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FIBROUS OBLITERATION OF APPENDIX: A MIMICKER OF APPENDICITIS- CASE SERIES

Pathology

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ABSTRACT

Acute appendicitis is the most common appendiceal disease leading to the surgical excision of the organ. We report four cases of fibrous obliteration of appendix / appendiceal neuroma presented as acute appendicitis on ultrasonography (USG). Due to similar clinical symptoms and USG findings, fibrous obliteration of the appendix can easily be mistaken as acute appendicitis.

KEYWORDS

Appendicitis, Fibrous obliterans, Ultrasonography, Appendix, Abdominal pain.

INTRODUCTION

Fibrous obliteration has been considered a neuroma, which is known as precursor for carcinoid; but neuroma is identified as one of the cause of appendiceal fibrous obliteration.[1] The first case was described by Pierre Masson in 1928.[2] The pathogenesis of this remains unknown. But it is believed to be a part of the aging process. It begins in the distal portion and extends proximally, resulting in the loss of normal appendiceal mucosa and Peyer patches, and finally replaces the mucosa and submucosa by fibrous tissue.[1,3,4] This condition mimics acute appendicitis. Fibrous obliterans is uncommon to clinicians and radiologists and often underdiagnosed clinically. Histopatholoical evaluation of appendix plays a very important role in diagnosis of this condition. Here we report four cases of fibrous obliteration of the appendix received over a period of one month.

CASE REPORTS

Case 1

A 38-year-old male with abdominal distension and pain for 2 week came to surgery department. On physical examination, patient's abdomen had tenderness in right lower quadrant and was mildly distended. Laboratory investigations were done including complete blood count (CBC) and C-reactive protein which came to be normal. Ultrasonography showed inflamed appendix. He underwent laparoscopic appendicectomy. Intraoperatively, the appendix appeared to be inflamed. There were no adhesions noted among the bowel. The surgeon diagnosed the case as acute appendicitis. This appendix was sent to Department of Pathology for histopathological evaluation. Postoperative period of patient was unremarkable.

Grossly, the appendix was grey white in color measuring 4cm in length and 1 cm in width diameter. Cut surface showed obliterated lumen with white material (**Figure 1a**).

On microscopy, the appendix was observed to lack normal appendiceal mucosa and lymphoid follicles. Lumen was displaced by fat infiltration, spindle cells in loose fibrous stroma with chronic inflammatory cells predominantly lymphocytes and eosiniphils. Hypertrophic nerve bundles also seen.(**Figure 1b).**

Case 2

A 27-year-old female came to outpatient department of surgery, with complaint of abdominal pain for 1 month on and off, which increased in last 5 days. She also had complaints of vomiting, diarrhoea and poor appetite for one month. On physical examination, patient had pain which was localized to right lower quadrant of the abdomen. Patient was afebrile. The full blood count and other laboratory investigations were done which were normal. Abdominal ultrasonography showed distended appendix and no fluid collection in the peritoneal and pelvic cavities. Patient underwent laparoscopic appendicectomy and resected appendix was sent to Pathology department for histopathological examination. Postoperative period was unremarkable.

Grossly, the appendix was dilated , measuring 4 cm in length and 0.8 cm in width diameter. External surface was grey in color. On cutting lumen was obliterated. (**Figure 2a**)

On microscopy**,** mucosal lining of appendix was diminished. It was replaced by fibrosis. The fibrotic area had neural differentiated cells showing wavy-spindle nuclei (**Figure 2b,2c**).

Case 3

A 42-year-old female visited emergency department with complaints of abdominal pain, generalized malaise, vomiting and anorexia for past seven days. The pain increased suddenly in last 2 days. On physical examination she had localized tenderness in right lower quadrant of the abdomen. Patient had mild fever for 2 days. There was no significant past and family history. Patient was not taking any medicine. Patient was admitted under surgery department and underwent routine laboratory and radiological investigations. Laboratory investigations showed mild increase in total leukocyte count (TLC). Abdominal ultrasonography showed inflamed appendix. No other systemic disease was noted. Patient underwent laproscopic appendecectomy and surgeon diagnosed this case as appendicitis. This appendix was then sent to Pathology department for histopathological examination. Postoperative period of patient was unremarkable.

Grossly appendix measuring 3 cm in length and 0.3 cm in width diameter and mesoappendix measured 0.5 cm. External surface was grey in color. Cut section showed obliterated lumen with whitish material(**Figure 3a**).

On microscopy wall of appendix and the mucosa was flattened at places, with proliferative spindle cells arranged in fascicles. The cells were elongated, spindle with wavy nuclei (**Figure 3b**).

Case 4

A 38-year-old female came to emergency with complaints of abdominal pain, vomiting on and off and decrease appetite since one week. On physical examination pain was localised to right lower quadrant of abdomen and there was mild distension of abdomen. Routine laboratory investigations including blood count and renal function tests were normal. Abdominal ultrasonography was suggestive of subacute appendicitis. Patient underwent laparoscopic appendicectomy and intraoperatively, appendix was inflamed. Appendix was resected and sent to Pathology department for histopathological examination. Postoperative period of patient was unremarkable.

Grossly, appendix was mildly dilated and measuring 3 cm in length and 0.5 cm in width diameter. External surface was white in colour. Cut surface showed obliterated lumen (**Figure 4a**).

On microscopy, mucosa of appendix was flattened and showed cells which had elongated spindle, wavy-nuclei with scant cytoplasm. Areas of fibrosis were noted. Serosal surface was unremarkable (**Figure 4b**).

All four cases were reactive towards S-100 immunostaining (**Figure 5**).

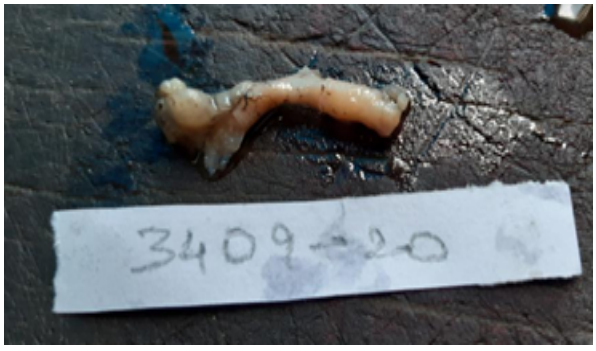

Figure 1: (a) On gross appendix, grey white in colour and measuring 4 cm in length.

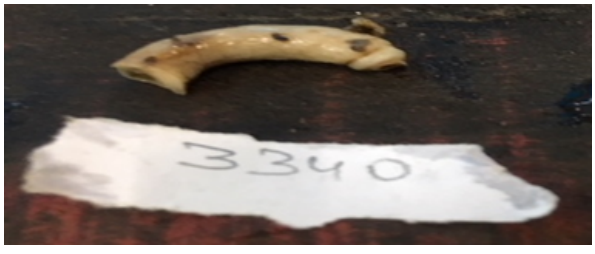

Figure 2:(a) On gross appendix, grey in colour and measuring 4 cm in length.

Figure 3: (a) On gross appendix, grey white in colour, measuring 3 cm in length.

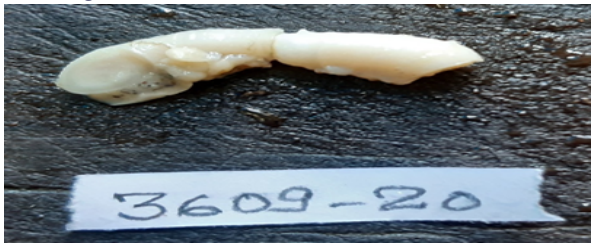

Figure 4: (a) On gross appendix white in colour, measuring 3 cm in length.

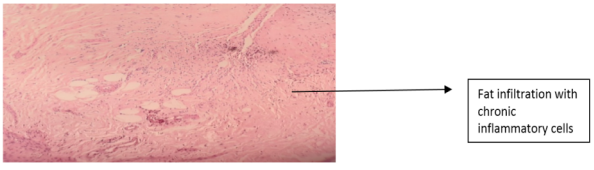

Figure 1: (b) Photomicrograph of appendix showing fat infiltration and fibrosis of appendiceal mucosa (H&E X100).

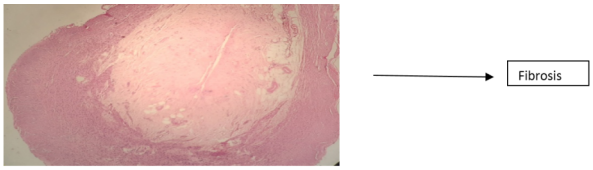

Figure 2:(b) Photomicrograph of appendix showing fibrosis of lumen (H&E X40).

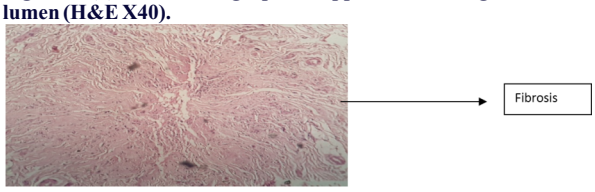

Figure 2: (c) Photomicrograph of appendix showing fibrosis of lumen (H&E X100).

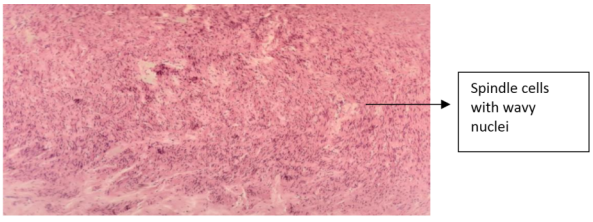

Figure 3: (b) Photomicrograph of appendix showing spindle cells with wavy nuclei and scant cytoplasm (H&E X100).

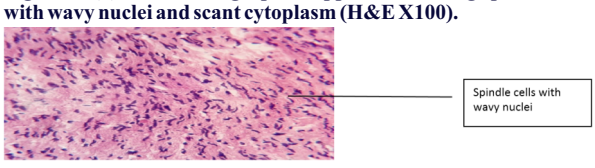

Figure 4: (b) Photomicrograph of appendix showing spindle cells with wavy nuclei and scant cytoplasm (H&E X400).

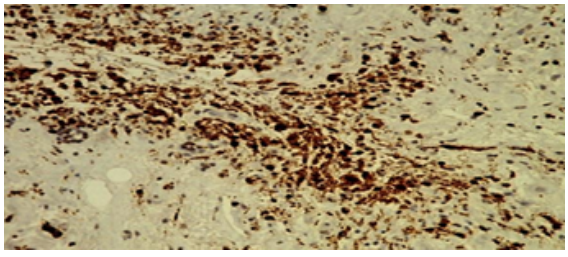

Figure 5: Photomicrograph of appendix showing positivity for S-100 protein (IHC X400).

DISCUSSION

Fibrous obliteration of appendix is a proliferative lesion and not a tumor. It was first described by Masson in 1928, he studied 1200 resected specimen of appendix. Some of these specimen were diagnosed as fibrous obliteration. These diagnosed cases when examined under electron microscopy, were found to have serotonin and somatostanin secretory granules in their cytoplasm. These granules are positive for S-100 and neuron specific enolase. Two types of cells were noticed which were making bundles of cells i.e. the cells with neurosecretory granules and Schwann cells.

Pathogenesis of Fibrous obliteration is unknown. Various studies stated that it is secondary to inflammation, which give rise to neuro-endocrine cells hyperplasia. It is frequently accompanied with fat and connective tissue and infiltration of eosinophils.[5]

Its incidence is high, and in literature it was found to be 51.2%.[6]

The presence of fibrosis is considered as a characteristic of end stage in this process. Because of the repeated, minimal and subclinical attacks of inflammation during this process, it triggers the lesion, with symptoms mimiking appendicitis.[7,8] Occlusion of distal end by fibrosis due to neuronal hyperplasia may eventually lead to carcinoid tumor.[3]

Some carcinoid adenocarcinoma cells also express similar serotonin secretary granules. Thus the possibility of neuroma transforming into neuroendocrine tumor can be postulate.

A study done by Ziari k et al[9] showed that after examining 2752 cases of appendicectomy, 157 cases were diagnosed with pathological conditions. Fibrous obliteration were seen in 80.2% of these 157 cases. Another study done by Yilmaz et al[10] on 338 living donor of liver transplant, showed that nine of them had fibrous obliteration of appendix.

Many cases of fibrous obliteration mimics appendicitis like symptoms, but some remains silent. Clinically patients present with abdominal pain, vomiting, diarrhoea and repeated attack of acute appendicitis. In literature it is found to be more in males then females, and in adolescents then adults.[11,12] Clinical history, physical examination and complementary examination are not able to preoperatively differentiate fibrous obliteration of appendix from acute appendicitis. Some authors stated in their studies that it should be suspected in patients with recurrent pain in right iliac fossa (RIF), this situation is similar to our cases.[13]

On gross, mostly the lumen of appendix is obliterated, but in intramucosal lesions the lumen remains patent. There are three microscopic histological patterns of neurogenous hyperplasia of appendix: first is obliteration of lumen (also known as appendiceal neuroma), second is mucosal hyperplasia and third is submucosal hyperplasia. Appendiceal neuroma consist of a proliferation of fusiform cells in a myxoid background comprising of connective tissue, fatty tissue and eosinophils. These fusiform cells are positive for S-100 protein and neuron-specific enolase.[14,15,16,17]

There are various condition which causes neurogenic appendicopathy like Schwannoma, Perineuroma, Ganglineuromatosis, Mucosal neuroma of MEN 2B syndrome, well differentiated neuroendocrine tumor, Neurofibroma of Von Recklinghausens disease which we should carefully look for.[13,18]

Fibrous obliteration as already discussed is considered as precursor to carcinoid. It shows immunoreactivity to S-100 protein and Neuron specific enolase.[8,18]

The treatment of this condition is surgical resection of appendix by either open or laparoscopic procedure.

CONCLUSION

Although fibrous obliteration of appendix is frequently encountered by pathologist, it is an uncommon entity among clinicians and radiologists. Its imaging features and the clinical features described in these case reports would be helpful in distinguishing fibrous obliteration from acute appendicitis and avoiding emergency operations of patients. Diagnosis of this lesion requires histopathological examination of an appendicectomy specimen.

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CONFLICT OF INTEREST

There are no conflicts of interest.

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