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Case Report

When anatomy mimics: broad ligament mimicry of neurofibroma

Niteen Ghorpade*, Tushar Rahane, Bindu Choudhary

Department of Obstetrics and Gynaecology, Inspiria Laparoscopy and IVF Research Centre, Rahata, Maharashtra, India

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***Correspondence:**
Dr. Niteen Ghorpade,
E-mail: niteenghorpade9@gmail.com

ABSTRACT

Schawannoma aka neurilemmoma are encapsulated tumour which arise from the nerve sheaths. Pre sacral tumours are rare tumors which arise from the pre sacral space. It can be diagnosed with modality like magnetic resonance imaging (MRI). Sometime ultrasonography findings for broad ligament fibroid can mimic with neurofibroma. Patient with complaint of pain in abdomen should be evaluated. Surgical removal is treatment of choice for that.

Keywords: Schawannoma, Presacral tumours, Broad ligament fibroid, Surgery, Pelviology

INTRODUCTION

Schawannoma aka neurilemmoma are encapsulated tumour which arise from the nerve sheaths. These are benign peripheral nerve sheath tumours with presence of neuronal components consisting of Schwann cells and fibroblast. The most common site is eight nerves where it presents as intra cranial acoustic neuroma, cases of schwannoma arising from other locations have also been reported. The overall incidence of neurogenic tumor in presacral region accounts for 10 % and this lesion can be misdiagnosed as an adnexal or uterine mass on imaging.[1] We here in presenting a case report detailing pelvic mass identified as broad ligament fibroid which turn out to be pre sacral schwannoma during surgery.

CASE REPORT

A 32 years old nulligravida female presented with complaints of heaviness lower abdomen, with history of irregular cycles for last few months and difficulty in urination in last 15-20 days. She had no other pressure symptoms, systemic presentation or neurological deficit.

On examination, patient vitally stable; BP-110/80 mmHg; p-76/min; patient had normal BMI. Abdomen and speculum examination was unremarkable. On bimanual examination-uterus normal size, anteverted, freely mobile, pod fullness felt with a separate firm regular mass of about 10×8 cm on post surface of uterus. Pelvic ultrasound confirmed the findings and suggested broad ligament fibroid of 8×8 cm involving right posterior wall. All haematological and biochemical investigations were normal.

Intraoperative, uterus with adnexa was normal, a separate mass identified in presacral region measuring approximately 8×8 cm, relatively fixed involving S1, S2 and S3 level with bilateral displacement of ureters. Bilateral retroperitoneal dissection done, ureters and bilateral iliac vessels exposed and encapsulated mass removed in total. In bag morcellation done and sample retrieved. The post operative recovery was smooth and patient was discharged on day 2. On follow up no neurological deficit seen.

On histopathology, it showed tumor composed of cells arranged ix interlacing fascicles and bundles. The cells are spindle shaped having elongated, wavy nuclei and abundant eosinophil cytoplasm. At places myxoid areas are noted. Intervening stroma shows mononuclear

inflammatory infiltrate comprising of lymphocytes and histiocytes and no evidence of malignancy seen.

Immunohistochemistry confirmed schawannoma with positive S-100 and SOX 10 and negative CD 34, desmin.

Figure 1 shows a tumor composed of cells arranged in interlacing fascicles and bundles. The cells are spindle-shaped having elongated, wavy nuclei and abundant eosinophilic cytoplasm. At places myxoid areas are noted. Intervening stroma shows mononuclear inflammatory infiltrate comprising of lymphocytes and histiocytes. There is no evidence of malignancy.

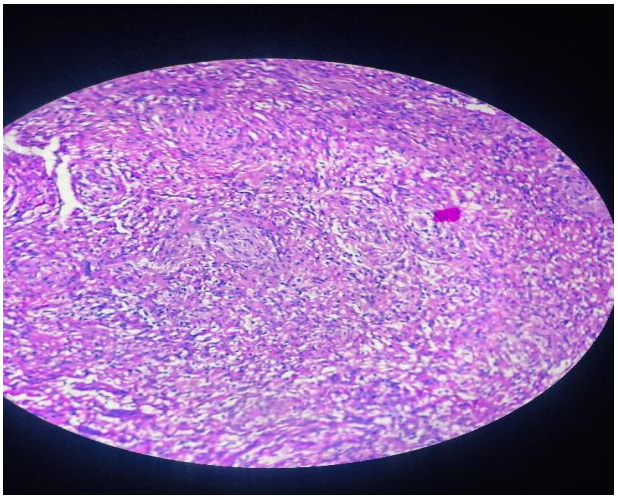

Figure 1: Histopathological slide-Shwanomma/neurofibroma.

DISCUSSION

Pre sacral tumors are rare tumors which arise from the pre scarla space. Pre sacral space is a narrow space which extends from posterior mesorectum upto the surface of sacrum covered by presacral peritoneum of Waldeyer. The upper margin is the reflection of peritoneum and the lower end is supra levator space. Laterally it is limited by iliac vessels along with its branches and ureters.[2]

Congenital tumors are the most common re sacral tomors comprises of 55-70 % of all the cases followed by neurogenic tumors i.e., 10%.[3]

Most of the neurogenic tumors are benign e.g, neurobifroma, neuprilemoma (schwannoma) and glioneuroma. Malignant neurogenic tumours (peripheral nerve sheath tumours) are reported as neuroblastoma, neurofibrosarcoma, malignant schannomas, ganglioneuroblastoma, ependymoma and neurogenic sarcomas. Out of these the most common neurogenic tumour is neurilemoma.

Schwannoma are benign encapsulated peripheral nerve sheath tumors which are usually asymptomatic for a long time or produce nonspecific symptoms such as lower abdominal pain, diarrhoaea, constipation. Due to their resemblance with leiomyoma, they can be mis diagnosed as fibroid uterus.

According to Klimo et al these shwanomma has classified into three types- type 1 – tumors confined to sacrum, type 2 tumors originate within the sacrum but erode the wall of the sacrum and extend into the adjacent spaces and type 3 tumors are located predominantly in the presacral or retro peritoneal area.[4]

Neuroimaging is very helpful in delineating these unusual sites of tumors and their extent. Magnetic resonance imaging (MRI) being more specific than CT provide more detail about the relevant anatomical involvement.

Schwanommas are heterogenous at MR imaging with low signal intensity on T1 weighted images and high signal intensity on T2 weighted images along with cystic areas and a pseudo-capsule.

CONCLUSION

Surgical removal is the treatment of choice for pre sacral tumors even if they are asymptomatic. A clear anatomy of retroperiteal space is essential for surgical exploration. Anterior(abdominal) approach has been used for type 1, posterior(sacral) approached for type 3 and combined approaches have been described for surgical removal of type 2 tumors. Anterior approach can be transperitoneal or extraperitoneal. Few cases of laparoscopic removal of schwannoma have also been reported recently. Laparoscopic magnification and expertise help in removal of these tumors with excellent haemostasis.

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