Presternal Scrofuloderma: Tip of the Iceberg Unveiling Multifocal Extrapulmonary Tuberculosis - A Rare Case Report

Ravindranath Brahmadeo Chavan¹, Vasudha Abhijit Belgaumkar¹,², Nitika S. Deshmukh ³ and Vijay Deepak Joshi¹

¹Department of Skin and VD, B.J. Government Medical College, Pune, India  
²Corresponding author: Department of Skin and VD, B.J. Government Medical College, Pune, India. Email: doc_vasudha@yahoo.co.in

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Abstract

Introduction: Scrofuloderma, a type of secondary cutaneous tuberculosis (TB) commonly arising from the underlying cervical or axillary lymph node foci via contiguity, is frequent in the pediatric population. The origin of scrofuloderma from the underlying sternal tubercular osteomyelitic focus is exceptionally unusual. It can rarely be a part of the convoluted riddle of multifocal TB, particularly in the setting of human immunodeficiency virus (HIV) infection.

Case Presentation: We hereby report a 16-year-old immunocompetent female who presented with chronic, sinus-like lesions with bridging scars on the upper chest, clinically confounded with infected keloid and deep fungal infection. She was later diagnosed with scrofuloderma arising from sternal osteomyelitis compounded with multifocal tuberculosis (sternal, mediastinal, cutaneous, lumbar vertebral) and received antitubercular treatment.

Conclusions: This case highlights the importance of harboring clinical scepticism supported by thorough systemic investigations while diagnosing these innocuous mimickers occurring at rare sites, even in immunocompetent individuals.

Keywords: Scrofuloderma, Sternal Tuberculosis, Multifocal Tuberculosis

1. Introduction

Extrapulmonary tuberculosis (TB) constitutes 15 - 20% of the total TB load in immunocompetent patients (1). Cutaneous involvement represents 1 - 2% of these cases (2, 3). The prevalence of cutaneous TB in India is 0.1 - 0.9%, according to various sources (3-5). Scrofuloderma is a type of secondary cutaneous TB arising from an endogenous source that spreads by contiguity from underlying lymph nodes, bones, or joints to the skin. It usually occurs on the neck in children, with cervical lymph nodes being the most common focus (6). The involvement of the skeletal system is unusual, which is observed in 6 - 10% of extrapulmonary TB cases (7). Approximately 60 - 80% of skeletal TB cases show the involvement of spine or weight-bearing joints, with sternal osteomyelitis being extremely rare in 1% of cases, even in endemic countries (7). We hereby report an immunocompetent adolescent female presenting with chronic non-healing sinus-like lesions and bridging scars mimicking infected presternal keloids. She was diagnosed with scrofuloderma with multifocal TB on a thorough evaluation.

2. Case Presentation

A 16-year-old unmarried female, hailing from an urban shantytown, came to our hospital with complaints of a chronic, non-healing, gradually progressive lesion on the upper part of the chest since one year ago. It had started as a pea-sized reddish swelling on the upper chest, clinically confounded with infected keloid and deep fungal infection. She was later diagnosed with scrofuloderma arising from sternal osteomyelitis compounded with multifocal tuberculosis (sternal, mediastinal, cutaneous, lumbar vertebral) and received antitubercular treatment.
It can involve lymphoreticular, gastrointestinal, musculoskeletal, cutaneous, and nervous systems, as well. Cutaneous tuberculosis can be categorized on the basis of route of infection as exogenous and endogenous, which is further classified as hematogenous, lymphatic, and contiguous and also on the basis of bacillary load as multibacillary and paucibacillary (6). Scrofuloderma is the most common form of cutaneous TB among children, whereas lupus vulgaris is the most frequent variant in adults (6). The proportion of scrofuloderma was found to be 53.3% by Kumar et al. and 36.9% by Vashisht et al. [cited in Kumar and Kumar (6)]. It is a multibacillary form of secondary skin tuberculosis that spreads via a contiguous route. It more commonly arises in the neck from underlying cervical TB lymphadenitis, also known as scrofula (Latin word: scrofa - a brood sow). Nasopharyngeal TB or the practice of consuming unpasteurized milk in rural areas can be causative factors for the involvement of cervical nodes (6). Other lymph node groups affected include inguinal, axillary, submandibular, epitrochlear, and supratrochlear. Around 66% of cutaneous TB cases show systemic foci (6). Other than lymph nodes, the bones, joints, testes, breast, and lacrimal glands are the other underlying foci of scrofuloderma (6). Rarely, hepatic and intestinal TB can also be a focus of skin infection (6). Clinical presentation in childhood is characterised by the presence of multiple, chronic asymptomatic, subcutaneous cervical swellings, which later undergo softening, and ulceration forming discharging sinuses. These ulcers have bluish, undermined edges. Healing occurs with bridging, cribriform, or puckered scars. Adult scrofuloderma is a more localized entity. Skeletal TB accounts for 6 - 10% of extrapulmonary TB cases (7). It most frequently involves spine or weight-bearing joints. Sternal affection is seldom seen, representing only 1% of skeletal TB cases (1,7), while sternal osteomyelitis constitutes 0.3% of osteomyelitis cases (1,7). It may be primary or usually found as an extension from hilar lymph nodes or hematogenous or lymphatic dissemination from other sites. Sternal TB has also been reported in the pediatric age group after Bacille Calmette Guerin (BCG) vaccination (9). Gallouj et al. have reported a similar case of scrofuloderma leading to the discovery of sternal TB (10). However, the finding of multifocal systemic TB (defined as the presence of two or more lesions in extrapulmonary sites with or without pulmonary involvement) (8) renders our case unique. Multifocal TB accounts for less than 10% of TB cases, and its association with skin involvement is rare, with only a few reported cases (8). Risk factors for extrapulmonary dissemination are female gender, malnutrition, low socio-economic status (all present in our case), HIV infection, and other immunocompromised states (11).
**Figure 1.** Dusky, rubbery plaques with discharging sinus and bridging scars on manubrium sterni.

**Figure 2.** Anteroposterior and lateral chest radiographs showing manubrial erosive lesion (red arrows).
Figure 3. Sagittal magnetic resonance imaging (MRI) of spine showing the 4th lumbar vertebral sclerosis.
Diagnosis of multifocal TB is challenging, owing to delayed presentation, clinical polymorphism, and the diversity of organs affected. Skin involvement is dominated by scrofuloderma and gummas (8). Skin is also a site easily accessible for clinicopathological examination. Two pathomechanisms can give rise to multifocal TB: (1) hematogenous dissemination, and (2) initial hemolymphatic seeding of multiple organs with subsequent reactivation (8). Numerous investigations, including screening (Mantoux test induration > 10 mm), imaging studies (X-ray, computed axial tomography, MRI, ultrasonography), cytological studies (fine needle aspiration cytology and histopathology), IS-6110 polymerase chain reaction (high sensitivity), and culture (gold standard) are necessary for the establishment of the diagnosis of multifocal TB. Our case had the clinical presentation of bridging scars with sinuses over the upper chest (manubrium sterni), which is an extremely unusual site for scrofuloderma (7). Lesional morphology resembled an infected keloid (for which the patient had been earlier unsuccessfully treated) further compounding the diagnostic conundrum and suggesting multiple differentials like deep fungal (lobomycosis) or atypical mycobacterial infection, actinomycetoma, and nocardiosis. Furthermore, such lesions occurring over intertriginous regions may simulate hidradenitis suppurativa. It was only on the ba-
sis of strong clinical suspicion and thorough radiological and histopathological investigations that a final diagnosis of multifocal TB (sternal, mediastinal, cutaneous, and lumbar vertebral) was reached. A multidisciplinary consultation was sought wherein conservative management was recommended for skeletal TB in addition to the antitubercular treatment (ATT). Indian’s new revised national tuberculosis control program (RNTCP) guidelines for extrapulmonary TB, recommend the use of FDC of ATT (Table 1) (12). Multifocal TB should always raise the suspicion of underlying HIV infection and immunosuppression (8). The presence of multidrug-resistant (MDR) or extensively drug-resistant (XDR) TB poses great therapeutic difficulty with prolonged treatment courses and poorer prognosis.

Although our patient was immunocompetent, the diagnosis was delayed due to atypical site of cutaneous presentation (upper chest instead of neck or axilla), rare focus (sternal TB), and more importantly, morphological resemblance to keloid with the presence of non-specific subtle symptoms and signs.

3.1. Conclusion

Involvement of skin and sternum in multifocal TB is rare. Accurate recognition of the myriad faces of cutaneous TB is essential, particularly in high burden countries, as skin provides a window of opportunity for diagnostic interventions. Scrofuloderma might just be the tip of the iceberg, warranting a meticulous evaluation to unveil hitherto undetected multifocal systemic tuberculosis.
Table 1. The Doses of the Fixed-Dose Combination of Antitubercular Treatment According to the Weight Bands (New Revised RNTCP Guidelines for Pulmonary and Extrapulmonary New/Untreated Cases) (12)

<table>
<thead>
<tr>
<th>Weight Category (kg)</th>
<th>Intensive Phase (2 months); HRZE (75, 150, 400, and 275 mg)</th>
<th>Continuation Phase (4 months); HRE (75, 150, and 275 mg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>25 - 39</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>40 - 54</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>55 - 69</td>
<td>4</td>
<td>4</td>
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<tr>
<td>≥ 70</td>
<td>5</td>
<td>5</td>
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</tbody>
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Abbreviations: H, isoniazid; R, rifampicin; Z, pyrazinamide; E, ethambutol.

Footnotes

Authors’ Contribution: Acquisition of data, VB and VJ; Analysis and interpretation of the data, VB, ND, and VJ; Drafting of the manuscript, VJ and VB; Critical revision of the manuscript for important intellectual content, RC and VB; Administrative, technical, and material support, RC and ND.

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References


