Scrofuloderma in a 1-year-old girl with severe malnutrition: a case report

Madeleine Ramdhani Jasin, Fahreza Aditya Neldy, Darmawan Budi Setyanto, Gufron Nugroho, Valerie Sunhaji, Maria Francisca Ham

Scrofuloderma can be rarely find in children. The condition needs to be considered as a differential diagnosis if we find a characteristic skin lesions in lymph node regions with associated systemic manifestations suggestive of TB. Late diagnosis and inappropriate treatment can lead to poor prognosis and antimicrobial resistance. In this report, we present a case of a severely malnourished 1-year-old girl with scrofuloderma. [Paediatr Indones. 2023;63:511-6; DOI: https://doi.org/10.14238/pi63.6.2023.511-6].

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Tuberculosis (TB) remains a global burden and a leading cause of mortality due to infectious diseases worldwide. In 2020, 845,000 new TB cases (312 per 100,000 population) were diagnosed, with 96,000 deaths, including 4,700 deaths of TB with HIV. Tuberculosis typically affects the lungs, but it can also affect other organs, a condition termed extrapulmonary TB. In 2020, 16% of all TB cases had extrapulmonary manifestations. The first case of TB affecting the skin was reported in 1826. There are many types of cutaneous TB; one of the most common is scrofuloderma, more often found in children and young adults. Scrofuloderma starts with a lesion in a lymph node, bone, muscle, or tendon that spreads to the skin. The lesion progresses very slowly and is usually not painful. Therefore, many patients wait to seek treatment until an advanced stage, leading to poor prognosis.

A one-year-old girl presented with a chief complaint of an enlarged lump on the right side of her neck. The mass was initially noticed one month prior to admission and grew in size before eventually rupturing a week prior to admission. In the previous six months, she had also experienced recurrent prolonged fever, weight loss of 2 kg, and lethargy, but no history of cough. She had no known contact with anyone diagnosed with TB or chronic cough, and her vaccination status was complete.

Physical examination of the right cervical region revealed a superficial, irregularly shaped ulcer with livid edges, reverberating walls of approximately 7x6 cm, and wound seepage (Figures 1 and 2). She was severely malnourished with a weight of 7.8 kg and height of 73.7 cm; her weight-for-height z-score was between -2 to -3. Other physical examination findings were within normal limits. Chest x-ray examination showed consolidation in the upper-middle-lower field of the right lung (Figure 3).
The patient’s tuberculin test was negative and she underwent biopsy of the right cervical mass. Histopathology showed chronic granulomatous inflammation with extensive caseous necrosis, a dense form of histiocyte and epithelioid cells, some datia Langhans cells, and lymphocytes, including a pattern of neutrophils and plasma cells (Figure 4).

The patient was diagnosed with scrofuloderma and started on antituberculous drugs (isoniazide, rifampicin, pyrazinamide, and ethambutol) immediately. During follow-up, there was significant improvement of the scrofuloderma; ulcers slowly resolved (Figure 5). She also gained more than 2 kg within two months of treatment. Antituberculous treatment was continued for 6 months and the patient was followed up in the clinic monthly.

Discussion

Symptoms of childhood TB are usually non-specific. Children with symptoms of prolonged fever for more than 2 weeks, weight loss or difficulty gaining weight, chronic cough, enlarged lymph nodes, and/or history
Figure 3. Chest X-ray shows right lung upper-middle-lower field non-homogeneous consolidation, accompanied by an atelectasis component with minimal tracheal deviation to the right.

Figure 4. Figure 4. (1) Extensive necrosis impresses the plant area. (2) A solid release of (A) histiocyte and (B) epitheloid cells. (3) Datia langhans cells. (4) (A) neutrophils, (B) Datia langhans cells, (C) epitheloid, and (D) histiocytes.
of close contact with adults with active TB should be evaluated for TB disease. This is done by culturing expectorated sputum, collected by sputum induction or gastric lavage. If bacteriological confirmation is negative or a specimen was not obtained, the Indonesian Pediatric Society TB scoring system can be used to diagnose TB clinically. Using the scoring system, those with scores of >6 are diagnosed clinically with TB disease and treatment can be initiated. However, in extrapulmonary TB, bacteriological confirmation for extrapulmonary specimens should be obtained, such as from specimens from enlarged adenopathy and/or ulcers to diagnose scrofuloderma or TB lymphadenopathy, cerebrospinal fluid to diagnose TB meningitis, or skin biopsy for cutaneous TB.\textsuperscript{6-9}

Scrofuloderma is a severe clinical form of secondary cutaneous tuberculosis. It is commonly characterized by an ulcer with a reverberated wall, as well as bluish-red nodules covering lymph nodes, bones or joints. The ulcer leads to disruption of affected tissue, which would then be replaced with granulation tissue, followed by suppurative discharge via sinus tracts.\textsuperscript{10} Scrofuloderma, often called tuberculosis colliquative cutis, results from the direct spread of tuberculosis lesions from nearby infected organs or endogenous spread by contiguous extension. The most common sites of lymph node involvement are the neck, axilla, and/or inguinal areas.\textsuperscript{11}

Cutaneous tuberculosis accounts for merely 1-2% of extrapulmonary tuberculosis.\textsuperscript{12} Scrofuloderma, one of the most common types (84%) of cutaneous tuberculosis, mostly infects children and adolescents.\textsuperscript{2,13} The diagnosis of scrofuloderma is usually confirmed by needle aspiration or excisional biopsy and demonstration of acid-fast bacilli by microbiological staining and suggestive histopathology.\textsuperscript{14} The tuberculin skin test may reveal a negative or positive result, as its sensitivity varies from 33% to 96% and its specificity is only 62.5%.\textsuperscript{15} Tuberculous chancre, miliary tuberculosis, and tuberculosis orificialis commonly show negative cultures, while cultures from scrofuloderma and lupus vulgaris cases are commonly positive.\textsuperscript{15} Our patient had a negative tuberculin test, possibly due to anergy, the inability to react appropriately to tuberculin skin tests due to a weakened immune system, even though the patient was already infected with TB. This condition can be found in patients with malnutrition, other infections, dehydration, long-term corticosteroid use, or HIV.\textsuperscript{16}

Cutaneous tuberculosis is classified based on morphology, route of spread, and patient immune status. This disease has six major infection routes such as direct transmission to the skin from organs under the skin, direct inoculation of the skin around the genital orifice, hematogenous transmission, direct transmission of mucosal lymphokines, and microbes that enter the skin directly.\textsuperscript{17,18}

According to the Indonesian Pediatric TB
Guidelines, scrofuloderma treatment includes four anti-TB drugs, consisting of rifampicin, isoniazid, pyrazinamide, and ethambutol for an intensive phase of two months, followed by continued rifampicin and isoniazid for the next four months. The drug of choice should be personalized according to weight and the presence of contraindications. Ulcers should be covered with wet gauze and an additional 1:5,000 dilution of potassium permanganate solution. Our patient received isoniazid, rifampicin, pyrazinamide, and ethambutol for two months and was observed every two weeks at the Respirology Clinic during this intensive phase. In the maintenance phase, she visited the Respirology Clinic monthly until 6 months of therapy was completed.

The prognosis of scrofuloderma is good; no mortality data has been reported. The cure rate for scrofuloderma is dependent on several things, such as immunity status, adherence to treatment, and self-hygiene. Many studies have reported that after receiving treatment for six months, most lesions heal appropriately with no active pus, although some skin lesions are slower to heal and may leave an unsightly scar. After one week of antituberculous therapy, our patient’s wound showed significant improvement, promoted also by the parents’ compliance in administering the antituberculous drugs, as well as their care for the wound with proper hygiene.

From this case, we learn that scrofuloderma needs to be considered as a differential diagnosis in children with characteristic skin lesions in lymph node regions with associated systemic manifestations suggestive of TB. Although rare, scrofuloderma can be found in children. Obtaining tissue specimens from the specific extrapulmonary sites is therefore necessary. Late diagnosis and inappropriate treatment can lead to poor prognosis and antimicrobial resistance. Prompt diagnosis and treatment will result in good clinical improvement.

References


