

Scrofuloderma: A diagnostic dilemma in primary care

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SUMMARY

A 35-year-old lady presented at the Klinik Kesihatan Bandar 32 Bera, Pahang with a one-month history of multiple cervical swellings and ulcers over her neck area. The lesions began as papules and later progressively developed into nodules and pustules. She also had low grade fever associated with weight loss for one month duration. Chest x-ray revealed normal findings and sputum direct smear for acid fast bacilli was noted to be negative. Histopathologic finding of skin biopsy revealed central epidermal necrosis surrounded by granulomatous tissue forming an abscess and histiocyte infiltrates, confirming the diagnosis of Scrofuloderma. In view of the report of the fine needle aspiration cytology (FNAC) of the cervical lymph nodes suggestive for tuberculous lymphadenitis, the patient was given anti-tuberculosis therapy. Fortunately, six months later, the ulcers began to solve and heal gradually.

INTRODUCTION

Global Burden of Disease Study has reported that tuberculosis (TB) is the second leading cause of death in the world.¹ TB is a chronic granulomatous infectious caused by *Mycobacterium tuberculosis*, an acid-fast bacillus whose route of transmission is via the inhalation of airborne droplets. The most common extra pulmonary tuberculosis is tuberculous lymphadenitis. Cutaneous tuberculosis (CT) on the other hand is a very rare case accounting for only 1-2% of TB cases worldwide.² Scrofuloderma is a form of cutaneous tuberculosis normally affecting children and young adults. Scrofuloderma also known as *tuberculosis colliquativa cutis* which occurs due to the direct spread from endogenous source who have been infected with *M.tuberculosis bacilli*, predominantly lymph nodes and other structures such as joint, tendon, bone and synovial fluid are infected.³ Suppurative ulcers are commonly found in cutaneous tuberculosis because of severe and chronic inflammation. Among the areas that most commonly affected are neck, chest and axilla.³

Clinical characteristics of Scrofuloderma generally starts with multiple or solitary painless lymph node enlargement.³ Over time, the basal part eventually become softened forming cold abscess. The abscesses will become suppurated and finally rupture, creating a linear and irregular ulcers. The differential diagnosis of CT comprise of atypical mycobacterial infection (NTM), sarcoidosis, verrucous vulgaris, blastomycosis, leprosy, and tertiary syphilis.⁴ Treatment of Scrofuloderma is similar to the treatment of pulmonary tuberculosis i.e. by using oral anti-tuberculosis regimen containing isoniazid, rifampicin, pyrazinamide and ethambutol.⁵

CASE REPORT

A 35-year-old woman presented to the health clinic, Klinik Kesihatan Bandar 32 Bera, Pahang with one-month history of multiple cervical swellings and ulcers over her neck and upper chest region. Examination revealed that there were initially two lesions around her neck, which began as papules but gradually increased in size and progressed to pustules leading to ulcerations with the drainage of pus. She also had low grade fever with weight loss and loss of appetite for one month. There was no history of haemoptysis, prolonged cough, trauma, or any similar presentations among family members. She had sought medical treatment a few times previously and was prescribed several courses of antibiotics. However, the lesions did not improve and became worse progressively.

Physical examination revealed multiple ulcers with suppurative surface, along with nodule measuring 3 cm x 0.5 cm as shown in Figure 1. Around her neck, some crusts were found measuring 2 cm x 0.5 cm typical of Scrofuloderma. There were also mobile and painless lymph nodes enlargement in the right and left anterior cervical region measuring 2x2cm.

In view of her complaints of weight loss and lack of appetite, we proceeded with investigations for tuberculosis. Chest X-ray was performed but noted to be normal. However, tuberculin skin test revealed positive finding with 15mm induration. However, direct smear of sputum specimen was noted to be negative for acid fast bacilli. In view of chronicity of the presentations and persistent lesions despite being given several courses of antibiotic at primary care, we referred the patient to the Dermatology Clinic in Hospital Tengku Ampuan Afzan, Kuantan for shared care. Skin biopsy was performed in which the histopathology results showed epidermal necrosis in the central region surrounded by granulomatous tissue with Langhan's type of giant cells peripherally, forming abscess with histiocytic infiltrate around the lesion. These reports confirmed the diagnosis of Scrofuloderma. The swabs from the discharging fluid were tested positive for acid fast bacilli. Furthermore, fine needle aspiration cytology (FNAC) of the enlarged cervical lymph nodes was done which was suggestive for tuberculous lymphadenitis.

Once the final diagnosis of Scrofuloderma was confirmed, the patient was started on standard regimen of anti-tuberculosis drugs containing Rifampicin (R), Isoniazid (H), Pyrazinamide (Z), and Ethambutol (E) for two months continued by Rifampicin (R), Isoniazid (H) for the next four months. During the second month, our patient showed remarkable

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Fig. 1: Multiple ulcers with suppurative surface and crusts over her neck area.



Fig. 2: Pictures show healing ulcers with scar tissue following six months post anti-tuberculosis therapy.

improvement as the cervical lymph nodes enlargement were reduced in size significantly. In the fourth month, the ulcers in the neck region began to resolve gradually leaving scar tissue. Overall, she showed a remarkable satisfying response with the anti-tuberculous therapy.

DISCUSSION

The prevalence of tuberculosis (TB) is projected to be around 9.6 million worldwide. It has been challenging time for clinicians in diagnosing cutaneous tuberculosis due to varieties of presentations that are like other cutaneous lesions. CT was recognized by Beyt at al. in 1981.⁴ It was described that the most common type of cutaneous TB is scrofuloderma which is predominantly found in children.

However, in our case it was found in a 35 year old adult. Typically, it occurs commonly in the neck, chest, and axilla region. Scrofula is an old term representing tuberculosis infection of lymph nodes in the neck, known as cervical tuberculosis lymphadenopathy.⁴ It is frequently the result of primary tuberculosis infection of the lymph node. *M. tuberculosis* bacilli can disseminate through lymph node and blood (hematogenously). Scrofuloderma represents a condition manifested by a bluish-red nodule overlying an infected lymph gland, joint or bone that ruptures to form an undermined ulcer with a granulating tissue at the base. Irregular adherent masses, densely fibrous tissue and discharging sinuses can occur as a result of the disease progression.^{3,4}

Chest X rays are mandatory to rule out systemic TB. Tuberculin sensitivity usually is marked but has a very low specificity. Histopathological examination is confirmatory, which reveals the presence of tubercular granulomas with epitheloidal cells, Langhan's giant cells and lymphocytes. According to various reports, only a small percentage of histopathological specimen's stain positive for acid fast bacilli. A combined effort of using the available clinical, radiological, and microbiological modality to reach early diagnosis can go a long way to avoid misdiagnosis and unnecessary delay in the treatment, especially in cases, without the pulmonary involvement. Various other conditions can clinically mimic scrofuloderma and should be correctly identified and differentiated. Differential diagnosis for discharging sinuses can be atypical mycobacterial infection due to mycobacterium scrofulaceum and *M. avium intracellulare*, actinomycosis, sporotrichosis, botryomycosis, nocardiosis.⁴ Other common conditions at primary care level that may presented with similar lesions include chronic eczema and recurrent impetigo. However, these conditions are not associate with alarming symptoms such as weight loss and loss of appetite which are present in our case. Therefore, the decision to have a shared care with the physician or dermatologist is indeed important in this case.

The treatment of cutaneous TB is crucial and universal measures should be taken to tackle any concomitant illness causing immunosuppression; and people in close contact with the patient such as the family members should undergo testing for TB. World Health Organization (WHO) and Clinical Practice Guidelines Management of Tuberculosis by Ministry of Health Malaysia recommends treatment for cutaneous tuberculosis which is anti-tuberculosis regimen containing Rifampicin (R), Isoniazid (H), Pyrazinamide (Z) and Ethambutol (E) for two months known as intensive phase followed by Rifampicin (R) and Isoniazid (H) for the next four months which is the maintenance phase.⁵ This treatment generally uses directly observed treatment short course (DOTS) approach to ensure compliance and successful treatment.

CONCLUSION

We report here a rare case of Scrofuloderma in a 35 year old lady which was successfully treated with anti-tuberculosis therapy. This form of CT is quite challenging to diagnose due to its similarity to many other skin lesions as it may be misdiagnosed. Therefore, a thorough history taking, complete examination and various relevant investigations should be carried out for early diagnosis and initiation of treatment for comprehensive recovery. In addition, the importance of completing the treatment should be emphasised to the patient to ensure successful therapy.

REFERENCES

1. Ganesan A, Kumar G. Scrofuloderma: A rare cutaneous manifestation of tuberculosis. J Indian Acad Oral Med Radiol 2017; 29(3): 223.
2. Soeroso NN, Harina EG, Yosi A. A very rare case of scrofuloderma with multiple cervical lymphadenitis tuberculosis. Respir Med Case Rep 2019; 27: 100842.
3. Gönül M, Gül Ü, Kılıç A, Soylu S, Demiriz M, Kubar A. Coexistence of tuberculosis verrucosa cutis with scrofuloderma. Turkish J Med Sci 2008; 38(5): 495-Multiple ulcers with suppurative surface and crusts over her neck area.9.
4. Beyt Jr BE, Ortals DW, Santa Cruz DJ, Kobayashi GS, Eisen AZ, Medoff G. Cutaneous mycobacteriosis: analysis of 34 cases with a new classification of the disease. Medicine (Baltimore) 1981; 60(2): 95-109.
5. Ministry of Health Malaysia, Academy of Medicine Malaysia, Malaysian Thoracic Society, World Health Organization. Clinical Practice Guidelines Management of Tuberculosis (3rd Edition). Vol. (5)2, Ministry of Health Malaysia. 2012. 109 p.