Case Report

BENIGN RETROPERITONEAL SCHWANNOMA

Balaramaraju S.¹, Saravana Kumar¹, Rajasekaran S.¹, Balasubramainan C.², Maheswaran K.V.¹, Balagurunathan¹ and Baskaran¹

¹Department of General Surgery, Sri Lakshmi Narayana Institute of Medical Sciences (Affiliated to Bharath University, Chennai), Osudu, Pondicherry
²Department of General Surgery, Sree Balaji Medical College and Research Centre, (Affiliated to Bharath University, Chennai), Chennai

*Author for Correspondence

ABSTRACT

Benign retroperitoneal schwannoma is rather an unusual presentation of an infrequent lesion. Because of their unusual location in the retroperitoneum and absence of clinical features until the tumour enlarges to a size capable of causing compression of intra-abdominal or intrapelvic organs the diagnosis of these cases are often delayed. Most often they are diagnosed incidentally on evaluation of coexistant and sometimes irrelevant symptomatology. We hereby report a case evaluated for mittelschmirz, provisionally diagnosed as a case of extra adrenergic paraganglioma and therein excised – the histopathology however turned out to be that of a schwannoma.

Keywords: Retroperitoneal Schwannoma, Tumour, Adrenal Paraganglioma

INTRODUCTION

Benign retroperitoneal schwannoma is rather an unusual presentation of an infrequent lesion the diagnosis of a benign retroperitoneal tumour is mainly one of exclusion, but ultimately based on histology. Most often they are diagnosed incidentally on evaluation of coexistant and sometimes irrelevant symptomatology such as mittelschmirz in this case.

CASES

A 35 year old lady had presented with mittelschmirz in a peripheral hospital where she evaluated with ultrasonogram of the abdomen and CT abdomen; both of which picked up a mass lesion 4X5 cms anteromedial to right kidney suggesting the possibility of a benign adrenal tumour and so was referred to our institution. Her biochemical profile was normal except for an elevated urinary Vannilly Mandeleic acid (34.1 ng/day). MRI suggested the possibility of an extra adrenal paraganglioma (Figure 1). Laparotomy revealed a solitary, solid, well encapsulated firm tumour of size 5X4.5 cms (Figure 2) nestled between the right kidney laterally, the IVC medially and the right renal vein superiorly. The tumour was excised in – toto. There were no cardiovascular events encountered in the intra – operative/post – operative period.

The cut section of the tumor composed of firm pale white homogenous solid tissue punctuated with few haemorrhagic areas. The histopathological evaluation confirmed the diagnosis of a schwannoma (Figure 3) with no evidence of malignancy.

DISCUSSION

Schwannoma is a benign encapsulated nerve sheath tumour composed of cells with immunohistophenotyping of Schwann cells. They are characteristically solitary, circumscribed, slow growing and painless lesions eccentrically located on proximal nerves or spinal nerve roots. However about 3% of schwannomas can occur in the retroperitoneum constituting approximately 4% of all retroperitoneal tumours (PatoeskaI et al., 2002). There are less than 100 cases reported in world literature (Chew et al., 2005). Schwannomas do not exceed a diameter of 5 to 6 cms, but larger size tumours upto a diameter of 28cms have been reported. The presenting symptoms are vague, poorly localized pain and
discomfort, accompanied by non specific digestive disturbances. There have been various instances of retroperitoneal schwannoma mimicking hepatic tumour in caudate lobe (Nakemura et al., 1997); presenting as an adrenal lesion; and a pancreatic cystic tumour (Hsiao et al., 1998). Hence the diagnosis of a benign retroperitoneal tumour is mainly one of exclusion, but ultimately based on histology. Malignant transformation although extremely rare is usually observed in cases with underlying von-Recklinghausen’s disease. Large size of the tumour; presence of symptoms; irregular margins and the absence of calcification are predictors of malignant tumours.

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Although there are no specific radiological features associated with schwannomas; following descriptions are reported as highly suggestive:

1. A well demarcated round/oval mass with heterogenous contrast enhancement due to cystic/haemorrhagic changes.
2. Calcification and cystic changes in a large tumour.
3. Iso/slightly hyperintense signal intensity on T1 weighted images compared with muscle and high signal intensity on T2 weighted images similar to fat.

Compared to CT, MRI seems to have more specificity and can better delineate the tumour from adjacent anatomical structure (Pui et al., 1998). Particularly the acquisition of a fat suppression sequence (besides standard T1 & T2 W images) on which the schwannoma will maintain its high signal allowing for a delineation from a pure lipomatous tumour.
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Surgical excision is recommended by most authors for primary treatment of solitary schwannoma with the preservation of associated nerve if identified. Open laparotomy has been the traditional approach in retroperitoneal and pelvic schwannomas. Once completely excised; recurrence of benign schwannomas is not expected unless the post operative histology confirms malignancy of the tumour. In such an unexpected event of proven malignancy one should probably consider re-resection if a wide margin has not been achieved originally, followed by adjuvant therapy. On cut section; they are usually firm to hard tan white to yellow glistening tissue with areas of cyst formation and focal haemorrhage. The nerve is often identified. Histopathology show biphasic pattern of compact ANTONI A alternating with sparse areas of ANTONI B. positive immunohistochemical staining results with S-100 protein; neuron specific enolase and microfilament proteins support the diagnosis of schwannoma. Because of several advantages like rapid recovery with short hospital stay and better cosmetic results over traditional surgical technique; laparoscopic resections have also been performed for retroperitoneal schwannomas (Melvin, 1996). Retroperitoneal schwannomas can also be safely resected through limited incision by endoscopic minilaparotomy (Kageyama et al., 2002).

Conclusion

Being rare in occurrence and diverse in presentation, neurogenic tumours of the retroperitoneum pose a diagnostic challenge. However the diagnosis can be confirmed by histology and surgical excision remains the mainstay of treatment.

REFERENCES


