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PATIENT CARE

Pristine Normal Looking Appendix on Diagnostic Laparoscopy-A Management Dilemma

Kasturi Rangan Sarathy* *and Abdus Samee*

Pennine Acute NHS Trust, United Kingdom

***Corresponding author:** *Kasturi Rangan Sarathy, Pennine Acute NHS Trust, United Kingdom*

Introduction

Abdominal pain especially in the right iliac fossa (RIF) or right lower quadrant (RLQ) is one of the most common presentations as surgical emergencies [1]. The cause for right lower quadrant abdominal pain encompasses a wide spectrum of diseases from a functional pain to an acute emergency where an immediate surgical intervention is definitely warranted. Acute appendicitis is the most common surgical emergency globally [2]. The clinical presentation of appendicitis is right lower abdominal pain and associated anorexia and fever. However, all patients do not have these classical symptoms. The diagnosis is sometimes guided with use of adjunct imaging, clinical scoring systems and rise in inflammatory markers in blood investigations. The sensitivity and specificity of these tests is not 100% in diagnosing appendicitis. The decision making to offer a surgery and appendicectomy is sometimes completely based on clinical judgement. During the surgery, the operating surgeon would be left in a dilemma if he encounters a normal looking appendix whether to proceed with an appendicectomy and no other pathology could be identified for patient's symptoms.

What Is Appendicitis Fibrous Obliterans?

Appendicitis fibrous obliterans is one of the rarely encountered histological diagnoses after an appendicectomy. There are many names to describe this condition. They are neurogenic appendicitis, appendiceal neuroma, neuronal hyperplasia of the appendix and neurogenic appendicopathy [3]. The diagnosis was made on the histology on the appendix specimen which surgeons thought to have performed a negative appendicectomy.

What is the Incidence of Appendicitis Fibrous Obliterans?

The incidence of appendicitis obliterans ranges between 0.16% to 0.81% in adult population as per different studies [4,5] while incidence in paediatric population is about 0.25%. The disorder is more common in second decade of life rather than first decade and is more common in female population [6]. A study showed that on reviewing 128 specimen of appendix which was considered grossly normal, four specimens showed the presence of appendicitis obliterans [7].

Why Are There Different Names to Suggest the Same Condition?

The reason for the other names is that appendicitis obliterans is considered the final stage where the entire lumen of appendix is completely obliterated. The precursor neuronal hyperplasia takes place within the lumen eventually leading to complete obliteration. In view of the same, the names are interchangeably used through-out this paper.

What is the Clinical Presentation?

The abdominal pain of appendicitis fibrous obliterans is very similar to acute appendicitis. However, other systemic symptoms like fever, anorexia might be variable. History and clinical examination do not make it possible to distinguish appendicitis fibrous obliterans with acute appendicitis [8]. Though the diagnosis of appendicitis fibrous obliterans is entirely incidental, one

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of the study mentions that Computerized Tomography scans (CT scan) identifies similar finding of intra-luminal linear enhancing structure and fatty accumulation of appendix in two of the patients who had appendicitis fibrous obliterans on histology. The CT finding is however not specific in diagnosing appendicitis fibrous obliterans [9].

What is the Histological Picture of Appendicitis Fibrous Obliterans?

The appendicular lumen is replaced by spindle cells in fibromyxoid background interspersed with chronic inflammatory cells, neuro-endocrine cells, hypertrophied nerve bundles, adipose tissue and collagen. There is loss of normal lymphoid follicles, mucosa and crypts [3].

How Does the Progression of Disease Occur?

The initial stage begins with intra-mucosal neural tissue hyperplasia often with co-existent submucosal and muscular nerve growth [10]. Mast cells and other inflammatory cells were found closely associated with neural tissue in both mucosa and submucosa [11]. The final progression to fibrotic stage occurs where the lumen of the appendix becomes eventually obliterated. The lesion is believed to be because of repeated, minimal, subclinical inflammatory attacks during this process and the symptoms can mimic appendicitis.

What is the Cause of Patient's Symptoms?

Neurogenic appendicopathy involves proliferation of nerve fibres. This proliferation along with increase in neuropeptides such as vasoactive intestinal peptide and Substance P may mimic symptoms of acute appendicitis [12]. The nerve fibres of appendiceal neuromas (fibrous obliteration of the appendix) harbor serotonin and substance P may promote muscular spasticity and abnormal peristalsis leading to abdominal pain in some patients [13].

How Should the Patients Be Managed with Normal Looking Appendix and Persisting Symptoms on Diagnostic Laparoscopy?

The patients whose symptoms are consistent with appendicitis but lack evidence of inflammation during the surgery must be subjected to appendicectomy. As per a systematic review of literature, a study concluded that neurogenic appendicopathy is defined on basis of presence of three criteria. They are: 1) Clinical presentation similar to acute appendicitis; 2) Absence of acute inflammation of histology; 3) Presence of spindle or Schwann cells on histochemistry. The study concludes that laparoscopic appendicectomy is safe and successful treatment option in this scenario [14].

Discussion

Although fibrous obliteration of the appendix is seen in young population, it usually occurs as a part of the aging process. It results in mucosa and submucosa of the appendix replaced by fibrous tissue and thereby loss of Peyer's patches and normal mucosa of the appendix [15]. The lesion on light microscopy is seen infiltrated with eosinophils, entrapped fat and connective tissue [16].

Proliferating nerve fibres, secretion of neuropeptides and eventual obliteration of appendicular lumen results in abdominal pain. The on-going mild inflammation during this process would result in lack of other symptoms of acute appendicitis. As the pathology occurs in the mucosal and sub-mucosal layers of appendix with the final progression to obliteration of the appendicular lumen, the external appearance during a laparoscopy procedure would be grossly normal. There are no currently available investigations to rule out appendicitis fibrous obliterans. In our opinion, it would be a worthwhile effort to do an appendicectomy on a patient with persisting symptoms.

A study that identified an incidence of 0.16% of neurogenic appendicopathy followed up all patients for a mean period of 73.2 ± 28 months concluded that all patients who underwent appendicectomy remained asymptomatic [5].

Case Resolution

Our patient was a twenty-five year-old lady who presented with classical clinical features of appendicitis. The blood investigations were essentially within normal range. The patient was initially admitted for observation as the clinical signs were not suggestive for acute appendicitis. The patient continued to have symptoms and hence was taken into theatre for diagnostic laparoscopy and an appendicectomy. Diagnostic laparoscopy failed to identify the cause of patient's symptoms. Patient underwent an appendicectomy and became symptom free thereafter. The specimen was sent for a histological examination by naming it as normal looking appendix.

The histological examination was reported as appendicitis fibrous obliterans. The patient was followed up and continued to remain asymptomatic following the surgery.

Education into Practice

Consider if a patient presented with right lower abdominal pain with a clinical suspicion of appendicitis and normal blood investigations, radiological imaging and grossly normal appearing appendix on laparoscopy, it would still be worthwhile doing an appendicectomy if no other causes of patients symptoms were identified.

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