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Retroperitoneal Gastrointestinal Type Schwannoma Presenting as a Renal Mass

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ABSTRACT

Retroperitoneal schwannomas are extremely rare, and unreported in Urology. Often thought to be malignant from imaging the diagnosis is often delayed until Histology. We report a case of retroperitoneal schwanoma thought to be a malignant renal mass. Seventy three year old lady presented with abdominal pain. Imaging showed a mass attached to the renal pelvis thus she underwent a radical nephrectomy. Histology reported retroperitoneal schwannoma. Malignant forms are rare however treatment for these is surgical excision. Awareness of the existence of these tumors may help in avoiding unnecessary radical surgeries by opting for biopsy preoperatively.

Introduction

Schwannomas are usually solitary, sometimes multiple benign tumors of nerve sheath composed solely of schwann cells. They grow slowly and push nerve fibers aside. They may arise in relation to named peripheral nerves, nerve roots or trunks.

A group of rare tumors occurring in the gastro-intestinal (GI) tract have similar histology and are known as GI type schwannomas. However these tumors have been identified as somewhat separate from conventional schwannomas on the basis of their histology, immunohistochemistry, and latterly, their genetic profile.[1] Very rarely GI type schwannomas have been recorded outside the GI tract.[2,3]

We report a case of retroperitoneal GI type schwannoma thought to be a malignant renal mass. To our knowledge this only the third report of a GI type schwannoma in the retroperitoneum.

Case report

A 73 year old female presented with vague abdominal pain, without any systemic symptoms. There were no urinary symptoms or urological history. Past history included hypertension, asthma and the patient was an ex smoker.

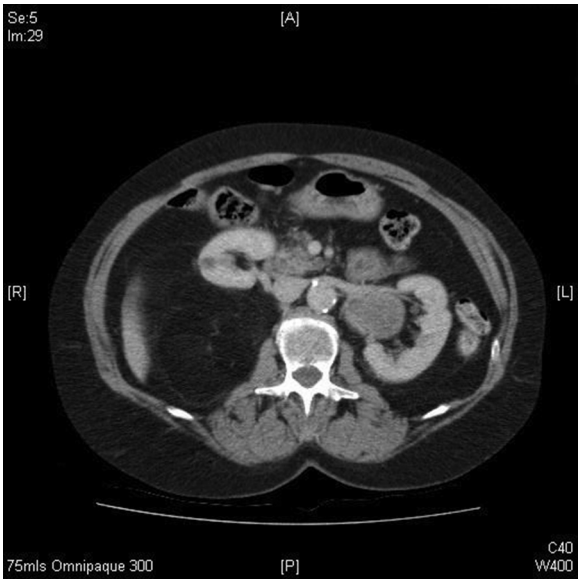

Figure 1. Axial CT showing mass lesion at the left renal hilum and a right retroperitoneal fatty mass.

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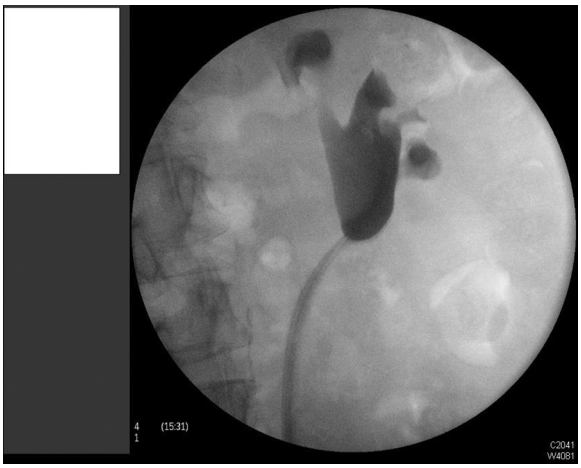

Figure 2. Retrograde showing filling defect in left renal pelvis.

An abdominal ultrasound showed an echo poor mass in the region of the left renal pelvis confirmed as 45 mm on computed tomography (CT). This scan also showed a right retroperitoneal mass, with a fatty appearance suggestive of a large lipoma or low grade liposarcoma (Fig. 1). A retrograde pyelogram showed smooth filling defect affecting the medial aspect of the renal pelvis and flexible ureteroscopy confirmed a bulge on the medial wall of the renal pelvis with normal mucosa, consistent with an extrinsic mass (Fig. 2). Ureteric brushings and biopsies were inconclusive.

Following discussion at MDT initial management was interval imaging. At computed tomography at 10 months the mass had increased to 54 × 41 mm with no evidence of renal vein invasion or metastases. MDT decision was radical nephrectomy for a potential renal malignancy. A left open nephrectomy was performed.

Macroscopically there was a pale circumscribed mass, 49 × 35 mm at the renal hilum, extrinsic to the kidney. On cut surface the lesion had a pale yellow surface.

Histology of the mass showed a spindle cell lesion with a monomorphic appearance. The nuclei were slender, slightly tapering with minimal atypia and the cells were separated by a light, uniform, collagenous stroma. There was an inflammatory cell component present, mainly lymphocytic and a prominent lymphoid cuff around the lesion, such that the initial impression was of a tumor within a lymph node. Immunohistochemistry showed strong positivity for S100 protein and GFAP. There was no staining for cytokeratin, SMA, desmin or C-kit (CD117). Staining for EMA revealed a population of slender cells surrounding the lesion consistent with perineurium, and staining for neurofilament protein showed axons running through the lesion, confirming origin of the lesion within a nerve. Fluorescence–in-situ hybridization studies showed no evidence of the MDM2 gene. The morphological and immunohistochemical features were consistent with a schwannoma of so called gastrointestinal type (Fig. 3).

Discussion

Schwannomas form one of the two major types of benign peripheral nerve sheath tumors, neurofibromas being the second. They are often found in nerve roots in the cranium and spine. In the periphery they often occur in association with a named nerve but can occur at almost any site. Schwannomas occur relatively commonly in the posterior thorax but less frequently in the retroperitoneum.[4]

Schwannomas are composed entirely of Schwann cells leading to their characteristic immunoprofile: i.e. strong S100 positivity.

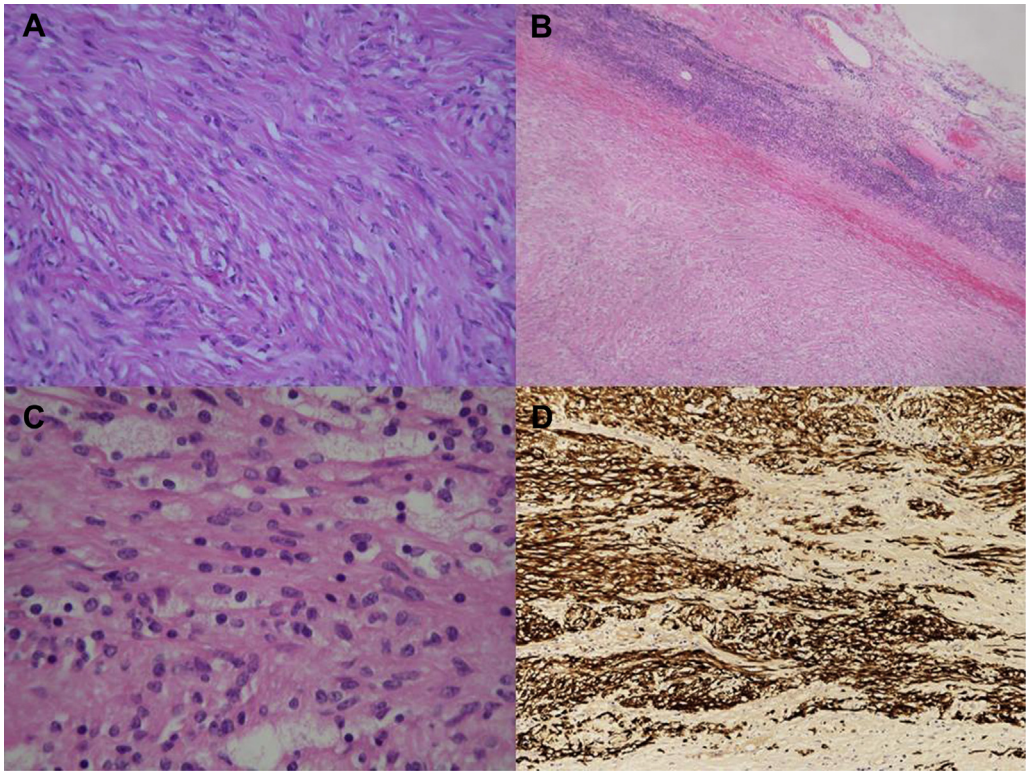

Figure 3. High power view showing delicate tapering spindle cells (A) (original magnification ×200). Edge of the lesion showing tumor (lower left) with prominent lymphoid cuff (upper right) (B) (original magnification ×50). Areas with inflammatory cells including lymphocytes and foamy histiocytes were present (C) (original magnification ×200). Strong staining for S100 protein (D) (original magnification ×100).

They have a characteristic biphasic appearance with alternating hypercellular and hypocellular areas known as Antoni A and B areas respectively. Nuclear palisading, known as Verocay bodies, is distinctive.

Over recent years it has been recognized that schwannomas occurring in the GI tract appear somewhat different: the biphasic appearance, palisading and certain other features, such as hyalinized vessels, are not present, neither is the nuclear morphology the same. GI schwannomas also have a characteristic inflammatory infiltrate, a lymphoid cuff and a dense stroma unlike that of conventional schwannomas or neurofibromas. Although GI schwannomas are not separately categorized in the WHO classification of soft tissue tumors there is evidence that, at a genetic level, they may be more closely related to neurofibromas than conventional schwannomas. Unlike conventional schwannomas, GI type schwannomas do not show mutation of the NF2 gene on chromosome 22.[1] In contrast there is loss of heterozygosity of the NF1 gene on chromosome 17 in around 50%. Like conventional schwannomas, GI type schwannomas are benign.

In the retroperitoneum the commonest spindle cell neoplasms are dedifferentiated liposarcoma and leiomyosarcoma. The former is a pleomorphic sarcoma whilst the latter shows positivity for the muscle markers. Dedifferentiated liposarcoma arises in its precursor lesion, atypical lipomatous tumor (ALT), whose synonym is well differentiated liposarcoma in the retroperitoneum. This lesion has a different biology at different sites; in the periphery ALT has a low propensity for dedifferentiation (and malignancy) whilst in the retroperitoneum it has a high propensity. CT scanning in this case revealed a fatty retroperitoneal mass on the contralateral side to the lesion in question, thus dedifferentiated liposarcoma was a consideration. However the molecular hallmark of this tumor, amplification of MDM2 gene on chromosome 12q15 was absent, excluding this possibility.

Another differential diagnosis for spindle cell neoplasms involving the kidney and retroperitoneum is sarcomatoid renal cell carcinoma. However in this case the lesion was clearly separate from the kidney, non-pleomorphic, cytokeratin negative and displayed S100 positivity, excluding this differential.

Gastrointestinal stromal tumors can rarely occur in the retroperitoneum and this was thus a consideration in the current case but was again ruled out by immunohistochemistry (CD117 and DOG1 negative).

Conclusion

Having excluded differential diagnoses by morphology, immunohistochemistry and molecular means we have identified a perirenal lesion in the retroperitoneum as a GI type schwannoma. Gastrointestinal type schwannoma is a rare diagnosis, most descriptions being based on case reports. Very rarely this tumor has been described in sites outside the GI tract.[5] To our knowledge only two cases of retroperitoneal GI type schwannoma have been described previously, at least one of which also presented as a perirenal mass diagnosed after nephrectomy.[2]

Conflict of interest

All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest, or non-financial interest in the subject matter or materials discussed in this manuscript.

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