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Case Report

Retroperitoneal pelvic schwannoma in pregnancy: a case report

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ABSTRACT

Solitary nerve sheath tumor such as Benign schwannomas arising in the pelvic retro peritoneum is infrequently reported. Retroperitoneal location accounts for 0.3-3.2% of primary schwannomas. We report a case of benign retroperitoneal pelvic schwannoma in pregnancy that was incidentally diagnosed when it presented with Preterm premature rupture of membranes and mechanical obstruction for labour. She underwent caesarean section and delivered a healthy baby. She was evaluated in the postoperative period by computerized tomography (CT) imaging studies and CT guided fine needle aspiration cytology (FNAC) was not diagnostic. Complete surgical excision of the tumor was achieved in the postpartum period. The adjacent vascular and urinary channels sustained no injuries and she had no neurologic deficit. Histology revealed spindle cell neoplasm composed of interlacing fascicles and sheets of spindle cell with focal areas of nuclear palisading and thick walled blood vessels. Immunohistochemistry was positive for S 100 suggesting schwannoma. Retroperitoneal location of schwannomas is rare and surgery is curative. Prognosis is good, since recurrence is rare.

Keywords: Broad ligament myoma, Obstructed labor, Retroperitoneal schwannomas

INTRODUCTION

Schwannomas are rare, benign tumor derived from the neurilemma or nerve sheath. It predominantly occurs in females between second and fifth decades of life.¹ It is seen commonly in the soft tissues of head, neck, mediastinum, retroperitoneum and extremities. Retroperitoneal location accounts for 0.3-3.2% of primary schwannomas.² It is asymptomatic till it reaches a size capable of causing compression of intra abdominal or intra pelvic organs. This makes the preoperative diagnosis hard to ascertain and diagnosis is usually confirmed by histopathological examination.

We report a case of benign retroperitoneal pelvic schwannoma that was incidentally diagnosed in late

pregnancy when it presented with mechanical obstruction to labour.

CASE REPORT

25 years old primigravida was referred to our hospital at 34 weeks with preterm prelabor rupture of membranes (PPROM) with a broad ligament fibroid. Her antenatal period was unremarkable till date. On examination, she was healthy looking with no abnormal systemic findings. Abdominal examination revealed a 34 weeks viable fetus. Pelvic examination revealed firm mass protruding through pouch of douglas into vagina fully occupying its upper half and completely obscuring cervix. Ultrasound report revealed single live fetus with broad ligament fibroid 9 x 8 cms occupying pouch of douglas.

In view of mechanical obstruction to labour, caesarean section was done under regional anaesthesia. She delivered preterm boy baby of 2.26 kg with good APGAR score. A large, firm, fixed, highly vascular tumor was found in pelvic retroperitoneum. In view of its high vascularity, biopsy was deferred intraoperatively.

She was evaluated in the postoperative period by CT imaging with contrast which revealed a large heterogeneously enhancing solid mass of size 99 x 108 x 102 mm in pouch of Douglas displacing cervix and uterus anteriorly and sigmoid colon to right, compressing both common iliac vessels. CT guided FNAC was reported as myo fibroblastic tumor (Figure 1).

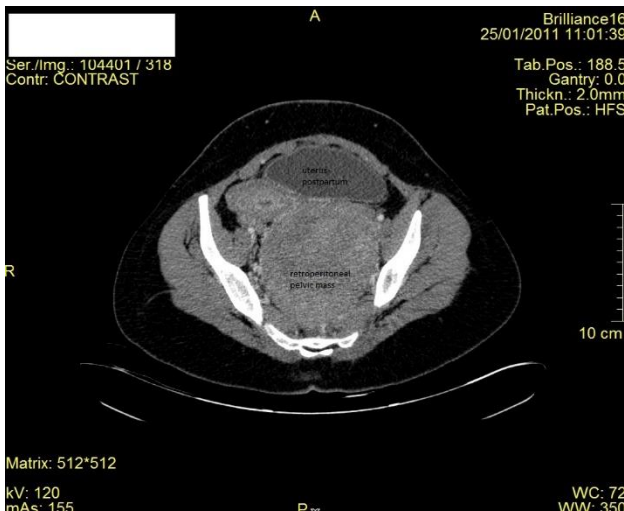


Figure 1: CT Abdomen and Pelvis – intravenous contrast showing large heterogeneously enhancing solid mass of size 99 x 108 x 102 mm in pouch of Douglas displacing cervix and uterus anteriorly and sigmoid colon to right, with vessels along the capsule of the mass and compressing both common iliac vessels.

Exploratory laparotomy was performed after four weeks. A large well encapsulated, deep seated tumor was found retroperitoneally in the presacral region. It was enucleated and the tumor was removed in toto after ligating the feeding vessels and pericapsular venous plexus. She received two units packed red blood cells intraoperatively. Postoperative period was uneventful.

Microscopically, it was a spindle cell neoplasm composed of interlacing fascicles and sheets of spindle cells with focal areas of nuclear palisading and thick walled blood vessels. Most of the tumour was infiltrated by lymphocytes and plasma cells. There was no obvious nuclear pleomorphism or abnormal mitotic activity.

Mitotic index was < 2 / High power field (HPF). Immuno Histo Chemistry was strongly positive for S 100 and negative for Smooth muscle actin (SMA) and CD 117

that confirms the nerve sheath origin (Figure 2 and Figure 3).

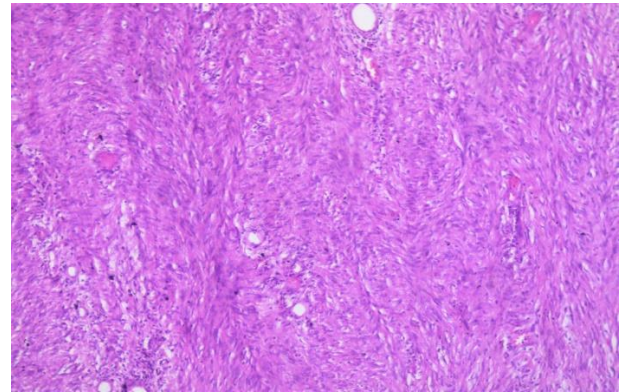


Figure 2: Histology showing spindle cell neoplasm composed of interlacing fascicles and sheets of spindle cells with focal areas of nuclear palisading and thick walled blood vessels.

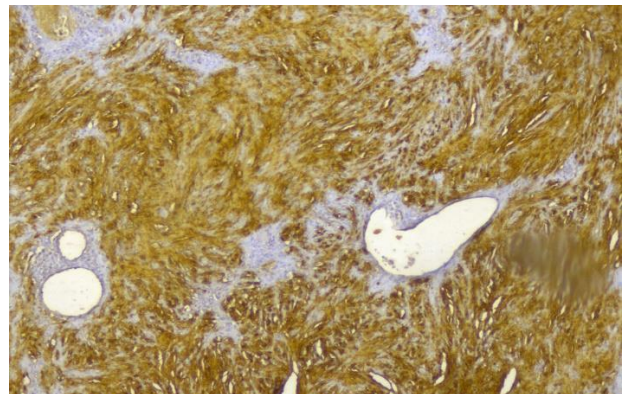


Figure 3: Immunohistochemical study showing positivity for S100.

After discussion in the tumour board, patient was reassured about the benign nature of the tumour and was advised follow up. She is asymptomatic for past six years with no evidence of recurrence.

DISCUSSION

Schwannomas are solitary, slow growing, nonaggressive tumor.³ Pelvic schwannoma is rare and accounts for less than 1% of all benign schwannoma.⁴ Most are diagnosed incidentally or present with vague, nonspecific symptoms. Our case was diagnosed when she presented with PPRM and mechanical obstruction to labour. Retroperitoneum has a potential free space for the tumor to enlarge without producing any symptoms and these reach a large size before being discovered. They may present with pain in the lower back, bladder or bowel symptoms or heaviness in the abdomen.

Lack of typical imaging features in ultrasound, CT, MRI makes preoperative diagnosis difficult.⁵ Abdominal

ultrasound is a simple, noninvasive method in which they appear as solid, hypoechoic lesions. Areas of necrosis and cystic degeneration are noted in large tumors.⁶ CT and MRI are widely used as imaging modalities in the evaluation of retroperitoneal soft tissues tumors. Although target and fascicular signs are characteristic radiological features of schwannomas in CT, they are not frequently seen in retroperitoneal schwannomas.⁷ They appears as well demarcated, hypodense, homogeneous masses. Areas of heterogeneity are noted in case of tumor necrosis.

MRI is the imaging modality of choice for the soft tissue tumors. Schwannomas appears hypo intense on T₁ weighted images and hyper intense on T₂ weighted images. However, it is incapable of reliably distinguishing between benign and malignant tumor.⁸

Schwannomas can rarely be almost entirely cystic, the differential diagnosis includes retroperitoneal pseudo cyst, abscess or lymphocele. When schwannomas are mostly solid, differential diagnosis includes neurofibroma and lymphoma, ovarian tumour, degenerating fibroid. Histological analysis provides the definitive diagnosis for schwannomas. Immuno Histo Chemistry shows positivity for S100.⁹

Wide surgical excision is the treatment of choice though its complex.¹⁰ Surgery can be done by laparotomy or laparoscopically or robotic assisted resection.^{11,12} Care must be taken in attempting removal since hyper vascularity can complicate the excision. Sufficient amount of blood products should be readily available. Even death has been reported due to uncontrollable haemorrhage in literature. Majority of the problems encountered with tumor resection are due to the close proximity of the tumor to major blood vessels rather than due to its own vascularity. Prognosis is good since postsurgical recurrences are rare if adequate clearance is performed during the primary surgery.¹³

CONCLUSION

Retroperitoneal location of schwannomas is rare and surgery is curative. Retroperitoneal schwannoma may masquerade as broad ligament fibroid or ovarian tumor in women. Role of conventional imaging is limited and high index of clinical suspicion is needed. Proper preoperative evaluation, multimodality treatment improves the outcome.

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Ethical approval: Not required

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