Clinical Case Report

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Hemorrhagic, calcified, and ossified benign retroperitoneal schwannoma

First case report

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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 $6 \times 6 \times 4.8$ 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 schwanommas 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

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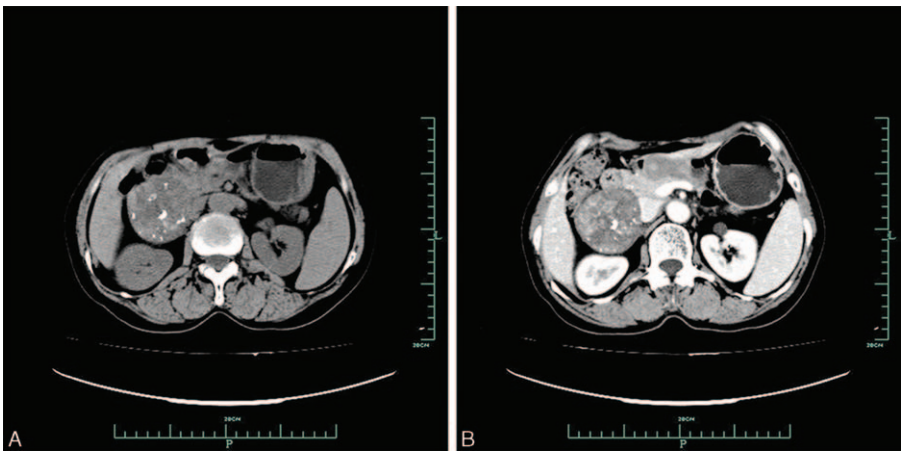

Figure 1. An unenhanced computed tomography (CT) scan showed a well-defined mass (arrow) in the area among duodenum, right liver, and kidney, 5 cm in diameter. The mass was mainly low-density, while regions of high density were visible (A). On the contrast-enhanced CT, the mass was mildly and inhomogeneously enhanced (arrow) (B).

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 (4–10), 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, whereas regions of high density were visible, compatible with calcification and/or ossification (Fig. 1A). Duodenum and pancreatic head were pushed by the mass. On the contrast-enhanced CT, the mass was mildly and inhomogeneously enhanced (Fig. 1B). On MRI, the mass appeared hypointense on T1-weighted images (Fig. 2A) and inhomogeneous hyperintense on T2-weighted images (Fig. 2B). According to these imaging results, a retroperitoneal neurogenic tumor was primarily considered.

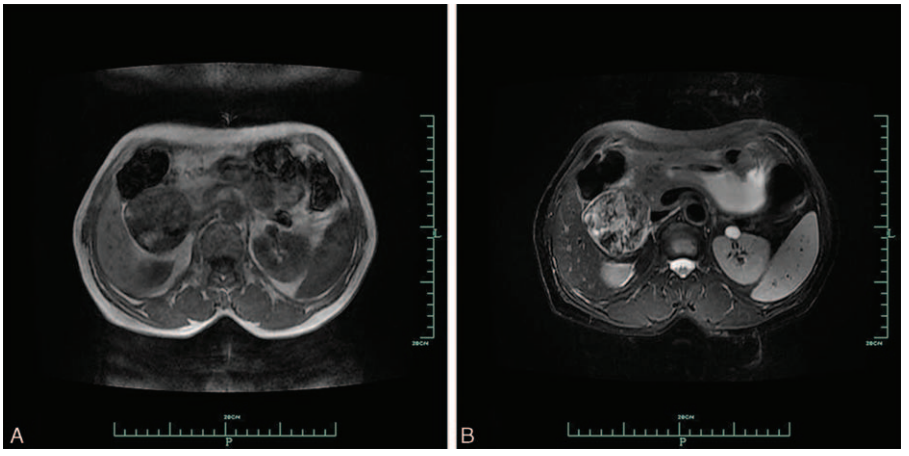

Figure 2. On magnetic resonance imaging, the mass appeared hypointense on T1-weighted images (arrow) (A) and inhomogeneous hyperintense on T2-weighted images (arrow) (B).

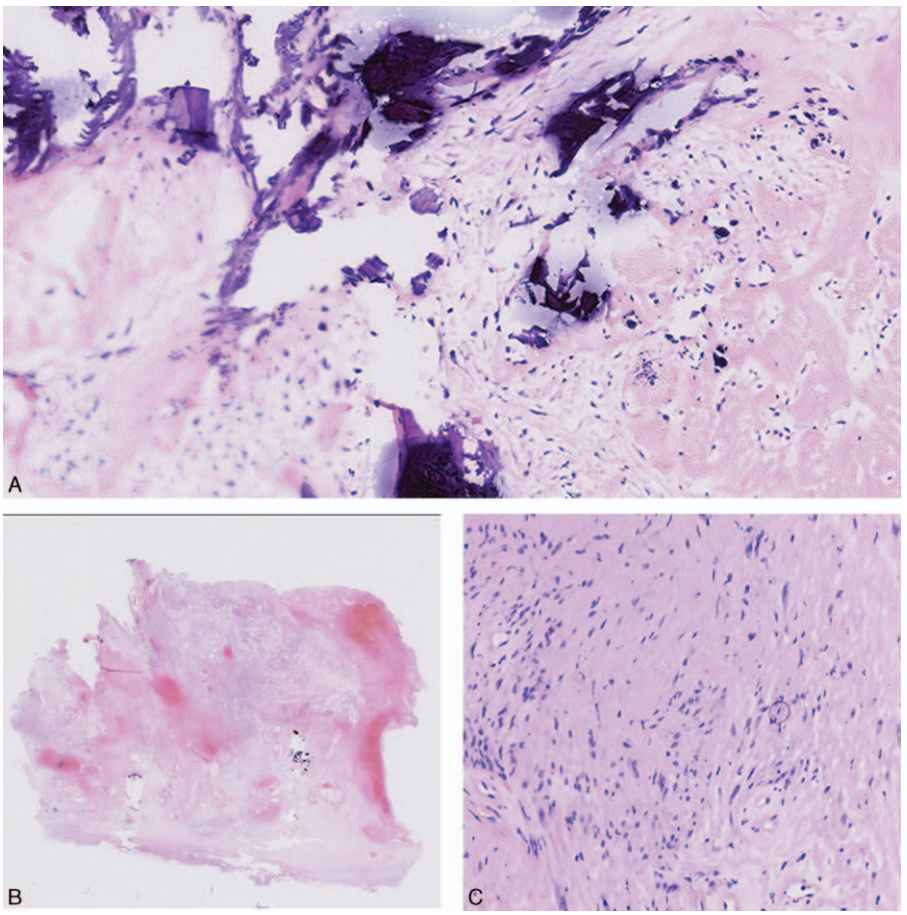

Figure 3. Microscopically, some areas of the tumor were calcified, ossified (A) (H&E stain, magnification power: ×200) and hemorrhagic (arrow) (B) (H&E stain, magnification power: ×5). The tumor mainly consisted of spindle-shaped cells with palisading arrangement. Both hypercellular and hypocellular areas were visible. Atypical cells or signs of malignancy were not showed (C) (H&E stain, magnification power: ×200).

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.

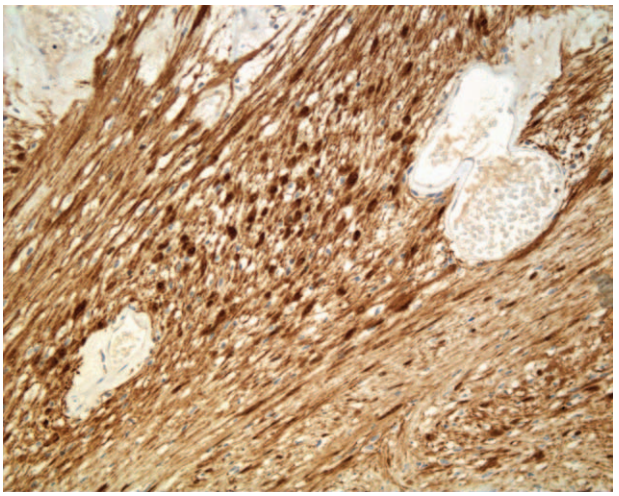

Figure 4. Immunohistochemical investigation showed the protein S-100 was strongly positive (H&E stain, magnification power: ×200).

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 shown (Fig. 3C). Immunohistochemical investigation showed the protein S-100 was strongly positive (Fig. 4), whereas smooth muscle actin (SMA), CD34, and CD117 negative. Finally, the tumor was diagnosed as a benign retroperitoneal schwannoma with hemorrhage, calcification, and ossification changes. After surgery, the patient recovered smoothly and left the hospital 6 days later. She was followed up for a period of 21 months, during which she was well with no evidence of tumor recurrence.

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

head, neck, and extremities are the most common sites.[12] Schwannomas in the retroperitoneum are rare, comprising only 1% to 3% of all schwannoma and almost 1% of retroperitoneal neoplasms.[13] Secondary degenerative changes including cyst formation, calcification, hemorrhage, and hyalinization can sometimes be shown. However, ossified retroperitoneal schwannoma was only reported in a malignant one[7] and we report the first ossified benign retroperitoneal schwannoma. Since the retroperitoneal space is rather large and flexible, most of the patients were asymptomatic and the diagnosis of retroperitoneal schwannomas was often delayed. Accurate diagnosis of the lesion is challenging even with multiple preoperative imaging modalities.[14]

Definitive diagnosis of schwannoma is determined by histopathological and immunohistochemical examinations of surgical specimens. Microscopically, a typical schwannoma is composed of compact cellular area (Antoni type A area) and loose, hypocellular myxoid area with microcystic spaces (Antoni type B area).[15] Secondary degenerative changes of the tumor can sometimes be shown, including cyst formation, hyalinization, hemorrhage, and calcification. However, ossification degeneration is extremely rare.[16] In the present case, the tumor was hemorrhagic, calcified, and ossified. Immunohistochemically, schwannomas stainings are strongly positive for proteins including S-100, vimentin, and CD 56, whereas negative for desmin, smooth muscle myosin, SMA, CD 34, and CD 117.[17] The positive S-100 protein is essential for the diagnosis of a schwannoma.[18]

The preoperative accurate diagnosis of retroperitoneal schwannomas is difficult. Imaging modalities have certain diagnostic value but lack specificity.[19] Some radiological characteristics may be helpful for physicians to differentiate benign tumors from malignant tumors and contribute to orienting their approach. On unenhanced CT scan, a typical schwannoma is depicted as a well-defined and inhomogeneous low-density mass on CT images with encapsulation and/or cystic degeneration. However, in the present case, regions of high density were visible, compatible with calcification and ossification. Calcification and ossification also have been reported in some other retroperitoneal lesions including serous cyst,[20] liposarcoma,[21] and mucinous cystadenocarcinoma,[22] so it is hard to distinguish retroperitoneal schwannomas from these lesions just according to that phenomenon. The clinical value of the phenomenon maybe that ossification cannot only occur in malignant schwannoma, but also the benign one. Schwannomas with high Antoni A areas appear inhomogenous because of increased lipid content. Antoni B areas of schwannomas appear cystic and multiseptated and show low density because of loose stroma and low cellularity.[23] On contrast-enhanced CT, well-enhanced areas correspond to Antoni A because of the increased vascularity and unenhanced areas correspond to Antoni B because of less vascularity.[19,24] Typical MRI findings of schwannomas show hypointense on T1-weighted images and inhomogeneous hyperintense on T2-weighted images.[25] Retroperitoneal schwanommas are usually shown as well-defined hypodense lesions by ultrasound (US) and blood flow signals cannot be detected within the lesions by Color Doppler US. Larger lesions may be heterogeneous, hyperechoic, and contain prominent cystic components, but these signs are nonspecific. Endoscopic ultrasound-guided fine needle aspiration (EUS-FNA) may greatly contribute to obtaining a precise preoperative diagnosis. In one report, 3 cases of asymptomatic retroperitoneal tumors were diagnosed as benign schwannomas by EUS-FNA.[26]

Surgical operation is the optimal treatment for retroperitoneal schwannomas.[23] In the present case, by laparotomy, we found the mass located in the retroperitoneum and a complete resection of the tumor was performed. After surgery, the prognosis of the patient is generally good. However, probably because of incomplete excision, recurrence is the most frequent complication and is reported in 5% to 10% cases.[27]

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 schwanommas generally have good prognosis and low risk of tumor recurrence.

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