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Case Report

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Retroperitoneal schwannomas

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Summary

Background: Retroperitoneal schwannomas are very rare and are usually found incidentally.

Cases Report: Two rare cases of retroperitoneal schwannomas are reported. Both were incidentally found during US scans for non-specific epigastric pain and were initially diagnosed as non-secreting retroperitoneal tumors. The diagnosis was confirmed by CT scan. In both patients the tumors were resected. The definitive diagnosis was possible by histopathology.

Conclusions: Although the preoperative assessment of a retroperitoneal tumor may be indicative of a retroperitoneal schwannoma, the definitive diagnosis is possible only by histopathology after surgical removal of the tumor.

key words: **schwannoma • retroperitoneum • adrenal gland**

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Background

Schwannomas are rare tumors originating from the Schwann sheath of the peripheral or cranial nerves. They are usually benign tumors and in 95% of the cases they originate from the peripheral nerves [1]. Visceral schwannomas are very infrequent. Retroperitoneal schwannomas are even more infrequent, while only a few cases of adrenal schwannomas have been reported.

Case Report

Case report 1

A 59-year-old woman presented with continuous epigastric pain over the last 2 months. A non-homogenous tumor (8×7 cm) near the aorta, the inferior vena cava, and the porta hepatis was found by ultrasound examination of the abdomen. The right kidney was ectopic, located in the pelvis. CT scan showed the presence of a well-circumscribed solid mass with non-homogeneous density located at the site of the right adrenal (Figure 1). The tumor was heterogeneously enhanced after IV administration of contrast material, due to regions of necrosis. The MRI of the abdomen confirmed the above findings. The tumor was a non-secreting one. The patient underwent resection of the right adrenal. By histopathology the tumor was found to be a schwannoma of the right adrenal (Figure 2).

Case report 2

A 61-year-old man presented with mild epigastric pain. The CT abdominal scan revealed a large (7.5 cm) well-circumscribed heterogeneous, mostly cystic tumor with irregular inner septa and thick wall, located in the left retroperitoneal area anterior to the left adrenal gland and adjacent to its lateral limb (Figure 3). The MRI confirmed the heterogeneous structure of the tumor and showed enhancement of the solid parts and hemorrhagic elements within it (Figure 4). The patient underwent resection of the tumor, which was found to be a schwannoma by histopathology (Figure 5).

Discussion

Schwannomas occur more frequently in the head, neck, stomach, and limbs. Only a few cases have been reported in the retroperitoneal area and particularly in the adrenal [2]. Schwannomas are usually benign, slow-growing, encapsulated tumors, and are rarely malignant. Malignant schwannomas are frequently associated with von Recklinghausen syndrome or other types of neurofibromatosis [3]. So far less than 30 cases of adrenal schwannomas have been reported in the literature. Most of them have been preoperatively diagnosed as non-secreting adrenal tumors. They originate from the Schwann cells of the nerve fibers innervating the adrenal medulla [4,5].

Most adrenal schwannomas are incidental findings. They vary in size from a few mm to 15 cm in diameter. Some patients experience minor symptoms like abdominal or back pain, or hematuria. They are usually non-secreting tumors, although 1 noradrenalin-secreting retroperitoneal schwannoma has been reported [6].

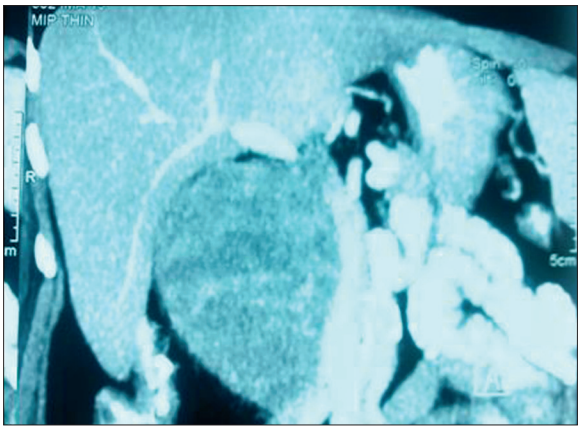

Figure 1. Computed tomography (CT) shows a large (8×7 cm) well-circumscribed solid mass with inhomogeneous densities, compressing the inferior vena cava.

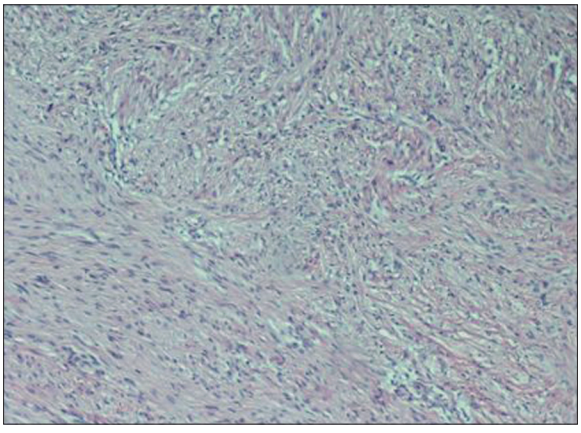

Figure 2. Cellular area of Schwann cells intermingled with wavy collagen bundles.

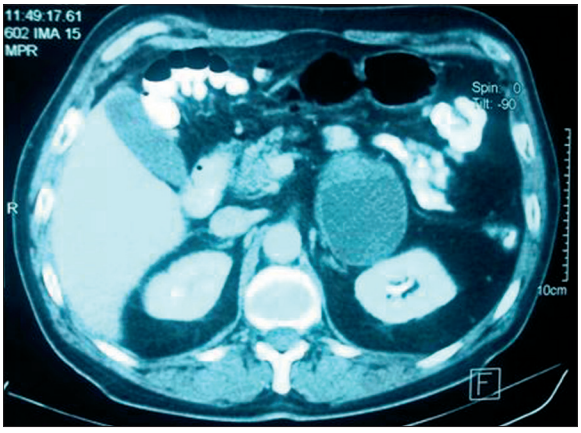

Figure 3. The CT abdominal-scan revealed a large (7.5 cm) well-circumscribed heterogeneous, mostly cystic tumor with irregular inner septa and thick wall located in the left retroperitoneal area anterior to the left adrenal gland and adjacent to its lateral limb (Figure 3).

CT-scan usually reveals a well-circumscribed, homogeneous, round or oval mass, with slight enhancement [7]. Cystic degeneration or calcification may rarely be present in long-standing schwannomas and may be suggestive of the diagnosis [8].

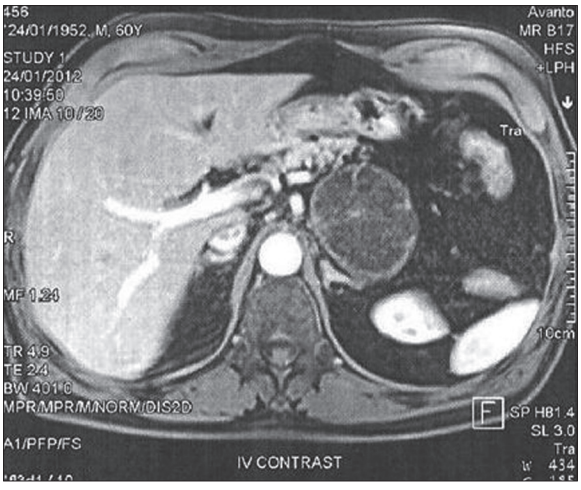

Figure 4. The MRI confirmed the heterogeneous structure of the tumor and showed enhancement of the solid parts and hemorrhagic elements within it.

Histopathology makes possible the definitive diagnosis [9]. Two distinct patterns of adrenal schwannomas have been described: Antoni type A has elongated spindle cells arranged in irregular streams and is compact in nature, and Antoni type B has a looser structure with cystic spaces mixed within the tissue. Large tumors often show central cystic degeneration. Tumors with degenerative atypia, hyalinization and regressive changes have been called ancient schwannomas. The presence of cystic structures within a retroperitoneal tumor is frequent in schwannomas and suggests the diagnosis [7]. These 2 types may coexist in the same specimen, but 1 predominates. Immunohistochemistry is essential because tumor cells are positive for S-100 and vimentin. Although it is unusual, in some cases positive calretinin excludes a possible S-100 positive neurofibroma [10].

Conclusions

Although the preoperative assessment of a retroperitoneal tumor may be indicative of the presence of a retroperitoneal schwannoma, definitive diagnosis is possible only by histopathology after surgical removal of the tumor.

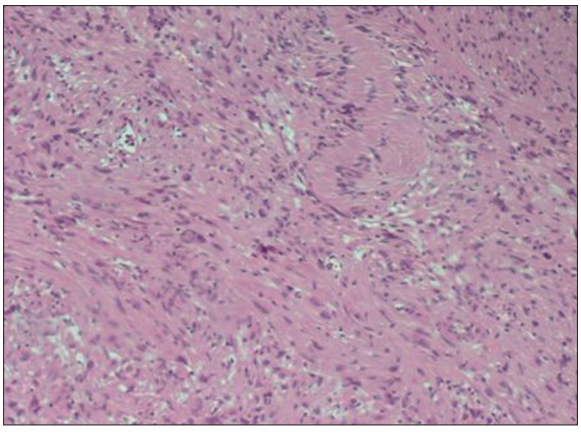

Figure 5. Antoni type A schwannoma with mild nuclear atypia.

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