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Case Report

Xanthogranulomatous appendicitis: Uncommon histological variant of a common entity

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has a benign course, and can be cured by surgical resection.⁴ Due to the rarity of this condition, we report a case of xanthogranulomatous appendicitis in 21-year-old lady, who presented with acute pain abdomen and operated as a case of an acute appendicitis.

Case report

A 21-year-old, nullipara, developed acute pain in the right iliac fossa and presented to the surgical Out Patient Department. On evaluation she was found to have tenderness in the McBurney's point along with rebound tenderness. She was managed symptomatically but the pain did not subside. Her ultrasonogram (USG) abdomen and routine blood tests were inconclusive. She was clinically diagnosed as a case of acute appendicitis and an appendectomy was performed. Per operatively the appendix appeared inflamed. No gangrenous change or perforation was noted. We received the specimen of appendix, which measured 6 × 3 × 2 cm in size. The external surface appeared congested and dull. Cut surface showed congested mucosa with few yellow colored areas. The lumen was patent. No fecolith or parasite was seen (Fig. 1A).

The Hematoxylin and Eosin (H&E) stained sections from the appendectomy specimen showed focal mucosal ulceration. Large areas of the mucosa and sub mucosa were replaced by collections of histiocytes with abundant granular eosinophilic cytoplasm admixed with variable amounts of lymphocytes, plasma cells and occasional eosinophils. Few reactive

Introduction

Xanthogranulomatous inflammation (XGI) is a rare form of chronic inflammation characterized histologically by presence of high number of foamy histiocytes admixed with lymphocytes and plasma cells, first reported in the genitourinary tract.¹ It can involve any organ, but the most common sites are kidney and gallbladder.^{2,3}

Although acute appendicitis is a very common surgical condition, xanthogranulomatous appendicitis is a rare phenomenon. Only few cases have been reported so far in the literature in which XGI involved the appendix and the patient presented as a case of acute appendicitis. Xanthogranulomatous appendicitis may mimic a locally advanced cancer but

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Fig. 1 – (A) Gross photograph of the appendectomy specimen with abnormal yellowish area, shown by the arrow. (B) Photomicrograph (Hematoxylin and Eosin stain: 100×) showing large areas of the mucosa and sub mucosa were replaced by collections of histiocytes. (C) Photomicrograph (Hematoxylin and Eosin stain: 400×) showing histiocytes with abundant granular eosinophilic cytoplasm. (D) IHC Photomicrograph showing strong cytoplasmic positivity of CD68 in the histiocytes.

lymphoid follicles were also seen (Fig. 1B and C) There were occasional foreign body-type multinucleated giant cells. Special stains were done. No acid-fast bacilli were seen on Ziehl–Neelson stain. Von Kossa (for calcium) and Perl’s stain (for iron) were done to rule out presence of Michaelis Gutmann bodies, which shows positivity for both iron and calcium stains. Immunohistochemistry (IHC) was done with CD 68 (Dako; Monoclonal mouse Anti Human CD 68 – clone PG – M1) which showed strong positivity in the foamy cells (Fig. 1D). These cells were negative for Pan Cytokeratin (CK). Finally based on H&E, special stains and IHC findings, a diagnosis of xanthogranulomatous appendicitis was offered. Post-operative period was uneventful and presently the patient is asymptomatic.

Discussion

XGI is a pathologic entity with unique and characteristic macroscopic and microscopic features. Typical findings include bright yellow or golden yellow mass-like lesions on macroscopic examination associated with abscess cavities, micro-abscesses, and large numbers of lipid-laden macrophages; as well a minor component of chronic and acute inflammatory cells on microscopic examination.⁵

Other lesions containing foam cells should be distinguished from XGI. Malacoplakia is characterized by an inflammatory and destructive xanthomatous proliferation with the presence of Michaelis–Gutmann bodies or calcospherites (concentrically layered intracytoplasmic inclusion) Small localized xanthoma deposits without parenchymal destruction or xanthomas with prominent foam cell features must also be considered in the differential diagnosis.⁶ Some time it is not possible to differentiate XGI from an infiltrative cancer because XGI might present as a mass-

like lesion with an extension of fibrosis and inflammation to the surrounding tissues, mimicking an infiltrative cancer.^{7,8} The exact pathogenesis of XGI is not well known. There are many hypotheses regarding the pathogenesis of XGI including defective lipid transport, immunological disorders such as disturbed chemotaxis of polymorphonuclear cells and macrophages, a specific immune response toward *Proteus* and *Escherichia* infections, and lymphatic obstruction.^{2,4} XGI probably represents a chronic inflammatory process in which host and microorganism interact that leads to tissue destruction and localized proliferation of macrophages containing large amounts of lipid which are the characteristic histological features of the disease.¹

Xanthogranulomatous inflammation of the appendix is rare. Microscopic examination of XGI usually reveals a nodular or diffuse collection of foamy histiocytes, intermixed with varying amounts of other inflammatory cells, such as multinucleated giant cells, lymphocytes, plasma cells, and eosinophils, as well as fibrosis. Occasionally, cholesterol clefts, granulation tissue, and necrotic debris are observed with reactive lymphoid hyperplasia. The cholesterol clefts were not seen but reactive lymphoid hyperplasia was seen in our case. XGI causes destruction and effacement of the normal structures of the involved organ and could be misinterpreted as a locally invasive cancerous lesion.^{7,8} Guo and Greenson⁹ compared histopathology of all interval appendectomy specimens within a 4-year period and compared them with a control group of patients who had acute appendicitis and underwent routine acute appendectomy. Eight (36.4%) of the interval appendectomy cases had XGI compared with none in the acute appendicitis group ($P < 0.0001$).

To conclude, the aim of presenting this case was to highlight the rarity of XGI of the appendix, presenting as an acute appendicitis.

Conflicts of interest

All authors have none to declare.

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