

Letter To The Editor

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A Rare Case Of Congenital Angiokeratoma of Fordyce

To the Editor. - Angiokeratoma of Fordyce is a benign vascular dermatosis characterized by dilated vessels of the superficial dermis associated with epidermal hyperplasia and mostly occurs on scrotum. Angiokeratomas often arise in the second or third decade but are commonly diagnosed in elderly men. Congenital form of the disease reported in the literature is extremely limited [1]. Herein, we report a rare case of a congenital angiokeratoma localized to the scrotum.

An otherwise healthy 5-year-old boy presented with a history of a lesion which developed shortly after birth on the scrotum. Dermatological examination revealed a violaceous crusted plaques and red-to-reddish blue papules at the scrotum (**Figure 1**). His history revealed that the lesion was enlarged and dark areas on the surface had occurred in the years.

Angiokeratoma of Fordyce most commonly occurs as multiple asymptomatic, 2 to 5 mm, dark keratotic papules with scaly surface located on the scrotum. However lesions may occur on shaft of penis, labia majora, inner thighs, or lower abdomen. The pathogenesis of disease is not exactly known; the speculated pathogenesis is localized venous hypertension [2]. Histologically, the lesions are characterized by ectasia of superficial dermal blood vessels and hyperkeratosis. Angiokeratomas are harmless surface vascular lesions that can usually be left alone. However, the disease was reported as a cause of intense scrotal bleeding [3]. As the black spots sometimes resemble melanoma, a skin biopsy may be performed to rule out malignancy and allay any fears. If bleeding becomes a concern or treatment is requested for cosmetic purposes, they can be removed. Surgical options include excision, laser therapy, cryotherapy or electro cautery.

A study conducted with 1552 Japanese males found that angiokeratomas were most common among people older than 40 years. The reported prevalence of the disease increases with age, from 0.6% in adolescent males to 17% in those older than 70 years [4]. However in a PubMed search we found only one case report describing isolated congenital angiokeratoma of Fordyce [1]. The clinical presentation of this case is unique, congenital onset characteristics that have been reported only rarely in the literature.

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Figure 1. Violaceous crusted plaques and red-to-reddish blue papules at the scrotum

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