Perianal Eccrine Syringofibroadenoma
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Abstract
Eccrine syringofibroadenoma (ESFA) is a rare tumor of eccrine ductal differentiation with variable clinical findings and characteristic histological features. Reactive ESFA represents a reactive epithelial transformation in association with other inflammatory or neoplastic dermatoses such as chronic skin ulcers,[1] burn scars,[2] lepromatous or diabetic neuropathy, erosive palmoplantar lichen planus, nevus sebaceous, and preexisting malignant tumors such as squamous cell carcinoma. Till date, 75 cases have been described in literature, none of which were noted in the perianal region.[3]

Key Words: Deep bacterial infection, eccrine syringofibroadenoma, perianal

What was known?
Eccrine syringofibroadenoma is a rare eccrine ductal tumor known to occur due to reactive epithelial proliferation. It is mostly seen in chronic inflammatory conditions. Most common sites include extremities and trunk.

Introduction
Eccrine syringofibroadenoma (ESFA) is a rare tumor of eccrine ductal differentiation with variable clinical findings and characteristic histological features. Reactive ESFA represents a reactive epithelial transformation in association with other inflammatory or neoplastic dermatoses such as chronic skin ulcers,[1] burn scars,[2] lepromatous or diabetic neuropathy, erosive palmoplantar lichen planus, nevus sebaceous, and preexisting malignant tumors such as squamous cell carcinoma. Till date, 75 cases have been described in literature, none of which were noted in the perianal region.[3]

Case Report
A 31-year-old married male presented with painless, multiple, flesh-colored nodules and plaques on the perianal region that had persisted for 6 months. He reported that the skin lesion developed few months after a single episode of a painful swelling which was associated with pus discharge. The patient took treatment from local doctor following which swelling and discharge subsided. A month later, the patient had relapse of the painful lesions along with multiple painless swellings which gradually increased in size and were slow responsive to oral antibiotics. There was no history of unprotected sexual exposure other than spouse. There was no associated lymphadenopathy.

Cutaneous examination revealed multiple, moist coalescing, firm, flesh-colored nodules in the perianal region [Figure 1a]. There were also multiple atrophic linear scars seen around medial aspect of both gluteal folds [Figure 1b].

The patient was referred to the surgical department and showed no evidence of any hemorrhoids or pilonidal sinuses on evaluation. Upper GI endo scopy and colonoscopy examination was normal. Stool routine analysis and culture were normal. Pus culture from the discharge showed Staphylococcus aureus.

Routine blood investigations were within normal limits including serum iron and ferritin levels. The patient was also negative for HIV and venereal disease research laboratory serological tests. The patient however had slight elevation of blood sugar levels and had not been investigated prior for diabetes.

Two biopsies were taken, one from single fleshy nodule and other was taken from a painful nodule. The patient was started initially oral doxycycline initially for 2 months followed by a course of oral amoxycillin+

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2 Indian Journal of Dermatology | Volume 62 | Issue 5 | September-October 2017

clavulanic acid after pus culture sensitivity for the next 2 months following which lesions showed partial regression. No similar lesions were reported among family members.

On histopathological examination, thin anastomosing strands of uniform, small, epithelial cells arising from the epidermis to the dermis were observed [Figure 2a]. The cells were embedded in a cellular fibrous stroma and exhibited a latticed pattern characteristic of ESFA. Luminal structures were observed within the strands, and there were no cytological abnormalities [Figure 2b]. Due to financial constraints, we could not perform immunohistochemistry.

In the second biopsy from the painful nodule, we visualized dense suppurative neutrophilic inflammation in the dermis with overlying sinus wall formation in the epidermis [Figure 2c and d].

Discussion

ESFA is an uncommon tumor of eccrine glands that was first described by Mascaro in 1963. ESFA usually manifests as a solitary nodule on the extremities. Other sites of occurrence include the face and trunk. Clinical findings are variable, ranging from solitary nodules to multiple papules, nodules, and plaques. Five clinical subtypes have been reported: (1) multiple ESFA associated with hidrotic ectodermal dysplasia, (2) multiple ESFA without associated cutaneous features, (3) unilateral linear ESFA, (4) solitary ESFA, and (5) reactive ESFA.

The diagnosis of ESFA is based on its characteristic histopathological findings. The findings typically show proliferation of anastomosing strands and cords of monomorphous epithelial cells in a reticular pattern with eccrine duct formations embedded in a fibrovascular stroma.

The histopathological differential diagnosis includes fibroepithelial tumor of Pinkus, tumor of the follicular infundibulum, pseudoepitheliomatous hyperplasia, reticulated seborrheic keratosis, squamous cell carcinoma, and artifacts of histologic processing.

Considering our patient’s history of initial development perianal painful nodules with discharge followed by linear atrophic scars and eruption of multiple fleshy papules, ESFA following deep bacterial infection was the most likely diagnosis in this case. The suggested pathogenesis of reactive ESFA includes repeated eccrine duct trauma resulting in eccrine duct remodeling and repair.

Hidradenitis suppurativa was one of the close differentials considered before evaluation, but there was a history of painful nodules with discharge without cysts (blind boils) formation. Furthermore, the duration of the lesion was short, and also, there was no other site like axillae involved.

The clinical course of ESFA is mostly benign. However, malignant transformation to eccrine syringofibrocarcinoma and the association with squamous cell carcinoma have been reported.

Perianal nodules should be evaluated thoroughly, and other causes such as hemorrhoids, Crohn’s disease, perianal warts, and neoplasms must be ruled out. Deep bacterial infection can be a cause of persistent perianal nodules in set of immunosuppression like in our case, diabetes mellitus.

Out of the 75 cases of ESFA reported till date, to the best of our knowledge, none of the cases arose in the perianal region making this the first case with this unique presentation.

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Nil.

Conflicts of interest

There are no conflicts of interest.
What is new?
Perianal nodules presenting as syringofibroadenoma is rare presentation of this uncommon tumour. No such prior associations have been documented prior to this report.

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