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Case Report

BENIGN RETROPERITONEAL SCHWANNOMA

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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 mittelshmirz, provisionally diagnosed as a case of extra adrenal 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 turmours (Patocskai *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.

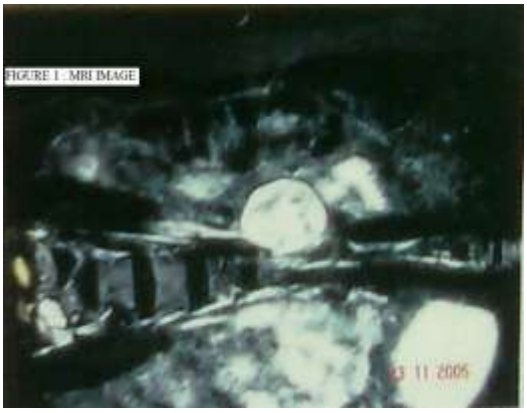

Figure 1: MRI showing the possibility of an extra well adrenal paraganglioma

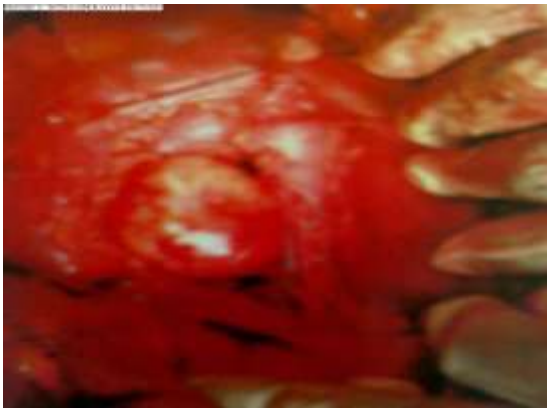

Figure 2: Laparotomy revealing a solitary, solid, encapsulated firm tumour of size 5X4.5 cms

Figure 3: The histopathological evaluation confirming the diagnosis of a schwannoma with no evidence of malignancy

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.

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

Chew BH and Knudson BE (2005). A retroperitoneal GI schwannoma presenting as a perinephric mass. *Canadian Journal of Urology* **12** 2555-2556.

Hsiao WC, Lin PW and Chang KC (1998). Benign retroperitoneal schwannoma mimicking a pancreatic cystic tumor: case report and literature review. *Hepatogastroenterology* **45** 2418-2420.

Kageyama Y, Kihara K, Ishizaka K, Okuno T, Kawakami S, Fujii Y, Masuda H, Suzuki M, Hyochi N, Arai G, Saito K and Sakai YJ (2002). Endoscope – assisted minilaparotomy (endoscopic minilaparotomy) for retroperitoneal schwannoma: experience with three cases. *Japanese Journal of Clinical Oncology* **32** 177-180.

Melvin WS (1996). Laparoscopic resection of a pelvic schwannoma. *Surgical Laparoscopy & Endoscopy* **6** 489-491.

Nakemura M, Takeyasu K, Takayama T, Shimada K, Yamamoto J, Kosuge T, Sakamoto M, Yamasaki S, Shimizu S and Makuuchi M (1997). Retroperitoneal schwannoma mimicking hepatic tumor in the caudate lobe. *Japanese Journal of Clinical Oncology* 27 282-284.

Patocskai EJ, Tabatabaian M and Thomas MJ (2002). Cellular schwannoma: a rare presacral tumour. *Canadian Journal of Surgery* **45** 141- 44.

Pui MH, Yang ZY and Li ZP (1998). Computed tomography of abdominal neurogenic tumours. *Australasian Radiology* **42** 183-187.