

radiation may play a role in the onset of this tumour. Our patient, who had never used tobacco or alcohol, presented with a tongue carcinoma even before cutaneous malignancies developed, as described also by others (6).

Malignant neoplasms on the anterior third of the tongue can be cured by prompt surgical excision in an early stage. Frazell & Lucas found a 5-year survival of 45.8% in patients with squamous cell carcinoma of the anterior third of the tongue (10). Because in our case radical excision of the tumour was refused, therapy with an oral retinoid (etretinate) was given tentatively. Treatment of advanced squamous cell carcinomas of the skin with isotretinoin has been described to be moderately effective (9). Initially the tongue tumour showed regression during therapy with etretinate. This regression might have been due to factors other than retinoid treatment, such as less inflammation, because of better hygienic care.

Therapy with indomethacin and prednisolone has been described to be effective for cutaneous squamous cell carcinomas in XP patients (7). In a group of 9 XP patients with squamous cell carcinoma of the skin, 3 patients showed complete regression of the tumour and 5 partial regression. One patient did not respond to treatment. In our patient too the squamous cell carcinoma of the tongue did not respond to this therapeutic approach.

REFERENCES

1. Kraemer KH, Lee MM, Scotto J. Xeroderma pigmentosum: cutaneous, ocular and neurologic abnormalities in 830 published cases. *Arch Dermatol* 1987; 123: 241-250.
2. Kraemer KH, Lee MM, Scotto J. Early onset of skin and oral cavity neoplasms in xeroderma pigmentosum. *Lancet* 1982; *i*: 56-57.
3. Plotnick H. Xeroderma pigmentosum and mucocutaneous malignancies in three black sibs. *Cutis* 1980; 25: 311-313.
4. Cleaver JE, Zell B, Hashem N, et al. Xeroderma pigmentosum patients in Egypt. II. Preliminary correlations of epidemiology, clinical symptoms and molecular biology. *J Invest Dermatol* 1981; 77: 96-101.
5. Harper JL, Copeman PWM. Carcinoma of the tongue in a boy with xeroderma pigmentosum. *Clin Exp Dermatol* 1981; 6: 601-604.
6. Wade HW, Plotnick H. Xeroderma pigmentosum and squamous cell carcinoma of the tongue. Identification of two black patients as members of complementation group C. *J Am Acad Dermatol* 1985; 12: 515-521.
7. Al-Saleem T, Sabri Ali Z, Qussab M. Skin cancers in xeroderma pigmentosum: response to indomethacin and steroids. *Lancet* 1980; *ii*: 264-265.
8. Bootsma D, Mulder MP, Pot F, Cohen JA. Different inherited levels of DNA repair replication in xeroderma pigmentosum cell strains after exposure to ultraviolet irradiation. *Mutat Res* 1970; 9: 507-516.
9. Lippman SM, Meyskens FL. Treatment of advanced squamous cell carcinoma of the skin with isotretinoin. *Ann Intern Med* 1987; 107: 499-501.
10. Frazell EL, Lucas JC. Cancer of the tongue. *Cancer* 1962; 15: 1085-1099.

Familial Sarcoidosis: High Ethnic Prevalence

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We report three sisters of Irish extraction who all developed sarcoidosis. The cases emphasize both the familial and ethnic preponderance of sarcoidosis which has not been adequately emphasized in the dermatology literature.

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CASE REPORTS

Sister 1

Presented in 1961, with lethargy, enlarged axillary and cervical lymph nodes and chest radiograph showing bilateral hilar lymphadenopathy. She was diagnosed as having sarcoidosis and over the next few months the lymphadenopathy resolved spontaneously. Further details were not available as her hospital records had been destroyed.

Sister 2

Presented in 1980, aged 56, with a one-year history of an asymptomatic, 3 cm, purplish plaque on the left cheek. She

was otherwise well, with no other abnormalities on examination. Histology of the skin lesion revealed multiple non-caseating granulomata in the dermis, with negative stains for fungi and acid-fast bacilli. A Kveim test showed granuloma formation and a Mantoux was negative to 10 IU tuberculin. Radiographs of the hands showed a cyst in one terminal phalanx, but no chest abnormality. Serum calcium and angiotensin converting enzyme were normal. After an initial response to potent topical steroids, over recent months the patient has developed disfiguring lupus pernio and systemic therapy is being contemplated.

Sister 3

Presented in 1987, aged 61, with two similar asymptomatic, smooth, pinkish plaques, 3 cm in diameter on the forehead and extensor surface of the left arm, which had been present for 2 years. She was otherwise well, with no other abnormalities on examination. Histology of a skin lesion revealed multiple non-caseating granulomata in the dermis, with stains for fungi and acid-fast bacilli negative. A Kveim test produced a 3 mm papule at 6 weeks, biopsy of which was refused. A Mantoux was negative to 10 IU tuberculin. Chest radiograph showed calcification of the paratracheal and hilar glands of an "eggshell type" at the left hilum, with the lung fields clear, reported by a respiratory physician as most likely to represent old glandular sarcoid. Serum calcium and angiotensin converting enzyme were normal. Treatment of the plaques of sarcoidosis with intralesional steroid achieved a satisfactory response.

DISCUSSION

Familial sarcoidosis was first described in 1923 by Martenstein (1), since when there have been several other reports. The largest series reported by the British Thoracic and Tuberculosis Association (2) noted a preponderance of like-sex over unlike-sex pairs and of monozygous over dizygous twins concordant for sarcoidosis. The interval between the time of onset of the affected individuals exceeded 2 years for the majority of the 62 families studied. Analysis of mode of onset, chest radiographs and pattern of extrathoracic involvement has failed to reveal any diagnostic clues which might hallmark familial sarcoidosis (3). Familial, like non-familial sarcoidosis shows a wide variation in prevalence between ethnic groups. In United

States' blacks and the Irish, two groups known to have a high prevalence of sarcoidosis, conservative estimates of the probability that an index case will have a sibling with sarcoidosis are approximately 10% (3, 4), i.e. much higher than chance alone would predict.

The aetiology of familial sarcoidosis is obscure, theoretically resulting from genetic or environmental factors. The weight of evidence favours the former (5), though attempts to demonstrate genetic linkage by HLA marker studies indicate that the HLA system acts as a marker for the expression and not the development of sarcoidosis (6).

As therapy for sarcoidosis has only been shown to affect long-term prognosis in the rare complications of hypercalcaemia and uveitis (7), we would disagree with the suggestions that siblings of affected patients should be screened for sarcoidosis (5). We report these cases to stress the importance of enquiring about family history when considering sarcoidosis in predisposed, ethnic groups, to improve diagnostic acumen and explain apparently unrelated symptoms in relatives.

REFERENCES

1. Martenstein H. Knochveränderungen bei Lupus pernio. *Z Haut Geschlechtskr* 1923; 7: 308.
2. British Thoracic and Tuberculosis Association. Familial associations in sarcoidosis. *Tubercule* 1973; 54: 87-98.
3. Sharma CP, Neville E, Walker AN, James DG. Familial sarcoidosis: a possible genetic influence. *Ann NY Acad Sci* 1974; 287: 386-400.
4. Hendings VE, Weston D, Young RC, Hackney RL. Familial sarcoidosis with multiple occurrences in eleven families; a possible mechanism of inheritance. *Ann NY Acad Sci* 1976; 278: 377-85.
5. Brennan NJ, Crean P, Long JP, Fitzgerald MX. High prevalence of familial sarcoidosis in an Irish population. *Thorax* 1984; 39: 14-18.
6. Turton C. High prevalence of familial sarcoidosis in an Irish population. *Thorax* 1984; 39: 479.
7. Scadding JG, Mitchell DN. *Sarcoidosis*. 2nd ed. London: Chapman and Hall, 1985: 597.