Case Report

Appendicular schwannoma: Case report of a rare differential of a right iliac fossa mass

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ABSTRACT

Schwannomas are slow growing, mostly benign tumors arising from the Schwann cells of the nerve sheath. They usually arise from the nerves of the head-and-neck region. They are rare in the gastrointestinal tract. The stomach is the most common site for gastrointestinal schwannomas. Appendicular schwannomas are even a rarer entity that may present as a right iliac fossa mass. The final diagnosis is by histopathology and immunohistochemistry. Treatment is essentially surgical. Such entities may present as diagnostic dilemmas presenting as the not-so-common cause of iliac fossa mass and hence deserve reporting and discussion. We report our experience of treating a case of appendicular schwannoma in a 70-year-old lady with a short discussion on its characteristics and management.

Key words: Appendicular mass, Appendicular neuroma, Appendicular schwannoma, Gastrointestinal schwannoma

Schwannomas are slow growing, mostly benign tumors arising from the Schwann cells of the nerve sheath. They usually arise from the nerves of the head-and-neck region. They are rare in the gastrointestinal tract and account for 2–6% of all submucosal tumors of the intestine. Stomach is the most common site for gastrointestinal schwannomas (60–70%) [1]. Schwannoma arising from the appendix is exceedingly rare with a handful of cases reported in the literature [2]. According to recently published articles, as few as only 14 cases have so far been reported in worldwide literature [3].

Considering the rarity of such an entity, and in the interest of discussing the diagnostic and therapeutic dilemmas, we present a case of appendicular schwannoma in a 68-year-old lady presenting with a large iliac fossa mass with a short review of the literature.

CASE REPORT

A 68-year-old lady presented with features of abdominal pain for 4 months. The pain was not well localized and intermittent in nature. It was increasing in frequency over time and was associated with a few episodes of vomiting over the past 2 weeks. There was no complaint of hematochezia, constipation, or loose motion.

The patient was average built, ECOG 2, clinically no pallor or icterus. Examination revealed a palpable lump in the right iliac fossa and mild distension of the abdomen with increased bowel sounds. The lump was firm in consistency, mildly tender on deep palpation with a smooth surface and ill-defined margins.

Blood examination for a complete blood count, urea and electrolytes, liver function test, coagulation profile, serum glucose level, CEA, and CA125 19.9 was all under normal limits. A contrast-enhanced computed tomography of the abdomen revealed a 5 × 7 cm mass arising from the cecum with non-visualization of the appendix (Fig. 1). There was no evidence of lymphadenopathy, omental deposits, or ascites.

With the progressive nature of obstructive symptoms, the patient was taken up for surgery on an emergency basis. Laparoscopic evaluation of the abdominal cavity revealed a large cecal mass. The appendix could not be separately appreciated (Fig. 2). The patient underwent a laparoscopy-assisted right hemicolectomy. The post-operative period was uneventful and the patient was discharged in stable condition on post-operative day 5.

On gross examination, the appendix was replaced by a 7.5 × 4.5 × 3.5 cm mass adhered to the cecum. Sections showed lesional components comprising both hypo and hypercellular areas with cells showing nuclear palisading (Verocay bodies) consisting of narrow, elongated, wavy cells with tapered ends, interspersed with collagen fibers and ill-defined cytoplasm. Mitosis was rare. A total of 47 lymph nodes were retrieved and all were unremarkable. Immunohistochemistry revealed a strong

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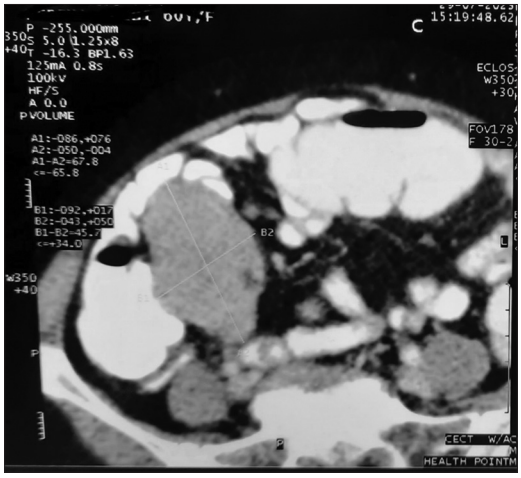

Figure 1: Contrast-enhanced computed tomography of the abdomen revealed a 5 × 7 cm mass arising from the cecum with non-visualization of the appendix

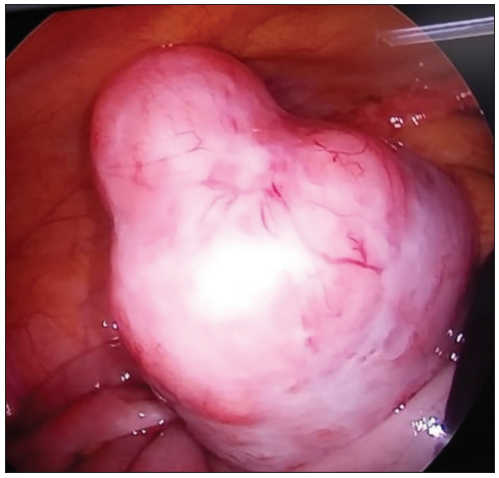

Figure 2: Laparoscopic evaluation of the abdominal cavity revealed a large cecal mass

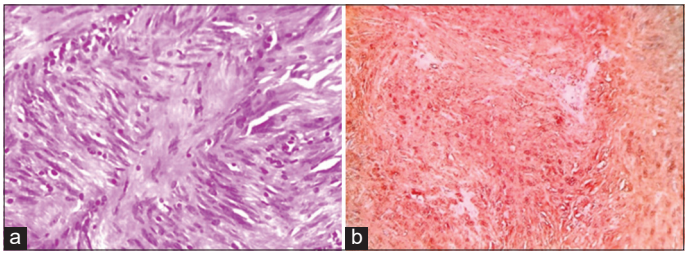

Figure 3: Immunohistochemistry revealed a strong and diffuse (a) S-100 protein staining and (b) negative staining for CD34

and diffuse S-100 (Fig. 3a) protein staining, non-specific DOG 1 uptake, and negative staining for SMA, cytokeratin, and CD34 (Fig. 3b), thus confirming the diagnosis of schwannoma arising from the appendix.

DISCUSSION

Schwannomas are mainly slow-growing benign tumors that originate from Schwann cells arising from the peripheral nerve sheath. Hence, they may originate wherever peripheral nerves are present and are usually found in the nerves in the head, neck, and flexor surfaces of the extremities [4]. Verocay was the first to describe gastrointestinal schwannomas in 1910. He described two histological growth patterns: "Antoni A" and "Antoni B." While Antoni A pattern is characterized by long spindle cells that create a palisading pattern, and when well developed, this palisading feature can create the so-called "Verocay bodies." The Antoni B pattern is characterized by a loose arrangement of cells, with varying degrees of myxoid and hyaline degeneration [5].

Typical computed tomography 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. Differential diagnoses of gastrointestinal schwannomas include GIST, leiomyomas, and NET G1 [6]. Another modality of differentiating these tumors is EUS. 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]. 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]. In our case, since we did not have a prior tissue diagnosis and due to the emergency nature of the procedure, a standard right hemicolectomy was performed.

Schwannomas arising in the appendix are rare and in approximately 30–50% of cases present with the clinical features of appendicitis. In the rest, the symptoms are non-specific, making other differentials of GIST or NETs as differentials [7]. Gastrointestinal schwannomas exhibit a good prognosis following complete surgical excision.

In our review of the literature, we found only a handful of cases with varied presentations. In one of the recent case reports, Jeong reported a similar case of subepithelial cecal mass presenting as acute appendicitis in an 80-year-old lady. Treatment was similar in nature, treated by hemicolectomy with routine follow-up [8]. In earlier reported cases, patients presented with non-specific symptoms with definitive diagnosis made in the histopathology of surgical specimens. Definitive treatment was always surgical [9].

CONCLUSION

We report the case of appendicular schwannoma, presenting as a right iliac fossa mass, with features of obstruction which we managed by laparoscopic-assisted right hemicolectomy. Given

the rarity of such cases, reporting and discussion of this entity remain crucial for considering such entities in the differentials, translating diagnostic findings, and planning treatment.

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