Genital Crohn's disease in a 6-year-old boy

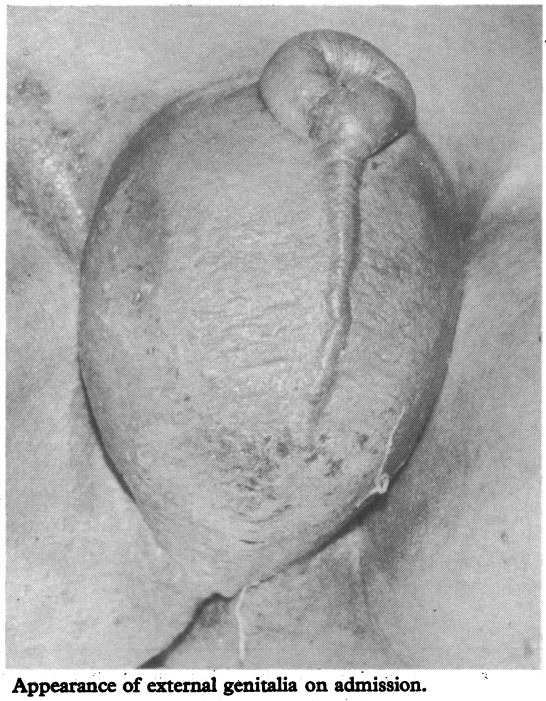
The male external genitalia may occasionally be affected in Crohn's disease; this almost invariably occurs as ulceration extending forwards from the perianal and perineal area on to the scrotum.[1] We describe the case of a 6-year-old boy who presented with diffuse infiltration of the scrotum and foreskin due to Crohn's disease.

Case report

The patient had previously been admitted to another hospital at the age of 4 years for investigation of malaise, angular cheilitis, intermittent rectal bleeding, and painful defecation. Examination had shown several anal fistulae and an anal fissure. The fissure had been excised, and histological examination showed granulation tissue containing non-caseating epithelioid cell granulomas. Crohn's disease had been provisionally diagnosed. Spontaneous improvement occurred, and further investigation was deferred. After remaining well for 18 months, the boy developed recurrent swelling of the external genitalia. Episodes of increased swelling were accompanied by fever and tenderness with erythema of the scrotum and foreskin. The development of phimosis during one of these exacerbations necessitated circumcision. Histology of the excised foreskin showed numerous non-caseating epithelioid cell granulomas beneath a thinned epidermis, compatible with a diagnosis of Crohn's disease.

When admitted to this hospital at the age of 6½, the boy's height and weight were both on the 50th centile. An angular cheilitis was present, and there were granular, oedematous lesions on the labial gingival mucosa. The scrotum and foreskin were grossly swollen and indurated (see figure). The overlying skin was red and hot, and a few small vesicles and exudative lesions were present. Inguinal lymphadenopathy was absent, and physical examination was otherwise normal. Except for a raised erythrocyte sedimentation rate (34 mm in first hour) all haematological and biochemical investigation results were normal. There was no radiological evidence of large or small bowel disease. The Mantoux test (1/1000) gave a negative result. Nose, throat, and scrotal skin swabs grew Lancefield group A β-haemolytic streptococci.

Rectal examination and sigmoidoscopy showed an anal fissure and a polypoid lesion in the anal canal; otherwise the appearances of the anal canal and rectum were normal. Biopsy of the anal fissure showed pronounced inflammation of the lamina propria with a dense infiltrate, mainly of plasma cells and lymphocytes. Histology of the anal polypoid lesion showed a similar diffuse subepithelial inflammatory infiltrate, together with prominent lymphoid aggregates containing germinal centres and non-caseating epithelial cell granulomas with giant cells. Biopsy specimens of the rectal mucosa showed patchy mucosal and submucosal inflammation with oedema and scattered non-caseating epithelioid cell granulomas. Biopsy of the gingival lesion showed a non-specific subepithelial chronic inflammatory infiltrate.



Appearance of external genitalia on admission.

Crohn's disease was diagnosed and treatment started with oral prednisolone 30 mg daily. Before he started on steroid treatment there had been some reduction of the genital swelling with bed rest, scrotal support, and a course of oral phenoxymethylpenicillin to eradicate the β-haemolytic streptococci. Subsequently the genital swelling decreased rapidly until, three weeks after starting oral corticosteroids, the genitalia appeared normal. They have remained so, apart from minor fluctuations, while the dosage of prednisolone has been reduced over five months to 10 mg on alternate days. Because of a tendency for early recolonisation of the skin by the same organism, phenoxymethylpenicillin is being given prophylactically (250 mg twice a day).

Comment

The diagnosis of Crohn's disease in this patient rests on the histological finding of non-caseating epithelioid and giant-cell granulomas in the prepuce, in association with similar findings in the rectum and anal canal. It is supported by the concurrent presence of typical lesions of Crohn's disease in the perianal area and gingival mucosa. The involvement of the external genitalia appears to be of a type that has not previously been described.

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[1] Mountain, J C, *Gut*, 1970, **11**, 18.

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Hazard of chemical sympathectomy

Since Diez[1] first described lumbar sympathectomy in 1924 it has proved most effective for treating ischaemia of the leg, especially in patients not suited for arterial reconstruction.[2] Such patients usually have widespread disease affecting coronary and cerebral vessels and may therefore be at considerable operative risk. For this reason chemical sympathectomy, by percutaneous injection of an aqueous solution of phenol, has become popular in several centres. We have now performed over 50 injections during the past three years using the technique described by Reid *et al.*[3] We report our first serious complication.

Case report

A 70-year-old woman presented with increasing dyspnoea, ankle swelling, and pain in her right foot. She had been treated for some months with steroids for suspected fibrosing alveolitis, and had secondary polycythaemia. Right ventricular failure was present on admission. No pulses were palpable in the right leg and there were pregangrenous changes in the toes of her right foot. Control of her cardiac failure, and treatment with venesection, intravenous naftidrofuryl (Praxilene), and dextran 40 produced some improvement initially in her right foot. Later, however, deterioration with severe rest pain prompted us to carry out chemical sympathectomy. Two injections of 2·5 ml of 6·7% aqueous phenol were made in the region of the right sympathetic chain at the levels of the third and fourth lumbar vertebrae. No blood or cerebrospinal fluid was obtained on aspirating at intervals before and during the injection. She made no complaints until two and a half hours after the procedure, when she commented that she had some weakness and anaesthesia of her legs. Examination confirmed that a satisfactory right lumbar sympathetic block had been achieved, but there was also anaesthesia and paresis on the right, corresponding to the nerve roots from L3 downwards, and on the left from L5 downwards. Only very limited neurological improvement took place during the 32 days before she died from bronchopneumonia.

The necropsy report by Drs A Gordon and A Busuttil stated that no macroscopic abnormality was observed in the cord or meninges. On microscopical examination there was demyelination and some axonal degeneration affecting predominantly the dorsal roots from T11 to the conus. The demyelination tended to be most definite at the periphery of each root. There