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**Case Report** 

# Pelvic schwannoma mimicking as an adnexal mass

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## **ABSTRACT**

Schwannoma (neurilemmomas), a peripheral nerve sheath tumour generally occurs on the head and neck region, their pelvic occurrence is much rare. Retroperitoneal schwannomas are the rarest of all retroperitoneal tumours. We report a 45 year woman with a schwannoma in the retro peritoneum that was preoperatively diagnosed as an adnexal mass.

**Keywords:** Pelvic schwannoma, Retroperitoneum, Cystectomy

### INTRODUCTION

Schwannomas are rare peripheral nerve tumours arising from Schwann cells of the nerve sheath and they are usually located in the head and neck. Their pelvic occurrence is much rarer, accounting for 1–3% of all schwannomas. Pelvic schwannomas arise mostly from the sacral nerve or the hypogastric plexus.<sup>1</sup>

Computed tomography (CT) and other imaging modalities may help obtain information about tumour size, location and relationships with neighbouring tissues, but they do not give a definitive diagnosis. Because of nonspecific clinical and imaging findings, preoperative diagnosis may be difficult leading to misdiagnosis and prolonged morbidity.<sup>2</sup> The prognosis is excellent, excision being curative in nearly every instance.<sup>2</sup> A successful resection of a presacral schwannoma near the left internal iliac vessels and ureter was reported.

#### CASE REPORT

A 45 year old woman presented with vague abdominal pain since 6 months. There was no history of bowel or bladder disturbances. On examination, she was stable. A cystic mass of around 10x10 cm felt in the left iliac and hypogastric region with restricted mobility. Per vaginal

examination revealed that the cervix was deviated to the right, hitched up behind pubic symphysis; size of the uterus could not be made out due to the mass. A hard fixed mass was felt in the posterior and left fornix.

Tumour markers were all within normal limits. Ca 125: 8.3U/ml, Ca 19-9: 18.4U/ml, Alpha Feto Protein: 1.25ng/ml, LDH: 140 IU/L, Beta HCG < 0.1mIU/ml. Transvaginal ultrasound revealed a mixed echogenic mass in the left adnexa measuring 9.4x10 cm. CT of abdomen and pelvis revealed a well-defined cystic lesion with thin internal septae in the pelvis on the left side, displacing left internal iliac artery anteriorly (Figure 1).

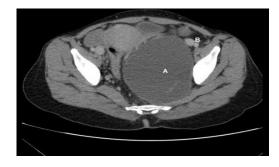


Figure 1: CT Pelvis showing (a) well defined cystic mass in the pelvis on left side, displacing the (b) left internal iliac artery anteriorly.

Patient underwent laparotomy. Intraoperatively, a huge retroperitoneal cyst of around 10x8 cm was found. Uterus, bilateral tubes and ovaries were normal. We proceeded with cystectomy. With careful dissection using sharp and blunt manoeuvres, the cyst was dissected from surrounding structures such as internal iliac vessels and ureter. As the base of the mass was reached, a sudden gush of bleeding occurred from the collateral vessel. Vascular clamps were applied to arrest bleeding ensuring that ureter was not included in the clamp. Vascular surgeons help was sought. Total blood loss was around 2.5 litres. The patient received total of 5 units of packed red blood cells, 2 units of fresh frozen plasma during and after surgery. She was managed postoperatively in intensive care unit (ICU) with ionotropic support. Patient recovered well postoperatively and was shifted out of ICU on second postoperative day.

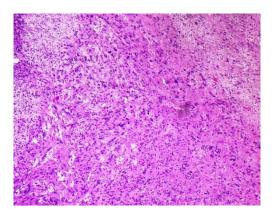


Figure 2: Tumour with hypo cellular and hypercellullar areas composed of spindle shaped cells with wavy nuclei H&Ex200.

Histopathology showed, the cyst wall composed of spindle shaped cells with Antoni A and B areas and few bizarre nuclei, suggestive of cystic ancient schwannoma (Figure 2 and 3).

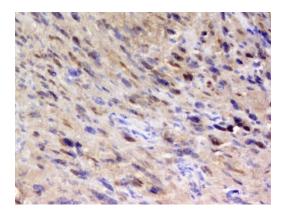


Figure 3: S-100 positivity in tumour cells IHCx400.

On 6th postoperative day patient complained of numbness of left lower limb and inability to dorsiflex her left foot. Nerve conduction study showed left common perineal nerve neuropathy, developing postoperative foot drop which was managed conservatively using short course of prednisolone and foot drop splint. Patient is now on regular post-operative follow up since 6 months and doing well.

#### DISCUSSION

Benign nerve sheath tumours are of two types, schwannomas (more common) and neurofibromas. Schwannomas are most frequently present in patients aged 20 to 50 years with male: female ratio of 2:3. They comprise 5% of all benign soft tissue tumours and have a predilection for the head and neck and flexor surfaces of the upper and lower extremities. Deeply seated schwannomas predominate in the posterior mediastinum and retro peritoneum.<sup>3</sup> Schwannomas are usually solitary, slow growing, and nonaggressive neoplasms.

Pelvic schwannoma is rare and accounts for less than 1% of all benign schwannomas.<sup>4</sup> Although, origin along nerves of retroperitoneal space is not uncommon, these tumours rarely appear as pelvic masses. In our cases, it may have originated from peripheral fibres of sacral plexus.

Majority of these tumours are asymptomatic and are often found incidentally or present with vague, nonspecific symptoms. They are usually detected as an abdominal mass on routine examination as in this case. The predominantly cystic nature of the tumour was demonstrated by ultrasound and CT scan examination but the exact nature of the tumour could not be diagnosed.

Retroperitoneal schwannomas are usually larger and have a high tendency of undergoing spontaneous degeneration and haemorrhage. Because the retro peritoneum is flexible and non-restrictive, these tumours frequently attain a large size before discovery. Because of their slow growth and anatomic location, pelvic schwannomas remain asymptomatic until they reach large size and cause mass effect. This can lead to pain in lower back and pelvis, and heaviness with urinary and digestive symptoms caused by bladder and bowel compression. Sciatic nerve impingement and pain caused by compression has also been reported.<sup>5</sup>

There is no gold standard, non-invasive diagnostic method for schwannomas. Ultrasound, CT and MRI are nonspecific in most cases. Li et al. reported that only 15.9% were identified preoperatively by these imaging modalities in a review of 82 retroperitoneal schwannomas. Because of their larger size, these tumours are likely to manifest secondary degenerative changes. Complete surgical excision is the treatment of choice for schwannomas.

Some authors have reported massive blood loss intraoperatively as a result of proximity of tumour to a pelvic venous plexus as was evident in our case also.

Once suspected, it is better to reserve adequate blood for transfusion in case of emergency.

Once a definitive diagnosis of schwannoma is made, after complete surgical excision, follow-up with pelvic sonography is adequate for further management. Our patient is symptomatically relieved after complete excision and is on regular follow-up.

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