Hemorrhagic, calcified, and ossified benign retroperitoneal schwannoma

First case report

Shao-Yan Xu, MD\textsuperscript{a,b,c,d}, Ke Sun, MD\textsuperscript{e}, Hai-Yang Xie, MD\textsuperscript{a,b,c,d}, Lin Zhou, MD, PhD\textsuperscript{a,b,c,d}, Shu-Sen Zheng, MD, PhD\textsuperscript{a,b,c,d}, Wei-Lin Wang, MD, PhD\textsuperscript{a,b,c,d},

Abstract

**Background:** Schwannomas are mesenchymal tumors arising from the neural sheaths of peripheral nerves. They can almost develop in any part of the body, while head, neck and extremities are the most common sites. Occurrence in the retroperitoneum is rare. Schwannomas can show secondary degenerative changes including cyst formation, hyalinization, hemorrhage, and calcification, whereas the ossified retroperitoneal schwannoma was only reported in a malignant one.

**Case summary:** We first present a benign ossified retroperitoneal schwannoma in a 61-year-old female. The mass was found by a routine health examination. Computed tomography (CT) and magnetic resonance imaging (MRI) showed a well-defined mass in the area among duodenum, right liver, and kidney. Definitive preoperative diagnosis of the mass was difficult. By laparotomy, the mass was found in the retroperitoneum. We completely removed the tumor and gross specimen showed a mass with a capsule and 6 × 6 × 4.8 cm in size. Microscopic examination showed the tumor is composed of spindle-shaped cells with degenerative changes of hemorrhage, calcification, and ossification. Immunohistochemically, S-100 protein was strongly positive. Finally, the mass was diagnosed as a hemorrhagic, calcified, and ossified benign schwannoma in the retroperitoneum. The patient was followed up for a period of 21 months, during which she was well with no evidence of recurrence.

**Conclusion:** We report the first case of a benign retroperitoneal schwannoma with secondary degenerative changes including hemorrhage, calcification, and ossification. Precise preoperative diagnosis of the tumor is challenging even with multiple preoperative imaging modalities. After complete resection, patients with benign retroperitoneal schwannomas generally have good prognosis.

**Abbreviations:** CT = computed tomography, EUS-FNA = endoscopic ultrasound-guided fine needle aspiration, MRI = magnetic resonance imaging, US = ultrasound.

**Keywords:** calcification, case report, hemorrhage, ossification, retroperitoneal schwannoma
1. Introduction

Schwannomas are neurogenic tumors originating from the Schwann cells in nerve sheaths.[1] They can occur in patients at all ages with equal frequency in male and female.[2] More than 90% Schwannomas are benign and occupy about 5% of benign soft-tissue neoplasm.[3] Almost every location of human body can be involved and the most common sites are head, neck, and extremities.[4] However, only 1% to 3% of schwannomas were found in the retroperitoneum and account for almost 1% of all retroperitoneal tumors.[5,6] Patients with retroperitoneal schwannomas are normally asymptomatic and the tumors are usually found incidentally. Surgical operation may be the optimal treatment for retroperitoneal schwannomas. Secondary degenerative changes of schwannomas can sometimes be shown including cyst formation, hyalinization, hemorrhage, and calcification. However, ossified retroperitoneal schwannoma was only reported in a malignant one so far.[7] In the present study, we first present a ossified benign retroperitoneal schwannoma in a 61-year-old female, who was cured by a complete excision of the tumor.

2. Case report

On July 25, 2014, a 61-year-old female was referred to our hospital because of a lesion found in the duodenum by a routine health examination in the local hospital. The abdomen was flat and soft with no mass palpable. His family history had no significant disease. Laboratory results were as follows: WBC 2.60 × 10^9 cells/L (4.0–10), neutrophils 1.8 × 10^9 cells/L (2.0–7.0), monocytes 0.06 × 10^9 cells/L (0.12–1.00). Tumor markers and other laboratory results were normal.

An unenhanced CT scan showed a well-defined mass in the area among duodenum, right liver and kidney, 5 cm in diameter. The mass was mainly low-density, while regions of high density were visible (A). On the contrast-enhanced CT, the mass was mildly and inhomogeneously enhanced (arrow) (B).

According to these imaging results, a retroperitoneal neurogenic tumor was primarily considered.
We performed a laparotomy and found a mass surrounded by a fibrous capsule located in the area among duodenum, front of vena cava, and lower edge of right liver. The mass was linked with the retroperitoneal adipose tissue by a pedicle. We completely removed the mass and intraoperative frozen pathological examination suggested a soft tissue tumor with degenerative changes of hemorrhage, calcification, and ossification. Macroscopically, a mass with capsule was 6.0 × 6.0 × 4.8 cm in size and yellowish-white in color. Microscopically, some areas of the tumor were calcified, ossified (Fig. 3A), and hemorrhagic (Fig. 3B). The tumor mainly consisted of spindle-shaped cells with palisading arrangement. Both hypercellular and hypocellular areas were visible. Atypical cells or signs of malignancy were not showed (C) (H&E stain, magnification power: ×200).

3. Discussion

Schwannomas are neoplasms that originate from Schwann cells of the nerve sheaths.[8] Malignant schwannomas are very rare and usually associated with von Recklinghausen disease.[9] Most schwannomas are benign and show either monosomy 22 or loss of 22q material.[10] They usually affect adult patients aged 20 to 50 years[11] and arise from almost anywhere in the body, while...
4. Conclusion

Schwannomas in the retroperitoneum are rare and ossified benign retroperitoneal schwannoma has not been reported. We present the first hemorrhagic, calcified, and ossified benign retroperitoneal schwannoma. It is a huge challenge to obtain a precise diagnosis before operation because of nonspecific clinical and imaging characteristics. Definitive diagnosis of schwannoma is determined by histopathological and immunohistochemical examinations of surgical specimens. After complete resection, patients with benign retroperitoneal schwannomas generally have good prognosis and low risk of tumor recurrence.

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