1-5% of all spinal schwannomas[1] . There are three defined types of sacral schwannomas. These are retroperitoneal or presacral, intraosseous and spinal schwannomas.Benign presacral nervesheath tumors represent up to 10% of all presacral tumors.[2] Patient commonly complain of pain and paresthesias due to spinal schwannomas extending to extraspinal tissues.

Case Report

A 42 yr male admitted with complaint of vague lower abdominal pain, chronic constipation, with radiating pain in both lower limbs. Patient had a history of partial excision of the some abdominal mass 20 yrs back. CT scan of the abdomen showed evidence of a well defined homogenous enhancing soft tissue mass in the pelvis of size 10.5 cm in the presacral region on the right side displacing rectum to the left side and prostate and bladder anteriorly with moderate hydronephrosis of right kidney due to obstruction on lower end of ureter.USG guided FNAC from the presacral mass showed spindle shaped tumor cells with mild pleomorphic hyperchromatic nuclei. Patient was operated by anterior abdominal approach and the mass was removed completely preserving the sacral nerve root fibres. Mass was around 10 cm in size and cut section was predominantly solid, firm, yellowish grey with focal translucent and hemorrhagic areas, cystic and friable areas seen. Microscopy showed encapsulated spindle cell tumors with areas of variable cellularity, hyalinization and cystic change with foci of calcification. Tumor cells expressed S-100 protein and desmin and immune negative for CD-34, SMA and myogenin. Impression was benign peripheral nerve sheath tumor. Postoperatively patient recovered well with no neurological deficit. Patient had no recurrence after two years of followup. **[Figure- 1, 2, 3]**

Discussion-

Present study collected many related studies in peer reviewed journal in which diagnosis of presacral nerve sheath tumor or schwannoma was made. The principal cells comprising of peripheral nerve sheath tumors are Schwann cells (endoneurium ectoderm cells) with perineural cells and the fibroblasts.[3] Most common symptom reported was radicular pain in bilateral lowerlimbs followed by neurological dysfunction, chronic pelvic pain[4],lower abdominal mass, backache, lower abdominal pain on bowel movement[5], dysuria in decreasing order. Intrapelvic tumors are often diagnosed at a later stage. Neuroimaging is very helpful to delineate this unusual site and the extent of the tumor development and to determine the appropriate surgical intervention. Most common modality used for imaging is MRI. Ultrasonographic guided needle and aspiration biopsy is usually not recommended because this provides an inadequate amount of tissue containing highly pleomorphic cells which is difficult to interpret and may associated with complications such as infection or hemorrhage from hypervascular tumor. Presacral and retroperitoneal schwannomas are extremely unusual and their surgical approach is challenging. These tumors can be approached by either anterior abdominal approach or inferior presacral approach. Most common surgical approach chosen was anterior abdominal approach . A clear understanding of retroperitoneal anatomy is essential for safe removal of such tumors. Complete resection is preferred to prevent local recurrence and malignant transformation. Successful laparoscopic resection for a high level retroperitoneal solitary tumor have been reported. Combined and posterior approach is the most appropriate for larger intra pelvic tumors. The S1 to S3 nerve roots should be preserved to retain the function of bowel and bladder. As previous reported studies, there were 35 females and 65males with mean age of 46 yrs. Average size of tumor reported was 11cm (range 3-25cm). Surgical approach chosen was anterior approach in 37 cases, posterior approach in 33 cases, combined approach in 30 cases. Laparoscopic removal done in 3 cases [6, 7, 8]. In one case pelvic exenteration was done. In one case sacral bone window approach through the base of sacrum by resection of L5 transverse process was taken [9]. In one case angiographic embolisation [10] followed by Low anterior resection of rectum with colostomy was done. Histopathology in most of the cases was Schwannoma .In three cases malignant peripheral nerve sheath tumor was reported. In one cases triton tumor[11] was reported which is malignant peripheral nerve sheath tumor with rhabdomioblastic differentiation. In three cases ancient schwannoma [12] (schwannoma with degenerative changes)[13] , in two cases cystic schwannoma[14] and nine cases neurofibroma was reported. One case of sacral schwannoma mistakenly treated as irritable bowel syndrome before diagnosis of presacral schwannoma was made on imaging.[5] In four cases, tumor was incidentally detected by imaging for other reasons.

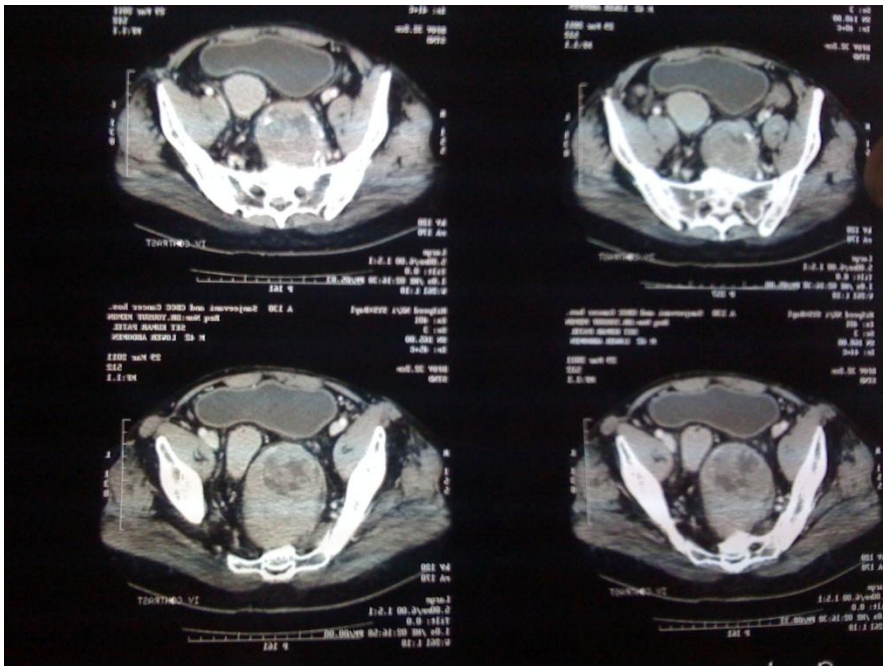

Fig 1 – Axial view of lower abdomen showing presacral mass

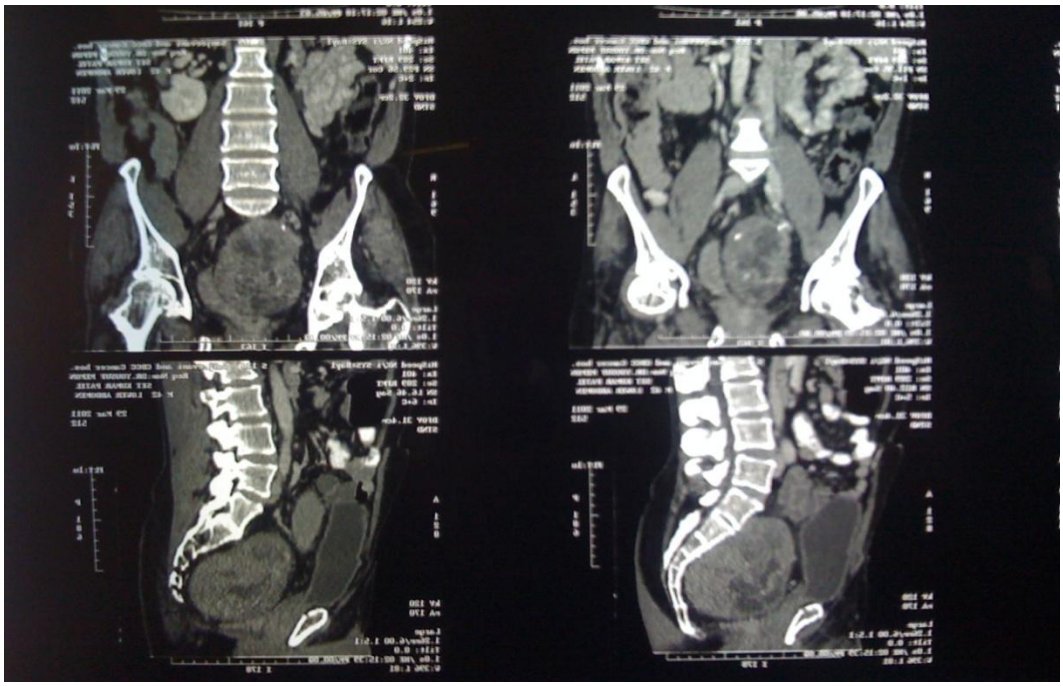

Fig 2 – CT showing sagital view of lower abdomen

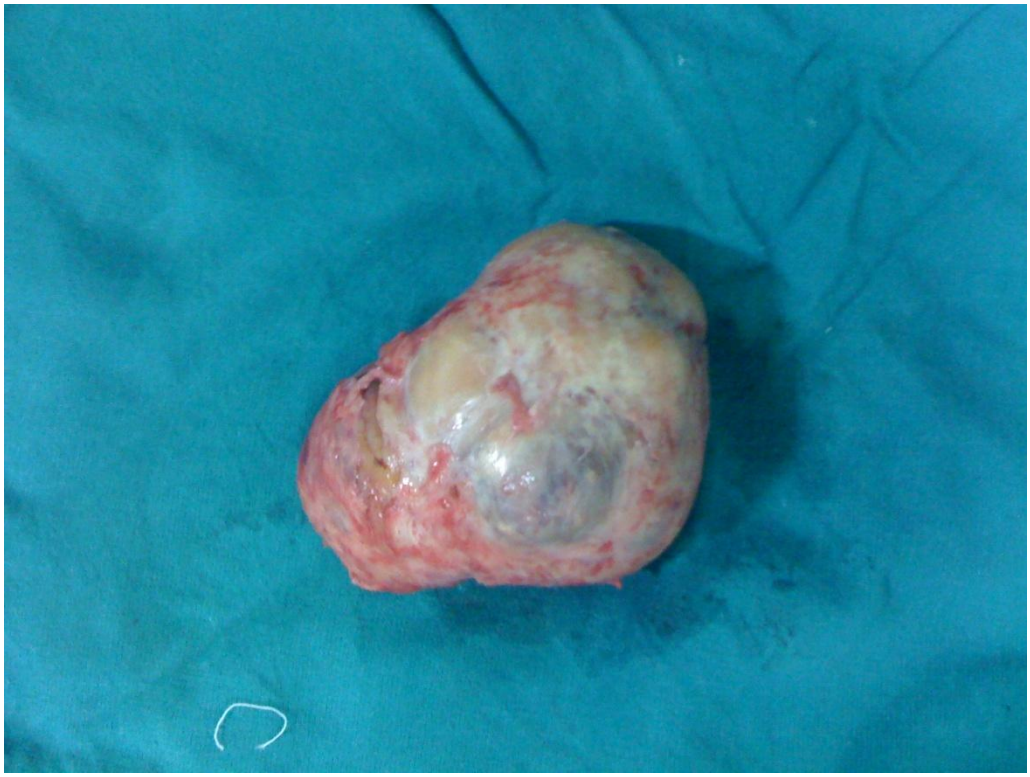

Fig 3 – Showing excised presacral mass

Conclusion

Presacral and retroperitoneal schwannomas are extremely unusual and their surgical approach is challenging. A clear understanding of retroperitoneal anatomy is essential for safe removal of such tumors. Complete resection is

preferred to prevent local recurrence and malignant transformation. Finding of the present study will be useful for surgeons to choose appropriate modalities of diagnosis and management of these types of cases.

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