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Myxoid Degenerated Giant Rectal Schwannoma Masquerading as Tailgut Cyst: A Rare Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors contributed to the study conception and design. Material preparation and data collection were performed by authors SLH, KR, BHS, BP and SG. The first draft of the manuscript was written by author SLH and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript

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Case Study

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ABSTRACT

Background: Colorectal schwannomas are rare tumors often detected incidentally and frequently misdiagnosed on preoperative evaluation. They can grow to a large size and create a significant mass effect. A rare case of large rectal Schwannomas mimicking tailgut cyst and managed by a laparoscopic approach is described in the present report.
Case Presentation: A 48- year-old gentleman presented with urinary retention and constipation. Examination revealed an ill-defined hard mass in the left iliac fossa extending to the pelvis. Imaging revealed a 18x12x 9 cm cystic lesion suggestive of a tailgut cyst. The patient underwent Hand-assisted laparoscopic excision of the lesion. The tumor showed spindle cells with myxoid degeneration on microscopic examination and S-100 positivity in immunohistochemistry suggestive of Schwannoma. His postoperative course was uneventful, and there is no evidence of tumor recurrence at eight months of follow-up.

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Conclusion: Rectal Schwannoma with myxoid degeneration can mimic tailgut cyst and pose a diagnostic challenge. The hand-assisted laparoscopic approach is feasible for excision of large rectal schwannomas.

Keywords: Giant rectal schwannoma; myxoid degeneration; tailgut cyst; Hand-assisted laparoscopic excision.

1. INTRODUCTION

Schwannomas are extremely rare tumors of the nerve sheath, developing from Schwann cells, which produce insulating myelin sheath to cover peripheral nerves. In the gastrointestinal tract, they present as spindle cell tumors originating from Auerbach's myenteric plexus. It accounts for approximately 2-6% of all mesenchymal tumors. Gastrointestinal tract schwannoma frequently occurs in the stomach (83%) followed by the small bowel (12%) [1]. The rectum is a rare site for schwannoma. Schwannoma is often detected incidentally, and a definite diagnosis is made on the pathological examination of the surgical specimen and immunohistochemistry. Complete resection is the preferred treatment to reduce recurrence [2]. The open approach was commonly used to treat rectal schwannoma. The present report describes a rare case of myxoid degeneration of Schwannomas mimicking tailgut cyst managed by laparoscopic approach. As myxoid degeneration is a rare pathological feature, radiological and clinical diagnosis becomes challenging. Also, the large size of the tumor poses mechanical constraints during laparoscopic surgery.

2. CASE REPORT

A 48-year-old gentleman initially presented with urinary retention for five months and constipation for two months duration. There was no history of vomiting, abdominal pain, melena, bony pain, breathlessness, or chest pain. The patient was moderately built and nourished with good

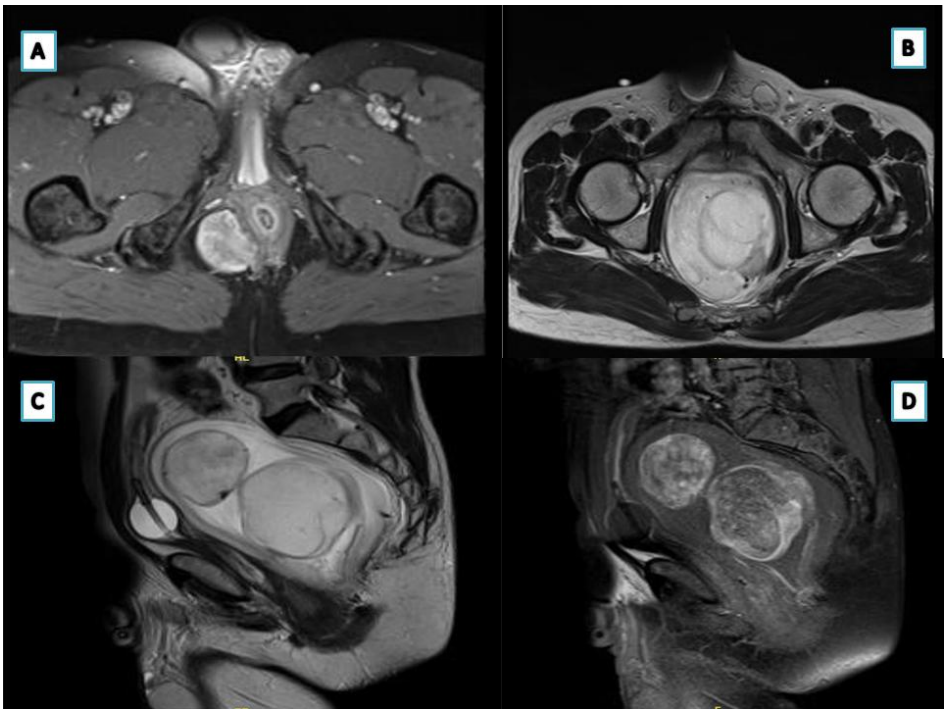

Fig. 1. MRI pelvis: Axial T2 phase showed [A] cystic lesion originating from the posterior lateral part of the rectum. [B] Large cystic lesion compressing the rectum. [C] The sagittal section, T2 phase shows a large cystic lesion with two daughter cysts seen within the cyst, compressing the urinary bladder anteriorly. [D] Sagittal section with contrast shows vascularity inside tumor

performance status. Abdominal examination revealed an ill-defined mass in the left iliac fossa extending into the pelvis. Rectal examination revealed a hard mass extrinsically compressing the rectal wall in the anterior and lateral aspects. Hemogram was within normal limits, and serum carcinoembryonic antigen (CEA) level was not elevated (CEA 1.8 ug/L). MRI pelvis revealed 18x 12x 9 cm lobulated T2 hyperintense and T1 hypointense cystic lesion in the pelvis compressing and displacing the rectum anterolaterally to the left. Anteriorly, the lesion was abutting the prostate and urinary bladder (Fig. 1). A provisional diagnosis of tailgut cyst was made based on the imaging findings, and the patient was planned for laparoscopic excision.

The intraoperative evaluation revealed a large tense lesion occupying the space between bladder and rectum, limiting laparoscopic dissection. Attempted aspiration of the cyst failed due to thick gelatinous content (Fig. 2).

An intraoperative frozen section was sent from the cyst wall, which didn't show any atypical cells suggestive of malignancy. Hence, a hand-assisted laparoscopic cyst excision was performed. The cyst was completely removed in a piecemeal fashion and the lowermost extent of the cyst was noted at the level of the pelvic floor. Microscopic examination revealed a gelatinous area with sheets of benign spindle-shaped cells arranged in short fascicles admixed with focal myxoid change and thick-walled blood vessels. Tumor cells showed diffuse positivity for S100 and negative for CD117. The microscopic and immunohistochemistry findings were suggestive of benign Schwannoma with myxoid degeneration (Fig. 3). The patient is asymptomatic at eight months follow-up with no clinical evidence of recurrence.

3. DISCUSSION

The gastrointestinal autonomic nerve tumors (GANTs) were first described and defined by Herrera et al. in 1984 [3]. Schwannomas belong to GANT, which is a subcategory of gastrointestinal stromal tumor (GIST) [4, 5]. Schwannomas located in the colon and rectum are unrelated to Von Recklinghausen's disease [1]. They can appear in any age group but are most frequently seen in the sixth decade of life [6]. This neurogenic tumor is usually benign and has indolent growth. Although they are generally asymptomatic, they may present with bleeding, obstruction, tenesmus, and rectal pain [1]. In the present report, the patient presented with features of lower urinary tract obstruction and constipation because of a large tumor compressing the bladder neck, and rectum.

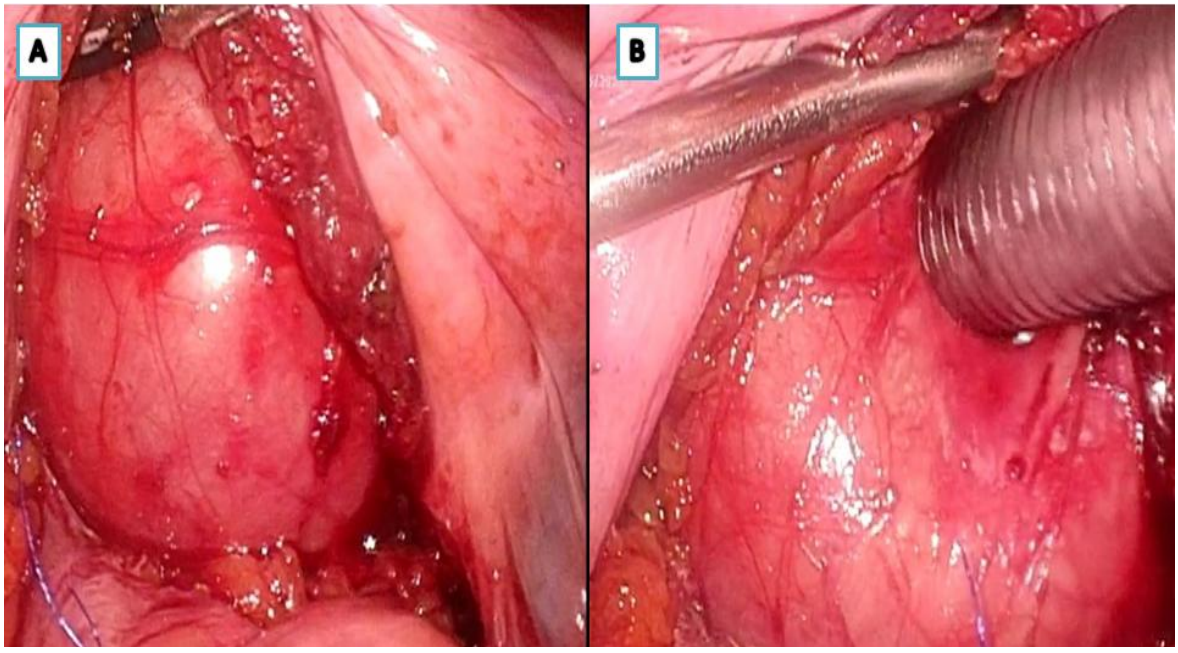

Fig. 2. [A] The laparoscopic intraoperative picture showed a large cystic lesion. [B] Attempting of aspiration revealed thick gelatinous material, which was not amenable to aspiration

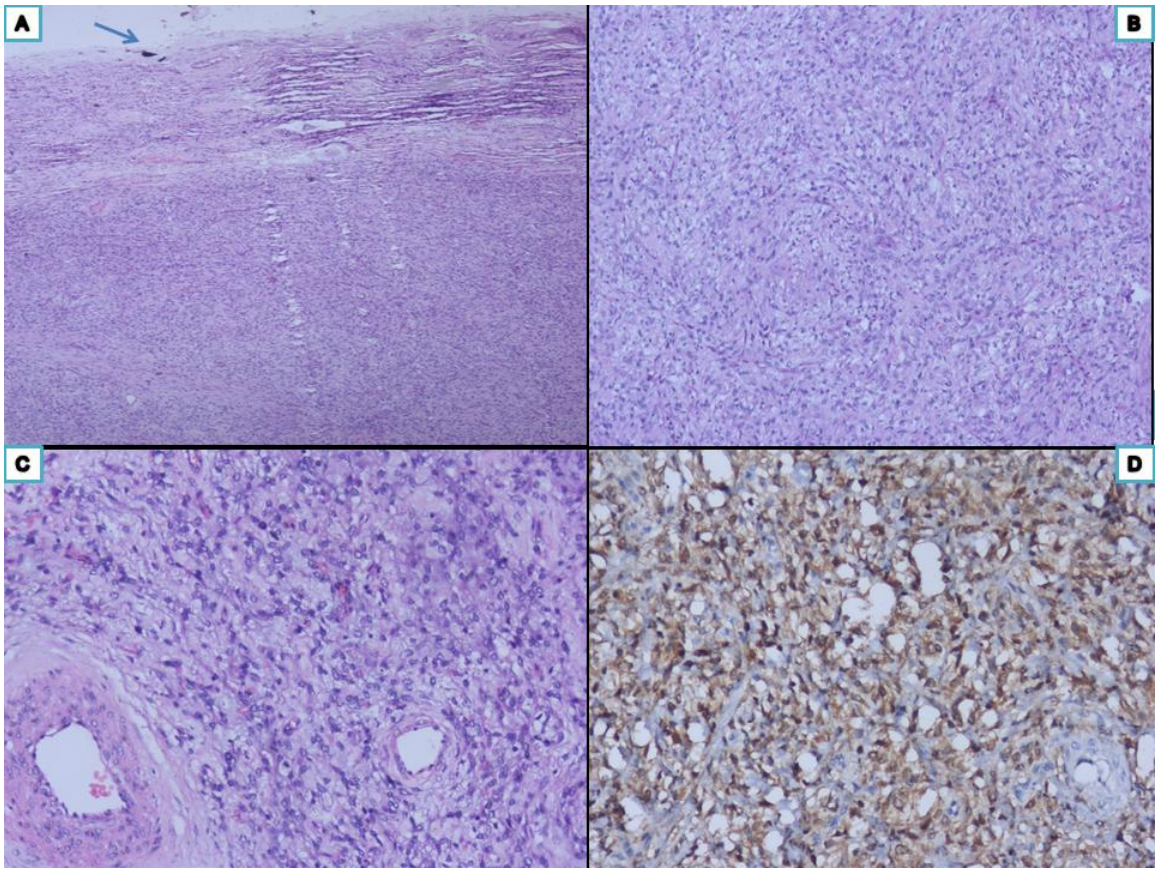

Fig. 3. Histopathological sections show [A] tumor with overlying capsule [arrow] [H&Ex40]. [B] Sheets and fascicles of tumor cells [H&Ex100]. [C] Tumor cells are round to oval in shape with minimal nuclear atypia along with adjacent thick-walled vessels [H&Ex100]. [D] Immunohistochemistry with S 100 shows strong nuclear and cytoplasmic positivity [DABX100]

MR imaging features that suggest the diagnosis of a neurogenic tumor include the presence of a fusiform-shaped mass with an entering and exiting tail representing the host nerve [7]. Typically, the Schwannomas of the gastrointestinal tract are homogenous. However, in our case, the imaging revealed a heterogeneous cystic lesion, so a preoperative diagnosis of a tailgut cyst was made [8]. Furthermore, schwannomas are often diagnosed on histopathological examination of operative specimens and immunohistochemistry, as a preoperative diagnosis is difficult [9]. Macroscopically, schwannomas are well-delimited, solid tumors that range in size from less than 1 cm to a very large tumor up to 28 cm that presents with an increase in abdominal girth [10, 11]. Longstanding tumors may develop degenerative changes such as hemorrhage, calcification and fibrosis, but cystic degeneration is rare [12-14]. Microscopically, schwannomas are encapsulated neoplasms and two histological growth patterns have been described: Antoni A and Antoni B. In Antoni A type, there is a dense growth of fusiform cells, compactly arranged in palisades to form Verocay bodies. In Antoni B, the fusiform cells are more loosely distributed with rounded or elongated nuclei, with a greater quantity of myxoid stroma and xanthomatous histiocytes [6, 9]. Immunohistochemistry plays an essential role in diagnosis with diffuse positive immunostaining for S-100 and Leu7 antigen is suggestive of Schwann cell origin [15]. Myxoid degeneration observed in the present patient is a rare finding in Schwannoma.

Complete surgical resection is the best therapeutic option for Schwannoma. Lymph node dissection is not recommended because of less risk of malignant transformation [4]. The surgical approach varies with the size and location of the tumor. When the tumor is diagnosed preoperatively, the endoscopic approach is feasible for small tumors [16, 17]. Minimally invasive surgery like laparoscopic wedge resection and laparoscopic right hemicolectomy

were reported for colonic schwannomas [18, 19]. However, an open approach is frequently used for large rectal schwannomas. Recently, Feifei et al. reported the feasibility of robot-assisted retro rectal schwannoma excision in 12 patients. However, the largest size of the tumor in that series was 4.6x 3.6x 4.1 cm. [20]. In the present report, an 18x 12 cm tumor resected using the hand-assisted laparoscopic method.

4. CONCLUSION

Rectal Schwannoma with myxoid degeneration can mimic tailgut cyst and pose a diagnostic challenge. Degenerative changes and the rarity of the tumor are important reasons for misdiagnosis in the case of rectal Schwannoma. Immunohistochemistry helps in the definitive diagnosis of Schwannoma in doubtful cases. Surgical resection is the mainstay of treatment for Schwannoma. A hand-assisted laparoscopic method is a feasible approach for the excision of large tumors with a low risk of malignancy.

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The Institute Research Ethics Committee approval was obtained.

CONSENT

Informed consent was obtained from the patient.

The authors affirm that human research participants provided informed consent for the publication of the images.

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COMPETING INTERESTS

The authors have no relevant financial or non-financial interests to disclose. They declare that they have no conflict of interest.

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