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Case Report

Appendiceal schwannoma presenting as acute appendicitis ☆

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ABSTRACT

Schwannomas are nerve sheath tumors that rarely occur in the gastrointestinal tract. In the gastrointestinal tract, schwannomas are mostly found in the stomach and small bowel. Herein, we describe a case of appendiceal schwannoma that caused appendix obstruction and developed acute appendicitis. An 83-year-old woman was admitted to the emergency department with 3 days' history of abdominal pain. She had tenderness and rebound tenderness in the lower right quadrant. Computed tomography revealed a 1.3 cm mass in the appendix orifice, with associated distal appendiceal dilatation and wall thickening. The patient underwent emergency surgery (laparoscopic partial cecectomy). Histopathological examination confirmed that the mass was a schwannoma and was associated with acute suppurative appendicitis. Our case is significant in that it adds to another rarely reported case of appendiceal schwannoma. Moreover, it is important to recognize the presence of an appendiceal tumor associated with acute appendicitis.

Introduction

Schwannomas are benign tumors derived from the Schwann cells of the nerve sheath. These neoplasms usually affect the nerves in the head, neck, and flexor surfaces of the extremities [1]. Gastrointestinal schwannomas are uncommon and account for 2%-6% of all submucosal neoplasms of the gastrointestinal tract [2,3]. Appendiceal schwannomas are extremely rare. Here, we present a case of appendiceal schwannoma associated with acute appendicitis.

Case report

An 83-year-old woman presented to the emergency department of our hospital with 3 days' history of abdominal pain. The pain was exacerbated by breathing or body movement, and worsened over time. The patient was under medications for hypertension. Physical examination revealed tenderness and rebound tenderness in the lower right quadrant. Laboratory examinations indicated elevated inflammatory markers (white blood cells, 14.93 × 10^3/uL [reference range; 4-10 × 10^3/uL]; segmented neutrophils, 88.0% [reference range; 50.0%-75.0%]; serum C-reactive protein, 11.4 mg/dL [reference range; 0-0.3 mg/dL]). Computed tomography (CT) was performed to distinguish the surgical abdomen. On the axial CT scans, we observed an ovoid, well-demarcated, homogeneous

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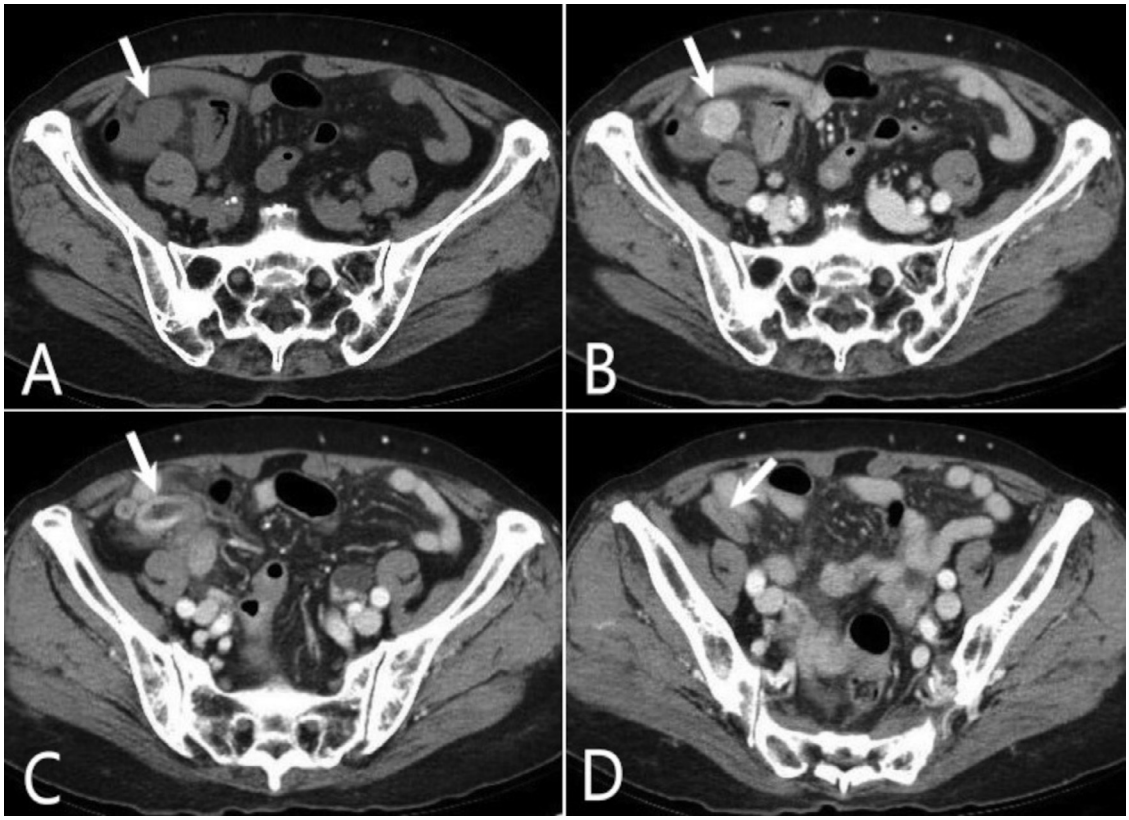

Fig. 1 – Axial computed tomography scans. (A, B) A well-defined ovoid homogeneously enhancing mass measuring 1.3 cm is seen at the cecal tip. (C, D) Distal appendix is dilated, with wall thickening, and surrounding fat infiltration.

enhancing mass measuring 1.3 cm at the cecal tip (appendix orifice) (Figs. 1A and B). It was associated with distal appendiceal dilatation, appendiceal wall thickening, and surrounding fat infiltration (Figs. 1C and D). On coronal and sagittal CT scans, the mass was located in the submucosal layer, and had normal overlying mucosa (Fig. 2A, arrowheads). Therefore, we presumed the mass to be a subepithelial tumor, similar to a neuroendocrine neoplasm, accompanied by acute appendicitis. The patient underwent an emergency surgery (laparoscopic partial cecectomy). Intraoperatively, the surgeon observed that the colon was mildly adhered to the peritoneum, a notable cecal mass, appendicitis, and turbid ascites in the pelvic cavity. The gross specimen revealed ill-defined yellowish myxoid mass measuring 1.3 cm × 1.0 cm at cecal tip. The appendix was a suppurative change measuring 8 × 1 cm. Histopathological examination revealed that the mass consisted of proliferating spindle cells that formed Verocay bodies in specific regions. Verocay bodies are components of Antoni A, which are dense areas of schwannomas located between palisading spindle cells. Two nuclear palisading regions and an anuclear zone comprised one Verocay body (Fig. 3A). Immunohistochemistry revealed a strong and diffuse S-100 protein staining (Fig. 3B). Pathological examination revealed that the mass was benign schwannoma associated with acute suppurative appendicitis. The postoperative course was uneventful and the patient was discharged.

Discussion

Schwannomas are mainly slow-growing benign tumors that originate from Schwann cells arising from the peripheral nerve sheath. These tumors could be found wherever peripheral nerves are seen, and usually affect the nerves in the head, neck, and flexor surfaces of the extremities [1]. Schwannomas are rare in the gastrointestinal tract; account for 2%-6% of all submucosal tumors of the intestine, and stomach is the most common site (60%-70%) [4]. Schwannomas of the large intestine are very rare. However, the incidence of accidental discovery of schwannomas of the large intestine has increased with the development of endoscopic examination techniques [5]. Nonetheless, appendiceal schwannomas are extremely rare, with only a few cases reported [2,5,6].

Gastrointestinal schwannomas appear as submucosal tumors. Among the primary gastrointestinal submucosal tumors, gastrointestinal stromal tumor (GIST) and neuroendocrine tumor (NET) G1 may evolve into malignancy; therefore, preoperative diagnosis is important. However, these tumors are difficult to distinguish despite advances in imaging. Recently, endoscopic ultrasonography (EUS) and endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) have improved the diagnosis of gastrointestinal submucosal tumors [5].

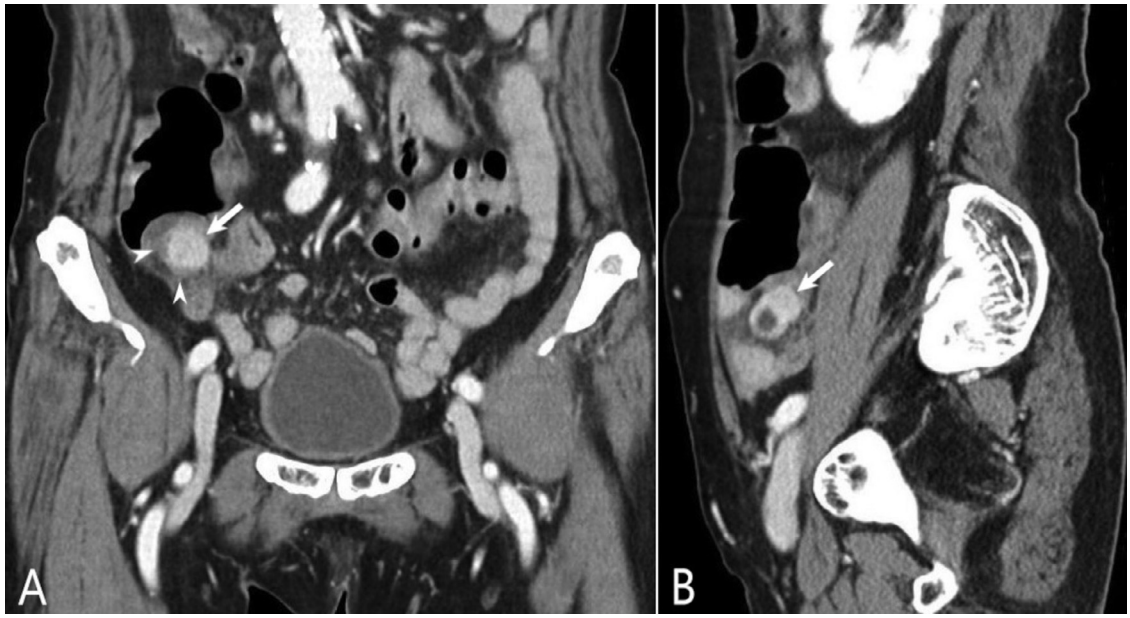

Fig. 2 – Contrast-enhanced coronal and sagittal computed tomography scans. (A, B) The mass is located in submucosal layer, and has normal overlying mucosa (A, arrow heads).

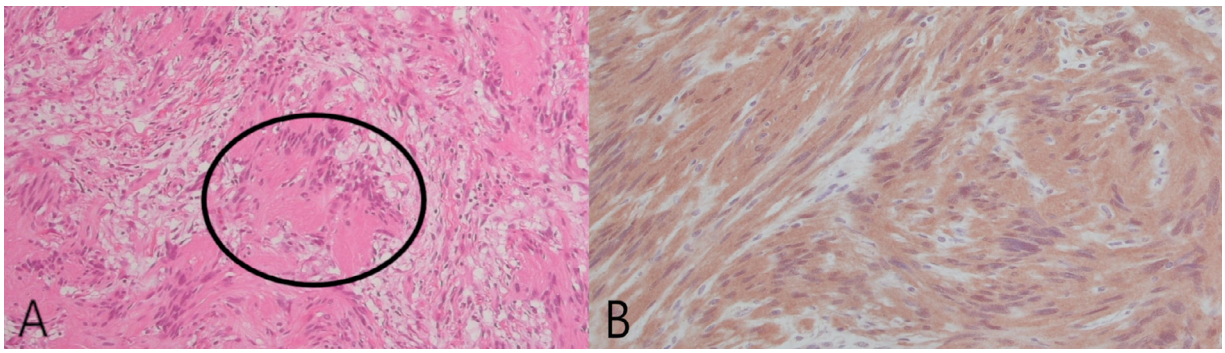

Fig. 3 – Histopathological findings. (A) Microscopic examination reveals proliferation of the spindle cells, which form the Verocay body (circle, hematoxylin and eosin stain, ×100). (B) Immunohistochemistry demonstrates strong and diffuse staining of the S-100 protein (×100).

In most patients, symptoms of gastrointestinal schwannomas are nonspecific. Our patient had tenderness and rebound tenderness in the right lower quadrant; consequently, this was a symptom of acute appendicitis, not of schwannoma itself. In fact, in approximately 30%-50% of appendiceal tumors, patients present clinical symptoms and signs of appendicitis [7].

According to Bucher et al., patients treated for an appendiceal tumor have a higher rate of associated colon cancer than the normal population; therefore, the study recommended colonoscopy in patients with incidentally discovered appendiceal tumors [2,8]. Therefore, it is important to recognize whether there is an appendiceal tumor associated with appendicitis.

Typical CT findings of gastrointestinal schwannomas are well-demarcated, round to oval, and homogeneously enhancing masses. On magnetic resonance imaging, schwannomas are usually hypointense and hyperintense on T1- and T2-weighted images, respectively. There are several reports of gastrointestinal schwannomas with increased fluorodeoxyglucose (FDG) uptake on (^{18}F) FDG positron emission tomography, even in appendiceal schwannomas [9].

Differential diagnoses of gastrointestinal schwannomas include GIST, leiomyomas, and NET G1. There are reports that on EUS, gastrointestinal mesenchymal tumors (schwannoma, GIST, and leiomyomas) originate in the fourth layer (muscularis propria), and NET G1 originates from the second to third layer (lamina propria–submucosa) [5,10]. However, we could not distinguish gastrointestinal mesenchymal tumors using radiological images alone.

The final diagnosis was based on histopathological examination. Schwannomas are composed of Schwann cells with compact (Antoni A) and loose cellular areas (Antoni B). Immunohistochemically, tumor cells strongly and diffusely expressed the S-100 protein.

Complete surgical resection is the standard treatment for schwannomas. Besides, the effects of chemotherapy and radiotherapy remain unclear. Lymph node dissection is not recommended for gastrointestinal schwannoma [5,10].

Gastrointestinal schwannomas exhibits good prognosis following complete surgical excision.

Conclusion

Our case is significant in that it adds to another rarely reported case of appendiceal schwannoma. Moreover, it is important to recognize whether there is an appendiceal tumor associated with acute appendicitis.

Patient consent

Consent for publication has been obtained.

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