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Case Report

Retroperitoneal Schwannoma- A Case Report

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Abstract: Schwannoma (neurilemmomas) is a peripheral nerve sheath tumor generally occurs on the head, neck and trunk. Retroperitoneal schwannomas are the rarest of all retroperitoneal tumors. We report a 30 year woman with a schwannoma on the retroperitoneum that was preoperatively misdiagnosed as a adnexal mass, with a brief review of the literature.
Keywords: Schwannoma, Neurilemmoma, Retroperitonium

INTRODUCTION

Schwannoma (neurilemmomas), a peripheral nerve sheath tumor generally occurs on the head, neck and trunk. Retroperitoneal schwannomas are the rarest of all retroperitoneal tumors [1]. The majority of symptoms caused by the tumor are due to its mass effect. Surgical resection is enough to treat the tumor. Schwannoma is usually a benign tumor and risk of recurrence and malignant transformation [2] is low. Hereby we are reporting a 30 year woman with a schwannoma on the retroperitoneum that was preoperatively misdiagnosed as a adnexal mass.

CASE REPORT

A 30 year old woman presented with abdominal pain since 6 months. There was no history of bladder or bowel disturbances. On general physical and per abdominal examination, no abnormality was detected. Per vaginal examination revealed a normal sized anteverted, anteflexed ,mobile uterus with a non tender, firm, slightly fixed mass of 5x5cms size with smooth surface was felt in the right fornix, left fornix was free.

Her routine haematological tests and serum LDH, beta hCG, CA-125 and AFP were within limits. Ultrasonography revealed a complex mass measuring 5x6cms in the right adenexa with dense internal echoes. CT scan was suggested for further evaluation which revealed a well defined cystic lesion measuring 5x5x5cms with homogenous internal debris in the right adenexa. No calcified areas were detected.

With a provisional diagnosis of endometriotic cyst of the right ovary, laparotomy was decided. The abdomen was opened through the lower midline incision. Uterus and bilateral adenexae were normal. There was a retroperitoneal round, firm, solid, smooth walled mass (6x6x5cms) in the pelvis, on the right side of the right sacro-iliac joint, attached to the underlying structures. The peritoneum over the mass was opened, and complete excision of the mass was done, with blunt dissection. Bleeding from the underlying vessels was controlled by sutures. Abdomen was closed in layers .Patient stood the surgery well and the post operative period was uneventful. The patient was asymptomatic on 3 months follow up.

Histopathological examination of the specimen revealed a capsulated mass showing cellular areas with spindle shaped cells, in a pallisading pattern. Less cellular areas showed scattered spindle cells. Areas of degeneration were also seen. These features were consistent with diagnosis of schwannoma (Fig 1).

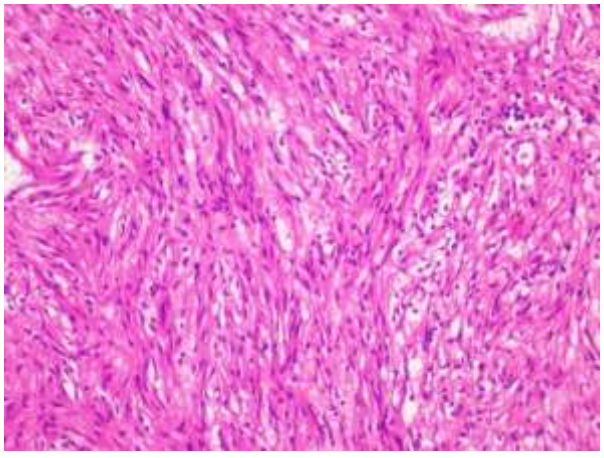

Fig.1: Loose and compact areas in schwannoma.The spindle cells show wavy nuclei, at places palisading is noted (H&E stain X10)

DISCUSSION

Benign nerve sheath Tumors are of two types- schwannomas (more common) and neurofibromas. Schwannomas are most frequently present in patients aged 20 to 50 years. They comprise 5% of all benign soft tissue tumors 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 [3]. Schwannomas are usually solitary, slow growing, non-aggressive neoplasms.

The majority of these tumors are asymptomatic and are often found incidentally or present withvague, non specificsymptoms.They are usually detected as an abdominal mass on routine examination as in this case. The predominantly cystic nature of the Tumor was demonstrated by ultrasound and CT scan examination but the exact nature of the Tumor could not be diagnosed. Malignant transformation is very rare .Malignant schwannomas are large in size and highly aggressive Tumors. They are painful, and may cause many different symptoms depending on the location and size [4].

On gross appearance, schwannomas are well circumscribed, encapsulated tumors, which frequently undergo cystic degeneration. Histologically, the nuclear palisading may be a striking feature. Typical schwannomas are composed of inter mixed Antoni A components (cellular and arranged in short bundles or interlacing fascicles) and Antoni B areas (less cellular and organized with more myxoid components). The cellular variant, which includes most large retroperitoneal and pelvic shwannomas, has a uniform spindle cell appearance without Antoni A or B areas. Characteristically, all schwannomas show uniform and intense staining for S 100 protein [5].

The common pathological variants of schwannomaare: (a) Conventional schwannoma is histologically benign and sometimes may cause destruction of surrounding osseous structures. (b) Ancient schwannoma, usually large deeply located (e.g. retroperitoneum) is a variant, displays prominent degenerative changes like cyst formation, calcification, haemorrhage and hyalinization and behave as ordinary schwannomas. (c) Cellular schwannoma, usually found in the deeper tissues (retroperitoneum or mediastinum), histologically simulate malignant peripheral nerve sheath Tumor (MPNST). Mitotic figures may be present. The thick fibrous capsule may show dense lymphocyte infiltrate. Desmin is negative. (d) Plexiformschwannoma, is particularly in a cellular form, and when occurring in childhood, simulates MPNST, and, (e) Melanoticschwannoma, a rare variant, and usually occurs in middle aged adults and commonly arises from the posterior spinal nerve roots. These cells contain abundant melanin pigment. Hence, it is often mistaken for melanoma.Less common variants are epitheloid schwannoma, glandular schwannoma, neuroblastoma–like schwannoma and schwannoma-perineurioma [6-7].

Hughes MJ *et al.* [3], in their largest radiological series of abdominal or pelvic schwannomas, showed that a smooth well defined border, ovoid or spherical shape and location in the pre sacral region or lower retro peritoneum at the pelvic brim, are frequent findings in primary abdominal or pelvic schwannomas ; 11 of 13 cases showed all these features. Evidence of degeneration was common, with 8 schwannomas showing cystic change and 3 showing areas of calcification. The cystic schwannomas tended to be larger, with a mean diameter of 10.5 cm.

Pre-operative diagnosis of these Tumors is difficult. With ultrasound guided fine needle aspiration, computed tomography, and magnetic resonance imaging, the accuracy of the diagnosis could be improved. Recommended treatment is surgical excision. Malignant schwannomas are insensitive to chemotherapy and radiation, resulting in poor prognosis. Immunohistochemical positivity for S-100 protein is very useful for the diagnosis [8]. A combination of clinical, pathological and immunohistochemial studies help in their diagnosis [9].

CONCLUSION

We report this patient with retroperitoneal pelvic schwannoma because of its rare location, vague symptoms and diagnostic dilemma.

REFERENCES

1. Ooi LL, Mack PO; Benign Retroperitoneal Neurilemmoma--a case report. Ann Acad Med Singapore, 1990; 19(3): 410-412.
2. Song JY, Kim SY, Park EG, Kim CJ, Kim do G, Lee HK *et al.*; Schwannoma in the retroperitoneum. J Obstet Gynaecol Res., 2007; 33(3): 371-375.
3. Hughes MJ, Thomas JM, Fisher C, Moskovic EC; Imaging features of retroperitoneal and pelvic schwannomas. Clin Radiol., 2005; 60(8): 886-893.
4. Tauro LF, George C, Kamath A, Lobo G, Shetty P, Hegde BR; Giant schwannoma in the pelvic retroperitoneum. Journal of Clinical and Diagnostic Research, 2008; 2: 1210-1214.
5. Weiss SW, Goldblum JR editors; Benign Tumors of peripheral nerves. In Enzinger & Weiss's soft tissue Tumors. Mosby, Messouri, USA, 200 :1146-1168.
6. Kurtkaya-Yapicier O, Scheithauer B, Woodruff JM; The pathological spectrum of Schwannomas. Histol Histopath., 2003; 18(3): 925-934.
7. Saad AG, Mutema GK, Mutasim DR; Benign cutaneous epitheloidschwannoma: Case report and review of literature. Am J Dermatopathol., 2005; 27(1): 45-47.

8. Ortiz Rey JA, Alexsandro da Silva E, Rico Gala S, AntónBadiola I, San Miguel Fraile P, Zungri Telo E, de la FuenteBuceta A. A retroperitoneal cellular schwannoma. Actas Urol Esp., 1999; 23(5): 455-458.

9. Nakashima j, Ueno M, Nakamura K, Tachibana M, Baba S, Deguchi N; Differential diagnosis of primary benign and malignant retroperitoneal Tumors. Int J Urol., 1997; 4: 441-446.