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Case Note

Ancient cystic pelvic schwannoma presenting as a right iliac fossa mass

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Abstract
Schwannoma is rare in pelvis. Ancient schwannoma is rarer histological subtype of schwannoma. We report a very rare case of pelvic schwannoma presenting with right sciatica and right iliac fossa pain. Clinical and imaging findings were suggestive of Peritoneal Hydatid cyst. The tumour was resected completely with marked clinical improvement. Histopathological examination showed Ancient cystic schwannoma.

Key words: Benign pelvic Schwannoma, Benign retroperitoneal tumour, Giant ancient Schwannoma, Presacral Cystic Schwannoma

Benign Schwannomas are generally slow growing and painless tumours originating from Schwann cells of peripheral nerve sheaths and predominantly occurs in females. Due to its non-specific clinical and imaging findings, preoperative diagnosis is very difficult. Needle biopsy is diagnostic and is able to differentiate between benign and malignant schwannoma. As they are encapsulated and non invasive tumours, local excision is considered the treatment of choice. Once completely excised, recurrence of benign schwannoma is not expected. Adjuvant therapy is therefore not necessary.

Case history
A 53 year old man presented with history of tingling and numbness of right sole for 4 years and pain in the right lower abdomen for 6 months. Clinical examination revealed a firm, non mobile, 10x10 cm, painless lump in right iliac fossa. The upper pole was reachable but lower pole could not be reached. On compressing the lump patient complained of paresthesia on right sole and lateral aspect of right leg (Tinel sign).USG and CT findings were suggestive of peritoneal hydatid cyst (fig 1).

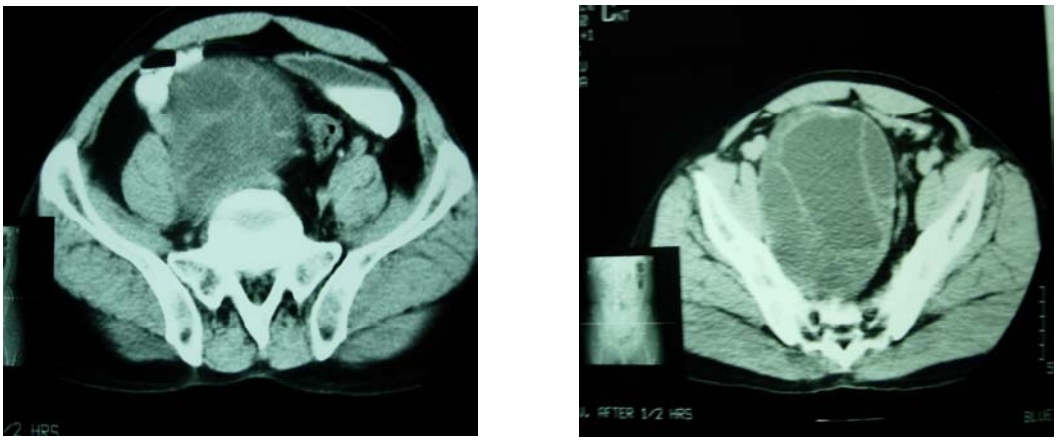

Fig.1 Large multiloculated cystic lesion (12.4 X 12 X 18 cm) in right lower abdomen and pelvic cavity indenting bladder to some extent.

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With a diagnosis of a peritoneal hydatid cyst, transabdominal/ transperitoneal approach was used to excise the mass. The mass was removed en-block by blunt dissection. Per operative findings revealed highly vascular mass compressing the right sacral plexus which could be the cause for right sciatica.

Cut section showed numerous cysts with hemorrhagic areas (fig 2). Histopathological examination revealed Ancient Cystic Schwannomna which showed Antoni A (fig 3a) and Antoni B (fig 3b) bodies with degenerative changes.

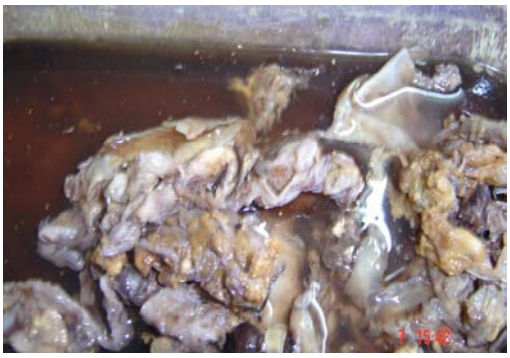

Fig.2: Cut section showing numerous cystic areas.

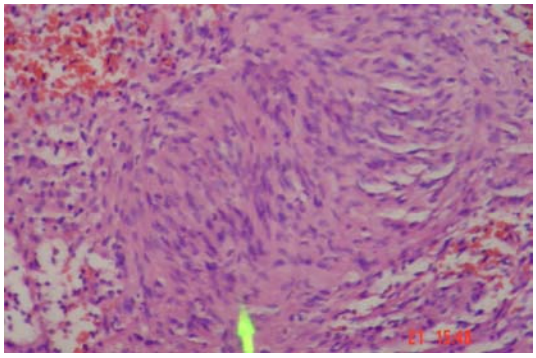

Fig.3a: Histology showing typical pallisading arrangement of spindle cells (Antoni A/Verocay body)

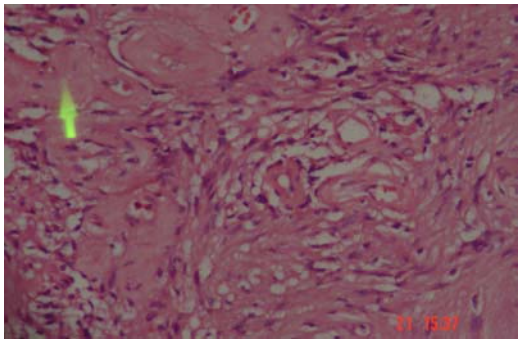

Fig.3b: Less cellular myxoid areas (Antoni-B)

Postoperative recovery was good except for mild hypoesthesia in S1–S2 dermatome and Grade IV motor weakness in plantar flexion of ankle and toes.

Discussion

Schwannoma (Neurilemmoma) is a benign neoplasm arising from the myelinated nerve sheaths. Malignant Schwannoma despite their name never arise from malignant degeneration of schwannomas. Instead they occur de novo or from transformation of a plexiform neurofibromatosis. This fact provides the basis for their association with Neurofibromatosis 2.[13]

Pelvic Schwannoma is rare and accounts for less than 1% of all benign Schwannoma. The Ancient variant is much rarer and so far only 2 cases have been reported in the literature[2].As these are slow growing and painless tumour, symptoms occur late unless they are large enough for pressure effect. This leads to discomfort and pain in the lower back, abdomen, and pelvis or heaviness with urinary and digestive symptoms from bladder and rectal compression. However, nerve impingement with sciatic pain down the lower limbs as noted in our case is less common[2]. Severe motor weakness is usually associated with malignant schwannoma.[3]

CT and MRI are widely used as imaging modalities in the evaluation of retroperitoneal soft tissue tumours. But CT fails to reproduce adequately the stroma heterogenicity, a main characteristics of

ancient cystic schwannoma (as haemorrhage, cyst formation, and calcification) as compared to MRI. Still MRI is not reliable for differentiating benign from malignant schwannoma[1]. However, central enhancement (due to highly packed cellular component-Antoni A surrounded by hypocellular area Antoni B) in CT and MRI, with target sign in MRI accounts for benign Schwannoma[3].

Definitive diagnosis is based on histological analysis of biopsy specimen which can further differentiate between benign and malignant disease. Histologically Schwannoma consists of compact cellular region with disposition in Verocay body-Antoni A or loose hypocellular with myxoid predominancy- Antoni B (Fig 3a and 3b). Ancient Schwannoma shows in addition features of degenerative modifications as cysts, calcification and haemorrhage.

Immunohistochemical staining can further aid in the diagnosis as benign schwannomas show diffuse immunoreactivity for S-100 protein (neural protein within Schwann cells that is lacking in neurofibromas)[1,3]. Further, Ki-67 indices were reported in 5-65% of malignant schwannomas; in contrast, it was less than 5% in benign schwannomas.

Since, tumour recurrence or malignant transformation almost never occurs in benign schwannomas, local tumour excision should be regarded as the treatment of choice[1,2,7]. Malignant transformation although extremely rare, is usually only observed in cases with underlying von-Recklinghausen's disease[6]. Hence, therefore it is important to survey the patient for stigmata of von- Recklinghausen's disease at first presentation[1,6]. Retrospectively, our patient had no any stigmata of von-Recklinghausen's disease. Should the postoperative histology confirm malignancy, local recurrence after marginal excision has to be expected in up to 72%, whereas recurrence after wide surgical margin resection has been reported only in 11.7%[1,5]. In such an unexpected event of proven malignancy one should probably consider re-resection if wide margin has not been achieved originally[1].

Care must be taken in attempting removal of retroperitoneal and intrapelvic schwannomas. Haemostasis can pose problem if the tumour capsule is adherent to the presacral venous plexus[1]. Sufficient amounts of blood products have to be readily available including fresh frozen plasma and platelets. The anaesthetist should be made aware that a high volume of blood loss might be encountered. Carpenter et al reported one intraoperative death related to uncontrollable haemorrhage from severing the right common iliac artery during a difficult dissection[5]. In a case report by Foote et al the attempt to excise a large retroperitoneal schwannoma was abandoned because of danger of uncontrollable hemorrhage[11]. In our case blood loss was about 800ml. If malignancy can safely be excluded, laparoscopic piecemeal excision should be considered as an alternative treatment as recurrence is unlikely.[1]

Conclusion

Neurological symptoms in the lower limbs (sciatica) and pain in the lower abdomen may be the only presenting features in pelvic schwannoma, causing delay in diagnosis and treatment. MRI is the imaging modality of choice in demonstrating tissue heterogenicity and anatomic location of the tumour. However, needle biopsy should be regarded as the diagnostic gold standard as none of the currently available imaging modality provides sufficient confirmation to exclude malignancy. Tumour excision in toto is considered the treatment of choice but can be hazardous if the tumour is adherent to the presacral venous plexus. Hence, piecemeal excision should be given consideration, provided enough evidence about the benign nature of the tumour is available. Defining nerve of origin may not be always possible and a minor degree of neurological deficit is therefore to be anticipated.[1]

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