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CASE REPORT

Xanthogranulomatous lesion in recurrent appendicitis

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Abstract Acute appendicitis (AA) is the most common acute surgical condition of the abdomen. Most resected specimens have been reported to have marked cellular infiltration, predominantly by neutrophils. By contrast, the occurrence of a xanthogranulomatous (XG) lesion is extremely rare. To date, only a few cases have been reported in the literature. Moreover, its clinical implications remain to be evaluated. Here, we report the case of a 16-year-old boy who presented with a typical history of AA for 2 days. He had no sepsis or local peritonitis. He experienced a similar attack 1 year ago, which was successfully and conservatively managed at our center. An interval appendectomy was performed 3 months later as requested by his parents. Furthermore, he responded well to the antibiotics administered. Intraoperatively, the appendix appeared fibrotic with a small lump at its base. Some adhesion was noted between the appendix and the omentum. His postoperative recovery was uneventful. Moreover, the histopathological examination of the appendix revealed features typical of an XG lesion. In addition, we conducted a literature search to establish the clinical implications of XG lesions.
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1. Introduction

Acute appendicitis (AA) is the most common condition requiring surgery. Most cases present with acute and singular events.[1] Microscopically, mucosal ulceration together with numerous acute inflammatory cells, mostly neutrophils, is observed in the muscularis propria along with necrosis, congestion, and perivascular neutrophilic infiltrate. Moreover, recurrent and chronic appendicitis do occur, which typically have different histological features.[1]

Xanthogranulomatous (XG) inflammation (XGI) is a rare form of chronic inflammation manifested by lipid-laden macrophages admixed with lymphocytes, plasma cells, neutrophils, and often multinucleated giant cells.[2] XGI was first reported in the genitourinary tract (kidney);

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however, it can affect all organs, particularly the gallbladder and ovaries.[3] Furthermore, involvement of the appendix is a rarer phenomenon, with only 11 cases reported to date. We explored the possible association among the reported cases and evaluated the clinical implications of XG lesions.

2. Case Report

Our patient was a 16-year-old boy who presented with a history of migratory abdominal pain typical of AA. He had a similar presentation 1 year ago, which was successfully and conservatively managed. During that time, the diagnosis was made on the basis of clinical signs and an Alvadro score of 8/10. No imaging was performed. During his recent admission, he had minimal abdominal signs (mildly tender over the right iliac fossa with rebound) and no sepsis. Therefore, his Alvadro score was only 6/10.

He was planned to undergo an open appendectomy for recurrent AA, which was postponed on request of his parents. Moreover, his symptoms almost completely abated after receiving antibiotics for 1 day. An interval appendectomy was performed 3 months later. Intraoperatively, his appendix appeared fibrotic with a small lump at its base. Omental adhesion was noted around the appendix.

The surgery was uneventful and he was discharged 1 day later. The histopathological findings of the appendix fulfilled the criteria for XGI (Figures 1 and 2). Numerous lymphoid follicles with prominent germinal centers were observed at the mucosa with an area of fibroblastic proliferation. In addition, foamy histiocytes, giant cells, and lymphoid aggregates were observed on its serosal aspect.

3. Discussion

XGI is a chronic inflammatory process that leads to tissue destruction and localized proliferation of macrophages containing a large amount of lipids, which is the characteristic histological feature.[3] XG appendicitis (XA) is represented by the prominent histiocytic component of clusters of xanthoma-like cells.[4] The exact etiology of XA remains uncertain; however, it may be associated with defective lipid transport, immunological disturbances, infection by low-virulence organisms, and lymphatic obstruction.[2,3]

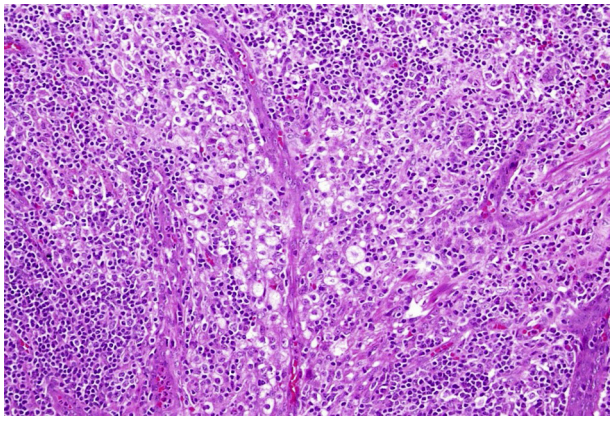

Figure 1 Microscopic findings show numerous lymphoid follicles with prominent germinal centers. (Hematoxylin and eosin staining: magnification, 40×).

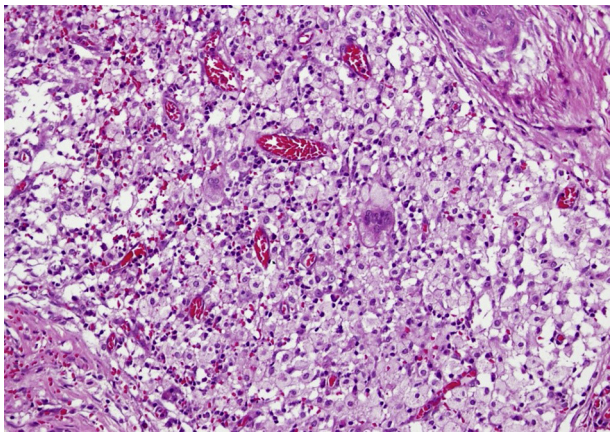

Figure 2 Microscopic image of xanthogranulomatous appendicitis shows numerous sheets and nests of foamy histiocytes with few giant cells. (Hematoxylin and eosin staining: magnification, 100×).

On the basis of a reported series (Table 1), XA majorly occurred in adults with a mean age of 47.9 years (83%, 21−78 years). Only two cases have been reported in the pediatric age group (<18 years), including our case. Most of them (92%) presented with right lower quadrant abdominal pain suggestive of being appendiceal in origin, although the duration of pain varied from hours to months. Nonetheless, those reported by Birtch et al[5] in 1993 were excluded because they had performed an incidental appendectomy during laparotomy for urinary diversion.

Several factors have been proposed that may precipitate XA, including organ obstruction, suppurative inflammation, hemorrhage, and local hypoxia.[2,6] The spectrum of appendicitis observed excludes the possibility of a single pathophysiology of XA. A review was conducted to further strengthen the prior concept. Five cases of XA (42%) were observed during surgery for complicated appendicular mass, and three of them were related to abscess. A similar conclusion was made approximately 30 years ago by Birtch et al[5] and McVey and McMahon.[7] They believed that XG responses may be associated with long-standing inflammation and mass formation.[2,4,8,9]

In contrast to the proposal of Guo and Greenson,[6] a comparable number of patients (4, 36%) experienced an XG response following an immediate surgery for AA. Moreover, two of the four patients had a relatively normal-looking appendix intraoperatively.[8] We believe that they may have had chronic or recurrent appendicitis, which could explain the nature of the inflammation observed on histopathological examination. Moreover, Rao et al[1] observed in their series that 10% of patients with proven appendicitis had one or more recurrent episodes of identical symptoms.

Table 1 Summary of all cases of xanthogranulomatous appendicitis reported in the literature.

	Age (y)/sex	Authors/y of publication	Main presenting symptoms	Duration of symptoms	Time of appendectomy. — Immediate for AA. — Delayed. — Interval — Incidental	Imaging and findings (if done) USG or CT	Technique of appendectomy (open, laparoscopic or laparotomy)	Diagnosis (intraop) — AA (suppurative). — AA (perforated/ gangrenous) —Appendicular mass —Normal	Remarks
1	16/ male	Present case	RIF pain	1 d	Interval appendectomy following recurrent appendicitis	Not done	Open appendectomy (done 3 mo after attack)	Acute appendicitis	Fibrotic appendix with adhesion intraoperatively.
2	73/ female	Altay et al[11]/2014	RIF pain	Acute (? duration)	NM	USG: heterogenous mass at RIF CT: appendicolith with mass-like cystic lesion	Laparotomy with limited right hemicolectomy, hysterectomy and partial cystectomy	Complicated appendicular mass/abscess	Endometrial abscess also noted during surgery
3	50/ male	Kocchar et al[9]/2014	RIF pain and fever	7 d	Delayed appendectomy for appendicular mass (abscess) (failed conservative management)	USG: inflamed appendix with echogenic mesentery CT: appendicular abscess	Laparotomy and right hemicolectomy and mucous fistula	Appendicular mass (abscess)	Post operatively developed septicemia with MOF and died
4	78/ male	Mado et al[4]/2013	RIF pain	2 mo	Delayed appendectomy	CT showed an irregular multicystic mass near caecum	Laparotomy and ileocecal resection	4 cm growth at tip of appendix	
5	11/ male	Al-Rawabdeh et al[2]/2013	Abdominal pain and vomiting	1 d	Immediate appendectomy for AA	CT showed an enlarged appendix without inflammation	Laparoscopic	Normal (no fecalith within)	Pink-tan appendix intraoperatively
6	21/ female	Singh et al[3]/2013	RIF pain/ fever	NM	Immediate appendectomy for AA	USG abdomen - normal	NM	Acute appendicitis	Congested mucosa with few yellow colored areas. No fecalith within lumen
7	39/ male	Chuang et al[13]/2005	RIF pain/ fever	NM		CT: diffuse cecal and ileal thickening	Laparotomy and hemicolectomy	Appendicular mass ? tumor	XA may mimic locally invasive cancer
8	30/ female	Martinez et al[12]/2011	RIF pain/ fever	NM	Immediate appendectomy for AA	Not done	Laparoscopic appendectomy	Acute appendicitis	

9	37/ female	Munichor et al[8]/2000	RIF pain/ fever	Hours	Immediate appendectomy for AA	NM	Laparotomy and appendectomy	Acute appendicitis Appendix appeared slightly dilated and removed	HPE: Fibrotic appendix at tip and dilated proximally. Inspissated fecalith filling the lumen.
10	40/ female	McVey and McMahon[7]/ 1994	RIF pain and tender mass	NM	Interval appendectomy	USG showed appendicular mass	NM	Normal	Operation done 2 mo after conservative management of appendicular mass
11	51/ male	Birtch et al[5]/1993	Background multiple sclerosis with recurrent UTI	4 y	Was not suspected (incidental appendectomy)	NM	Laparotomy: Incidental appendectomy together with supravesical urinary diversion and ileal conduit	Appendix surrounded by fibrinous adhesion. Large inspissated fecalith within lumen	
12	60/ female	Birtch et al[5]/1993	Right flank pain	NM	Immediate appendectomy (was not suspected before surgery)	CT: large soft tissue mass at right lateral pelvis	Laparotomy: retrocecal abscess surrounding the appendix	Fibrotic appendix, covered with fibrinous exudate	

AA = acute appendicitis; CT = computed tomography; HPE = histo-pathological examination; MOF = multi-organ failures; NM = not mentioned; RIF = right iliac fossa; USG = ultrasound; UTI = urinary tract infection; XA = xanthogranulomatous appendicitis.

Although fecalith is the most common cause of AA, only three patients (25%) had it during surgery.[10] Typically, the fecalith becomes inspissated. The other two cases of XA (16%) seen were following an interval appendectomy for recurrent AA and appendicular mass.[7] This finding was in contrast to that reported by Guo and Greenson[6] in 2003. They reviewed all interval appendectomy specimens and reported that 36% of them (8 out of 22 patients) had XA features; XA features were not observed in the acute group.[6]

Excluding three cases where the imaging method was not mentioned (Martinez et al[12], Munichor et al[8], and Birtch et al[5]), computed tomography was the most common (50%) imaging method used because of its high sensitivity for detecting mass. Nevertheless, consistent radiological features for XA could not be validated. Similarly, colonoscopic examination was not useful as demonstrated by Mado et al.[4] Seven patients (58%) underwent laparotomy and half of them required hemicolectomies. Excluding Birtch's series for incidental appendectomy, four patients (36%) underwent additional visceral resection. Therefore, XA lesions may be invasive and infiltrative in nature and cannot be easily differentiated from malignant lesions during surgery. Most patients had an uneventful recovery, except for one patient who presented with appendicular abscess and died of multiorgan failure following laparotomy and hemicolectomy. Therefore, the overall mortality rate was 8.3%.

In conclusion, XGI can be considered as the unnatural healing process of appendicitis. Moreover, clinicians should consider this condition, particularly in high-risk cases such as those undergoing delayed surgery for a complicated appendicular mass and interval appendectomy. Intraoperatively, the invasive nature of XA mimics a locally advanced malignancy. An *en bloc* visceral resection may be required. 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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