Histoid leprosy with penile shaft lesions

Sir,

A 30-year-old male patient presented to our outpatient department, complaining of an asymptomatic raised skin lesion over the penile shaft of 8 months duration. The patient had been married for 2 years and denied any premarital or extramarital sexual contact. He however, gave a history of ulceration over the penis 10 years ago which resolved with treatment. Details of the same were not available. The patient was a known diabetic, not well controlled at time of presentation and was on treatment with oral hypoglycemics for it. He did not give a history of any other significant skin or systemic disorder in the past. Other salient points in the history included a feeling of nasal stuffiness over the past 7 months and mild numbness of the skin over the lower legs and feet over the past 5 months.

On clinical examination, there was a discrete, well-defined, shiny, erythematous plaque of about 3 × 3 cm over the distal part of the penile shaft, on the ventral aspect [Figure 1]. Multiple smaller plaques of a similar nature were seen over the nape of neck and upper back [Figure 2] and both ear lobes [Figure 3]. There was no significant lymphadenopathy. There was minimal impairment of fine touch and temperature over the penile lesion. There was no evident sensory impairment over the other skin lesions. Peripheral nerve examination showed the right supratrochlear nerve to be thickened and tender. Bilateral ulnar nerves; common peroneal nerves, and posterior tibial nerves also showed significant thickening. There was no evidence of motor system involvement. Based on the clinical examination and the history, a possibility of histoid leprosy was considered, which was confirmed by a biopsy which showed collections of elongated, spindle-shaped histiocytes in a storiform pattern [Figure 4]. The histiocytes demonstrated an eosinophilic cytoplasm, round nucleus, and prominent foamy change. Acid fast bacilli stain staining demonstrated a large number of solid staining bacilli, many having an elongated, slender morphology [Figure 5]. Other investigations, including HIV ELISA, VDRL, and lipid profile, were within normal limits.

The patient was put on multibacillary multidrug treatment as per World Health Organization guidelines and is under follow up.

We also thought of a possibility of lesions on the penis being...
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a possible route for sexual transmission of leprosy, especially considering the high bacterial load. However, the patient’s wife on examination at the time did not show any features suggestive of an active Hansen’s disease. An ear lobe smear taken from the patient’s wife was also negative for bacilli.

Histoid leprosy originally described by Wade is considered to be a variant of lepromatous leprosy. It is characterized by presence of well-defined; shiny nodular lesions. Histoid lesions can arise as part of relapse or occur de novo. Mucosal and genital lesions have been described in leprosy, including the histoid type. Arora et al. and Kumar et al. reported genital lesions in 2.9% and 6.6% of leprosy patients, respectively. In our case like in most previously reported penile lesions of leprosy, the location was on the distal third of the shaft. Previous authors have discussed the possibility of the proximal part of the shaft being relatively immune because of the higher temperature.

The histopathology of histoid leprosy is probably its most distinctive feature. The notable histopathological findings are the presence of spindle-shaped histiocytes in the dermal infiltrate arranged in intertwining and whorled patterns. They contain numerous bacilli, and the cytoplasm may be slightly vacuolated but lacks the marked vacuolation seen in lepra cells. The bacilli are intact and rod-shaped in contrast to the fragmented forms seen in conventional LL. The precise reasons for the slender bacillary forms or the spindly transformation of the histiocytes have not been elucidated. One of the earlier theories was that histoid transformation represented a proliferation of sulfone-resistant mutants of Mycobacterium leprae after the elimination of the susceptible ones by therapy. This, however, does not explain histoid leprosy that has evolved de novo.

To the best of our knowledge, there is no confirmed, documented evidence of sexual transmission of leprosy. We would like to raise the possibility of a genital lesion with a high bacillary load being a possible route for transmission of the disease via sexual intercourse. In our case, the patients’ wife did not have signs of active disease. However, we realize that considering the long incubation period and the natural immunity to leprosy it would be difficult to comment on the significance even if the patient wife did develop leprosy.

To conclude, we would like to stress on the need to think of the possibility of leprosy in patients presenting with nodules on the external genitalia, especially in countries endemic for leprosy.

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Cutis laxa with pulmonary artery stenosis

Sir,

A 9-month-old female patient presented with cough and breathlessness of 1 week's duration. She was found to have loose, sagging skin all over the body since birth. Child was born of non-consanguinous marriage and was delivered full-term. Her developmental milestones were normal and she had no past history of major illness. At the time of presentation she had moderate respiratory distress with a respiratory rate of 38/min, a pulse rate of 132/min, and temperature of 100°F. On examination, apart from right upper zone crackles, a diastolic murmur was heard.

The skin of the entire body was extremely loose, hanging in folds, soft and inelastic. The changes were remarkable on face, neck, and flexures. Drooping of corners of mouth and loose skin around eyes gave her "blood hound" facial appearance [Figures 1 and 2]. The lax skin could be easily pulled from underlying tissue and when released would return slowly to its initial position. No other apparent abnormality of nails, mucosa, or genitalia was evident. No cyanosis, edema, or purpura were observed. No hernia or palpable lymph nodes were present. Her motor and sensory reflexes were normal.

On investigation her total WBC count was 16,000/cmm with 84% neutrophils. Other investigations including ESR, liver function tests, and renal function tests were normal. The test for TORCH antibody did not show any elevated titre. X-ray chest showed right and left upper zone patchy opacity with normal heart shadow. X-ray skull, clavicle, and carpal bones were normal. USG abdomen did not show any abnormality. 2-D echocardiogram showed right pulmonary artery stenosis with normal valves suggesting acyanotic congenital heart defects. Skin biopsy was not done as clinical diagnosis was obvious and parents did not give consent for the biopsy.

Her pedigree chart was made which showed two distant relatives of the same generation affected with similar skin changes suggesting an autosomal recessive (AR) inheritance [Figure 3]. No other details regarding any systemic abnormality in other affected family members could be obtained.

Cutis laxa (CL) is an acquired or inherited skin disorder