Discussion

Initially thrombolytic therapy for mechanical valve obstruction was reserved for right-sided valves[5]; the risk of cerebral and/or systemic emboli derived from the dissolution of thrombus on aortic and mitral prostheses being considered too great. However, by 1988, 58 patients with left-sided thrombosed valves had been treated, with an 18% incidence of systemic emboli; massive irreversible cerebral damage was uncommon[6]. The overall mortality was 15% with 22% requiring early re-operation after thrombolysis in whom there was an operative mortality of 31%. Two-thirds of patients are successfully treated by thrombolytic therapy alone[6]. Streptokinase is the most commonly used thrombolytic agent but in this patient it was ineffective. Streptokinase treatment was not monitored with fibrinogen levels, laboratory measurements of thrombolysis have been shown to be poor predictors of the success of thrombolytic therapy for this condition[7]. The secondary use of r-tpa immediately following streptokinase therapy is uncommon; in two large case studies surgery was performed on all cases of thrombolytic failure[8,9] with a 36% mortality. Acute or sub-acute valve thrombosis can occur at any time post surgery but in two large studies the median time from implantation to dysfunction was 30 months[9] and 54 months[4]. This typical late thrombosis of disc valves has been attributed to a lapse in systemic anticoagulation, although 52% of patients with thrombosed valves are adequately anticoagulated[4]. The development of atrial fibrillation, albeit of short lived duration and in the presence of more than adequate anticoagulation, was almost certainly the event that precipitated the valve thrombosis; indeed atrial fibrillation and left atrial enlargement have been shown to be additional risk factors[9]. Acute prosthetic valve thrombosis is associated with a high mortality and treatment to date has been surgical. We believe thrombolytic therapy is an effective treatment and should be considered in such patients.

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Sarcoidosis-lymphoma syndrome

S Oliwiecki MRCP[1] **B Kotecha** FRCS[2]
T Kingston MD MRCP[1] **M P Rothera** FRCS[2]
[1]*The Skin Hospital, Chappel Street, Salford M60 9EP and* [2]*Hope Hospital, Eccles Old Road, Salford M6 8HD*

Keywords: sarcoidosis; lymphoma, non-Hodgkin's

Case presented to Section of Dermatology, 20 December 1990

Sarcoidosis is a multisystem disorder of unknown cause. It has been reported in association with lymphoma in the past - the sarcoidosis-lymphoma syndrome[1]. We report a case of cutaneous sarcoidosis in association with T-cell lymphoma of the palate.

Case report

The patient, a 49-year-old aniline dye worker first presented in July 1986 with a facial rash which was diagnosed as acne rosacea. The rash subsequently faded. He remained well until August 1989 when he developed painless nodules on the ears and hands.

A skin biopsy showed moderately well formed granulomata scattered through the dermis. A diagnosis of cutaneous sarcoidosis was made.

The skin lesions became more numerous, painful and ulcerated. Treatment with methotrexate 10 mg weekly was commenced. Three weeks later he presented to the accident and emergency department with severe right-sided facial pain and headache. At the same time he noticed a painful mass on the hard palate (Figure 1). He was admitted for investigation.

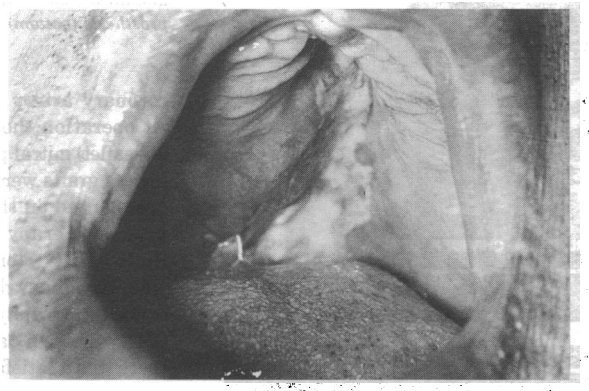

Figure 1. Lesion on hard palate

Routine blood count, urea, electrolytes, liver function tests, serum calcium and chest X-ray were normal. 24-hour urinary calcium excretion was raised at 9 mmol/day (normal 2.5-7.5 mmol/day). Angiotensin converting enzyme (ACE) level was raised at 77 iu/l (normal 19-55 iu/l). X-ray of facial sinuses showed opacification of the right maxillary sinus but no bony destruction.

A biopsy of the palatal mass was performed under local anaesthetic. Histological examination showed the presence of fungal hyphae within the specimen. A diagnosis of probable invasive aspergillosis was made, Actinomycosis was considered as a differential. He was treated with

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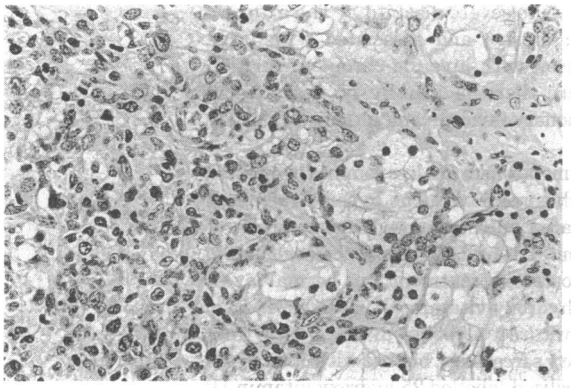

Figure 2. Photomicrograph of palatal lesion. Diffuse non-Hodgkin lymphoma consisting of small, medium and large cells (H&E, ×450)

amoxycillin 1 g three times a day, metronidazole 400 mg three times a day and itraconazole 200 mg daily. A second palatal biopsy taken 2 weeks later showed necrotic tissue only. The right maxillary sinus was drained under general anaesthetic, producing some relief of the patient's symptoms.

The palatal mass enlarged despite treatment and a palatal fistula developed. Further biopsies were taken under general anaesthetic. Histological examination (Figure 2) reveals the presence of a diffuse non-Hodgkin lymphoma consisting of small, medium and large cells. The small cells have irregular nuclei and stain positively with the T-cell markers UCHL-1 and CD3. A diagnosis of diffuse, high grade, T-cell non-Hodgkin lymphoma was made.

He was treated with radiotherapy and combination chemotherapy and responded well to treatment.

Discussion

The sarcoidosis-lymphoma syndrome was first described in 1986 by Brincker who reported 17 cases. Brincker calculated that lymphoma was 5.5 times more common in patients with sarcoidosis than expected. The syndrome tends to occur in older patients (the median age was 41 years, approximately 10 years higher than in sarcoidosis generally) and is associated with persistent disease activity, anergy and lymphopenia. All types of lymphoma are reported but Hodgkin's disease predominated. Other malignancies are also more common in patients with sarcoidosis[2].

The reason for this association between malignancy and sarcoidosis is unknown. However, sarcoidosis is known to be associated with significant immunological abnormalities such as an increased number of T-helper cells in granulomatous tissues, a decreased number of circulating T-helper cells and hyperactivity of the B-cell system[3]. It has long been known that patients with congenital or acquired immunodeficiency disorders and with certain immunoinflammatory diseases are more likely to develop lymphomas. It has been proposed that the increased mitotic activity of lymphocytes increases their risk of mutation and malignant transformation[4]. The mitotic activity of lymphocytes appears to be increased in sarcoidosis due to the inflammatory response in affected tissues[3] and this may partly explain the increased risk of lymphoma in patients with active sarcoidosis.

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Hypercalcaemia and sarcoidosis in infancy

S J Stanworth BA MRCP[1]
C T C Kennedy MA FRCP[2] **P A J Chetcuti** MRCP[1]
F Carswell MD FRCP[1] *Departments of [1]Paediatrics and [2]Dermatology, Bristol Royal Hospital for Sick Children, St Michael's Hill, Bristol BS2 8BJ*

Keywords: hypercalcaemia; sarcoidosis; infant

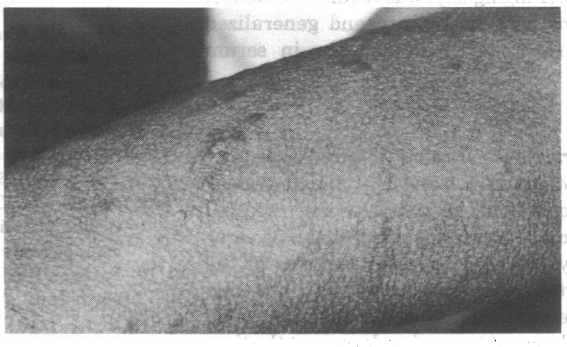

Figure 1. Numerous micropapules and several larger papules coalescing into plaques

Case presented to Section of Dermatology, 17 January 1991

Sarcoidosis is a rare disease in young children. Skin, joint and eye manifestations are the characteristic early features. Marked hypercalcaemia was found as an additional feature at diagnosis in the following case history of a West Indian infant.

Case report

A 9-month-old West Indian female infant presented to the Dermatology Clinic with a 5-month history of a flexural papular non-pruritic rash. Differential diagnoses at this stage included atopic eczema and lichen nitidus, but there was no improvement with weak topical steroid treatment. By 12 months of age the rash was more extensive and she had developed intermittent finger and ankle swelling, with periodic conjunctival redness and irritation. She had become reluctant to weight bear and consequently motor development was delayed. She was more irascible, with weight loss below the third centile.

There was a widespread hypopigmented and slightly scaly papular eruption over her trunk, limbs and face (Figures 1 and 2). There was diffuse swelling of the metacarpophalangeal, interphalangeal and ankle joints (Figure 2). Enlargement of the parotid glands was noted, but there was no splenomegaly, lymphadenopathy or lacrimal gland enlargement. A fluctuating pyrexia up to 38°C was recorded.

Investigations revealed a haemoglobin 11.9 g/dl, raised white cell count 20×10^9/l (neutrophils 48%, lymphocytes 44%, monocytes 8%). Serum calcium was elevated to

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