Retroperitoneal schwannomas of renal and pararenal origin: presentation of two case reports

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Abstract

Retroperitoneal schwannomas are a rare entity. They originate from the Schwann cells of the nerve sheaths and may be of renal or pararenal origin. We report on two patients with retroperitoneal schwannomas, who received surgery under the suspicion of renal cell carcinoma.

Introduction

Schwannomas are among the most frequent nerve sheath tumors, along with neurofibromas. They originate from the Schwann cells and in most cases exhibit benign growth. They either occur sporadically or in association with a neurofibromatosis.[1] They are usually found in the head/neck area or in the extremities. Only about 3% of all schwannomas are located in the retroperitoneal space.[2] We are going to give an account of two patients with a retroperitoneal schwannoma, one of them being of renal and the other of pararenal origin, who were treated in our department.

Case Report #1

During a routine checkup, a 60 years old, asymptomatic male patient without comorbidities was sonographically diagnosed with a space-occupying lesion in his right kidney. The computed tomography examination (Figure 1A) revealed a polycystic, centrally hypodense space-occupying mass with an axial perimeter of 11×6.5 cm and a craniocaudal perimeter of 9.5 cm which absorbed the contrast agent in a ring-like form. The diagnosis was *cranially approximating the liver and medially impressing the vena cava inferior*. It was not possible to exclude an infiltration of the vena renalis based on the morphology found in the imaging data. There was no evidence for distant metastases. The physical, laboratory and urine examinations were without pathological findings. No tumor cells were found in urine cytology.

Subsequently the kidney was surgically exposed via thoracoabdominal access. Intraoperatively, the existence of a large, centrally located tumor was confirmed. The frozen section biopsy showed tumor tissue consisting of spindle cells, but no particulars. Since a malignant tumor could not be excluded, a nephrectomy and a hilar lymphadectomy were conducted. The postoperative course was uncomplicated. The patient was released on the 9th day after the surgery.

Histologically, the tumor seemed to be composed of spindle cells. These possessed an eosinophile cytoplasm and spiral, slim nuclei. The structure resembled an Antoni A pattern. The immunohistochemical S100 staining showed a nuclear and cytoplasmatic reaction. The rate of proliferation in Ki67 immune staining was less than 5% in the tumor cells (Figure 2).

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Case Report #2

During a routine checkup, an asymptomatic micro-hematuria was discovered in a 69 year old female patient. During further sonographic investigation, evidence of a space-occupying lesion in the right kidney was found; therefore a computed tomography of the abdomen was conducted (Figure 1B). It revealed in the area of the right renal pelvis a tumorous space-occupying process of 6.5×3.8 cm with partially central colliquations, compressing the vena cava. The space-occupying process rested on the M. psoas without infiltrating it. An infiltration into the vena renalis or evidence for distant metastases were not detected. Known comorbidities were a coronary stenting following a STEMI 3 years before, an arterial hypertonia and a latent hypothyreosis. The physical, laboratory and urine examinations were without pathological findings. We were unable to confirm the presence of a micro-hematuria. No tumor cells were found in urine cytology.

Subsequently the right kidney was surgically exposed via a flank section. Intraoperatively, the tumor was found as described, touching the renal pelvis, the vena cava, the duodenum and the ureter without infiltrating these structures. The tumor was dissected and macroscopically removed completely. The kidney was left *in situ*. The frozen section biopsy showed a tumor consisting of spindle cells without proof of malignancy. The postoperative course was uncomplicated. The patient was released on the 7th day after the surgery.

Histologically, we found a cell-rich tumor encapsulated by connective tissue and with a lymphocytic mound with tumor cells running parallel to each other. The structure resembled an Antoni A pattern. The immunohistochemical S100 staining showed a nuclear and cytoplasmatic reaction. The rate of proliferation in the Ki67 analysis was low.

Discussion

Schwannomas are neurally differentiated tumors of mesenchymal origin which can be located cerebrally, spinally, peripherally or viscerally. Urogenital or retroperitoneal schwannomas are very rare. There are only a few case reports on their occurrence in the prostate, testicles or penis.[3,4] Their occurrence as incidentalomas in the adrenal gland is a rarity as well.[5]

Retroperitoneal schwannomas make up about 3% of all schwannomas. The earliest description of a renal schwannoma dates from the year 1956.[6] A total of 15 cases is described in the English-language literature published up to 2007.[7] In a recent publication, Verze *et al*. mention 27 cases covered in works written in English and other languages.[8] Out of these cases, 5 patients (19%) suffered from malignant growth, and 19 (70%) reported clinical symptoms such as flank pain, overall feeling of illness, and loss of weight.

Histologically, two growth patterns can be

observed. These are referred to as Antoni A, for a cell-rich structure with cell nuclei running parallel to each other, and Antoni B, for Schwann cells without a particular order in a myxoid stroma. Both cases presented here exhibit growth in the Antoni A pattern. The definitive diagnosis usually requires the immunohistochemical proof of neurogenic differentiation, with S100 used as the marker. Occasionally, retroperitoneal schwannomas can express cytokeratins. Various histological variants of the schwannoma are known, such as the degeneratively changed schwannoma, the cell-rich schwannoma, the plexiform schwannoma, the melanocytic schwannoma, the glandular schwannoma and the epitheloid schwannoma. It is therefore not unlikely that the morphology can make differential diagnosis difficult.

Preoperative diagnosis is also difficult: computed tomography and magnetic resonance findings resemble renal cell carcinomas in localization and signal behavior, which results in frequent misinterpretations of imaging results.[9,10] This was also the case with our patients described.

Due to the difficulty of histological diagnosis described above, preoperative fine-needle aspiration does not seem promising. As with the intraoperative frozen section biopsy, a positive statement on the type of the tumor cannot usually be made.[11] Often the final diagnosis can only be reached after the surgery. For these reasons therapy equals the total surgical resection. In swannomas of renal origin, depending on the localization of the tumor, a partial renal resection could be a theoretical option. Because of their size and the fact that most renal schwannomas are located centrally in the area of the hilus rather than peripherally however, kidney-preserving surgery is usually not possible. A recommendation for a surgical safety distance cannot be made due to the small number of cases.[12] If the patient is not eligible for a surgical procedure because of existing comorbidities, radio-frequency ablation could be an alternative method of therapy.[13] In case of a malignant differentiation it proved impossible in most reported cases to cure the patients by means of a postoperative radio- or chemotherapy, and the patients deceased some months after surgery.[14-16] There is, however, one reported case of a malignant renal schwannoma which remained relapse-free for a period of over 4 years following the total surgical resection, and without adjuvant therapy.[17] As in most of the cases described, our patients, too, were initially suspected to be suffering from renal cell carcinomas, and a nephrectomy and a retroperitoneal tumor resection, respectively, conducted. In both cases the ensuing histological analysis lead to the diagnosis with a schwannoma. Despite the lack of malignant differentiation we recom-

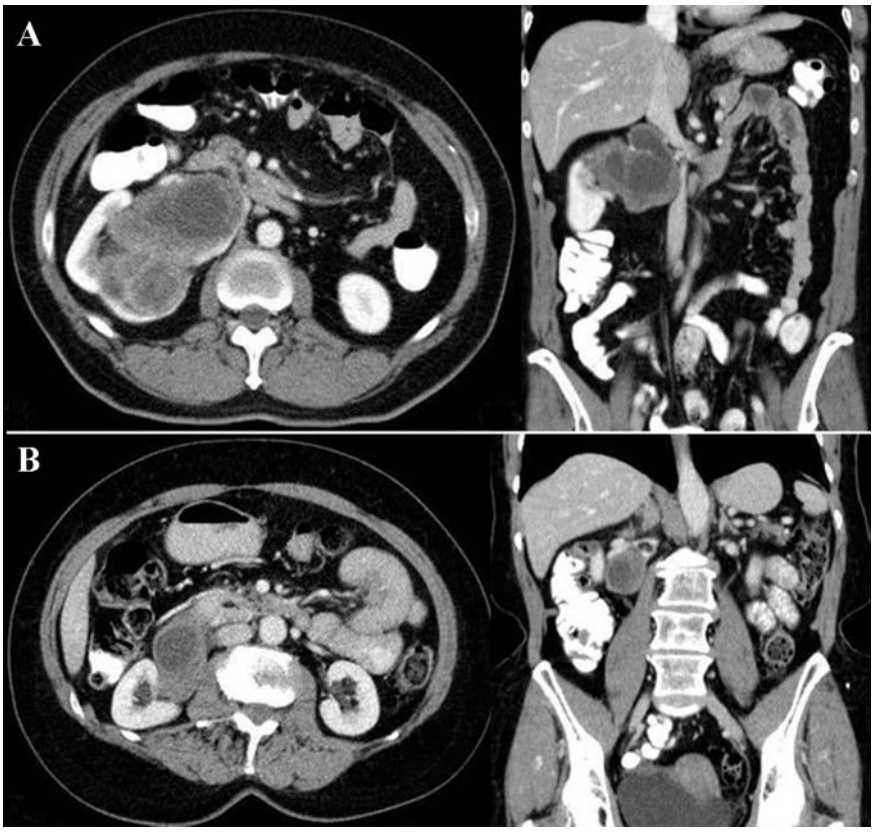

Figure 1. A) Computed tomography-scan showing a right renal mass with central hypodense areals, contrast enhancement and contact to the vena renalis and vena cava. B) Computed tomography-scan showing a 6.4×3.8 cm mass with origin of the renal hilum and contact to the vena cava.

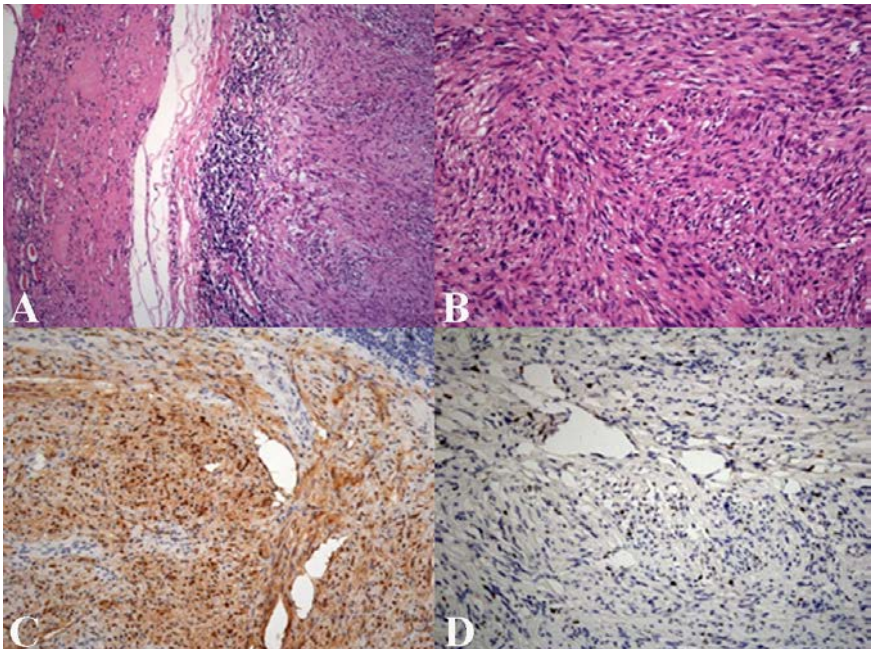

Figure 2. A) Spindle cells with Antoni A growth pattern and demarcation from the atrophic renal parenchyma (100×). B) Spindle cells show a palisading fashion. No signs of atypic cytologic changes (200×). C) Cells show a strong immunoreactivity with S-100 (100×). D) Immunoreativity with Ki67 shows a low proliferation rate (200×).

mended an MRT examination after 6 months in order to assess the postoperative status. Although there is a connection between nerve sheath tumors and neurofibromatoses,[1] this could not be confirmed in our patients.

Conclusions

Schwannomas are rare tumors which originate from the Schwann cells of the nerve sheaths. When located in the retroperitoneum, they may be of renal or pararenal origin. They are therefore often preoperatively misinterpreted as renal cell carcinomas. The histological diagnosis is also frequently difficult and can in most cases only be reached through an immunohistochemical analysis. Although most schwannomas exhibit benign growth, in some cases malignant degeneration may occur.

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