

Lupus Vulgaris in a Maltese Patient

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Introduction

The incidence of cutaneous tuberculosis (TB) in the developed world has fallen, in parallel with the decline of other forms of TB. Nevertheless cases still occur and their diagnosis may be difficult. We report a case of lupus vulgaris in a Maltese patient who went on to have an excellent response to antituberculous chemotherapy.

Case report

A previously healthy 45-year-old Maltese lady presented with reddish-brown swelling of her right ear lobe. This had been first noted by the patient about 5 years earlier and was asymptomatic but cosmetically upsetting. There had been no response to treatment with various topical antibiotics and corticosteroids and the abnormality was gradually extending. On examination there was soft, reddish-brown infiltration of the lobe and posterior aspect of the right ear (Figures 1a & 1b). There was no palpable lymphadenopathy and general examination and review of the rest of the skin were unremarkable.

Histological examination of an incisional skin biopsy showed granulomatous inflammation with large epithelioid granulomata with focal necrosis (but no frank caseation) and surrounding lymphocytic cuffing (Figure 2). No Leishman-Donovan bodies were visible and special stains for mycobacteria and fungi were negative. Skin cultures for mycobacteria and fungi were negative. Results of chest X-ray, full blood count, creatinine, liver enzymes and angiotensin-1-converting enzyme were within normal limits. HIV serology was negative. A Mantoux test was strongly positive (18mm induration) (Figure 3). A diagnosis of lupus vulgaris was made. On further questioning there was no history to suggest previous TB.

The patient had no respiratory symptoms and had never lived abroad. She had been vaccinated with Bacille Calmette-Guerin in childhood. Results of chest X-ray and Mantoux test in the other members of her household (husband and daughter) were normal or negative. The patient's ears had been pierced in childhood.

The patient was treated with standard antituberculous chemotherapy (rifampicin 600mg daily, isoniazid 300mg daily and pyrazinamide 2g daily for 2 months followed by rifampicin 600mg daily and isoniazid 300mg daily for a further 4 months). There was gradual improvement with a reduction in both the ear lobe swelling and reddish brown discoloration; some atrophic scarring eventually appeared in the previously affected area (Figures 1c & 1d).

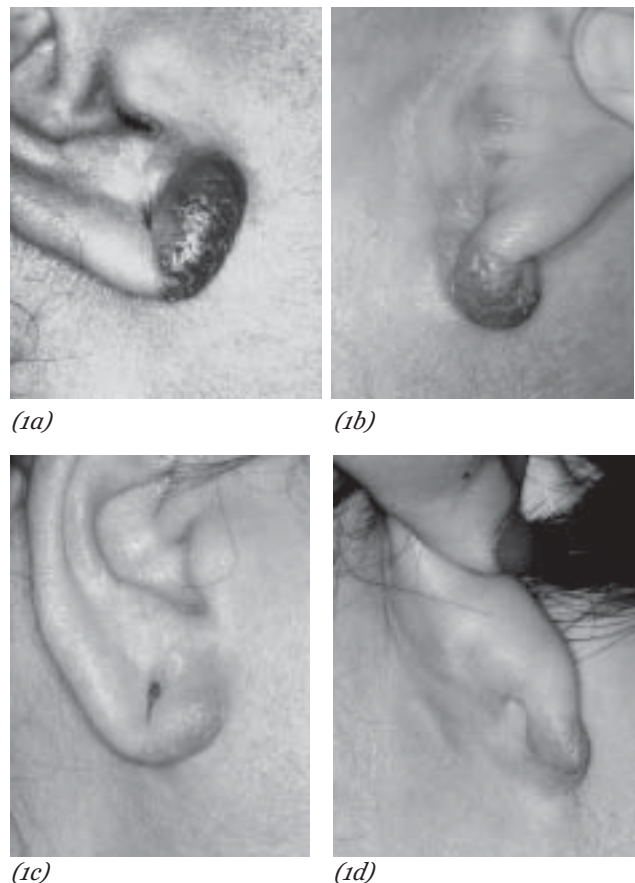


Figure 1: Reddish-brown infiltration of the lobe and posterior aspect of the right ear at presentation (a & b) and after completion of antituberculous treatment (c & d).

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Discussion

Lupus vulgaris is a slowly progressive form of cutaneous TB that occurs in individuals with a moderate or high degree of immunity to *Mycobacterium tuberculosis*.¹ It may follow haematogenous spread, extension from underlying infected joints or lymph nodes, or, rarely, local inoculation and has also been described as a complication of BCG vaccination.² It has been suggested that, in some cases, organisms disseminated at the height of a previous infection can remain latent in the skin for many years and then become reactivated by local trauma or a non-specific inflammatory process.³ Often, however, the origin in a particular case is obscure. In our case no obvious cause was apparent.

Lupus vulgaris is commoner in females than in males and may present at any age. A cool, moist climate appears to favour development of lupus vulgaris and this has been proposed as an explanation for the previous high prevalence in northern European countries. However, recent studies from South Africa and Northern India found that lupus vulgaris was the commonest form of cutaneous TB in those populations too.^{4,5} Other recognised forms of cutaneous TB include tuberculous chancre, warty TB, scrofuloderma, tuberculous abscess, orificial TB, acute miliary TB, tuberculous gumma and tuberculous mastitis. The term 'tuberculide' is used for a group of mainly symmetrical and disseminated eruptions that arise in response to an internal focus of TB and clear with antituberculous therapy. Examples of tuberculides include lichen scrofulosorum, papulonecrotic tuberculides, erythema induratum (Bazin's disease) and some cases of nodular vasculitis.

Cutaneous TB accounts for only a small proportion of cases of extrapulmonary TB. In one study from the United States cutaneous TB accounted for less than 1.5% of all cases of TB and was less common than TB of the bones or joints or genitourinary TB.⁶ In Malta in the period 1979-2001 a total of 565 new cases of TB were notified to the Public Health Department. Of these cases there were 168 patients (29.7%) who had extrapulmonary TB of whom 11 (1.9% of the total) had cutaneous TB (Figure 4).

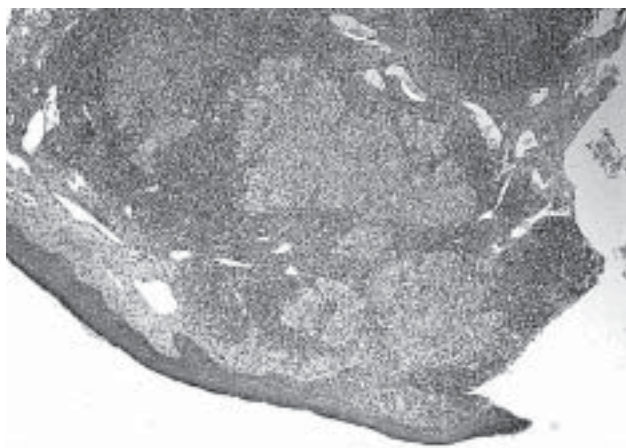


Figure 2: Histology: Dermal granulomatous inflammation with large epithelioid granulomata with focal necrosis (but no frank caseation) and surrounding lymphocytic cuffing. (Haematoxylin & Eosin, x 20)

The natural history of untreated lupus vulgaris is slowly but inexorably progressive. Extension may be continuous or irregular and is slower in younger patients.⁷ The characteristic lesion of lupus vulgaris is a soft, reddish-brown plaque of gelatinous consistency said to resemble apple-jelly, most commonly affecting the head and neck. Clinical variants include ulcerative, vegetating, tumour-like and papular forms.¹ Nasal, buccal and conjunctival mucosal involvement may also occur. The usual slow evolution, reddish-brown colour and softness of the lesions help to distinguish lupus vulgaris from other conditions including sarcoidosis, lupoid leishmaniasis, lymphocytoma cutis, discoid lupus erythematosus, tertiary syphilis, leprosy, deep mycotic infections and chronic vegetating pyoderma.⁸ However, clinical diagnosis of lupus vulgaris can be difficult; lesions on the face have been misdiagnosed as rosacea,⁹ Wegener's granulomatosis,⁹ squamous cell carcinoma,⁹ and even a port-wine stain¹⁰. It is not uncommon for the diagnosis of lupus vulgaris to be delayed for many years. In one recently published case, a plaque of lupus vulgaris on the buttock, probably present since childhood, was only diagnosed correctly when the patient was 87 years old!¹¹ A high index of clinical suspicion is therefore required, particularly in populations like Malta's where TB is relatively uncommon.

Fortunately, unlike other sites where TB may occur, cutaneous TB is not only visible but also easily amenable to biopsy. Histological features of lupus vulgaris are variable but typically include dermal epithelioid granulomata with scanty or absent caseation, multinucleated giant cells and peripheral lymphocytes.¹² Epidermal changes including ulceration, atrophy and acanthosis, depending on the clinical variety, may be present. Organisms are usually scarce and, as in our patient, culture is generally negative; in one series, *Mycobacterium tuberculosis* was cultured from lupus vulgaris lesions in only 6% of cases.⁷ Bacilli are not usually demonstrated on histological sections stained to show acid-fast bacilli (e.g. Ziehl-Neelsen stain). In recent years, polymerase chain reaction techniques that may detect *Mycobacterium tuberculosis* DNA have been developed (not yet available locally) that may markedly facilitate

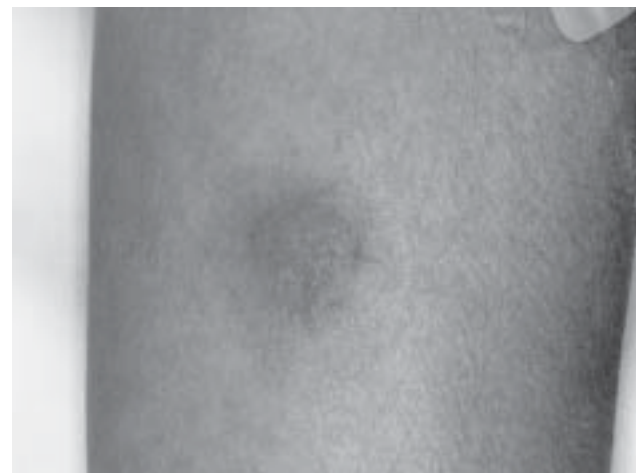
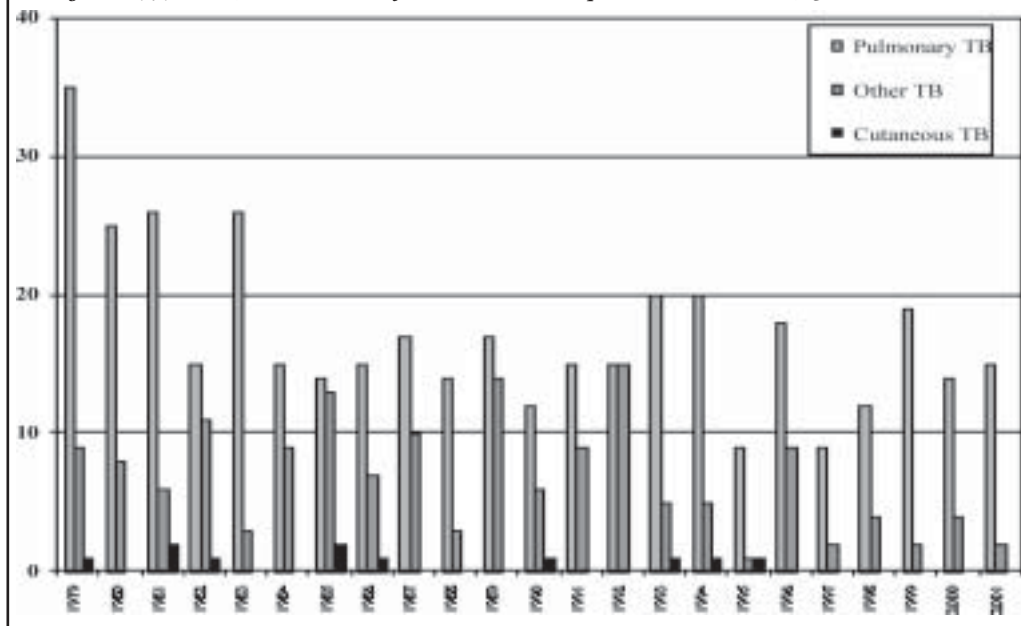


Figure 3: Positive Mantoux reaction.

Figure 4: Incidence of cutaneous, pulmonary and other forms of tuberculosis (TB) in Malta during the years 1979-2001; Data collected from the Health Department Chest Unit, Qormi Health Centre.



the diagnosis of cutaneous TB.¹³ Our patient had a strongly positive tuberculin (Mantoux) test. This is usual in lupus vulgaris and is an important aid to diagnosis. In sarcoidosis, by contrast, the tuberculin response is typically negative or weak.¹⁴

Local complications of lupus vulgaris may include tissue destruction (*lupus* – wolf, from the supposed resemblance to the bite of a wolf), ulceration, secondary infection and scarring. Rarely, cutaneous malignancy including squamous cell carcinoma and, less commonly, basal cell carcinoma may develop in the previously affected area.¹⁵

Lupus vulgaris is eminently curable and should be treated with a 6-month course of standard triple antituberculous therapy (rifampicin, isoniazid and pyrazinamide for 2 months followed by rifampicin and isoniazid for a further 4 months).¹⁶ Isolation of the patient is not required. Those affected should be investigated to exclude concomitant internal TB and, after treatment, should be followed up because of the risk of cutaneous malignancy developing in the scarred tissue. The consequences of failing to diagnose lupus vulgaris can be serious for the patient. Although the incidence of lupus vulgaris in the developed world has declined, cases are still reported with regularity. The possibility of lupus vulgaris should be considered in patients who have persistent/progressive granulomatous cutaneous lesions.

Acknowledgement

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