Crohn’s disease of the lung

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Abstract

Two years after developing colonic Crohn’s disease, a 17 year old boy presented with focal pulmonary consolidation. A lung biopsy specimen showed areas of non-caseating epithelioid granuloma. Although some respiratory abnormalities appear to be associated with inflammatory bowel disease, granulomatous disease affecting the lung has not previously been reported in a child.

Crohn’s disease appears to be a multisystem disorder, but granulomatous pulmonary involvement has not been previously reported. We present our experience with a teenage boy.

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Discussion

We believe this to be the first case of kala-azar to be diagnosed in a child in Scotland. Because of inexperience with the disease and a degree of scepticism regarding the diagnosis, treatment of the condition was delayed by three weeks during which time the girl was subjected to a number of unnecessary investigations. In these days of global travel, with so many British tourists spending their holidays in the Mediterranean, it is likely that more cases of leishmaniasis will be seen in Britain because of the long incubation period.

The condition should be included in the differential diagnosis of any child or young adult, presenting with massive splenomegaly, severe anaemia, and thrombocytopenia who has been in an endemic area up to three years previously. Negative leishmaniasis serology may not exclude the diagnosis during the initial stages of the disease as it may take a few weeks before becoming positive.

patchy consolidation in the left lower lobe and when symptoms failed to respond to treatment with oral amoxycillin, admission to hospital was arranged for further investigations to be performed. He was noted to be pale and unwell with a persistent cough and fever. Radiological examination confirmed that the consolidation in the posterior segment of the left lower lobe was still present. In addition, there was a small pleural effusion which, like sputum, was sterile on culture. After treatment with erythromycin there was some improvement in symptoms, but several weeks later he became worse, with a productive cough and dyspnoea on moderate exercise.

INVESTIGATIONS
Investigations showed him to be anaemic (haemoglobin concentration 99 g/l) with a raised erythrocyte sedimentation rate (90 mm in the first hour) and plasma orosomucoids (2 g/l, normal <0·5 g/l). Repeated microbiological examinations of sputum, blood, gastric washings, and urine proved negative. Mantoux testing (1/1000) was also negative. A radioisotope lung scan indicated a matched ventilation/perfusion defect in the left lower lobe, and a thoracotomy with open lung biopsy was performed. The histological findings were of multiple focal areas of non-caseating epithelioid granuloma containing multinucleated giant cells in the interstitium, peribronchiolar, and perivascular regions (fig 2). Staining for micro-organisms including acid fast bacilli was negative as was tissue culture for mycobacteria; a Kveim test was subsequently negative.

CLINICAL PROGRESS
Treatment with isoniazid 300 mg and rifampicin 450 mg each day was started. Chemotherapy was discontinued three weeks later when all microbiological cultures proved negative. Steroids were not used. Fever and radiological abnormalities completely resolved over a further three weeks. In the next five years three relapses of Crohn's disease occurred with abdominal pain, weight loss, and diarrhoea, but without respiratory symptoms.

Discussion
Although Crohn's disease appears to be a multi-system disorder,1 we know of no previous reports of granulomatous pulmonary involvement during childhood. Tuberculosis was initially considered the most likely diagnosis but was excluded by the absence of caseating granuloma, negative Mantoux tests, failure to identify mycobacteria histologically or on culture, and remission of symptoms after only a few weeks of antituberculous treatment. Sarcoidosis remains a possible alternative diagnosis, but is unlikely in view of the negative Kveim test. Chronic granulomatous disease was not investigated by tests of white cell function but would be very unusual in a boy who was well for the first 15 years of life.

Even though symptomatic pulmonary involvement in Crohn's disease may be rare, some studies have suggested that latent pulmonary involvement is a common finding.2 Inflammatory bowel disease has also been linked with respiratory disorders such as chronic bronchitis (in the absence of smoking) and bronchiectasis.3 Carbon monoxide transfer factor appears to be significantly reduced in patients with both Crohn's disease and ulcerative colitis when compared with control patients,4 although some workers have disputed this finding.5 It has been suggested that morphological and developmental similarities between bronchial and colonic epithelium may make both susceptible to damage from an unidentified humoral factor, or they might together be unduly sensitive to some external agent both inhaled and ingested.6 The findings in our patient indicate that when Crohn's disease is complicated by granulomatous reactions in organs other than the bowel, the lung too may be involved.

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