

Case Report

Scrofuloderma: a common type of cutaneous tuberculosis. A case report

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Abstract Scrofuloderma is a common type of cutaneous tuberculosis characterized by a bluish-red nodule overlying an infected lymph gland, bone or joint that breaks down to form an undermined ulcer with a granulating tissue at the base. Progression of the disease leads to irregular adherent masses, densely fibrous at some places while fluctuant and discharging at others. It heals with a characteristic puckered scarring at the site of infection. The disease is caused by *Mycobacterium tuberculosis* and common anti-tuberculous drugs are recommended for treatment. Many similar cases with additional features have been reported in foreign literature. We describe a case of this disorder along with review of literature.

Key words

Scrofuloderma, tuberculosis, Mycobacterium, anti-tuberculous drugs

Introduction

Cutaneous tuberculosis has a worldwide distribution.¹ Though, human disease with *Mycobacterium tuberculosis* and *M. bovis* usually spreads by droplets, and the portal of entry is often the respiratory tract, skin can also be involved primarily.^{2,3} Many types of cutaneous tuberculosis like lupus vulgaris, scrofuloderma, tuberculosis verrucosa cutis, tuberculous gumma, orificial tuberculosis etc. are seen in our population.^{1,2,4}

Scrofuloderma, also called ‘tuberculosis colliquativa cutis’ is a common form of cutaneous tuberculosis affecting children and young adults in which there is breakdown of skin overlying a tuberculous focus in the lymph node, bone or joint.^{1,4} Initially, there are firm painless, subcutaneous nodules that gradually enlarge and suppurate.^{1,3,4} These lead to ulcers

and sinus tracts with undermined edges and ultimately puckered scars.¹ Diagnosis is usually performed by needle aspiration biopsy or excisional biopsy of the mass and the microbiological demonstration of stainable acid-fast bacteria.³ PCR has a low sensitivity but high specificity.^{5,6}

The best approach for treatment of this disorder is with conventional anti-tubercular drugs while people in close contact with the patient, such as family members, should undergo testing for tuberculosis.³ The role of surgery cannot be denied.² The affected nodes can be treated with electrosurgery, cryosurgery and curettage with electrodesiccation as an adjunct measure, with pharmacological therapy as the primary method of treatment.³ We report a case of scrofuloderma, a commonly seen variety of cutaneous tuberculosis in our society.

Case report

A 25-year-old male, unemployed, resident of Fatehgarh, Lahore, presented at the Department

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of Dermatology Unit-I, King Edward Medical University/ Mayo Hospital, Lahore with complaints of multiple discharging sinuses on his left shoulder and scapular region and right upper arm for the last 6 months. Initially, the lesions started as papules that progressed to nodules and pustules leading to draining sinuses. He also had a history of low grade fever with evening rise, associated with rigors and chills. There was no history of trauma, cough, haemoptysis, anorexia and weight loss or similar disease in his family.

The patient was diagnosed as a case of cutaneous tuberculosis in another hospital at Lahore and was put on anti-tubercular therapy but he discontinued the treatment after 2-3 weeks. When he presented to us, he had swellings and redness on right little finger, left wrist and left elbow for the last 3 months. The discharge from the lesions was initially serous and then purulent in nature. He had generalized aches and pains with joint stiffness. He also had swellings on the dorsal and ventral surface of left hand, right elbow and left foot. There was no history of contact with a tubercular patient and nothing was positive on systemic enquiry. In family history, his parents, 5 brothers and 3 sisters, were all alive and healthy. He was non-smoker, non-addict and belonged to a poor class.

Physical examination revealed a young man with normal built and pale complexion. Lymph nodes examination showed a palpable left epitrochlear node, single, mobile, non-tender, 2cm in size with a firm consistency while no other nodes were palpable. Cutaneous examination revealed discharging sinuses on the right little finger, left wrist and left elbow (**Figures 1 and 2**). There were soft, fluctuant, erythematous swellings around the sinuses with a purulent discharge while firm, tender, erythematous subcutaneous nodules on the



Figure 1 Multiple discharging sinuses & swelling over right little finger



Figure 2 Discharging sinus (left elbow joint)



Figure 3 Swelling and discharging sinus (palmar aspect of left hand). Swelling on right little finger as shown in Figure 1.

ventral and dorsal aspect of left hand (**Figure 3**), right elbow and dorsal aspect of left foot near the lateral border. The right elbow and ankle joint were also found to be swollen. Multiple depressed atrophic scars were also present on the right arm and anterior aspect of right shoulder with striae distensae.

There was tenderness and difficulty in flexion/extension of all joints in the vicinity of affected areas. Respiratory system examination showed a decreased expansion in the upper chest while vocal fremitus was increased and the percussion note dull in the right upper chest. Auscultation revealed diminished breath sounds and an increased vocal resonance in the right upper zone. No significant finding was noted in gastrointestinal, cardiovascular and nervous system.

On laboratory investigations, the blood complete examination revealed TLC: 9800/mm³, Hb: 10gm/dl, ESR: 70 mm and a microcytic hypochromic picture while urine and stool examination had no abnormal finding. Gram staining and smear for Leishman-Donovan bodies were negative. The bacteriological examination of pus showed no acid-fast bacilli or other micro-organisms. Hepatitis B surface antigen, anti-HCV and HIV screening were negative. Mycodot test revealed negative IgG and IgM result. Microscopy for fungal spores was also negative. Sputum for AFB was positive and polymerase chain amplification showed a positive result with mycobacteria. Chest X-ray showed non-homogeneous shadowing in the right upper lobe, bordered by horizontal fissure with upward pull of hila due to chronic inflammatory process, most probably tuberculosis. X-ray left elbow joint revealed subtle cortical erosion around medial epicondyle, with overlying mild soft tissue swelling. X-ray of right hand showed destructive changes, with ill-defined sclerosis and lucencies and overlying soft tissue swelling at the proximal phalanx of little finger while x-ray of left hand revealed sclerosis with cortical thickening and infiltration of medullary cavity of third metacarpal and overlying soft tissue swelling. X-ray left foot showed cortical thickness with sclerosis and

infiltration of medullary cavity in the 5th metatarsal while X-ray right foot revealed irregularity at the medial aspect of shaft of 1st metatarsal. Abdominal ultrasonography revealed hepatomegaly with a fatty liver. Histopathology of skin revealed chronic non-specific inflammation while bone curettage showed granulomata composed of epithelioid cells with Langhan's giant cells and caseation necrosis. No malignancy was seen and the final report was suggestive of tuberculosis.

Currently, the patient is under observation with continued treatment on oral isoniazid, rifampicin, ethambutol, pyrazinamide, pyridoxine and injection streptomycin 1gm intramuscular daily for the last 6 months with some improvement.

Discussion

One-third of the world's population is infected with *M. tuberculosis* and global burden of the disease continues to grow.^{1,5,7} The organism responsible for tuberculosis was identified more than 100 years ago while a tuberculosis vaccine has been in use for over 60 years and chemotherapy for over 30 years.⁸ Despite all these, the disease still remains a major international health problem.^{3,5,7} The reasons may be malnutrition, low socioeconomic conditions and multidrug resistant strains of *M. tuberculosis*.^{3,9} In our case, swellings with draining sinuses, histopathology report, positive result on PCR testing and a good response to ATT favoured the diagnosis of scrofuloderma.

The condition has to be differentiated from some other similar clinical entities (**Table 1**). Atypical mycobacterial infections clinically mimic scrofuloderma.¹ *M. scrofulaceum* produces only a benign, self-limiting lymphadenopathy with no organ involvement.

Table 1 Differential diagnosis of discharging sinuses [1-3,6,10]

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- Atypical mycobacterial infection due to *Mycobacterium scrofulaceum* and *M. avium-intracellulare*
 - Actinomycosis
 - Sporotrichosis
 - Botryomycosis
 - Nocardiosis
 - Syphilitic gumma
 - Hidradenitis suppurativa
-

The infection is seen in children, mainly between the ages of 1 and 3 years. Submandibular and submaxillary nodes are typically involved and there are no constitutional symptoms. Primary skin disease caused by *M. avium-intracellulare*¹ has been reported in rare instances, presenting as single or multiple, painless, scaly yellowish plaques or subcutaneous nodules with a tendency to ulceration and a slowly progressive, chronic course. This infection also causes lung disease or, less frequently, osteomyelitis and may produce a cervical lymphadenitis with sinus formation that is clinically indistinguishable from scrofuloderma. Both of these conditions were ruled out on the basis of histopathology and positive PCR report for *M. tuberculosis*.^{2,5,6}

Actinomycosis is characterized by granulomatous and fibrotic lesions, which tend to break down and form abscesses that drain through multiple sinuses.¹¹ Typical “sulphur granules” occur in these abscesses.¹¹ The disease is caused by *Actinomyces israelii*, the anaerobic organism producing filamentous branching hyphae. The lesions occur primarily on the face and neck, especially on the lateral surfaces of neck beneath the jaw, less often on the chest wall or lower abdomen.¹¹ The absence of sulphur granules and negative culture report for *Actinomycetes* helped to delineate this condition. Where sporotrichosis is endemic, it must also be excluded.¹² The clinical features of this condition

are similar to scrofuloderma but a negative report for fungal hyphae and histopathology led to exclusion of this condition.

Botryomycosis is a chronic inflammatory condition due to a bacterial infection.¹³ The infection is more common in immunocompromised patients. The disease presents as nodules, sinuses, fistulae, abscesses and ulcers, leading to scarring. The lesion usually occurs on the extremities but less commonly affected sites are head, neck and buttocks.¹³ The granules (like sulphur granules) are formed and discharged from the lesions which on histologic examination are proven to be masses of cocci, mostly *Staphylococci*.¹³ The condition can be ruled out on the basis of absence of bacteria from pus or biopsy specimen. In *nocardiosis*, papulo-nodular lesions occur on the limbs and trunk leading to draining sinuses and the organisms are detected as gram-positive branched-filaments and branching at right angle is confirmatory.^{11,12}

Syphilitic gumma is a typical granulomatous lesion of tertiary syphilis mainly found in the skin and bone.^{12,14} Cutaneous lesions are rounded, red to flesh-coloured nodules which can occur anywhere. The nodules may break down to form punched-out ulcers leading to atrophic scars or can heal with no residue.^{12,14} In our case, there was no neurological or cardiovascular involvement and serological tests for syphilis were also negative.

The earliest lesion of hidradenitis suppurativa is a painful erythematous nodule in an apocrine gland-bearing site such as axilla, inguinal, perianal and even the areolar skin leading to abscess formation.¹⁵ Rupture and drainage of the abscesses soon occurs, with healing by deep fibrosis.¹⁵ As the process recurs again and again, there is an increase in scarring and sinus tract

formation with restriction of movement of the affected limb. Many patients with severe chronic disease show open comedones or blackheads, in the affected areas. The clinical picture and negative report for micro-organisms differentiated the condition from scrofuloderma.

The key elements in the diagnosis of this infection are a high index of suspicion, taking a history with an emphasis on exposure to any sufferer in the family or other potential sources and tissue biopsy for culture & histopathology.³ Therapeutic regimens include anti-tubercular treatment with four drugs for initial 2 months and then combination of two drugs regularly for a sufficient period up to 12-18 months, depending upon the clinical picture and investigations.^{3,4,6}

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