A 39 year old woman presented with moderate to severe Crohn’s disease of the left colon and also with intermittent severe right lower quadrant abdominal pain for 20 years. She had had multiple Crohn’s disease flares, which had been treated with steroids and antibiotics. However, her intermittent severe right lower quadrant abdominal pain persisted and intensified. It had been presumed, but unable to be proven, that her right lower quadrant abdominal pain was due to Crohn’s disease of the ileum. The patient had localized right lower quadrant abdominal tenderness without guarding or rebound. CBC demonstrated mild anemia and thrombocytosis. ESR and CRP were normal. Abdominal CT scan demonstrated evidence of left sided colitis and possible inflammatory changes in the right lower quadrant. Colonoscopy revealed mild to moderate left sided colitis, but no evidence of Crohn’s disease of the right colon or terminal ileum. The patient was treated with steroids and antibiotics, but continued to have very severe symptoms elsewhere which were not due to Crohn’s disease.

Background

*Primary tumors of the appendix are uncommon and are usually not included in the differential diagnosis when a patient presents with Inflammatory Bowel Disease (*IBD*) presents with either acute or chronic right lower quadrant abdominal pain (1,2).

*Appendiceal neuromas are proliferations of neural tissue and are categorized as one of the causes of fibrous obliteration of the appendix (3-7).*

*We present a patient with left sided colonic Crohn’s disease, who also had 20 years of intermittent severe right lower quadrant pain; the pain was thought to be due to Crohn’s disease involving the terminal ileum, but was due to an appendiceal neuroma (fibrous obliteration of the appendix).*

Case Presentation

A 39 year old woman presented with moderate to severe Crohn’s disease of the left colon and also with intermittent severe right lower quadrant abdominal pain for 20 years. She had had multiple Crohn’s disease flares, which had been treated with steroids and antibiotics. However, her intermittent severe right lower quadrant abdominal pain persisted and intensified. It had been presumed, but unable to be proven, that her right lower quadrant abdominal pain was due to Crohn’s disease of the ileum. The patient had localized right lower quadrant abdominal tenderness without guarding or rebound. CBC demonstrated mild anemia and thrombocytosis. ESR and CRP were normal. Abdominal CT scan demonstrated evidence of left sided colitis and possible inflammatory changes in the right lower quadrant. Colonoscopy revealed mild to moderate left sided colitis, but no evidence of Crohn’s disease of the right colon or terminal ileum.

The patient was treated with steroids and antibiotics, but continued to have very severe right lower quadrant abdominal pain. An exploratory laparotomy was undertaken, but revealed no evidence of Crohn’s disease involving the terminal ileum or right colon. Despite a normal appearing appendix, an appendectomy was performed due to the 20 year history of unexplained intermittent severe right lower quadrant abdominal pain. Pathology of the appendix revealed no evidence of active appendicitis or Crohn’s disease, but did demonstrate luminal fibrous obliteration, consistent with an appendiceal neuroma. Following the appendectomy, the patient felt markedly improved for the past four years, with complete resolution of her intermittent severe right lower quadrant abdominal pain.

Discussion

Appendiceal neuromas (fibrous obliteration of the appendix) contain spindle cells with eosinophilic processes within a fibrous or fibroadipose stroma (1 – 6). Appendiceal neuromas are rare, and in one study of 8,699 patients who underwent appendectomy, only 5 appendiceal neuromas were discovered out of a total of 103 appendiceal tumors (2). Serotonin and substance P have been found in the nerve fibers within appendiceal neumomas (fibrous obliteration of the appendix), and it has been hypothesized that these neuropeptides may promote muscular spasticity and abnormal peristalsis leading to abdominal pain in some patients (4 – 6). The association of appendiceal neuromas with clinical symptoms, such as abdominal pain, has been unclear (1, 2). Most appendiceal neuromas are found incidentally in asymptomatic patients, after pathological examination of the appendix reveals fibrous obliteration (7, 8). However, in chronic appendicitis the incidence of appendiceal neumomas has been reported to be as high as 51.2% (3). In some cases, fibrous obliteration of the appendix (appendiceal neuroma) is the only finding that would explain the symptoms of what clinically was thought to be chronic appendicitis (3 – 5). Most appendices removed from IBD patients are involved with chronic inflammation due to Crohn’s disease or ulcerative colitis (9). In one study of 41 children with Crohn’s disease or ulcerative colitis who underwent appendectomy, 33 out of 41 (80%) appendices were abnormal with pathologic changes consistent with inflammatory bowel disease (9). Non-specific changes including fibrous obliteration (appendiceal neuroma), exudate fibrosis, lymphoid hyperplasia and neuronal hyperplasia were observed in 8 of 41 (20%) of the appendices (9). Both carcinoid tumors and mucinous cystadenomas are also well described as occurring in the appendices of patients with IBD (10, 11).

Conclusion

*Clinically significant appendiceal neuromas may present with debilitating chronic intermittent right lower quadrant abdominal pain, which may be confused with Crohn’s disease.*

*Although Crohn’s disease may be present in one part of the intestine, severe symptoms elsewhere can occur which are not due to Crohn’s disease.*

*For 20 years, this Crohn’s disease patient had intermittent severe right lower quadrant abdominal pain not due to Crohn’s disease, but rather due to an appendiceal neuroma.*

*Since her appendectomy four years ago, the patient has experienced complete remission from her previous intermittent severe right lower quadrant abdominal pain.*

References