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CASE REPORT

Scrofula Mimicking Cutaneous Malignancy: A Rare Case Report

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ABSTRACT

Since a long time scrofuloderma (SCF) was thought to be a common disease of childhood and was attributable to *Mycobacterium tuberculosis*. In 1951, a new entity nontuberculous scrofuloderma was described and it is caused by atypical mycobacteria namely *Mycobacterium scrofulaceum*. The clinical picture closely mimics tuberculous scrofuloderma but diagnosis should be established through culture isolation and identification, because drug susceptibility may be different in these cases.

In this article, we report a case of a 22-year-old pregnant female patient who presented to us with scrofulaceous lesion on the right side of neck.

Keywords: Tuberculosis, Primary scrofuloderma, Scrofula, Nontuberculous mycobacteria.

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INTRODUCTION

Scrofula, a word derived from the Latin word *scrofa*, means a brood sow due to likeness of the cluster of nodes to piglets feeding from the sow. Scrofula is a term predominantly applied to skin involvement in tuberculous and nontuberculous mycobacterial infections (NTMs) affecting the cervical lymph nodes. In olden days, most of these lesions were thought to be caused by tuberculous mycobacteria but now a new kind of scrofuloderma attributable to atypical mycobacteria is described. Primary scrofuloderma is caused by *Mycobacterium scrofulaceum* (scotochromogens), *Mycobacterium avium* intracellulare (nonphotochromogens) and *Mycobacterium kansaii* (photochromogens). Primary scrofulderma is usually a benign, self-limiting lymphadenopathy with no organ involvement but clinical course may be different. Scrofuloderma is also called as tuberculosis colliquativa cutis, a common form of cutaneous tuberculosis affecting children and young adults frequently. In this, there is breakdown of skin overlying a tuberculous focus in the lymph node, bone, or joint and this is secondary scrofuloderma. Spontaneous healing can take place but it will take many years followed by hypertrophic scars. So hereby, a case is being reported for its rare presentation of scrofula in a young pregnant female patient which mimicked a cutaneous malignancy.

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CASE REPORT

A 22-year-old female patient reported to the department of ENT with a lump on the right side of neck from last 6 months. Swelling had been gradually increasing until it bursts open and become a discharging lesion (Fig. 1). On general examination, the patient was moderately built and nourished. There was no history of trauma, fever, cough, or weight loss. However, her medical history revealed that at the age of 6 months she had similar swellings in the neck and in the temple area and she took treatment for 4 to 5 months and swelling subsided leaving scars. One year back, she had developed similar swelling in the posterior part of the neck and she took treatment again for 1 month which also healed spontaneously leaving a scar. A further history revealed that she had a swelling in the right supraclavicular region which was gradually increasing in size and reached to stage of abscess formation. Patient consulted a local practitioner who drained the abscess which did not heal and patient finally presented with secondarily infected lesion, as shown in figure. On local examination, oval-shaped proliferative swelling of size 4 × 6 cm, granular surface with increased vascularity was seen in the right supraclavicular region (Fig. 2). On palpation swelling was firm in consistency, nontender and did not bleed on touch. On close examination, depressed atrophic scars (striae distensae) were also seen on face and neck (Fig. 3) seen keeping differential diagnosis of scrofula, squamous cell carcinoma, metastatic lymph node, actinomycosis, pyogenic granuloma, syphitic gumma, hedranitis suppurativa, and kichura disease, all the routine investigations including HIV were done. Erythrocyte sedimentation rate was raised. Montoux test was highly positive. No abnormality was detected in chest radiographs.

Fig. 1: Site of lesion

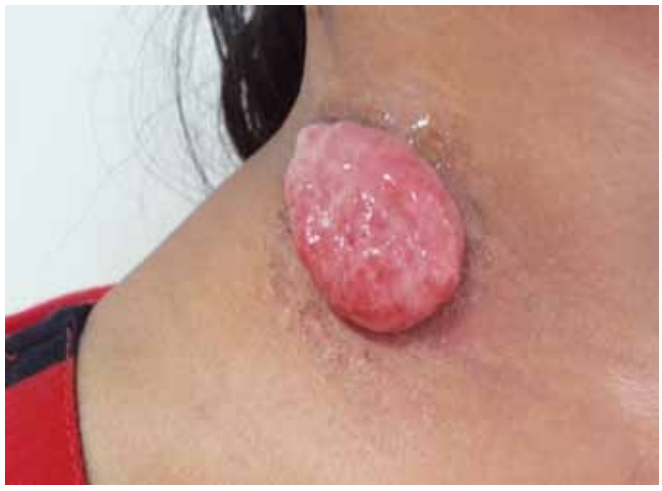

Fig. 2: Granular reddish surface

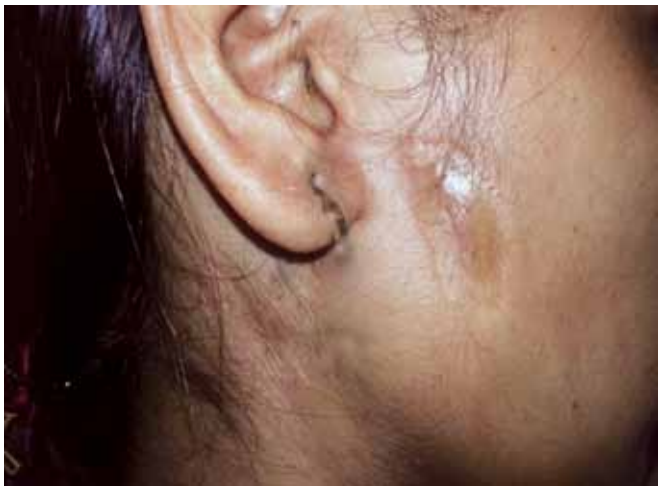

Fig. 3: Scar from previous lesion

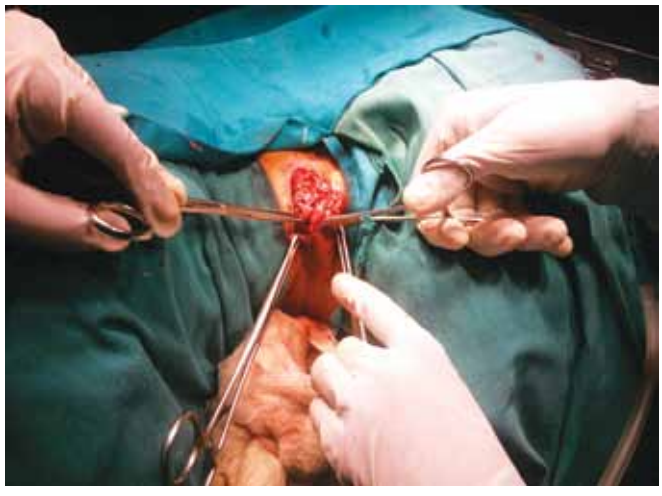

Fig. 4: Lesion being excised for excision biopsy

An ultrasound scan of the patient's neck reported the swelling 5 × 4 cm involving skin and it was highly vascular. The radiologist also made a note of few lymph nodes around it of varying sizes on the same side. The other side of the neck and other structures like thyroid and vessels were normal. Keeping in mind the antenatal status of the patient, since no definite therapy could be started wedge biopsy of the lesion was taken under local anesthesia for the diagnosis which, however, reported nonspecific inflammation and pathologist recommended excision biopsy. Antenatal check up of the patient including ultrasonography was within normal limits. Diagnosis of inflammatory pseudotumor of pregnancy was also kept in mind owing to exaggerated proliferative changes in response to pregnancy hormones.

Complete surgical excision of the mass (excision biopsy) was done under local infiltration (Fig. 4) and sent for histopathology. The report showed multiple epitheloid cell granulomas and plenty of Langhans giant cells, foci of caseation necrosis with dense infiltration of lymphocytes, histiocytes, plasma cells, and foci of granulation tissue.

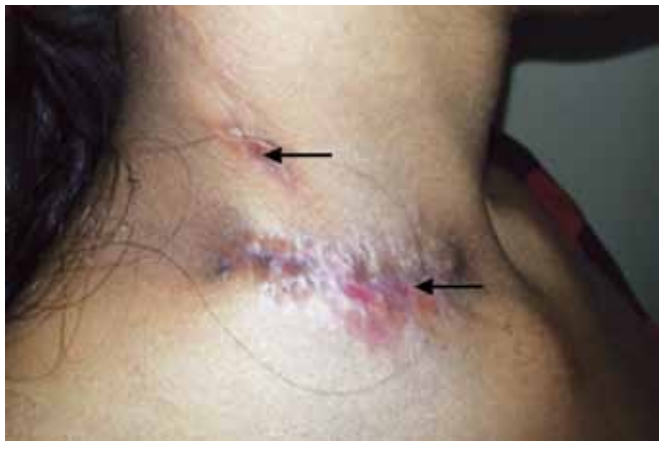

Fig. 5: Postoperative healing and scars from previous lesions (arrows)

Section was positive for acid fast bacillus which confirmed our clinical diagnosis of cervical tuberculous lymphadenitis with cutaneous involvement (secondary scrofuloderma). The patient was referred to the nearest DOTS center for treatment. Patient is on regular follow-up and lesion healed completely (Fig. 5).

DISCUSSION

The classic term scrofula derived from the Latin word glandular swelling scrofuloderma is also known as tuberculosis cutis colliquativa, a skin condition caused by tuberculous involvement of skin by direct extension usually from underlying tubercular lymphadenitis.[5] Hippocrates mentioned scrofulous tumors in his writing. The European kings of the middle ages imparted the royal touch to cure the 'King's evil' to which mycobacterial lymphadenitis referred. In second half of 20th century, scrofula became a less common disease in adults but remained common in children. The cutaneous involvement occurs often in children and in patients with low immunity. This risk increases further during pregnancy and early postpartum where immunity is somewhat compromised tuberculosis resurged again with appearance of AIDS and was known as 'cursed duet.'[3]

Tuberculous lymphadenitis most frequently involves the cervical lymph nodes followed in frequency by mediastinal, axillary, mesenteric, hepatic portal, perihepatic, and inguinal lymph nodes; while in primary scrofuloderma lymph nodes involved are either preauricular or submandibular. These present as a unilateral single or multiple painless growing masses.[1] Multiplicity, matting and caseation are three cardinal features in both types of scrofula. Jones and Campbell have classified peripheral tuberculous lymph nodes into following five stages:[1]

1. *Stage 1*: Enlarged, firm, mobile, and discrete nodes showing nonspecific reactive hyperplasia;
2. *Stage 2*: Large rubbery nodes fixed to surrounding tissue owing to periadenitis;
3. *Stage 3*: Central softening due to abscess formation;
4. *Stage 4*: Collar-stud abscess formation; and
5. *Stage 5*: Sinus tract formation.

Many reports suggest that usual port of entry in primary scrofuloderma is either eye/oropharynx or it is a primary involvement of skin and ultimately caseation or suppuration may occur. Approximately, 95% of adult scrofula cases are caused by mycobacterium tuberculosis, while the remaining 5% are caused by NTM, such as *M. avium* intracellulare, *M. scrofulaceum*, *M. kansasii,* and *M. chelonei*.[3] In children, this statistic is reversed, with NTM responsible for up to 92% of scrofula cases. Nontuberculous mycobacteria infections do not usually exhibit notable constitutional symptoms.[3] Diagnosis usually requires fine needle aspiration for histologic examination and culture of aspirate. The sensitivity of fine-needle aspiration cytology is 52.9% when used alone, but up to 82% when combined with polymerase chain reaction.[2] Once scrofula is diagnosed, it is important to determine the exact etiology as tuberculous or NTM as the latter is treated differently. The current standard drug regimen for sensitive *M. tuberculosis* consists of isoniazid, rifampicin, pyrzinamide, and ethambutol for the first 2 months, followed by isoniazid and rifampin for a total of 6 to 12 months. It is difficult to define a clear cut end point for assessing the efficacy of treatment of such extrapulmonary tuberculosis with delayed response to treatment.[4] The ATS/CDC/IDSA Committee (2003) indicated that therapeutic excision is not indicated except in unusual circumstances. Excision is reserved for NTM disease, for confirmation of diagnosis where there is suspicion as in present case and where there is residual disease after completion of ATT. In nontuberculous adenitis and some selected cases, surgery is more useful as it provides a rapid tissue diagnosis and confirms the bacterial type. Surgery increases the cure rate with excellent cosmetic result and a low complication rate. Surgical techniques include aspiration, incision and drainage, curettage, complete surgical excision of the lesion and the overlying skin. Treatment should be initiated during pregnancy as early as possible after diagnosis. Despite extensive necrosis, infiltration of adjacent fat planes is minimal that may help to differentiate this process from other infections or malignant nodal disease. There have been varying reports of exaggerated fibroblastic proliferative response in certain lesions (as seen in our case) due to pregnancy hormones and pseudotumor of pregnancy is one example of these kind of lesions.[6]

CONCLUSION

Atypical mycobacterial infections are uncommon and therefore there is a tendency to overlook the same. This case has been reported to emphasize the need for definite diagnostic differentiation between primary and secondary scrofuloderma by proper identification through isolation and culture, followed by institution of therapy as per drug susceptibility tests. Drug resistance strains may also be discovered and require intelligent handling in these cases. Although uncommon, scrofuloderma should be considered in cases of persistent lymphadenitis, particularly when there is cutaneous extension in patients from countries where tuberculosis is endemic.

REFERENCES

1. Mohapatra PR, Janmeja AK. Tuberculous lymphadenitis. J Assoc Physicians India 2009;57(6):585-590.
2. Iftikhar U, Nadeem M, Aman S, Kazmi AH. Scrofuloderma: a common type of cutaneous tuberculosis. J Pak Assoc Derma 2011;21:61-65.
3. Lin BH, Yeh HP. Nontuberculous scrofuloderma. Derm Sinica 1989;7:23-27.
4. Kvaerner KJ, Kvestad E, Orth M. Surgery required to verify atypical mycobacterial infections. Int J Pediatr Otorhinolaryngol 2001;61(2):121-128.
5. Golden MP, Vikram HR. Extrapulmonary tuberculosis: an overview. Am Fam Physician 2005;72(9):1761-1768.
6. Cantrell RW, Jensen JH, Reid D. Diagnosis and management of tuberculous cervical adenitis. Arch Otolaryngol 1975;101(1):53-57.