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Case report

Fibrous obliteration of the appendix mimicking acute appendicitis: a case report.

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Abstract

Background:

Fibrous obliteration of the appendix, also termed appendiceal neuroma, neurogenic appendicopathy, or neurogenic hyperplasia, is an uncommon benign lesion characterized by replacement of the appendiceal lumen by spindle-cell proliferation, fibrosis, and neural-type tissue. It is usually detected incidentally in appendicectomy specimens but may clinically and radiologically mimic acute appendicitis. The objective of this case report is to describe a rare case of fibrous obliteration of the appendix presenting as suspected acute appendicitis and to highlight the diagnostic value of histopathology and immunohistochemistry.

Case presentation:

A 40-year-old man presented with severe right lower quadrant abdominal pain for three days, localized right iliac fossa tenderness, rebound tenderness, mild fever, and neutrophilic leukocytosis. Ultrasonography suggested an inflamed appendix, while computed tomography showed a complex, thick-walled collection with air foci inferior to the caecum and periappendicular inflammatory changes, raising suspicion of appendicular abscess. Laparoscopic appendicectomy was performed. Gross examination revealed a grey-white appendix measuring 3.5 × 1.2 × 1 cm with congested vessels and an obliterated grey-white lumen. Microscopy showed luminal obliteration by spindle cells with wavy nuclei arranged in bundles, associated fibrosis, and inflammatory infiltrate predominantly composed of lymphocytes and eosinophils. S-100 immunostaining was positive in the spindle cells, confirming appendiceal neuroma/fibrous obliteration.

Conclusion:

Fibrous obliteration of the appendix is a rare benign lesion that can closely simulate acute appendicitis. Routine histopathological examination of appendicectomy specimens, supported by S-100 immunostaining when required, is essential for definitive diagnosis and exclusion of other appendiceal stromal or neurogenic lesions.

Take-away lessons:

Clinicians and pathologists should consider fibrous obliteration in appendicitis-like presentations with an obliterated lumen. Histopathology remains decisive, and immunohistochemistry helps avoid misclassification of this benign entity.

Keywords: *Appendiceal neuroma; Appendicitis; Fibrous obliteration; Neurogenic appendicopathy; Neurogenic hyperplasia; S-100 immunostaining*

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Introduction

Fibrous obliteration of the appendix, also termed appendiceal neuroma, neurogenic appendicopathy, or neurogenic hyperplasia, is a benign lesion in which the appendiceal lumen is progressively occluded by spindle-cell proliferation, fibrosis, and neural-type tissue [1]. Although it is often detected incidentally during histopathological evaluation of appendicectomy specimens, it can occasionally produce symptoms that are indistinguishable from acute appendicitis [2,3].

The lesion was initially described by Masson in 1928 in relation to nerve hyperplasia of the appendicular mucosa [4]. The exact pathogenesis remains uncertain. Repeated low-grade or subclinical inflammatory episodes have been proposed to induce neuroendocrine-cell hyperplasia and neural proliferation in the lamina propria and submucosa, eventually resulting in fibrosis and luminal obliteration [4,5]. Because preoperative clinical and radiological findings may overlap with acute appendicitis, histopathological evaluation is required for a reliable diagnosis.

The objective of this case report is to describe a rare case of fibrous obliteration of the appendix presenting with clinical and radiological features suggestive of acute appendicitis, and to emphasize the role of histopathological examination and S-100 immunohistochemistry in establishing the definitive diagnosis.

Case presentation

A 40-year-old man presented to the emergency department of NRI Institute of Medical Sciences in January 2024 with severe right lower quadrant abdominal pain for three days. The pain was continuous, started after an episode of alcohol intake, aggravated on walking, and was relieved in the supine position. On physical examination, localized tenderness was present in the right iliac fossa with rebound tenderness. Mild fever was also noted.

The patient did not report chronic smoking, regular alcohol intake, or use of specific long-term medication. Complete blood count showed mild neutrophilic leukocytosis, with a leukocyte count of 12,300 cells/cumm. Other routine investigations were within normal limits. Ultrasonography showed an inflamed appendix with a thickened periappendicular wall. Computed tomography of the abdomen revealed an irregular, complex, thick-walled collection containing air foci inferior to the caecum, with surrounding fat stranding and a few small-volume lymph nodes, suggesting an appendicular abscess.

Laparoscopic appendicectomy was performed with an intraoperative impression of acute appendicitis. The specimen was submitted to the Department of Pathology for histopathological evaluation. The excised appendix measured 3.5 x 1.2 x 1 cm. Grossly, it was grey-white with congested blood vessels. The cut surface showed an obliterated lumen with grey-white areas.

Microscopic examination of hematoxylin and eosin-stained sections showed obliteration of the appendiceal lumen by a proliferation of spindle cells with wavy nuclei arranged in bundles. The appendiceal wall showed inflammatory-cell infiltration, predominantly lymphocytes and eosinophils, with areas of fibrosis. The serosal surface was unremarkable. Immunohistochemistry showed S-100 positivity in most spindle cells, confirming the diagnosis of appendiceal neuroma/fibrous obliteration of the appendix.

Follow-up and postoperative condition

The postoperative period was uneventful. The patient had a stable recovery and was discharged in good clinical condition. Follow-up for six months did not reveal any postoperative complications or recurrence of symptoms. Fibrous obliteration of the appendix, also termed appendiceal neuroma, neurogenic appendicopathy, or neurogenic hyperplasia, is an uncommon benign lesion characterized by replacement of the appendiceal lumen by spindle-cell proliferation, fibrosis, and neural-type tissue. It is usually detected incidentally in appendicectomy specimens but may clinically and radiologically mimic acute appendicitis. The objective of this case report is to describe a rare case of fibrous obliteration of the appendix presenting as suspected acute appendicitis and to highlight the diagnostic value of histopathology and immunohistochemistry.

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Figures

Figure 1: Gross appearance of the appendix showing a grey-white specimen measuring approximately 3.5 cm in length.

Figure 2: Cut section showing grey-white areas with obliteration/absence of the appendiceal lumen.

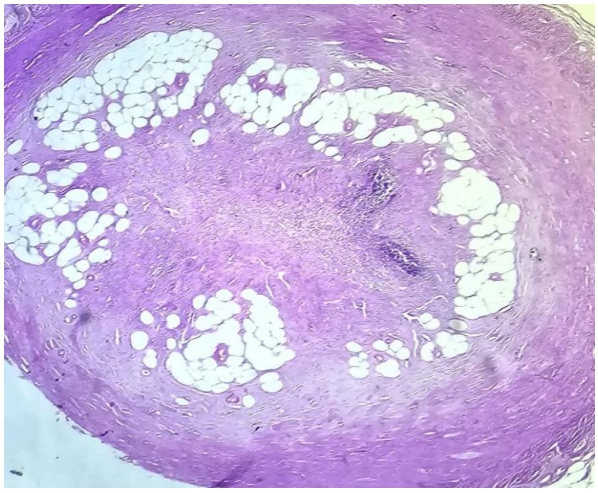

Figure 3: H&E-stained section at 4x magnification showing appendiceal lumen largely replaced by fibrous tissue, with adjacent adipose tissue.

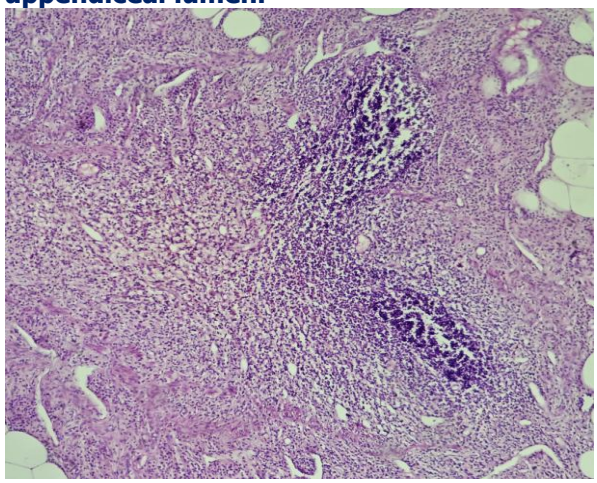

Figure 4: H&E-stained section at 10x magnification showing obliterated lumen with fibroblastic/spindle-cell proliferation and inflammatory infiltrate composed predominantly of lymphocytes and eosinophils.

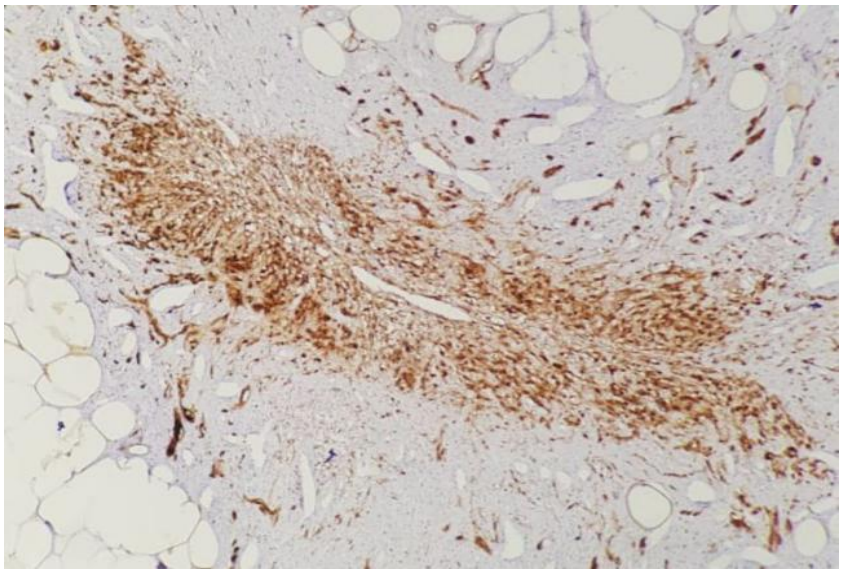

Figure 5: S-100 immunostain at 40x magnification showing positivity in spindle cells, supporting appendiceal neuroma/fibrous obliteration.

Discussion

Fibrous obliteration of the appendix is a rare benign condition that may be encountered during routine examination of appendicectomy specimens. It has also been described as neurogenic hyperplasia or appendiceal neuroma [4]. The lesion is more commonly reported in older individuals, although cases in younger patients have also been documented [6].

The proposed mechanism involves repeated subclinical inflammatory episodes that promote neuroendocrine and neural proliferation within the appendiceal wall. Electron microscopic studies have identified cells containing secretory granules, including serotonin and somatostatin, and immunohistochemical studies have shown reactivity with S-100 protein and neuron-specific enolase [2]. Schwann-cell proliferation, fibrosis, adipose tissue, connective tissue, and eosinophilic infiltrates may also be present [2].

The process usually begins at the distal appendix and may involve the appendiceal tip or the entire vermiform appendix [6]. Fibrosis is considered an end-stage feature of the process. Because luminal occlusion and recurrent low-grade inflammation can produce right iliac fossa pain, many patients present clinically with appendicitis-like symptoms [1]. Vomiting, diarrhea, recurrent abdominal pain, and repeated attacks resembling acute appendicitis have been described in some cases [7,8,9].

Preoperative differentiation from acute appendicitis is difficult because clinical examination, laboratory findings, ultrasonography, and computed tomography may all suggest inflammatory appendiceal disease. In the present case, the patient had right iliac fossa pain, rebound tenderness, neutrophilic leukocytosis, and radiological evidence of appendiceal inflammation, leading to laparoscopic appendicectomy. The final diagnosis was established only after microscopic examination and S-100 immunohistochemical confirmation.

Three histological patterns of neurogenic appendicopathy have been described: obliteration of the lumen, mucosal hyperplasia, and submucosal hyperplasia. The luminal obliterative pattern, also known as appendiceal neuroma, consists of fusiform or spindle cells in a myxoid to fibrous background, accompanied by connective tissue, fatty tissue, and eosinophils. Positivity for S-100 protein and neuron-specific enolase supports neural differentiation [10].

Important differential diagnoses include leiomyoma, gastrointestinal stromal tumor, schwannoma, perineurioma, ganglioneuromatosis, mucosal neuroma associated with multiple endocrine neoplasia type 2B, well-differentiated neuroendocrine tumor, and neurofibroma associated with von Recklinghausen disease [2,10]. Careful histomorphological assessment and appropriate immunohistochemistry are therefore required to avoid misclassification.

Conclusion

Fibrous obliteration of the appendix is an uncommon benign entity that can clinically and radiologically mimic acute appendicitis. Histopathological examination of appendicectomy specimens remains the definitive method for diagnosis. S-100 immunostaining is useful in confirming neural differentiation and in distinguishing this lesion from other appendiceal stromal tumors.

Take-away lessons

Fibrous obliteration of the appendix is a rare benign entity that may present with clinical, laboratory, and radiological features similar to acute appendicitis. Preoperative diagnosis is difficult because imaging findings may suggest inflammatory appendiceal disease or appendicular abscess. Routine histopathological examination of all appendicectomy specimens is essential for identifying this uncommon lesion. S-100 immunostaining is useful for confirming neural differentiation and excluding other appendiceal spindle-cell or stromal lesions.

Limitations

This report describes a single case; therefore, the findings cannot be generalized to all patients with appendicitis-like presentations. Preoperative diagnosis was not possible because clinical and radiological findings were nonspecific. A broader immunohistochemical panel could not be performed, although S-100 positivity supported the diagnosis. Long-term follow-up was limited to six months.

Recommendations

All appendicectomy specimens should undergo routine histopathological evaluation, even when the clinical diagnosis appears to be straightforward acute appendicitis. When spindle-cell proliferation with luminal obliteration is identified, immunohistochemistry should be considered to support the diagnosis and exclude clinically significant mimics.

Abbreviations

CBC - Complete blood count
CT - Computed tomography
FO - Fibrous obliteration
H&E - Hematoxylin and eosin
IHC - Immunohistochemistry
RIF - Right iliac fossa

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Conflict of Interest

The authors declare no conflict of interest.

Availability of Data

Data are available from the corresponding author on reasonable request.

Author contribution

Dr. P. Sandya Rani Guruvelli contributed to case documentation, histopathological description, literature review, and manuscript drafting. Dr. Sravanthi Gurugubelli contributed to diagnostic interpretation, literature review, and manuscript revision. Dr. Vattikuti Satya Veni and Dr. Kaki Hari Priya contributed to data organization, figure preparation, and review of the manuscript. Dr. Atla Bhagya Lakshmi supervised the work, reviewed the diagnosis, provided academic guidance, and critically revised the manuscript. All authors read and approved the final version of the manuscript and agree to be accountable for the integrity of the work.

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