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Case report

Appendiceal neuroma presented as acute appendicitis: A rare case report from Syria

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ABSTRACT

Introduction and importance: Appendiceal neuroma, also known as fibrous obliteration, is an exceptional benign lesion discovered in the vermiform appendix removed for indications of acute appendicitis.
Case presentation: We report a case of a 76-year-old woman who presented with signs and symptoms of acute appendicitis. An abdominal ultrasound revealed a mildly distended appendix with a peri-appendiceal fluid collection. Gross examination of the excised appendix showed an obliterated lumen of the distal part on serial cuts. Microscopically, the lumen is replaced by the proliferation of spindle cells into fascicles embedded in fibrous tissue. The diagnosis of appendiceal neuroma was reported and supported by the result of the S-100 immunohistochemical staining.
Clinical discussion: An appendiceal neuroma represents an incidental finding in appendices excised for other reasons. Clinically, there are no specific signs or symptoms, and it is discovered inadvertently. Occasionally, it can mimic acute appendicitis. This lesion is characterized by the proliferation of spindle cells that occlude the lumen of the appendix.
Conclusion: A histopathological evaluation is essential to achieve a definitive diagnosis of an appendiceal neuroma.

1. Introduction

Appendiceal neuroma, also known as neurogenic appendicopathy or neurogenic hyperplasia, is a rare tumor that obstructs the appendiceal lumen due to the proliferation of spindle cells [1]. It is more common in older patients. Appendiceal neuromas are typically seen incidentally by the pathologist during the evaluation of appendectomy specimens [2,3]. In this report, we describe a case of appendiceal neuroma presented with symptoms of acute appendicitis in a woman.

This case report has been reported in line with the SCARE criteria 2020 [4].

2. Case presentation

A 76-year-old woman patient presented to the emergency department at Tishreen University Hospital in 2022 with right lower quadrant abdominal severe pain for 4 days. The physical examination revealed localized tenderness by palpation at the right iliac fossa with evidence of

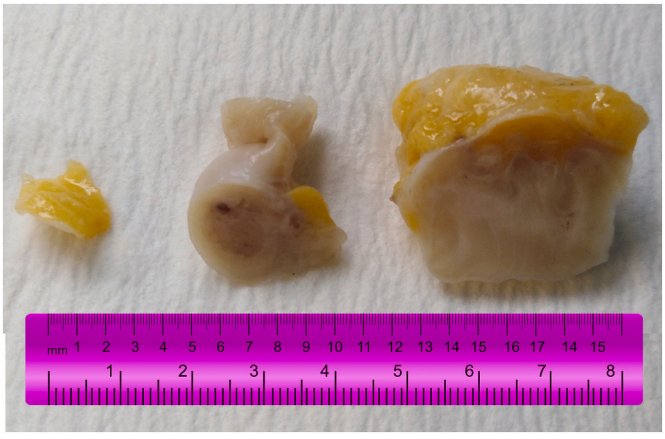

Fig. 1. The gross image of the excised appendix shows the lumen was absent in serial sections.

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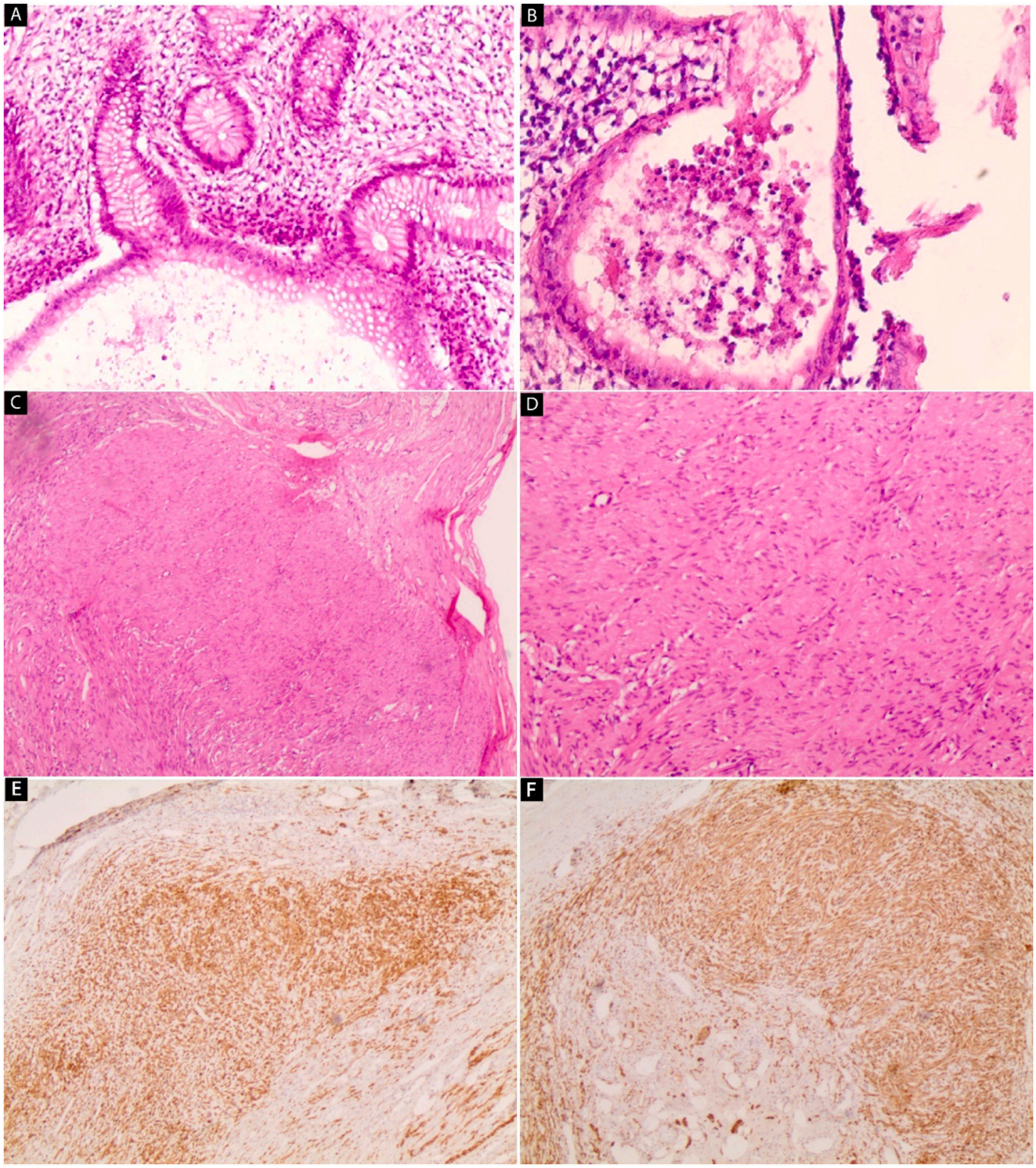

Fig. 2. Microscopic images of the vermiform appendix using H & E stain (A, B, C, and D) and IHC (E and F). (A) Mild crypt hyperplasia at the base (×100). (B) Infiltration of polynuclear cells (×200). (C) The lumen of the tip was obliterated by a proliferation of spindle cells arranged in bundles (×40). (D) The spindle cells have waved nuclei (×100). (E and F) S-100 staining is positive (×100).

rebound. Mild fever was present. The patient was a non-smoker and non-alcoholic. She did not use any specific drugs. She had no history of ganglioneuromatosis, neurofibromatosis, or MEN 2B syndrome. She underwent cholecystectomy. Laboratory tests showed white blood cells (6000 mm^3) with neutrophilia (79 %). Other routine blood values were within normal limits. An abdominal ultrasound showed a dilated vermiform appendix in the right lower fossa with a peri-appendiceal fluid collection. A diagnosis of acute appendicitis was made, and an appendectomy was performed. The specimen was sent to the pathology department. The excised appendix measured 3.6 cm × 1.2 cm × 1 cm. The external surface of the appendix was grey-white with congested blood vessels, and the lumen of the distal part was absent in serial sections (Fig. 1). Microscopic examination of the H&E-stained sections showed mild crypt hyperplasia in the appendix's base (Fig. 2A) with infiltration of polynuclear cells (Fig. 2B). The lumen of the appendix's tip was obliterated by a proliferation of spindle cells with wavy nuclei arranged in bundles (Fig. 2C, D). Most of these cells were positive for S-100 protein (Fig. 2E, F). The final diagnosis was appendiceal neuroma. The patient had a stable recovery, and she was discharged a few hours later.

3. Discussion

Appendiceal neuroma, also referred to as neurogenic hyperplasia, was thoroughly described by Masson over 50 years ago [5]. It is usually found in older individuals but is occasionally seen in young patients [6]. An appendiceal neuroma represents an incidental finding in appendices excised for other reasons [2,3]. The pathogenesis remains unclear. Several studies believe it may result from the proliferation of neuroendocrine cells due to frequent inflammatory attacks [3,7]. The process typically begins at the distal part and may affect only the tip or even the entire vermiform appendix [6]. Clinically, it is usually an incidental and

asymptomatic finding, but it can mimic appendicitis, as in this case [3,7]. The differential diagnosis of stromal tumors of the appendix includes leiomyoma, gastrointestinal stromal tumors, and neurogenic lesions [8]. Histopathologically, there was a proliferation of S100-positive nerve tissue. The process may be confined to the mucosa or may replace the entire lumen [6]. Immunohistochemical staining confirms the diagnosis of appendiceal neuroma, where the neoplasm cells are stained with S-100 protein [1,2,6]. The treatment of choice is surgical excision of the appendix [1,7].

4. Conclusion

An appendiceal neuroma is an exceptional entity that may be clinically presented as appendicitis. It was found incidentally during the examination of appendectomy specimens by the pathologist. Histopathological evaluation remains a reliable method for accurate diagnosis of this benign entity and for differentiating it from the other stromal tumors of the vermiform appendix.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Ethical approval

No ethical approval was needed for this case report.

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CRediT authorship contribution statement

Moatasem Hussein Al-janabi: study design, data collections, data analysis, and writing.
Shady Hasan: performed surgery.
Rana Issa: in reviewing the manuscript.

Declaration of competing interest

The authors have no conflicts of interest to declare.

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