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Acquired smooth muscle hamartoma associated with eosinophilic ulcers of the scrotum

Rania El -Tatawy, MD, Amany Abdel -Latif, MD, Laila Mansour, MD and Fatma Abd Rabboh, MD

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* Department of Dermatology and Venereology Faculty of Medicine, Tanta University, Accepted for publication in November 29, 2007 .

Case Report :

A 38 year-old Egyptian male presented to the Outpatient Clinic of Dermatology and Venereology Department of Tanta University Hospital in December, 2006 having skin lesions on the scrotum. He complained of tight scrotal skin with two episodes of ulceration and healing of 7 month duration. The ulcers were slightly painful with history of delayed healing. No history of other diseases including schistosomiasis or scabies or recent drug intake.

On examination, a patch of thickened skin was found on the left side of the scrotum with multiple, tender irregularly shaped ulcers of variable sizes (few mm - 4cm in diameter) with elevated indurated margins surrounded by erythematous zones with purulence emanating from the ulcers. They were scattered on the left side of the scrotum and the upper medial aspect of the left thigh (**Fig. 1a**). There was a weak positive pseudo Darier sign. No lymph node enlargement was found. General examination and routine laboratory investigations revealed nothing abnormal.



Fig 1a: Multiple ulcers on the left side of the scrotum and the thigh.

A four mm punch biopsy specimen was obtained from the border of the ulcer including the adjacent intact elevated margin. Hematoxylin and eosin (H & E) stained sections revealed ulcerated epidermis with the adjacent skin showing epidermal hyperplasia. The dermis showed invasive dense cellular infiltrate of mononuclear cells (MNCs) and numerous eosinophils in addition to vascular hyperplasia (**Fig 2a, b**). Bundles of smooth muscle fibers separated by interfascicular fibrosis and collection of fat cells were found in the reticular dermis (**Fig 2c**). Eosinophils infiltrated deep into subjacent smooth muscles dissecting through and separating the muscle fibers. Masson's trichrome stain confirmed the smooth muscle nature of the fibers (**Fig 2d**).

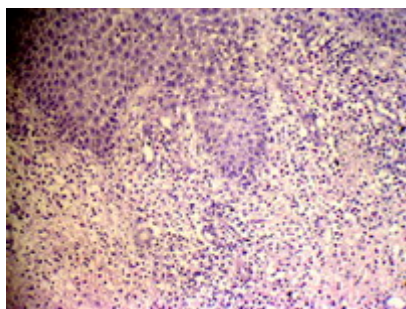


Fig 2a: Epidermal hyperplasia and dermal dense cellular infiltrate of mononuclear cells and numerous eosinophils (H & E X 160).

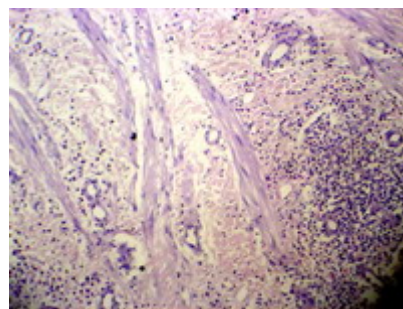


Fig 2b: Dense cellular infiltrate extending deep between smooth muscle, with vascular hyperplasia (H & E X 160).

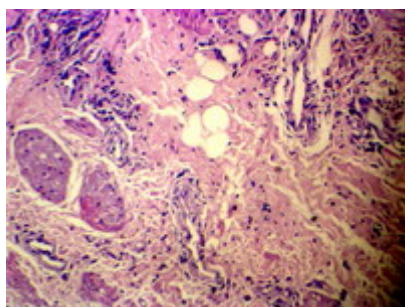


Fig 2c: Smooth muscle separated by interfascicular fibrosis and collection of fat cell (H & E X 160).

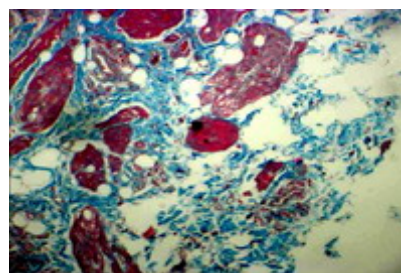


Fig 2d: Bundles of smooth muscles separated by collagen fiber and fat cells (Masson's trichrome stain(X 160)).

Immunohistochemical studies showed S100 positive cells (dendrocytes) in the dermis and increased number of Langerhans cells (LCs) in the epidermis adjacent to the ulcer (**Fig. 2e**). CD68 immunohistochemical stain demonstrated that the large MNCs included CD68 positive histiocytes

(Fig. 2f).

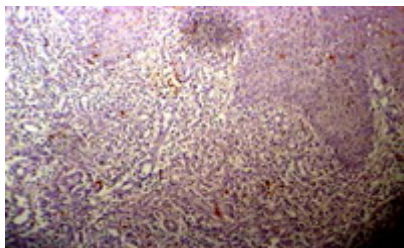


Fig. 2e: S100 immunoperoxidase stain showing focally clustered S100 +ve dermal cells and increased Langerhans cells in the epidermis (X 160).

Fig. 2f: CD68 immunohistochemical stain demonstrating that the large MNCs include CD68 +ve histiocytes(X 160).

The clinical, histological and immunohistochemical findings were in keeping with the diagnosis of acquired smooth muscle hamartoma with eosinophilic ulcers of the scrotum. The patient was treated with 3 intralesional injections of triamcinolone acetonide (5 mg/ml) at 2 weeks intervals. He got satisfactory improvement with healing of the ulcers with scarring. (**Fig.1b**).



Fig.1b: Scrotal ulcers healed with scarring after treatment with intralesional triamcinolone acetonide (5mg/ml)

Discussion:

Smooth muscle hamartoma (SMH) is a rare benign proliferation of smooth muscles first described by Stokes in 1923 [1]. Initially, it was known as congenital SMH but later, few cases of acquired variant have been reported [2,7]. Congenital SMH is usually characterized by

hypertrichosis, hyperpigmentation and induration. The lesions usually occur on the trunk and proximal extremities. Patches, plaques or less frequently papular follicular lesions may be noted. Transient piloerection or elevation of a lesion induced by rubbing (pseudo- Darier sign) is often seen [8]. It is believed that the pseudo- Darier sign may be produced by the prominent nerve fibers observed between smooth muscle bundles under electron microscopy [8]. Rarely, SMH occurs as acquired lesions without hyperpigmentation or hypertrichosis. Those acquired cases have been reported on the abdomen, shoulder, vulva and scrotum [2,7]. Histologically, SMH shows variably oriented discrete hyperplastic smooth muscle bundles in the mid to lower dermis with overlying epidermis showing variable acanthosis and hypermelanosis. Treatment of SMH is not necessary owing to the asymptomatic benign nature of the condition but involves surgical excision if desired [8].

Eosinophilic ulcer (EU) also known as traumatic ulcerative granuloma with stromal eosinophilia, Riga-Fede disease in infants or traumatic eosinophilic granuloma is an uncommon benign self limited lesion [9] poorly described in the dermatological literature [10]. It probably includes a spectrum of related disorders presenting as an ulcer with elevated indurated border affecting the tongue, oral mucosa or lip. EU typically presents by asymptomatic, mildly tender or painful, usually solitary non healing irregular ulcer occurring in either sex and in all ages [11].

Histological findings are characteristic and consist of eosinophil-rich cellular infiltrate accompanied by a population of large MNCs whose origins have been a matter of debate [9,11]. Immunohistochemical studies of these cells suggested a myofibroblast or histiocytic origin [10]. The large MNCs include two phenotypically distinct cell types: CD68-positive histiocytes and factor XIIIa-positive submucosal dendrocytes in varying ratios. Neutrophils, mast cells, occasional plasma cells and focally scattered S-100 positive histiocytes are also seen. An increased number of LCs may be identified in the epithelium adjacent to the ulcer [9,12].

The pathogenesis of EU is unclear. It was considered as a benign reactive lesion that is usually associated with history of trauma. Riga- Fede disease is a form of EUs that develops in infants as a result of chronic mucosal trauma from adjacent primary teeth [9]. Several investigators have proposed that EUs develop as a result of T-cell mediated immune response. In certain predisposed individuals, recurrent trauma may lead to alteration of tissue antigens or ingress of unknown factors (e.g. viral particles, toxic microbial products), which result in a hypersensitivity reaction. Activated T-lymphocytes produce a variety of lymphokines that are involved in eosinophilic maturation and may act as eosinophil chemotactic factors. Damage and degeneration of mucosal tissues may be due to proliferation of cytotoxic T-cells or toxic products released from degranulating eosinophils. Eosinophil secretory granules contain a number of highly cytotoxic proteins, including eosinophil cationic protein, major basic protein and eosinophil derived neurotoxin [9,12].

The histologic differential diagnosis may include angiolymphoid hyperplasia with eosinophilia, Langerhans cell histiocytosis and lymphoma. Eosinophilic ulcer can represent another histological simulator of CD30 positive lymphoproliferative disorders [10]. Treatment of EU includes removal of the traumatic irritant, palliative and corticosteroid treatment. Prognosis is excellent even with conservative treatment. However, if EU does not resolve after 2 weeks of treatment, biopsy is recommended. Occasionally lesions may have to be surgically excised [9,11].

Our case fulfilled the clinical, histological and immunohistochemical criteria for diagnosis of EU but what we found interesting is its occurrence on the scrotum not in oral mucosa as the previously reported cases. To the best of our knowledge, this is the first reported case of EU occurring in the scrotum. The clinical differential diagnosis of scrotal ulcers may include ulcers

related to trauma (physical, chemical, thermal), Behcet's disease, infectious agents (viral, bacterial, mycobacterial fungal), contact or systemic allergy (including drug reaction) neoplastic diseases, systemic diseases hemolytic and autoimmune, disorders, vasculitis), sarcoidosis and chronic granulomatous disease.

Another interesting finding in our case of eosinophilic scrotal ulcer is its association with acquired SMH of the scrotum. The relation between both conditions is unclear. In one study, it was found that eosinophils can stimulate the formation of myofibroblast-like cells [13]. On the other side, cultured human airway smooth muscle cells stimulated with IL-1 supported eosinophil survival through production of GM-CSF and were suggested to contribute to eosinophil accumulation in the airway [14].

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