PERIANAL DISEASE AS INITIAL MANIFESTATION OF CROHN'S DISEASE

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ABSTRACT

BACKGROUND

Crohn's disease is a chronic inflammatory disease, which involves almost all parts of body. Perianal manifestations maybe the initial presentation in these patients, but usually includes other systemic manifestations along with it. This study was conducted to determine presence of perianal disease as the initial manifestation in otherwise asymptomatic Crohn's patients.

MATERIALS AND METHODS

This study was conducted in Department of Gastroenterology in a tertiary care hospital. All patients presenting with perianal manifestations were evaluated with colonoscopy and other imaging (in selected cases) to identify Crohn's disease.

RESULTS

A total of 46 patients were evaluated for perianal disease, out of which 3 patients (6.5%) had evidence of Crohn's disease in colonoscopy. Major presentation in these patients were skin tags (n=27, 58.6%), anal fissures (n=15, 32.6%) and fistula (n=4, 8.6%). The major symptoms were pain in perianal region (n=38, 82.6%), bleeding PR (n=23, 50%), anal pruritus (n=8, 17.3%) and mucus discharge PR (n=7, 15.2%).

CONCLUSION

Perianal disease maybe the initial presentation in Crohn's disease. Judicious evaluation of these perianal lesions helps in early diagnosis of Crohn's disease.

KEYWORDS

Crohn's Disease, Perianal Disease, Fissure, Fistula.

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BACKGROUND

Crohn's Disease (CD) is a chronic inflammatory disorder that can affect any part of the gastrointestinal tract from the mouth to the anus. The disease is characterised by transmural inflammation that can be complicated by the development of fibrotic strictures, perforation, abscess formation and fistulisation.[1] CD ranges from mild, relapse-free disease to chronic recurrent disease resulting in significant morbidity.[1-3] Perianal Crohn's Disease (PCD) is defined as inflammation at or near the anus including tags, fissures, fistulae, abscesses or stenosis.[4] Perianal disease can be categorised as fistulising and non-fistulising. The symptoms of PCD include pain, itching, bleeding, purulent discharge and incontinence of stool.

In the adult literature, the reported incidence of PCD ranges from 25% to 80% and fistulising PCD from 17% to 43%.[5] The clinical course of PCD depends on the type, including skin tag, rectal stricture, fistulae or fissures and location of disease. Skin tags in general do not resolve completely with treatment, but remain present and benign. Fissures will often heal completely with minor medical therapy. Simple fistulising disease (involves a low intersphincteric or transsphincteric location, a single short tract, an internal opening lower and closer to the anal verge and the external opening near the anal verge without an abscess) heals spontaneously in 50% of cases. Complex fistulae include those with origin of the fistulous tract that is high intersphincteric, high transsphincteric, extrasphincteric or suprasphincteric. Rectovaginal and complex fistulae rarely heal without therapy.

Multiple abnormalities can occur in the perianal area in addition to Crohn's fistula. The development of large anal tags (often referred to as "elephant ears") is a clue that Crohn's disease is present especially when associated with gastrointestinal symptoms. Fistulas may hide between these large folds. Patients may also suffer from fissures that tend to involve the squamous lining of the anal canal and are usually very painful in contrast to the relatively painless large anal tags.[3,6]

The characteristic presentation in CD is abdominal pain and diarrhoea, which maybe complicated by intestinal fistulisation or obstruction.[6] However, the presentation maybe indolent and patients may present only with perianal

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disease. Identifying these cases is extremely important because this ultimately influences not only the medical, but also the surgical management and approach to therapy.

The data on occurrence of CD in patients who present with perianal disease alone and no bowel-related symptoms is minimal. The main aim of the study is to identify asymptomatic Crohn's patients with no bowel-related symptoms who present with perianal disease.

MATERIALS AND METHODS

This study was conducted in Department of Gastroenterology in a tertiary care center in north Kerala over a period of 1 year from January 2016 till December 2016. All patients who present with perianal disease in the form of skin tags, fissures, fistula and rectal stricture were included in the study. Patients who were evaluated for other diseases and incidentally detected perianal disease were also included in the study. Patients with prior diagnosis of CD by clinical and/or biopsy findings were excluded. Patients with prior history of surgery and malignancy were excluded. After enrollment in the study, informed consent was obtained and all patients underwent colonoscopy and other imaging like barium meal follow through, MR fistulogram and/or CT enterography in indicated cases depending on symptoms. Crohn's disease is diagnosed on the basis of a combination of clinical, laboratory, histologic and radiologic findings. In colonoscopy, various lesions like nodularity, ulcers, etc. were evaluated and if mucosa is normal, 5 biopsies were taken from normal ileum, cecum, colon and rectum. All biopsies were interpreted by a single senior pathologist of our institute. Patients were also followed up for 1 year after diagnosis of perianal disease and were assessed for any new symptoms and diagnosis of CD later on. Institutional research and ethics committee clearance was obtained.

RESULTS

A total of 46 patients were diagnosed with perianal disease in the study group. Among this, majority were males constituting to 76.08% (n=35). Major presentation in these patients were skin tags (n=27, 58.6%), anal fissures (n=15, 32.6%) and fistula (n=4, 8.6%). Among patients with fistula, one patient presented with de novo fistula and other 3 had an abscess, which eventually became a non-healing fistula. All patients had a complex fistula. Total duration of symptoms before the identification of perianal lesion was 32 ± 11 days.

The major symptoms of these patients were pain in perianal region (n=38, 82.6%), bleeding PR (n=23, 50%), anal pruritus (n=8, 17.3%) and mucus discharge PR (n=7, 15.2%). Weight loss was present in 8 patients (17.3%). Mean HB in these patients was 11.4 ± 1.7, mean ESR was 48 ± 16, mean CRP was 14 ± 4 and mean albumin was 3.3 ± 0.6. The clinical variables of patient are mentioned in table 1.

Among the study group, 46 patients underwent colonoscopy and lesions in the form of nodularity, aphthous ulcers, pseudopolyps and stricture was seen in only 3 patients. Clinical characteristics of patients with Crohn's disease are shown in Table 2. These 3 patients showed crypt distortion and non-caseating granulomas suggestive of Crohn's disease on biopsy. 14 patients had a combination of lesions, among which Crohn's proven patients had both fissure and fistula. Other 11 patients had combination of skin tag and anal fissure. Patients with fistula underwent MR fistulogram and all showed features of complex fistula. Only 3 patients who had biopsy proven CD showed features of small bowel enhancement and thickening.

	Patients Without CD (n=43)	Patients with CD (n=3)	Mean ± SD in Study Group
Age (in years)	33 ± 17	31 ± 4	39 ± 14
Mean duration of symptoms (in days)	35 ± 16	26 ± 4	32 ± 11
Symptomatology			
Perianal pain	76.7%	100%	82.6%
Bleeding PR	41.8%	66.6%	50%
Perianal pruritus	17.3%	-	17.3%
Mucus discharge PR	17.3%	33.3%	15.2%
Haematological Variables			
HB	11.2 ± 2.1	11.6 ± 0.6	11.4 ± 1.7
WBC count	8100 ± 2400	10800 ± 1200	9500 ± 2300
Albumin	3.5 ± 0.4	3.2 ± 0.2	3.3 ± 0.6
ESR	44 ± 22	58 ± 4	48 ± 16
CRP	8 ± 6	11 ± 2	14 ± 4

Table 1. Clinical Variables of Patients with Perianal Disease

Sl. No.	Symptomatology	Duration of Symptoms (in days)	Colonoscopic Findings
1.	Perianal pain, bleeding PR, anal fissure, skin tags	22	Ileal Ulcer, nodularity
2.	Perianal pain, mucus discharge, weight loss, anal fissure, fistula	28	Ileal ulcer, pseudopolyps, mild stricture at ascending colon
3.	Perianal pain, bleeding PR, anal fissure, fistula	30	Ileal ulcer, nodular and erythematous mucosa

Table 2. Clinical Variables of Patients with Crohn's Disease

DISCUSSION

This study was conducted to determine the prevalence of perianal disease in asymptomatic Crohn's patients and to prospectively follow up in these patients. A total of 46 patients presented with perianal disease in the study period, out of which 3 patients had CD constituting 6.5%. Published literature estimates the prevalence of perianal disease in CD to range from 3.8% to 80%.[3,4,7] Also, perianal disease maybe the first manifestation of Crohn's disease.[7] Our study showed that 6.5% of asymptomatic patients who presented with perianal disease had Crohn's signifying the importance of evaluation in these patients.

All patients with CD, with or without perianal complaints, should at minimum have an external examination of the perianal region. Radiographic modalities for diagnosis include Magnetic Resonance Imaging (MRI), Computerized Tomography (CT), Endoscopic Ultrasound (EUS) and fistulography. Among this, MRI is highly sensitive for fistulae and abscesses. MRI can show soft tissues separately with high distinction and images can be viewed in both coronal and sagittal planes to better delineate landmarks.[8]

Consensus recommendations from the European Crohn's and Colitis Organization support the combination of MRI and proctosigmoidoscopy evaluation in making a diagnosis of perianal fistula and for planning therapy.[9] In our study, we performed colonoscopy in all patients to determine the same and 3 (6.5%) patients had colonoscopic features of Crohn's disease and was useful in obtaining tissue diagnosis.

The major perianal manifestations in our study were skin tags, anal fissures and anal fistula. In one of study by Platell et al, 306 patients with Crohn's and perianal disease were evaluated. Among this, commonest presentations were perianal abscess (29.5%), anal fissure (27.6%) and low anal fistula (26.7%). Also, patients were likely to present with anal symptoms after they had been diagnosed as having intestinal Crohn's disease (46.1%).[10] 3 out of 46 patients with perianal disease were actually Crohn's in our study. This determines the masked presentation of Crohn's in the community. However, these patients are known to present with a flare or as full-blown disease in later life.

Although, natural course of disease is one of chronic relapsing one, diagnosis at the earliest shall improve immediate morbidity and long-term evaluation of these patients. Diagnosis of Crohn's has important clinical implications since without immunosuppressants, these lesions are unlikely to heal. Also, biologicals and biosimilars are the treatment of choice for patients with fistulising Crohn's disease. In a population-based retrospective study, 151 out of 398 fistulising Crohn's patient had only perineal fistula. Odds Ratio (OR) for likelihood of having luminal fistula disease, if perineal disease was present was 5.02. Also, it was shown that compared with luminal fistulas, perineal fistulas had a higher likelihood to have colonic (OR, 3.32; 95% CI, 1.59-6.90; P = 0.002) rather than isolated ileal involvement. This signifies the importance of colonoscopy in these perianal disease patients.

Crohn's colitis is much more frequently associated with an anal lesion than Crohn's disease of the small bowel (52 percent vs. 14 percent).[11] When an anal lesion is the manifesting sign, Crohn's disease will soon develop elsewhere in the intestine and thus it is important to follow up these patients to diagnose Crohn's at the earliest. Since these lesions frequently herald the onset of intestinal Crohn's disease, the physician must always be aware of the possibility of inflammatory bowel disease when dealing with suspicious anal lesions. Literature indicate that perianal manifestations of Crohn's disease pursue a relatively benign course and are rarely an indication for proctectomy.[12] But, this requires prompt identification of disease at the earliest and treating with judicious immunosuppressants.

CONCLUSION

Crohn's disease is an indolent disease with varying presentation. Perianal disease maybe the initial presentation in CD. 6.5% of patients were known to present with perianal disease in our study and determines the importance of screening with colonoscopy, so as to identify CD in these otherwise asymptomatic cases. Skin tags, anal fissures, abscesses, fistula and stricture are few of common perianal presentation of Crohn's disease. Prompt evaluation of these perianal lesions helps in early diagnosis of disease and shall prevent development of long-term complications secondary to Crohn's disease.

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