Rectal and gastric involvement in a case of sarcoidosis

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SUMMARY A case of sarcoidosis with involvement of the rectum is described. This is believed to be the first unequivocal case of large bowel sarcoidosis.

Involvement of the intestinal tract by sarcoidosis is rare. Scadding (1967) reports no record of involvement of the large intestine in established sarcoidosis; Mayock, Bertrand, Morrison, and Scott (1963) report no such involvement in 145 cases of histologically proven sarcoidosis. However, the observations of Palmer (1958) suggest that gastric involvement may not be as uncommon as previously thought. He noted histological evidence of sarcoid involvement in six out of 60 gastric biopsies in patients with other evidence of sarcoidosis but without gastrointestinal symptoms and with normal barium meal and gastroscopy findings.

We report a case of sarcoidosis with involvement of the rectum, stomach, liver, spleen, and lungs.

Case Report

A 29-year-old Jamaican male presented in January 1971 with a six-month history of cough, initially dry, but subsequently productive of mucoid sputum. There were no abnormal physical signs on examination but a chest radiograph showed bilateral hilar lymphadenopathy with scattered bilateral pulmonary infiltration. Investigation at this time showed a negative Heaf test, an ESR of 47 mm/hr, and an increase in the serum alpha and gamma globulins. Pulmonary function tests showed a restrictive pattern, FVC 2.54 litres (predicted 4.85 litres); FEV 90% (predicted 84%); peak flow 545 litres/min (predicted 560 litres/min) and a marked reduction of gas transfer (steady state method 10.6 ml CO/min/mmHg—predicted normal 22.5 ml CO/min/mmHg).

In the following five months the patient became

less well with cough, chest pain, night sweats, and weight loss. He developed a pleural rub with some increased opacity at the left base; there was also a

Fig 1 Mediastinal lymph node biopsy showing typical sarcoid granulomata

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Fig 2 Gastric mucosal biopsy from greater curve showing giant cell granulomata

Fig 3 Rectal biopsy showing submucosal collection of epithelioid cells forming a tubercle-like structure

further reduction in gas transfer. The sputum was again negative for acid-fast bacilli on both smear and culture. To confirm the diagnosis mediastinoscopy was performed and one of a number of enlarged paratracheal and parabronchial lymph nodes was biopsied. This showed a marked sarcoid reaction without necrosis (fig 1). No acid-fast bacilli were seen on direct smear nor isolated on culture. He was treated with oral prednisone for the next six months in varying dosage, but without obvious benefit. At this time, poorly localized abdominal pain became a prominent symptom and he was found to have developed an enlarged liver and spleen. Further investigations gave the following results: ESR 63 mm/hr, serum calcium 9.1 mg/100 ml, serum phosphorus 2.9 mg/100 ml. The alkaline phosphatase was raised (198 KA units/100 ml) as was the serum 5 nucleotidase (44 IU/litre) but bromsulphthalein retention was normal (3% of standard dose at 45 min). The prothrombin time was also prolonged; prothrombin ratio 1.4. A sickle cell test was negative; a Kveim test (K12 antigen, Colindale Public Health Laboratory) was also negative. Barium enema examination was normal but a barium meal on two occasions showed a fine diffuse irregularity of the gastric mucosa, especially on the greater curve. Gastroscopy revealed endoscopic gastritis but no ulceration and biopsy (greater curve) showed unequivocal giant cell granulomata without caseation or acid-fast bacilli (fig 2). The perineum appeared normal as was the rectal mucosa at sigmoidoscopy but a rectal biopsy at 9 cm revealed a submucosal collection of epithelioid cells forming a tubercle-like structure (fig 3) similar to that in the gastric biopsy. A barium follow-through examination and a jejunal biopsy were normal.

Comment

This patient has pulmonary sarcoidosis presenting with bilateral hilar lymphadenopathy and involvement of the lung parenchyma. Abdominal pain of indefinite character but often in the lower abdomen has been a prominent feature of this patient's illness. Histological evidence is presented of rectal and gastric involvement with sarcoidosis. The association between the abdominal symptoms and the intestinal sarcoidosis is not clear, but Scott, Smith, Cox, and Palmer (1953) report a patient with sarcoid involvement of the stomach and lachrymal glands in whom vague intermittent abdominal pain was prominent. Davies (1972) reports severe abdominal pain of several years' duration in a 46-year-old man who, 14 years previously, had had resolving bilateral hilar lymphadenopathy with pulmon-

ary infiltration. Laparotomy for the abdominal pain showed enlarged coeliac lymph nodes with sarcoid granulomata on microscopy. There was no evidence of Crohn's disease or of tuberculosis.

With the possible exception of a patient reported by Aaronson, Meir, and Ulin (1957), no previous well documented case of sarcoid involvement of the large bowel or rectum has been described. Three authors have described non-caseating granulomata found incidentally at laparotomy and claimed these to be evidence of sarcoidosis of the bowel (Raven, 1949; MacFarlane, 1955; Gourevitch and Cunningham, 1959). Similarly Allen, Batten, and Jefferson (1956) describe a patient with proven gastric sarcoidosis but infer caecal involvement on radiological grounds only. Aaronson *et al* (1957) reported the case of a 37-year-old negro female with generalized sarcoidosis, the diagnosis being based on radiographs of the chest, hands, and a skin biopsy. She presented with constipation and blood-streaked stools and barium enema revealed a lesion of the lower sigmoid colon which was thought to be an annular carcinoma. On resection the lesion was a localized stenosis with granulomatous inflammation of the mucosa and submucosa and diffuse submucosal fibrosis. The precise relationship between this lesion and previous intracavity uterine irradiation is speculative. This was claimed as a possible case of colonic sarcoidosis although Crohn's disease could not be excluded.

Many pathological conditions can cause granulomatous lesions in the alimentary tract (James and Sharma, 1967) and two granulomatous diseases can coexist as in the case of a 23-year-old white male with both sarcoidosis and typical regional ileitis

reported by Padilla and Sparberg (1972). Plainly all that is granulomatous is not sarcoidosis! We do not think that lesions of the bowel should be labelled sarcoidosis in the absence of good evidence of this disease process elsewhere. However, we feel that the rectal granulomata in our present case really do represent an unusual manifestation of sarcoidosis.

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