Idiopathic scrotal calcinosis

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Summary

Idiopathic scrotal calcinosis is a rare benign disease characterized by multiple, asymptomatic and painless nodules on the scrotum. Over 100 cases have been reported to date. We herein report our case with this rare disease and shortly review the literature.

Key words: Calcinosis, scrotum

Özet

İdiyopatik skrotal kalsinozis

İdiyopatik skrotal kalsinozis skrotum derisi üzerinde görülen, birden çok sayıda asemptomatik ve ağrısız nodüller ile karakterize benign bir hastalıktır. Bugüne kadar 100'den fazla olgu bildirilmiştir. Burada nadir görülen hastalığa sahip olgumuz sunularak, bu konudaki literatür bilgisi kısaca gözden geçirilmiştir.

Anahtar kelimeler: Kalsinozis, skrotum

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Introduction

Idiopathic scrotal calcinosis (ISC) is a rare benign condition which presents with multiple, asymptomatic and painless nodules on the scrotal skin wall. Various theories regarding the etiology and pathogenesis of this condition have been proposed although none has been widely accepted. Although calcinosis of the scrotum was first described by Lewinski in 1883 (1), its cause and nature have remained elusive. Hutchinson has emphasized that these calcific masses appear with no evidence of underlying cause (2), and finally the term "idiopathic scrotal calcinosis" used by Shapiro in 1970 has most commonly been accepted (3).

Histologically, ISC is characterized by the presence of calcium deposits that are variable in size within the dermis, often surrounded by a foreign body-type granulomatous reaction. Some cysts reveal calcification of their keratin contents with little evidence of active inflammation.

Because of its rarity and controversial pathogenesis of the disease, we present a case of ISC with multiple nodules over the scrotal skin.

Case Report

A 21-year-old man was referred to the outpatient clinic of urology service with one year history of painless multiple cystic bumps over both sides of the scrotum. The lesions were measured 0.5 to 2 cm in diameter and skin-colored to white-yellow and moveable beneath the skin surface. The patient stated that he did not experience itching, pain or discharge. The testes were normal. Blood chemistry including serum electrolytes, urea, creatinine, calcium, phosphate, uric acid

and alkaline phosphatase and parathyroid hormone levels were normal. The patient complained of poor cosmesis and wanted the lesions be removed, and the nodules were extirpated surgically under local anesthesia. The recurrence was not observed after a 6-month follow-up period. Histologic examination of an excised nodule stained with hematoxyline and eosin showed a thin layer of epidermis overlying a dermal lesion made of multiple cystic spaces of varying sizes. These spaces were distended by large amorphous granular basophilic material consistent with ICS (Figures 1,2).

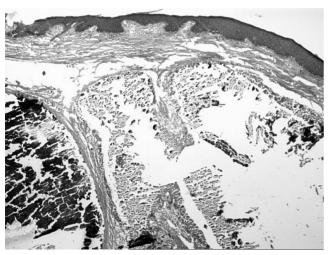


Figure 1. Histologic section of a nodule from scrotal skin shows a large circumscribed area of amorphous granular basophilic material in the dermis (Hematoxyline-Eosin, ×4)

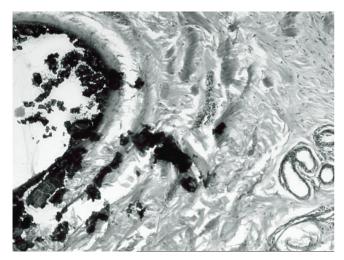


Figure 2. Focal collection of basophilic material (calcium) within the dermis of the scrotal skin in a dense fibrous stroma (Hematoxyline-Eosin, ×20)

Discussion

ICS is a rare condition characterized by multiple, slowly growing and distinct nodular masses embedded within the dermis of scrotal skin, and was first described by Lewinski in 1883 (1). The lesions have been attrib-

uted as sebaceous cysts, calcified steatocystoma, fibroma, atheroma and xanthoma. Shapiro et al. reviewed the histologic data and found no evidence of an epithelial lining, residual cysts and lipid or organisms, and concluded that the calcification was idiopathic introducing the term "idiopathic scrotal calcinosis" (2).

ISC occurs mainly between 20 to 40 years of age, and the youngest and the oldest patients were 9 and 85 years old, respectively (4). The pathogenesis is unclear and controversy exists as to whether the condition is idiopathic or the result of dystrophic calcification of preexisting epidermal cysts (5-8).

Akosa et al. (9) and recently Saad and Zaatari (4) have reported that scrotal calcinosis might result from inflammation of epidermal cysts followed by dystrophic calcification within the keratin of the cyst or dermis adjacent to a ruptured cell wall. Others support the theory of epidermal conclusion cysts playing a major role in the pathogenesis of the disease (5). In fact the idiopathic etiology is based upon the absence of the epithelial lining within the lesions (5,6,8).

Swinehart and Golitz also described this epithelial lining surrounding the calcified material and offered a model for ICS (10). Veress and Malik (11) and Feinstein et al. (12) have suggested that minor trauma plays an important role as the starting point of dystrophic calcification. And recently Pabuccuoglu et al. have reported that dartoic muscle degeneration and necrosis are the most important factors in the process of this pathology (13).

Most of the cases are asymptomatic and patients usually seek medical advice for cosmetic reasons as in our case. Surgical excision and histopathologic examination are necessary to confirm the diagnosis. Recurrence is unusual.

We think that the cause of the condition is most likely multifactorial. There is evidence supporting the calcification of the ruptured epithelial cysts, and the cause of ICS may be the degeneration process of the dartoic muscle. For the remainder of the cases, there is still difficulty in finding a preexisting lesion and the use of the term "idiopathic" may be appropriate.

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