Crohn's Disease of the Appendix

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We report a 35 year old patient who presented with right iliac fossa pain of two weeks duration. The patient underwent appendectomy. The histology of the appendix showed features of Crohn's disease. Relevant literature of Crohn's disease of the appendix has been reviewed.

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Crohn's disease is a chronic progressive granulomatous inflammatory disorder of the gastrointestinal tract. The peak incidence occurs between the second and fourth decades. The etiology is unknown. Crohn's disease may affect any part of the gastrointestinal tract from lips to anus[1]. The small bowel alone accounts for 15-30%, while 25-30% occur in the large bowel. The commonest site is the distal ileum[2].

Microscopically there is extensive transmural inflammation with granulomas in the wall in 50-70% of patients and in mesenteric lymph nodes in 25% of patients. Skip lesions occur in 15% of patient. Grossly, the lesion appears as hemorrhagic spots with elevated margins (aphthus ulcer), and with progression of the disease clefts and fissures with edematous submucosa occur which gives a cobblestone appearance[1,3].

Crohn's disease has many modes of presentation: Diarrhoea in 90% of patients, recurrent abdominal pain in 70%, constitutional symptoms in 60% and anorectal lesions in 15-25% of patients[2].

Treatment of Crohn's disease is usually medical with rest, low residue, milk-free and high protein diet, parenteral nutrition, drugs like prednisone and sulfasalazine in acute disease. In chronic phase azathioprine and mercaptopurine singly or with other drugs can be used. The indication for operation is usually for obstruction, internal fistula, abscess, and growth failure in children[2]. We report a case of Crohn's disease of the appendix, which is very rare .

THE CASE

A 35 year old Bahraini male was admitted to Salmaniya Medical Center (SMC) with a two weeks history of crampy right iliac fossa pain. There was no history of change in bowel habit, melena, hematochezia, vomiting, anorexia or weight loss. His medical, social and family history were unremarkable.

On physical examination, the patient was afebrile and did not appear acutely ill. His abdomen was soft with rebound tenderness and guarding in the right lower quadrant. Rectal examination was negative.

The laboratory values were unremarkable. The white blood count was 6.9 x 10^9/L with Neutrophils 63%, Lymphocytes 26%, Monocytes 6% and Eosinophils 4%. Urine analysis was unremarkable. Abdominal X-ray films were normal.

The pre-operative impression was appendicitis and the patient underwent appendicectomy. At surgery there was a phlegmonous mass involving the omentum and the appendix, which was markedly enlarged. The cecum and terminal ileum were grossly normal. Patient did well post-operatively and was discharged home on the 5th postoperative day.

Figure 1. Specimen of the appendix which markedly swollen with acute inflammation and multiple tubercules in the mucosa and submucosa

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Histologically, the specimen measured 7cm in length and 3cm in diameter; the appendix was markedly swollen with acute mucosal inflammation, a generalized lymphedema and lymphoid follicles were seen in all coats (Fig 1). In particular, the serosa showed the most lymphorrhagis. A couple of tubercles were seen in the mucosa and submucosa, with no AFB. The proximal part of the appendix was preserved except for acute suppuration in the lumen. These features favour the diagnosis of Crohn's disease of appendix. The patient was followed for a period of one year and half without any complaint. The patient was thoroughly investigated in terms of repeated U/S, CT Scan of abdomen, Barium follow through colonoscopy and biopsy. All the investigations were normal. The patient was still having an appointment for follow-up .

DISCUSSION

Crohn's disease limited to the appendix is uncommon[4,5]. The disease involves the appendix by extension of granulomatous disease from distal ileum (25%) or the cecum in (50%). The patient usually presents as acute or subacute appendicitis. However, appendiceal involvement is rarely the first manifestation of the disease[9,12].

The differential diagnosis of chronic granulomatous appendicitis can be either infectious, like Mycobacter TB, Schistosoma, Yersinia, Actinomyces, Campylobacter, Histoplasma capsulatum and some parasites. Most of these infections can be ruled-out by clinical findings, stains, cultures and serology[6,7]. The non-infectious form of Granulomatous appendicitis includes Crohn's disease, foreign material and Sarcoidosis[6,7]. The appendixes in Crohn's typically have a diameter in the range of 1.5 to 2cm[6,8].

In 1990 Ruiz et al reviewed 85 cases in literature, the average age of patients was 24 year and the ratio of male to female 2:1. Eighty five percent of patients presented with right lower quadrant pain, 27% of patient had palpable mass, and 86% of patient had radiological evidence of the disease. In most cases, pre-operative diagnosis was acute appendicitis in 68% of patients; or appendiceal abscess in 27% of patients. A simple appendectomy was the surgical procedure in 64% of the cases. Post-operative complications were rare, which included pelvic abscess, wound infection, fistula formation, bowel obstruction. toxic megacolon and hematoma. No post operative death was recorded. A follow-up (average 2.5y) revealed a recurrence rate of 16%[11].

We reviewed the medical records of Salmaniya Medical Center from January 1987 to November 1997 and only 20 cases including our case were reported to have Crohn's disease, all of them involving various parts of the intestine commonly ileocecal area except our case which involved the appendix only. Seventeen cases out of 20 were Bahraini and 3 were non-Bahraini. Only one of them presented as acute appendicitis.

CONCLUSIONS

Crohn's disease limited to the appendix is rare and it is commonly seen in the second and third decade of life. It clinically mimics appendicitis. These patients will need long term follow-up after appendicectomy. Small bowel follow-through, CT Scan abdomen and colonoscopy are required on a regular basis to recognize recurrence as early as possible.

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