# SHORT REPORTS

## Sarcoidosis and multiple myeloma: an association

An association between sarcoidosis and neoplasia has been suggested.[1] The development of malignant lymphoma has been reported in several patients with sarcoidosis,[2] but there have been few reports of patients with sarcoidosis and multiple myeloma.[2-4] We describe five patients with sarcoidosis who developed multiple myeloma.

### Case histories

The table shows details of five patients with sarcoidosis seen by us during 1969-86. In each case the diagnosis of sarcoidosis was based on both the clinical findings and histological evidence of non-caseating epithelioid cell granulomas. The mean age of the patients at the time of diagnosis was 55 (range 42-65). In four patients myeloma developed several years after the diagnosis of sarcoidosis; in one both diagnoses were made at the same time. The duration of sarcoidosis was more than two years in three patients (cases 1, 2, and 4), and sarcoidosis was considered to be active at the time of diagnosis of myeloma in two (cases 2 and 5). Two patients (cases 1 and 2) had been treated with steroids.

All patients met the diagnostic criteria for multiple myeloma. Four patients (cases 1-4) were treated with cytotoxic drugs and the other (case 5) was followed up without treatment. Four patients survived, but the other (case 3) died in 1985, four years after the diagnosis of myeloma.

### Comment

All our patients showed features consistent with sarcoidosis and were not thought to have had a sarcoidosis like reaction, such as has been described in association with malignant tumours, among them lymphoma and myeloma. In four of the cases the long time interval between the diagnoses (three to 12 years) increases the chance that these patients had two separate disease processes. In the one patient who presented with sarcoidosis and myeloma at the same time it is of course conceivable that both disorders were due to the same disease process.

The mean age of our patients at the time of diagnosis of sarcoidosis was appreciably higher than the average age at which sarcoidosis is diagnosed. We thus agree with Brincker that it is the chronic active type of sarcoidosis that develops in middle aged patients that is associated with the subsequent development of neoplastic disease.[2]

Proof of a causal rather than a merely coincidental association between sarcoidosis and myeloma may be obtained only by prospective epidemiological studies. Both diseases, however, are comparatively rare, the incidence of sarcoidosis in Finland varying from 5 to 21/100 000 and that of myeloma being about 3/100 000. On the basis of these figures, the expected number of cases of both sarcoidosis and myeloma in the Finnish population (somewhat below five million) during a 10 year follow up is less than 0·3, which suggests a very low chance of the two disorders occurring by coincidence.

Although the aetiology of multiple myeloma is unknown, evidence suggests that chronic stimulation of the immune system may be a predisposing factor, presumably in combination with genetic or viral factors.[5] Sarcoidosis is a chronic granulomatous disease, also of unknown aetiology, that has pathogenetic features that may predispose to the development of lymphoproliferative diseases, such as myeloma. In sarcoidosis activated T helper/inducer lymphocytes in the sites of inflammation cause a polyclonal stimulation of B lymphocytes, which may result in hypergammaglobulinaemia. Prolonged stimulation of B lymphocytes together with a disturbance in the regulation of the immune system might result in the development of an autonomous plasma cell clone producing a monoclonal immunoglobulin. Alternatively, a common primary immunological derangement or a common aetiological factor might underlie the development of both sarcoidosis and myeloma in certain people.

*Clinical data on five patients with sarcoidosis and multiple myeloma*

| Case No | Age* (years) and sex | Time between diagnoses (years) | Type of myeloma | Features consistent with sarcoidosis | Evidence for myeloma |
|---|---|---|---|---|---|
| 1 | 42 M | 6 | IgG λ | Fever, hilar adenopathy, positive result on Kveim testing | Bone marrow plasmacytosis >90%; monoclonal IgG 69·8 g/l; λ chain excretion 1·8 g/day; lytic bone lesions; reduction in IgA and IgM |
| 2 | 58 M | 6 | IgA κ | Hilar adenopathy, uveitis, parotid enlargement and biopsy findings | Bone marrow plasmacytosis 70%; monoclonal IgG 51·7 g/l |
| 3 | 56 M | 3 | IgG κ | Breathlessness, arthralgia, hilar adenopathy, lung parenchymal infiltration, scalene node biopsy findings | Monoclonal IgG 65·5 g/l; κ chain excretion 1·5 g/day; bone marrow plasmacytosis 10%; lytic bone lesions; reduction in IgA and IgM |
| 4 | 65 F | 12 | IgG λ | Mediastinal node enlargement and biopsy findings | Bone marrow plasmacytosis 30%, monoclonal IgG 42·6 g/l; λ chain excretion 3·0 g/day; lytic bone lesions; reduction in IgA and IgM |
| 5 | 60 F | None | IgG λ | Erythema nodosum, hilar adenopathy, bronchial mucosa biopsy findings | Bone marrow plasmacytosis 50%; monoclonal IgG 17·5 g/l; reduction in IgA and IgM |

*Age at onset of sarcoidosis.

1 James DG, Sharma OP. Overlap syndromes with sarcoidosis. *Postgrad Med J* 1985;61:769-71.
2 Brincker H. The sarcoidosis-lymphoma syndrome. *Br J Cancer* 1986;54:467-73.
3 Selroos O, Brander L, Virolainen M. Sarcoidosis and myeloma of lambda-type IgG. *Acta Med Scand* 1974;195:59-63.
4 Schafer AI, Miller JB. Association of IgA multiple myeloma and pre-existing disease. *Br J Haematol* 1979;41:19-24.
5 Isobe T, Osserman EF. Pathologic conditions associated with plasma cell dyscrasias: a study of 806 cases. *Ann NY Acad Sci* 1971;90:507-18.

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**Fourth Department of Medicine, Helsinki University Central Hospital, 00170 Helsinki, Finland**
TOM PETTERSSON, MD, senior physician

**Department of Medicine, Tampere University Central Hospital, Finland**
ELLI KOIVUNEN, MD, senior physician

**Lapland Central Hospital, Rovaniemi, Finland**
MIRJA ILVONEN, MD, internist

**Seinäjoki Central Hospital, Seinäjoki, Finland**
JUSSI JOUPPILA, MD, internist

**Härmä Hospital, Alahärmä, Finland**
ERKKI AALTO, MD, chest physician

**Finnish Leukaemia Group, Vasa**
CURT WASASTJERNA, MD, professor

Correspondence to: Dr Pettersson.

## Acute mesenteric ischaemia: a presenting feature of coeliac disease?

We describe two patients with acute mesenteric ischaemia requiring extensive small bowel resection. Both had a prolonged and difficult convalescence that was presumed to be due to the short bowel syndrome. Underlying coeliac disease was later diagnosed in both cases and presumably contributed towards their management problem.

### Case reports

CASE 1

A 37 year old woman presented with a two week history of abdominal pain and vomiting. Clinical examination showed features of peritonitis. At laparotomy there was small bowel infarction thought to be due to mesenteric venous thrombosis. The infarcted bowel was resected and primary anastomosis per-