Crohn's disease – is there a long latent period?

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Summary: Four cases of Crohn's disease developing in adult females, who had been in close contact during their teens, are described. It is suggested that this could indicate an infective causation for Crohn's disease with a long latency.

Introduction

The aetiology of Crohn's disease remains obscure despite substantial research endeavour. Environmental factors are considered important as there is a higher incidence in urban rather than rural dwellers (Kyle, 1971; Humphreys & Parks, 1975) and Crohn's disease is more common in North America and Northern Europe than in other parts of the world. There is some genetic predisposition with a small increased risk in siblings (Mayberry et al., 1980; Weterman & Peña, 1980). Laboratory attempts at animal passage looking for a possible transmissible agent have been successful in the hands of a few workers (Mitchell & Rees, 1970; Cave et al., 1975) but on the whole this approach has been disappointing and the evidence is as yet unconvincing. Epidemiological studies failed to show any direct case-to-case infection and Mayberry & Newcombe (1981) in a survey of nurses with Crohn's disease did not find any evidence that they had more exposure to Crohn's disease patients compared with other nurses. A search for time space clustering in the Nottingham area also yielded negative results (Miller et al., 1975). However, both of these workers concluded that an infection with a long latent period could not be excluded by the evidence available. We would like to report a small cluster of Crohn's disease in four adult women who had been in prolonged contact throughout their secondary school days.

Case reports

Four women presented with Crohn's disease in adult life. They had been brought up in a rural area and three of them attended secondary school together for seven years from the age of 11. The fourth had a close friendship and contact with the group in after-school activities. None of the girls gave any history of bowel symptoms in their teens and had no period of prolonged ill-health. On leaving school at 18 years, three of them moved out of the area for further education and subsequent employment, only returning after their Crohn's disease was established.

The ages at first presentation were 21, 23, 25 and 30 years. All of them had radiological and histological features of the disease. Three have needed resection of the terminal ileum and caecum and the fourth has had Crohn's disease involving the terminal ileum, transverse and part of the descending colon but has not required surgical intervention.

The three patients who required bowel resection had appendicectomies prior to the diagnosis of Crohn's disease and in one a review of the appendix histology was suggestive of Crohn's disease.

Case 1

This girl remained well until the age of 21 when she developed episodic diarrhoea, weight loss, abdominal pain with anorexia, and erythema nodosum. A barium meal showed Crohn's disease of the terminal ileum with involvement of the transverse and upper half of the descending colon. Rectal biopsy confirmed the diagnosis. The symptoms settled with parenteral steroids and over the subsequent 15 years she has had a number of exacerbations which have been helped by steroids. She is now off treatment and her weight is constant.

Case 2

This patient had an appendicectomy at the age of 21. At the age of 30 she presented with crampy pain in the right iliac fossa, diarrhoea, weight loss and malaise. Barium meal and barium enema at that time were normal but her symptoms gradually got worse and her weight continued to fall. Her diarrhoea became more persistent and she developed an anal fissure. In the
following year she came to surgery when she was found to have extensive Crohn's disease with narrowing and stricturing of the terminal ileum and involvement of the ascending colon. She had resection of part of the terminal ileum with the ileocaecal valve and ascending colon. Two years later she needed further surgery with further resection of the terminal ileum and the formation of an ileostomy. She has needed surgery on a number of occasions for abdominal and perianal fistulae. Currently she is fairly well controlled with sulphasalazine.

Case 3

When aged 22 this woman developed abdominal pain, diarrhoea and had an appendicectomy. Histology showed follicular aggregation of mononuclears with surrounding areas of caseation or necrosis suggesting either Crohn's disease or tuberculosis. The symptoms continued post-operatively. Barium meal and follow-through examination showed Crohn's disease of the terminal ileum. She was treated successfully with sulphasalazine and prednisone but had to discontinue sulphasalazine because of a rash. The following year she developed an intestinal obstruction and at surgery a fibrous band was divided in the terminal ileum although the bowel appeared to be normal. Two years later she had flare-up of symptoms with a palpable mass in the right iliac fossa and barium enema was suggestive of Crohn's disease. She subsequently had a number of exacerbations and 12 years from the onset she had a severe exacerbation requiring resection of the caecum and terminal ileum. Currently she is well maintained on a small dose of prednisone.

Case 4

At the age of 25 this woman presented with appendicitis and at operation Crohn's disease of the terminal ileum was found and the appendix was removed. One year later she had an acute exacerbation of Crohn's disease and needed resection of the terminal ileum, caecum and part of the ascending colon. She has remained reasonably well although she has had a number of minor exacerbations and currently she is on a small dose of steroids.

Discussion

Regional variations in prevalence and increased incidence in relatives are suggestive of an infective cause in Crohn's disease. Adult infection, however, is unlikely as there are only three or four known husband and wife pairs with the disease and a study has demonstrated that nurses who look after Crohn's patients are not at increased risk (Mayberry & Newcombe, 1981). Childhood infection with a long latency or some form of slow virus infection is a possibility. We feel that this small cluster could suggest this type of infection although this would indicate a latent period of at least four and perhaps greater than 11 years. There is some evidence in favour of other diseases such as sarcoidosis and Hodgkin's disease being caused by an infectious agent with a long latency and the failure to identify an infective agent in Crohn's disease may be because of such a latent period.

References


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