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Case Report

Xanthogranulomatous Appendicitis - An Unusual Presentation

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Abstract

Xanthogranulomatous inflammation is an uncommon entity. It has been described in various organs particularly in kidney and gall bladder, but it is rarely reported in appendix.

The present study report a case of Xanthogranulomatous appendicitis found incidentally in an appendicular lump in interval appendicectomy. The patient was admitted for three times. Intra operatively, there was presence of an ileocaecal lump with multiple mesenteric lymph nodes and serositis. As caecal cancer cannot be excluded, right hemicolectomy was performed. Microscopically, there was presence of dense chronic inflammatory infiltrate in the submucosa, muscularis propria and serosa of appendix comprising of lymphocytes, plasma cells and sheets of foamy histiocytes and few foreign body giant cells. Histiocytic nature of foamy cells was confirmed cytochemically and immunohistochemically.

It is important to diagnose Xanthogranulomatous appendicitis, as it might be associated to intestinal inflammatory diseases or may mimic a locally advanced cancer but has a surgical cure.

Keywords: Xanthogranulomatous; Appendicitis; Xanthoma Cells; CD 68

Introduction

Xanthogranulomatous inflammation is an uncommon entity. It has been described in various organs particularly in kidney [1] and gall bladder [2] but it is rarely reported in appendix. It can masquerade other diseases like cancer which may cause patient to undergo radical surgery [3]. The present study report a case of Xanthogranulomatous appendicitis found incidentally in an appendicular lump in interval appendectomy. The patient followed a chronic course with three times admission and showed that it is not possible to monitor it on a conservative treatment. XG responses may be associated with long-standing inflammation and mass formation. Also, it is a rare form of chronic appendicitis which may mimic other fulminant diseases, so before doing a radical surgery this differential diagnosis must be kept in mind. So we are reporting this case due to its rarity along with small review of literature.

Case Report

48 years male presented with a history of fever and pain in right iliac fossa for 15 days. There was no significant personal and past history. On palpation, there was a tender lump present in right iliac region. Her hemogram showed leucocytosis (16 X 10^9 µL). Ultrasonography abdomen revealed a dilated, non-compressible appendix suggestive of appendicitis, and adherent bowel loops. At that time, he was treated conservatively, and no antibiotic cover was given due to his refusal to treatment.

He was then discharged after symptomatic treatment. He was admitted for second time after 10 days with complaints of pain abdomen. At that time laprotomy was done which revealed presence of abscess and bowel adhesions. Abscess was drained and abdomen was then closed. He was then again discharged. He was again readmitted with similar complaints after 15 days. Further

investigations were asked for. On CT scan abdomen, thickening of caecum and part of ascending colon, matted bowel loops and free fluid in abdomen with septations were noted. Per operatively there was presence of an ileocaecal lump with multiple mesenteric lymph nodes and serositis. As caecal cancer cannot be excluded, right hemicolectomy was performed. Grossly, an appendicular mass was received with adherent ileum, caecum and colon. Serosa of appendix was dull with presence of dense adhesions. On cutting, lumen showed presence of fecolith. The wall of appendix was thickened measuring 0.8 cm at its maximum. No growth identified. Serosa of adherent ileum, caecum and colon showed presence of exudate and congestion.

On microscopic examination, appendiceal mucosa was partially ulcerated. There was presence of dense chronic inflammatory infiltrate in the submucosa, muscularis propria and serosa comprising of lymphocytes, plasma cells and sheets of foamy histiocytes and few foreign body giant cells. These foamy histiocytes or xanthoma cells were polygonal in shape with abundant clear, vacuolated cytoplasm and central nucleus. Peri-appendicular region showed vascular congestion and chronic inflammatory infiltrate along with few xanthoma cells (Figure 1). Immunohistochemically, CD68 positivity proved the histiocytic nature of these xanthoma cells (Figure 2). No Michaelis-Gutmann bodies were seen in sections stained with PAS and Von Kossa. Sections taken from ileum, caecum and colon showed non-specific chronic inflammation and serositis. The mesenteric lymph node resected showed reactive pathology. There was no evidence of granulomas elsewhere and no evidence of malignancy seen on multiple sections taken.

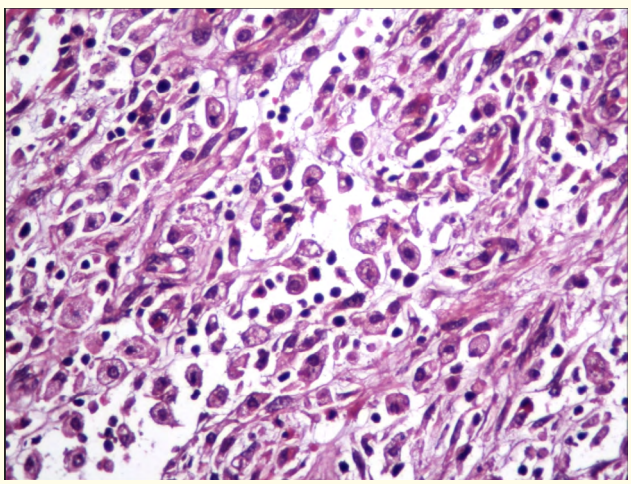

Figure 1: Xanthoma cells in the appendicular wall (H/E, 400X).

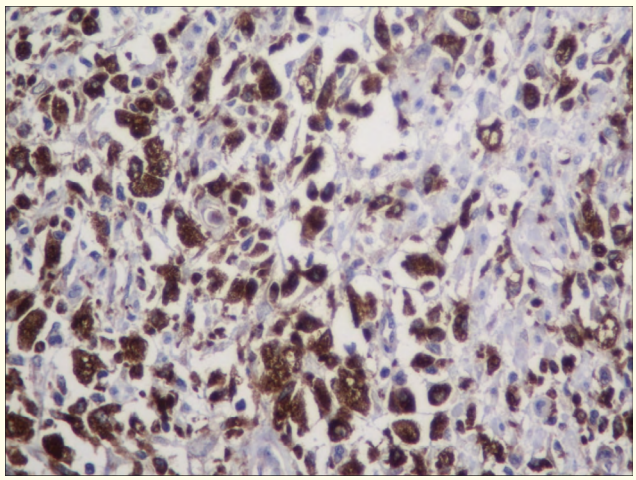

Figure 2: CD68 positivity in xanthoma cells (400X).

Discussion

Xanthogranulomatous inflammation is a rare type of chronic inflammation and has been reported in a number of organs, though rarely like in kidney [1], gall bladder [2], urinary bladder [3], thyroid [4], endometrium [5], adrenals [6], and fallopian tube [7]. There are very few reports of its occurrence in the appendix.

Cozzuto and Cabone in 1998 were the first to describe briefly the xanthogranulomatous process in appendix [8]. Birch., *et al*. reported 2 cases in 1993, both had fibrotic appendix with presence of chronic inflammation including foamy histiocytes in the wall [9]. One case showed presence of foamy histiocytes in the peri-appendicular region also similar to the present case.

Most cases reported were of adult age group, similar to the present case, only few reports have been found involving pediatric age group like Rawabdeh., *et al* [10] and Jusoh., *et al* [11].

McVey also reported similar case in 1994 in a 40 year old lady who had many granulomas including foamy cells in the wall of appendix [12]. Similar to the present case, Munichor., *et al*. in 2000, studied immunohistochemically as well as electron microscopically a case of xanthogranulomatous appendicitis [13]. CD 68 positivity confirmed the histiocytic nature of the xanthoma cells and electron microscopy showed the presence of electron lucent variably sized lipid droplets in these cells.

Guo., *et al*. in 2003 described about 8 cases of xanthogranulomatous appendicitis in their study of 22 cases of interval appendectomy specimens of chronic appendicitis [14]. Two recent reports in 2005 and 2011 have been described by Chuang.,

et al [3] and Martinez Garza., *et al*. [15] respectively. Former case had findings similar to the present case, as it also mimicked caecal cancer but was proved pathologically as xanthogranulomatous appendicitis.

Various pathogenetic mechanisms responsible for the xanthogranulomatous inflammation have been proposed including defective lipid transport, immunological disorders, infection by low virulence organisms, reaction to specific infectious agent and lymphatic obstruction [16]. Cozutto and Carbone [8] suggested the role of abscess formation in its causation and McVey [12] suggested its association with a long standing or recurrent inflammatory process.

Many recent studies have emphasized on its diagnostic importance like Maj singh., *et al* [17] and Omer., *et al.* [18] who described this entity as a rare histological variant of chronic appendicitis. Jusoh., *et al*. [11] also described a case having Xanthogranulomatous lesion in recurrent appendicitis, similar presentation as in the present case who underwent interval appendectomy.

Conclusion

Xanthogranulomatous inflammation is an uncommon entity. Whatever is the cause, ischemia, chronic obstruction and mucosal ulcerations play an important role in the development of this type of inflammation. Its diagnosis is based on histopathology and it is important to diagnose it as it might be associated to other intestinal inflammatory diseases or cancer but has a surgical cure. Thus, the invasive nature of XA mimicking a locally advanced malignancy may lead to a radical surgery, especially in cases with long clinical course. In addition, pathologists should be primed regarding the case for preventing misinterpretation, such as Crohn's disease, owing to their histological similarity.

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