XANTHOGRANULOMATOUS APPENDICITIS- AN UNCOMMON ENTITY

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ABSTRACT

BACKGROUND

Xanthogranulomatous inflammation is a clinical finding described commonly in kidney and gallbladder, but xanthogranulomatous appendicitis is rare and very few cases have been reported so far. We report a case of a 56-year-old lady who presented with acute pain in Rt lower quadrant of abdomen along with nausea. Ultrasonography was inconclusive and with clinical impression of acute appendicitis, the patient underwent appendicectomy. Histologically, a diagnosis of xanthogranulomatous appendicitis was made.

KEYWORDS

Appendicitis, Histiocytes, Xanthoma.

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BACKGROUND

Xanthogranulomatous inflammation is a rare form of chronic inflammation that can involve various organs but is most commonly reported in the kidney and gallbladder.[1] Histologically, it is characterised by presence of large number of foamy histiocytes admixed with lymphocytes and plasma cells.[2] Xanthogranulomatous inflammation of appendix is a rare phenomenon and few cases have been reported in literature.[1,3,4] In view of the rarity of this condition, we report a case of XA in a 56-year-old lady who presented with acute pain abdomen and nausea.

Case Report

A 56-year-old lady presented with complaints of acute pain in right lower quadrant of abdomen with nausea. On clinical examination, she was found to have tenderness in the McBurney's point along with rebound tenderness. There was no history of fever. Her ultrasonography was inconclusive. Routine blood test showed a white blood count of 14,000 cells/cu mm with 81% neutrophils, Hb 11.5 g/dL, platelet count 1.8 lakhs/mm^3 and an increased ESR level (32 mm/hour). Even though the ultrasonography report was inconclusive based on the clinical findings and blood report, she was clinically diagnosed as a case of acute appendicitis and an appendicectomy was performed. Per operatively the appendix was inflamed and did not show any gangrenous change or perforation. We received a specimen of appendix which measured 7 cm in length and 1 cm in diameter. The external surface appeared congested and dull. Cut surface showed presence of focal yellow coloured areas and congested mucosa. The lumen contained faecolith. The Haematoxylin and Eosin stained sections from the appendicectomy specimen showed patchy neutrophilic infiltration of mucosa and clusters of foamy histiocytes (Figure 1) along with foreign body type multinucleated giant cells, lymphocytes and plasma cells in the muscular and serosal layers (Figure 2). No epithelioid granulomas or Michaelis-Gutmann bodies were noted which rules out the possibility of Crohn's disease or malakoplakia. A diagnosis of xanthogranulomatous appendicitis was made.

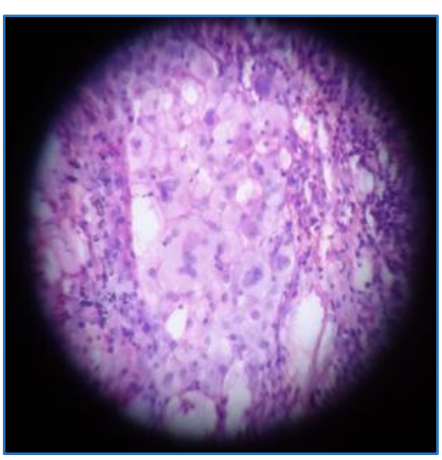

Figure 1. H&E stained section of appendix showing focus of xanthogranulomatous inflammation with xanthoma cells and chronic inflammatory cells

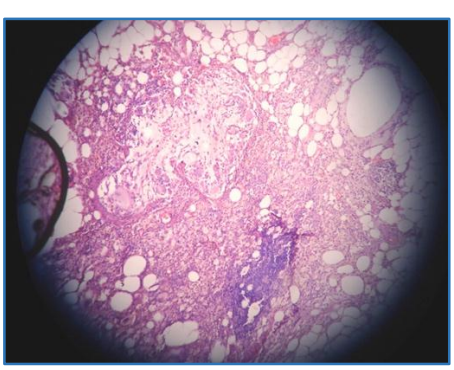

Figure 2. H&E stained section of appendix on low power view of numerous giant cells and chronic inflammatory cells

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DISCUSSION

Xanthogranulomatous inflammation is a rare form of chronic inflammation. Although it is commonly described in various organs such as kidney and gallbladder, xanthogranulomatous appendicitis is a rare phenomenon.[1] Osterlind initially described xanthogranulomatous inflammation in the kidney in 1944.[5] Birch et al published the first reported cases of XA in 1993.[6] After a year later, McVey et al added another case of XA to the literature.[7] An association of xanthogranulomatous response with longstanding appendiceal inflammation and the formation of appendiceal mass was suggested by both the authors. Few cases of involvement of appendix by xanthogranulomatous inflammation has been reported in literature.[1,3,4] Besides these two common sites it has also been reported in other organs as endometrium, fallopian tubes and sigmoid colon.[8,9]

Xanthogranulomatous inflammation affects women more frequently than men with a variable age of presentation. This finding has been reflected in various series[10,11] and till date majority of the case reports of XA published have been females. Histologically, it is manifested by collection of lipid laden macrophages admixed with mixed inflammatory infiltrate composed of lymphocytes, plasma cells, neutrophils and multinucleated giant cells with or without cholesterol clefts.[12]

The pathogenesis of xanthogranulomatous inflammation is unknown but some of the proposed aetiologies suggest a chronic inflammatory process which results in tissue destruction and localised proliferation of lipid containing macrophages.[2] Munichor et al suggested that a defective lipid transport, immunologic disturbances and infection by low virulence organisms and lymphatic obstruction may be the underlying pathogenic mechanism.[4] He examined the specimen of appendix under electron microscopy and showed the presence of electron–lucent lipid droplets of variable sizes in the xanthoma cells and in other cells as well.

Xanthogranulomatous inflammation (XGI) may also present with mass like lesion and cause destruction of the organ involved thus mimicking an invasive cancerous lesion.[1,13] This was shown in a case reported by Chuang et al who reported a case of a 39-year-old man who presented with fever, lower abdominal pain and mass in right iliac fossa. With the impression of cancer hemicolectomy was done. The histological examination revealed it as XA.[1]

When XGI presents as a mass with extension of fibrosis and inflammation to surrounding tissues, it is very difficult to differentiate it from advanced cancer or a local abscessed intestinal mass.[14]

Radiological diagnosis of XGI using USG and CT is challenging. Contrast–enhanced USG using Sonazoid contrast enhancing agent has increased the diagnostic accuracy in differentiating chronic inflamed gall bladder from a malignant lesion.[15] This explains the inconclusive USG report in our case which would have been precise if contrast enhancement was done.

Recently Kaushik et al have published a case report where they have highlighted the usefulness of an intraoperative imprint cytology for quick diagnosis to decipher the cause of the mass lesion so as to avoid extensive resection.[16]

The classic microscopic appearance of XA display presence of numerous lipid laden histiocytes, abundant hemosiderin, multinucleated giant cells admixed with cholesterol clefts and mixed inflammatory infiltrate composed of polymorphs, lymphocytes and plasma cells.[17]

Guo and Greenson noted a higher incidence of XA in interval appendectomies as compared to control group of patients who had acute appendicitis and underwent routine appendectomy.[18]

CONCLUSION

Xanthogranulomatous appendicitis is a rare clinical entity which might present with varied clinical presentation of acute or subacute abdominal pain or sometimes as a mass lesion which might lead to radical resection.

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