

Xanthogranulomatous Appendicitis with a Fulminant Course: Report of a Case

GAURAV KOCHHAR¹, SUDIPTA SAHA², MANOJ ANDLEY³, ASHOK KUMAR⁴, AJAY KUMAR⁵

ABSTRACT

Xanthogranulomatous inflammation is a well-described entity with involvement of various body organs. But the involvement of vermiform appendix in the disease process is quite rare with only few cases are reported in literature. This case report describes a 50-year-old man, who was diagnosed as a case of acute appendicitis with appendicular lump on the basis of clinical history, physical examination, and hematological and radiological investigations. Patient underwent surgical interventions twice. But, he succumbed to the disease. We are reporting this case in view of rarity of the disease and the fulminant course, which has not been described in any other reports.

Keywords: Appendicectomy, Xanthogranulomatous, Vermiform appendix

CASE REPORT

A 50-year-old male presented with the complaints of pain in the right lower abdomen for seven days along with history of fever and anorexia. On examination, there was approximately 8 x 6 cm lump present in right iliac fossa. Total leucocyte count was 24000/ mm³. Ultrasound abdomen showed inflamed appendix with echogenic mesentery. Clinical diagnosis of acute inflammatory appendicular lump was made and patient was started on expectant management. Despite of expectant management his clinical condition did not improve. CECT abdomen was done which suggested abscess formation [Table/Fig-1]. As there was no window for percutaneous aspiration, extra peritoneal drainage of abscess was done on fourth day of admission. Hundred ml of pus was drained. Appendicectomy was not attempted as small intestine, omentum and cecum was forming a mass. But patient developed small intestinal obstruction which did not resolved with conservative management. Patient was then taken up exploratory celiotomy on eighth day of admission. Intra-operatively, there was a larger ileo-cecal phlegmon with dense interloop adhesions and omentum was adhered to the phlegmon with dense interloop adhesions and omentum was adhered to the phlegmon. Small intestine was dilated proximal to the mass. Right hemicolectomy was done with ileostomy and mucous fistula. Postoperatively patient went into septicaemia and later on into multiorgan dysfunction. Despite of aggressive management he patient succumbed to the disease process. The histopathological report was suggestive of transmural involvement of appendix with infiltration by lymphocytes, histiocytes and giant cells. Foamy macrophages and plasma cells were also present, and diagnosis of xanthogranulomatous appendicitis was suggested [Table/Fig-2].

DISCUSSION

Xanthogranulomatous inflammation is a rare entity. Any organ of the body can be involved in the disease process with kidney and the gall bladder being the commonest ones [1]. Other than these two predominantly involved sites, other organs where xanthogranulomatous inflammation is reported are endometrium, epididymis, fallopian tubes, bone, skin, appendix, urinary bladder, thyroid and adrenal glands [2,3]. The differential diagnosis includes the conditions showing the presence of xanthomatous cell such as malakoplakia. The disease masquerades as malignancy due to presence of diffuse inflammatory and fibrotic changes [4].

The exact pathogenesis is not known; it probably represents a chronic inflammatory process leading to tissue destruction and

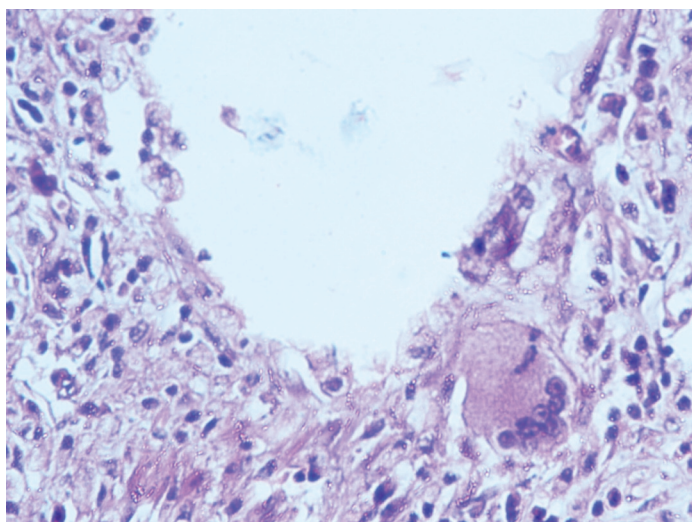
localized proliferation of macrophages containing large amounts of lipid which are the characteristic histological features of the disease [5]. Other speculations are defective lipid transport, disorders of neutrophilic chemotaxis, lymphatic obstruction and specific immune response towards *Proteus* and *E. coli* [2,6]. The disease has a typical histopathological appearance. On gross appearance there is presence of golden yellow mass with abscesses. On microscopy the lesion shows recruitment of acute and chronic inflammatory cells, lipid laden macrophages and foam cells [7].

Xanthogranulomatous inflammation of ileo-cecal region is a rare entity. Around ten cases of involvement of vermiform appendix have been reported in literature [8]. In a histopathological review of interval and emergency appendicectomies over a period of four years Guo and Greenson found xanthogranulomatous inflammation in 8 out of 22 (36.4 %) in interval appendicectomies group, while no such findings were seen in emergency appendicectomies group [9]. They also derived the conclusion that the xanthogranulomatous is characteristically associated with the chronic inflammatory states, which comprised of granulomas, xanthomatous cells, mural fibrosis, and transmural chronic inflammatory infiltrates. These changes may mimic as Crohn's disease.

Munichor et al., reported a case of an adult lady who had acute appendicitis and histopathology of the specimen revealed



[Table/Fig-1]: CECT showing a large collection in right iliac and lumbar region with collapsed colon, suggestive of appendicular abscess (marked by arrow)



[Table/Fig-2]: Histopathological slide showing transmural involvement of appendix with infiltration by lymphocytes, histiocytes and giant cells, foamy macrophages and plasma cells

xanthogranulomatous appendicitis. The electron microscopy showed presence of electron lucent droplets of various sizes in the xanthoma cells and other cell types. They derived inference that various cell types can be damaged by common mechanism which includes obstruction, haemorrhage and hypoxia [7].

Xanthogranulomatous appendicitis can also masquerade as other diseases. In a case reported by Chuang et al., a 39-year-old man presented with fever, right lower abdominal pain and hard lump.

Right hemicolectomy was done in suspicion of cancer. The final histopathological report suggested xanthogranulomatous appendicitis, inferring that this disease may simulate locally invasive right colonic cancer [10].

We are presenting this case of xanthogranulomatous appendicitis in view of its rarity. Also, the uniqueness in our case is was the fulminant course of the disease and poor prognosis which have not been mentioned in the previous reported case.

REFERENCES

- [1] Franco V, Aragona F, Genova G, et al. Xanthogranulomatous cholecystitis. Histopathological study and classification. *Pathol Res Pract*. 1990;186:383–90.
- [2] Oh YH, Seong SS, Jang KS, et al. Xanthogranulomatous inflammation presenting as a submucosal mass of the sigmoid colon. *Pathol Int*. 2005;55:440–44.
- [3] Gray Y, Libbey NP. Xanthogranulomatous salpingitis and oophoritis: a case report and review of the literature. *Arch Pathol Lab Med*. 2001;125:260–63.
- [4] Maeda T, Shimada M, Matsumata T, et al. Xanthogranulomatous cholecystitis masquerading as gallbladder carcinoma. *Am J Gastroenterol*. 1994;89:628–30.
- [5] Peterson RO. Kidney. In: Peterson RO, ed. *Urologic Pathology*. Philadelphia: J.B. Lippincott; 1986:40e46. Chap 6.
- [6] Munichor M, Kerner H, Cohen H, Bickel A, Iancu TC. Xanthogranulomatous appendicitis—an incidental finding of localized pathology. *Ultrastructural Pathology*. 2000;24:33–39.
- [7] Anadol AZ, Gonul II, Tezel E. Xanthogranulomatous inflammation of the colon: a rare cause of cecal mass with bleeding. *South Med J*. 2009;102:196–99.
- [8] Al-Rawabdeh SM, Prasad V, King DR, Kahwash SB. Xanthogranulomatous appendicitis in a child: report of a case and review of the literature. *Case Rep Med*. 2013;2013:498191.
- [9] Guo G, Greenson JK. Histopathology of interval (delayed) appendectomy specimens: strong association with granulomatous and xanthogranulomatous appendicitis. *American Journal of Surgical Pathology*. 2003;27:1147–51.
- [10] Chuang YF, Cheng TI, Soong TC, Tsou MH. Xanthogranulomatous appendicitis. *Journal of the Formosan Medical Association*. 2005;104:752–54.

PARTICULARS OF CONTRIBUTORS:

1. Senior Resident, Department of Surgery, Lady Hardinge Medical College, Delhi, India.
2. Assistant Professor, Department of Surgery, Lady Hardinge Medical College, Delhi, India.
3. Professor, Department of Surgery, Lady Hardinge Medical College, Delhi, India.
4. Professor, Department of Surgery, Lady Hardinge Medical College, Delhi, India.
5. Director Professor, Department of Surgery, Lady Hardinge Medical College, Delhi, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Gaurav Kochhar,
Room No. 331, House Surgeon Block, LHMC Campus, Shaheed Bhagat Singh Marg, New Delhi, India.
Phone : +919582935505, E-mail : gauravkochhar82@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Jan 26, 2014**

Date of Peer Review: **Jul 07, 2014**

Date of Acceptance: **Oct 27, 2014**

Date of Publishing: **Dec 05, 2014**