EUROPEAN JOURNAL OF BIOMEDICAL AND PHARMACEUTICAL SCIENCES
http://www.ejbps.com

ISSN 2349-8870
Volume: 4
Issue: 12
769-773
Year: 2017

RETROPERITONEAL ANCIENT SCHWANNOMA: A RARE DIFFERENTIAL FOR BROAD LIGAMENT FIBROID

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Article Received on 01/10/2017 **Article Revised on 21/10/2017** **Article Accepted on 11/11/2017**

ABSTRACT

Introduction: Broad ligament fibroid in not an unusual diagnosis in gynecological practice, but retroperitoneal ancient schwannoma (with degenerative features such as cysts calcifications) is rarely considered in differential diagnosis. This case of retroperitoneal schwannoma is being presented here for its rarity (incidence is 1-10% of all retroperitoneal tumors) and diagnosis difficulty. Schwannomas originate from Schwann cells of the peripheral nerve fibers. Preoperative diagnosis of retroperitoneal schwannoma is difficult. Prognosis of retroperitoneal benign schwannoma is extremely good. **Case Presentation {Material & Methods, results}:** A 25 yr old lady with complain of lower abdominal pain for 1 yr presented to private hospital where she underwent laparotomy when surgery could not be completed due to technical difficulty and patient was referred to us. On clinical examination on admission, a pelvic mass hard in consistency, immobile, non tender was palpated, suspicious of malignant pathology. Imaging (USG, CT and MRI) suggested broad ligament fibroid or retroperitoneal tumour and FNAC/ Tru cut biopsy suggested leiomyoma. On exploratory laparotomy in our hospital, a large retroperitoneal mass 10X10 cm was resected with the help of surgeons. Surgical specimen sent for frozen section was reported to be benign spindle cell tumour and HPE was reported to be benign ancient schwannoma. Postoperative course was uneventful. **Conclusion:** Retroperitoneal schwannoma is a rare tumour which may mimic broad ligament fibroid with difficulty in preoperative diagnosis and final diagnosis can only be made after HPE and immunohistochemical examination.

KEYWORDS: Retroperitoneal schwannoma, broad ligament fibroid, Histopathology, immunohistochemistry.

INTRODUCTION

Broad ligament fibroid is not an unusual diagnosis in gynecological practice, but retroperitoneal ancient schwannoma is rarely considered in differential diagnosis. Schwannoma is a neurogenic tumor arising from the schwann cells of the nerve sheath, usually seen between the third and sixth decades of life, with an equal predilection for men and women.[1] When schwannomas show degenerative changes including cyst formation, calcification, etc, these are termed "ancient" schwannomas.[2] They usually occur in head, neck and flexor surfaces of extremities, retroperitoneal space being the rare location accounting for 0.3-3.2% of all primary schwannomas and 0.3-6.0% of all retroperitoneal tumors.[3] Schwannomas are usually benign, but infrequently undergo malignant transformation such as when associated with Von Recklinghausen disease.[1] Clinical features are nonspecific similar to that of broad ligament fibroid and depend on the site of location in the retroperitoneum. Preoperative diagnosis of retroperitoneal schwannoma is difficult due to its location, non specific symptoms and lack of typical imaging features. Its prognosis is favourable following complete excision of the tumour. Here we report a similar case of a 25-year-old female with a benign retroperitoneal schwannoma which was preoperatively diagnosed as broad ligament fibroid.

CASE PRESENTATION

A 25 yr old lady presented with complaint of lower abdominal pain for 1 year and progressively increasing lower abdominal mass for the last 3 months to a private hospital where she underwent laparotomy when surgery could not be completed due to lack of multidisciplinary

team and patient was referred to us. There were no other presenting complaints. She was regularly menstruating. Bowel and bladder functions were normal.

On clinical examination, a fixed pelvic mass of 16-18 weeks size, hard in consistency, non tender was palpated, suggestive of malignant pathology. On per vaginal examination, cervix was found to be deviated to left, movement of cervix was not transmitted to mass. Normal size uterus was made out deviated to right side. Left side adnexal mass was felt suggestive of broad ligament fibroid. Biochemical investigations revealed normal white cell count, normal level of tumour markers (CA-125= 25.9U/ml, CEA= 3.5ng/ml). Mantoux test was also negative (6mm).

USG showed 8.4X7.9X10.6 cm heterogenous echo texture lesion in left adnexa with cystic areas and internal vascularity within. Left ovary was not seen separately. A normal sized uterus with endometrial thickness 7 mm was seen, displaced to the right side by the mass. A normal sized right ovary was seen. MRI pelvis revealed a well defined large solid mass lesion of size 10.1X7.9X9.5 cm in left adnexa with cystic areas within showing signal pattern distribution (hypointense on T1 and heterogeneously intermediate to low signal intensity on T2 images) with significant post contrast enhancement. Anterosuperiorly left ovary and broad ligament were visualised displaced anteriorly by the mass. Posteriorly the mass was seen abutting the sacrum, however sacrum appeared normal in morphology. Right ovary was visualised and appeared normal in morphology and size. Fat planes between adjacent structures were preserved. Owing to the location and signal characters, the presumptive diagnosis was broad ligament fibroid or neurogenic tumour.

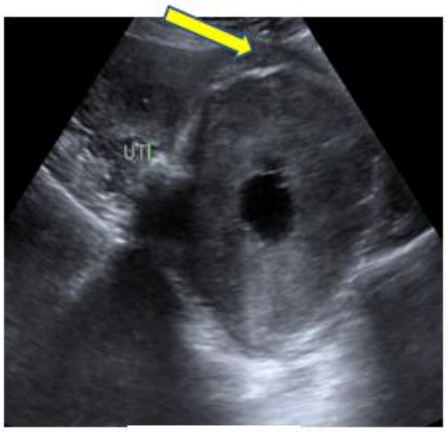


Fig 1

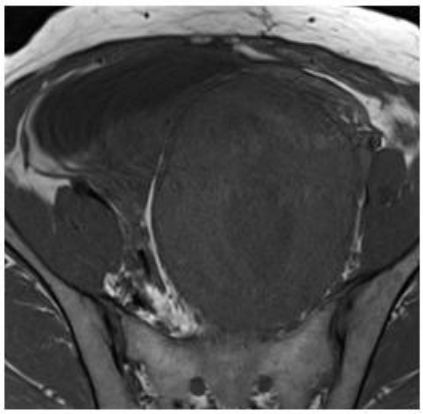
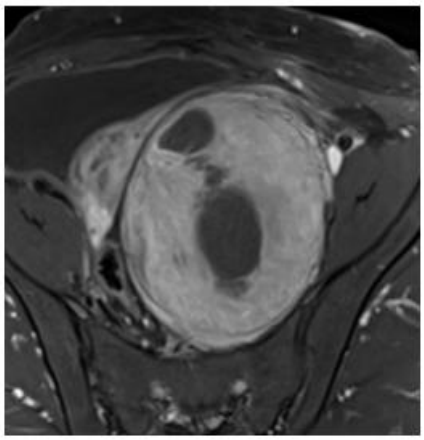


Fig 2



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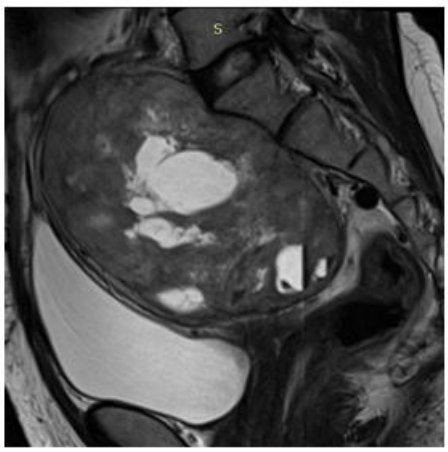

Fig 4

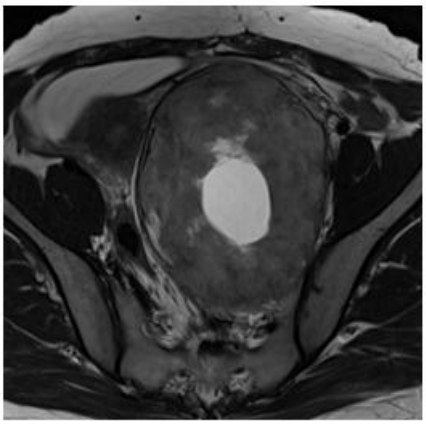

Fig 5

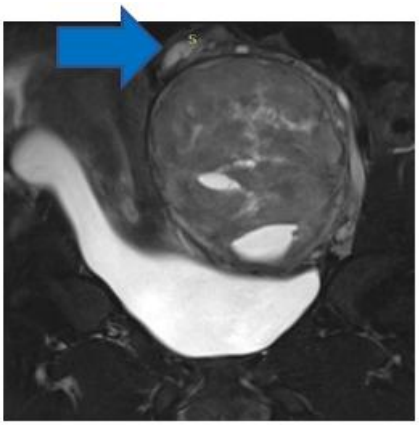

Fig 6

Pelvic schwannoma: Fig 1. Transabdominal USG reveals a solid-cystic left adnexal lesion, causing anterior displacement of round ligament (yellow arrow). Axial Fig 2, coronal Fig 3 and sagittal Fig 4 T2-weighted MR images show a well-circumscribed predominantly solid left adnexal lesion with multiple cystic areas and fluid-debris level in few of them. Planes with surrounding structures are maintained. Left ovary is seen separately (blue arrow). No obvious intraspinal extension is demonstrated. The lesion shows intermediate to hyperintense signal on T2W and isointense on T1W Fig 5 compared to adjacent skeletal muslces and myometrium. On T1-weighted post contrast Fig 6 image shows nearly homogenous enhancement.

FNAC/ Trucut biopsy suggested leiomyoma. To look for mass effects colonoscopy was performed and extrinsic compression at recto-sigmoid junction was seen. Cystoscopy was also performed and was found to be normal study.

Per-operatively 14X10 cm retroperitoneal mass was found adhered to left ovary and was separated. Tumour was excised completely along with the capsule with the help of the surgeons. Surgical specimen sent for frozen section was reported to be benign spindle cell tumour and hence surgery was not proceeded further. Around 3.9 litres blood loss occurred intraoperatively and 3 units PCV, 4 units FFP and 4 units platelets were transfused and the patient was kept in ICU for observation. Postoperative period was uneventful.

On gross examination of the specimen, it was a capsulated globular soft tissue measuring 14X10 cm. On cut section- whorled and yellowish areas were seen with some areas showing necrosis. On microscopy, cells were seen arranged in whorled pattern with round to oval elongated nucleus and indistinct cytoplasm with no mitosis or atypia. Focal areas show xanthomatous and necrotic changes. Immunohistochemistry showed positivity with S-100 and focally with SMA. Final diagnosis of Giant Ancient Schwannoma was made.

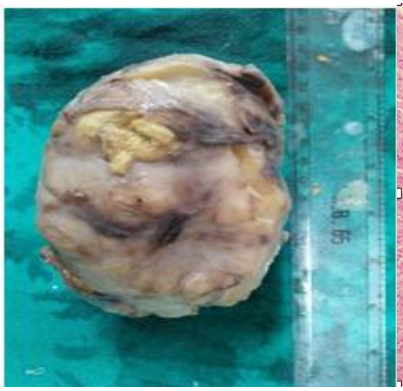

Fig. 7

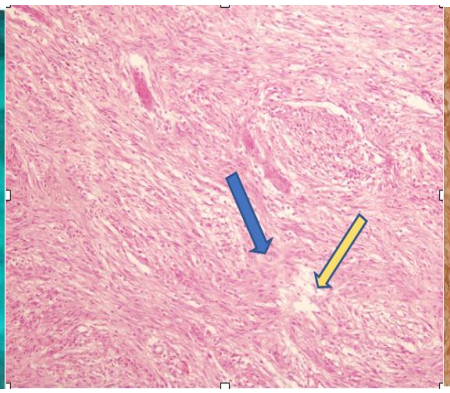

Fig. 8

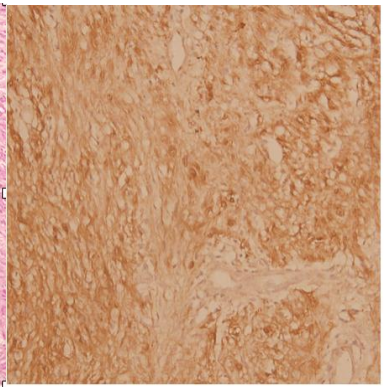

Fig. 9

Retroperitoneal schwannoma Fig 7. Gross specimen showing capsulated tumor mass containing fat and areas of necrosis Fig 8. Blue arrow showing benign spindle cells arranged in fascicles (Antoni A) with focal hypocellular areas shown by yellow arrow (Antoni B); H&E(200X) Fig 9. Spindle cells showing positivity with S-100; IHC-Streptavidin peroxidase(400X).

DISCUSSION

Schwannoma or neurilemmoma is a common tumour which is rarely found in retroperitoneal space. The most common symptom is slowly progressing abdominal pain. Pressure symptoms such as urinary or fecal incontinence and neurological symptoms in the lower extremities, are rarely observed.[4] [5]

A preoperative diagnosis is very difficult to make due to the lack of typical imaging features.[6] In general, MRI is regarded as the diagnostic modality of choice in the evaluation of retroperitoneal tumors as it allows better evaluation of the origin, extent and internal composition of these lesions. There are a few well-known imaging characters for schwannomas which are mainly target sign and fascicular sign. However, these typical signs are not seen frequently in retroperitoneal schwannomas. The "fascicular sign" stands for the the appearance of bundles which is a general property of neurogenic tumors. On the other hand, the "target sign" is the presence of hypointense center and hyperintense periphery on T2 weighted MRI. The lesion presented in this paper does not exert any of the above-mentioned typical diagnostic signs.[7] Its large size can also make it challenging for the radiologist to determine its exact location because a retroperitoneal schwannoma can displace the uterus and mimic an intra-peritoneal mass as in our case it was confused with broad ligament fibroid. The differential diagnosis should include retroperitoneal lymph nodes, ovarian tumours, broad ligament fibroid, paraganglioma, neurofibroma, malignant fibrous histiocytoma, lymphoma, liposarcoma.

USG/CT-guided biopsy is usually unreliable as evident in our case. This is especially so in large tumors in which cellular pleomorphism is present in areas of degeneration, which may thus be misinterpreted as malignant cells or presence of spindle cells may be misinterpreted as leiomyoma. Occasionally it may be helpful if the sample contains sufficient Schwann cells to visualize microscopically. Hemorrhage, infection and tumor seeding are possible risks of percutaneous biopsy.[7]

The ideal treatment of retroperitoneal schwannoma is complete excision.[8] Chemotherapy and radiotherapy have only limited roles but may have marginal added benefit in malignant schwannomas.[8]

Complete resection of the retroperitoneal schwannoma is challenging due to the presence of adjacent or circumscribing major vessels (that is, the aorta and its branches, the inferior vena cava and others) but it was possible in our case as it was encapsulated and not infiltrating the surrounding tissues.[8]

Although local resection is generally enough, metastatic cases have been reported after resection.[11] Therefore, follow up is important for this patient.

Final diagnosis is made on histopathological examination and immunohistochemical staining. On histological examination, two main microscopic patterns could be recognized: A highly cellular component (Antoni A area) and a myxoid component (Antoni B area). The predominance of one or the other reflects the heterogeneous findings at radiological investigation (CT and MRI). Almost all the schwannomas show intense immunohistochemical staining for S100 protein, confirming the neuroectodermal origin of the tumor cell as was positive in our case too.[12] The present study is therefore consistent with the majority of studies in supporting that the final diagnosis of retroperitoneal schwannoma can only be made based on the findings of postoperative histopathology.

CONCLUSION

Retroperitoneal schwannoma is a rare tumor which may mimic broad ligament fibroid and should be considered as a differential diagnosis. Preoperative diagnosis is challenging and final diagnosis can only be made after histopathological and immunohistochemical confirmation.

REFERENCES

1. Tortorelli AP, Rosa F, Papa V, Rotondi V, Sanchez AM, Bossola M, et al. Retroperitoneal schwannomas: Diagnostic and therapeutic implications. Tumori, 2007; 93: 312-5.
2. Bezzi M, Orsi F, Rossi P. Nonencapsulated ancient schwannoma of the renal sinus. AJR Am J Roentgenol, 1996; 166: 1498.
3. Kudo T, Kawakami H, Kuwatani M, Ehira N, Yamato H, Eto K, *et al*. Three cases of retroperitoneal schwannomadiagnosed by EUS-FNA. World J Gastrtoenterol, 2011; 17: 3459-64.
4. Song JY, Kim SY, Park EG, Kim CJ, Kim do G, Lee HK and Park IY: Schwannoma in the retroperitoneum. J Obstet Gynaecol Res., 2007; 33: 371-375.
5. Mastoraki A, Toska F, Tsiverdis I, Kyriazi M, Tsagkas A, Danias N, Smyrniotis V and Arkadopoulos N: Retroperitoneal schwannomas: Dilemmas in diagnostic approach and therapeutic management. J Gastrointest Cancer, 2013; 44: 371-374.
6. Hughes MJ, Thomas JM, Fisher C, Moskovic EC. Imaging features of retroperitoneal and pelvic schwannomas. Clin Radiol, 2005; 60: 886-93.
7. Goh, B.K., Tan, Y.M., Chung, Y.F.A., Chow, P.K., Ooi, L.L. and Wong, W.K., 2006. Retroperitoneal schwannoma. The American Journal of Surgery, 192(1): 14-18.
8. Daneshmand S, Youssefzadeh D, Chamie K, et al. Benign retroper- itoneal schwannoma: a case series and review of the literature. Urol- ogy, 2003; 62: 993–97.
9. Shoji F, Maruyama R, Okamoto T, Wataya H, Nishiyama K, Ichinose Y. Malignant schwannoma

of the upper mediastinum originating from the vagus nerve. World J Surg Oncol, 2005; 3: 65.
10. Zhao M, Li X, Wang J, Li W, Huang Z. Retroperitoneal schwannoma treated with percutaneous computed tomography-guided radiofrequency ablation. J Neurosurg Spine, 2012; 17: 173-6.
11. C. S. Wong, T. Y. Chu and K. F. Tam, "Retroperitoneal schwannoma: a common tumour in an uncommon site," Hong Kong Medical Journal, 2010; 16(1): 66–68.
12. Wong, C.S., Chu, T.Y. and Tam, K.F., 2010. Retroperitoneal schwannoma: a common tumour in an uncommon site. Hong Kong Medical Journal.