Lupus Vulgaris in a Pediatric Patient: A Clinicohistopathological Diagnosis

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Lupus vulgaris is the most common form of cutaneous tuberculosis which usually occurs in patients previously sensitized to *Mycobacterium tuberculosis*. We present a case of a 10-year-old boy who was diagnosed as lupus vulgaris clinically and histopathologically. He had well demarcated, irregularly bordered, pink, infiltrated plaques on his left cheek showing apple-jelly appearance on diascopy. The histopathological examination showed tuberculoid granulomas with Langhans type giant cells. The Mantoux reactivity was in normal limits, and no acid-fast bacilli was found in the lesion, either by direct stained smears or by culture. The lesions showed marked improvement on anti-tuberculosis treatment. We want to emphasize that histopathological examination has diagnostic value in lupus vulgaris in correlation with clinical appearance, when direct analysis or culture is negative.

Key-Words: Lupus vulgaris, histopathology, clinical appearance, culture.

Lupus vulgaris is a progressive form of cutaneous tuberculosis which is acquired either exogenously by direct inoculation of the bacilli into the skin or endogenously by hematogenous or lymphatic spread from an underlying infected focus in a sensitized host with a moderate to high degree of immunity to *Mycobacterium tuberculosis* [1]. It is characterized by plaque with apple-jelly nodule that extends irregularly with scar formation and tissue destruction [2]. Differential diagnosis of lupus vulgaris is also difficult and unreliable purely on clinical grounds, and histopathological and microbiological examinations are required [3].

We report a 10-year-old boy with lupus vulgaris involving the left cheek with emphasis on clinicohistopathological diagnosis.

Case Report

A 10-year-old immunized boy presented with a three year history of pink plaques appearing and progressing slowly on the left cheek. Clinical examination revealed well demarcated, irregularly bordered, slightly tender, pink, infiltrated plaques on the left cheek, extending from the preauricular area to the mandibular area (Figure 1). Apple-jelly colour was seen when examined by diascopy. There was no regional lymphadenopathy, and systemic examination revealed no abnormalities. No other family members had similar lesions.

Routine biochemical analysis, complete blood count, and urine microscopy were all normal, and the erythrocyte sedimentation rate was 8 mm/h. Venereal Disease Research Laboratory test (VDRL) and anti-human immunodeficiency virus (HIV) antibody test were negative. Chest radiograph and computed tomography findings were normal, and no sign of pulmonary tuberculosis was present. The purified protein derivative test (Mantoux test) showed normal reactivity with a 10 mm induration after 48 hours.

Histopathological examination of the incisional biopsy specimen showed normal epidermis with superficial focal parakeratosis, and noncaseating tuberculoid granulomas consisting of epithelioid histiocytes, plasmocytes, and Langhans giant cells in the papillary dermis (Figure 2A and Figure 2B). The tissue sections were negative for acid-fast bacilli (AFB) by the Ehrlich-Ziehl-Neelsen stain, and cultures of the biopsy material and blood were negative.

The standard short-course chemotherapy for treatment of cutaneous tuberculosis which involves the administration of three antituberculous drugs for the first two months (isoniazid 10 mg/Kg, rifampicin 10 mg/Kg, pyrazinamide 30 mg/Kg), followed by four months of isoniazid and rifampicin was started. Marked improvement of the lesions with atrophic scarring was seen by the end of six months treatment.

Discussion

Tuberculosis of the skin is caused by *Mycobacterium tuberculosis*, *Mycobacterium bovis*, and under certain conditions, the bacillus Calmette-Guérin (BCG), an attenuated strain of *Mycobacterium bovis*. Cutaneous tuberculosis represents 1.5% of all cases of extrapulmonary tuberculosis [2]. Classification has been attempted according to morphology and, more recently, the mode of infection or the immunologic state of the host [4]. Lupus vulgaris is the commonest form of cutaneous tuberculosis seen in most countries [5]. Beyt et al. have classified lupus vulgaris under both inoculation and hematogenous tuberculosis, but they have overlooked lymphatic spread [6]. But this classification does not reflect the immunological spectrum of the disease, nor does it take consideration of systemic organ involvement. For practical management, it is only necessary to know the extent of the disease and whether or not the tubercle bacilli can be detected from the lesions [5].

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Figure 1. Well demarcated, irregularly bordered, pink, infiltrated plaques on the left cheek, extending from the preauricular area to the mandibular area.

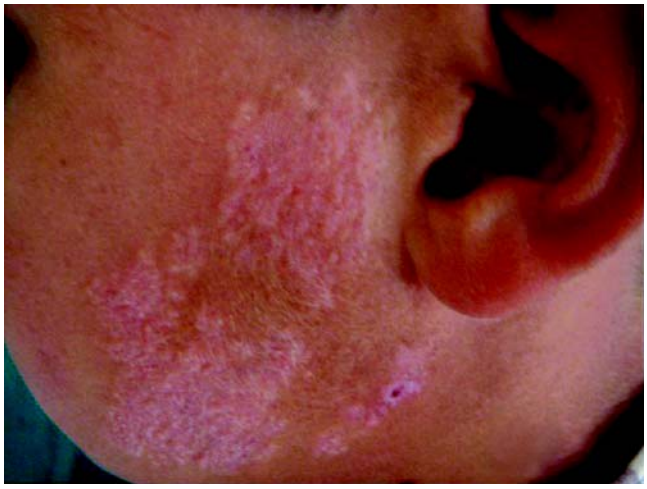
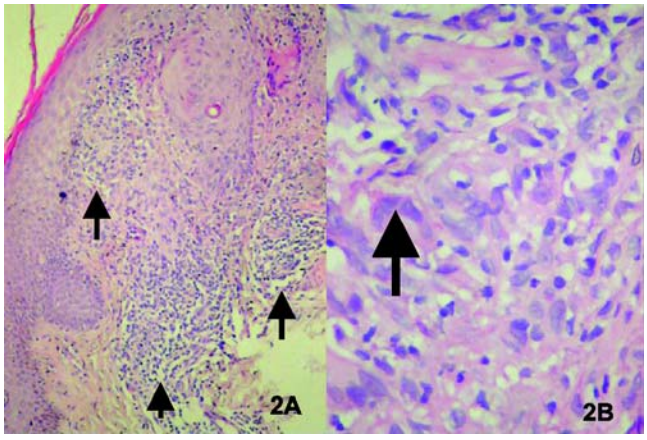


Figure 2. (A) Normal epidermis and superficial focal parakeratosis and noncaseating granulomas indicated with the arrows in the papillary dermis (hematoxylin-eosin stain x 100). (B) Langhans giant cell indicated with the arrow in the granuloma which mostly consist of epithelioid histiocytes and plasmocytes (hematoxylin-eosin stain x 400).



Clinical features of lupus vulgaris are the softness of the lesions, the brownish-red colour, and the slow evolution. Also the apple-jelly nodules revealed by diascopy are highly characteristic, but finding them may be decisive, especially in ulcerated, crusted or hyperkeratotic lesions. Lupus vulgaris is extemely chronic, and without therapy its course usually extends over many years. Although there are periods of relative inactivity, it is progressive and leads to considerable impairment of function and to disfiguration. The most serious complication of long-standing lupus vulgaris is the development of carcinoma [4].

Most cases of primary tuberculosis, particularly in children, are contracted on the extremities and on the face following scratches, bruising, lacerations, pin-pricks, impetigo, boils, piercing, tattoos, and circumcision [7]. Lupus vulgaris might occur at the site of BCG vaccination suggesting exogenous inoculation of the infection [8]. The secondary form appears in subjects that are already sensitized to previous tuberculosis infections, or by BCG. The secondary form in these patients can be as a result of exogenous reinfection or by endogenous reactivation of dormant or persistent *Mycobacterium tuberculosis* after a reduction of cell-mediated immunity [9].

Tissue culture is the gold standard for diagnosis and for monitoring the emergence of drug-resistance strains [10]. But, compared with pulmonary tuberculosis, the number of bacilli encounterd in cutaneous tuberculosis is low [11]. Because lupus vulgaris is a paucibacillary form of tuberculous infection, culture is often negative and the diagnosis is mainly based on the Mantoux test, the histopathological appearance, and the response to chemotherapy [8]. But the Mantoux test does not allow precise diagnosis, only indicates that the patient has had previous contact with *Mycobacterium tuberculosis*, and can lead to false negative results in case of anergy [7]. Our patient was immunized and showed a normal Mantoux reaction. Also this skin test has limited diagnostic value in developing countries due to high rates of exposure to mycobacteria and BCG vaccination [4].

Because *Mycobacterium tuberculosis* should be detected either by direct analysis or culture for a correct diagnosis, skin biopsy was taken from our patient. Ehrlich-Ziehl-Neelsen staining of the biopsy material was negative for acid-fast bacilli (AFB), and culture of the biopsy material was negative, either. The histopathologic examination showed the tubercles which are hallmarks of cutaneous tuberculosis. They consist of accumulations of epithelioid histiocytes with Langhans giant cells and varying amount of caseation necrosis in the center [4]. But the absence of caseation necrosis may not rule out the diagnosis of tuberculosis [12,13]. Although tuberculoid granuloma formation is highly characteristic of cutaneous tuberculosis, it is not pathognomonic. Deep fungal infections, syphilis, and leprosy can show similar histological features. But the clinical criteria helpful in the differential diagnosis are the softness of the lesions, the brownish-red colour, the slow evolution, and the apple-jelly nodules revealed by diascopy [4].

Polymease chain reaction (PCR) is a useful, rapid method that has become available in recent years in the diagnosis of lupus vulgaris and other forms of cutaneous tuberculosis, but its sensitivity is reduced when used with smear-negative specimens or paucibacillary samples [14,15].

Standard short-course chemotherapy for treatment of cutaneous tuberculosis involves the administration of three antituberculous drugs for the first two months (isoniazid, rifampicin, and pyrazinamide), followed by four months of isoniazid and rifampicin [16].

When cutaneous tuberculosis can be difficult to confirm, the diagnosis is only established retrospectively, after response to a therapeutic trial [10].

In summary, the diagnosis of cutaneous tuberculosis is based on clinical features, demonstration of acid-fast bacilli

on smear, tissue culture, skin biopsy, and in recent years, PCR. However, the yield from culture and PCR is often low and diagnoses may need to depend on clinical features, histopathological findings, and retrospective review of response to treatment [10].

The purpose of this case report was to emphasize that the diagnosis of lupus vulgaris depends chiefly on clinical suspicion and histopathological features when the acid-fast bacilli can not be found either by direct stained smears or by culture.

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