CASE REPORT

Scrotal Calcinosis, Dystrophic or Idiopathic, Does It Matter? A Case Report

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Scrotal calcinosis is a rare condition with controversy surrounding its unclear etiopathogenesis. Several schools of thought have emerged from recent publications supported by histological findings seen in the respective cases reported. The recommendation of surgery as management, and its outcome, however, have remained the same throughout history. This paper reports a 36-year-old male who presented with gradually progressing multiple pruritic nodules over the scrotum who underwent wide excision with favorable outcome on follow up. Histological examination confirmed the diagnosis of scrotal calcinosis and showed evidence of intact epidermoid cysts that have undergone dystrophic calcification.

Keywords: scrotal calcinosis, dystrophic calcification, epidermal inclusion cyst

Introduction

Scrotal calcinosis is a form of idiopathic calcinosis cutis.[1] It is a rare pathology manifested by multiple or solitary calcified nodules involving only the scrotal wall. There have only been 102 publications on this disease so far. In addition, there is only one reported case in the Philippines.[2] The etiopathogenesis of the disease is still controversial, with several publications showing different histopathologic evidences.[3-8] Despite this controversy, surgery remains to be the best management, showing similar outcomes among cases. This paper aims to review literature concerning the etiopathogenesis of this disease, report a rare case of scrotal calcinosis showing histopathologic findings consistent with dystrophy along with the outcome following excision, and to correlate the findings with the studies reviewed.

The Case

A 36-year-old male presented with a 10-year history of hard painless pruritic nodular lesions over the scrotum. The lesions gradually increased in number and size, which on manipulation expressed moist chalk-like substance. The patient did not have any constitutional symptoms or any other genital lesions nor any history of scrotal and testicular infections, trauma or injury and other comorbidities. He denied promiscuous sexual activity. On examination, there were multiple non-tender hard nodules occupying approximately 40% the patient's scrotum, the largest of which measured up to 2cm in diameter (Figure 1A).

The patient underwent wide excision of the lesions under regional anesthesia. Hard round lesions over the scrotal wall measuring approximately

0.5-2.0 centimeters were noted. Elliptical incisions were done over the lesions followed by dissection of the scrotal dermis from the dartos muscle (Figure 1B). Primary closure was done (Figure 1C). The patient was then discharged within the day after an unremarkable post-operative course.

Histologic results showed multiple intradermal cysts of varying sizes and shapes lined by flattened stratified squamous epithelium containing basophilic calcified deposits with chronic inflammatory infiltrates and occasional foreign-body giant cells. The overlying epidermis is unremarkable. No variations in nuclear size, lack of differentiation and disruption of cellular architecture were noted signifying no malignant process (Figure 2). No recurrence of lesions was noted 6 months after the procedure. There was no disfigurement of the scrotum over the operative site. The quality of life returned to the patient's pre-disease state.

Discussion

In general, scrotal calcinosis presents as multiple or solitary monomorphic nodule over the scrotal wall. The condition is benign and no reports of secondary malignancies arising from these lesions have been reported so far.[4,8,9-12] Most patients are asymptomatic but may present with pruritus, swelling, secondary skin infections as well.[4,8,9-11] Progression has been described as indolent, and most patients consult several years or even decades after they first notice their lesions due to bothersome symptoms or cosmetic concerns.[10,11,13,14]

Scrotal calcinosis was first described as a rare benign condition classified as a subtype of idiopathic calcinosis cutis along with familial tumoral calcinosis and subepidermal calcified nodules. Other subtypes include: 1) dystrophic calcinosis cutis, which is a manifestation of an underlying connective tissue disease such as systemic sclerosis, systemic lupus erythematous and dermatomyositis 2) metastatic calcification, secondary to abnormal serum calcium and phosphorus levels caused by hyperparathyroidism, malignancies and chronic kidney failure, and 3) iatrogenic calcification, occurring in patients receiving calcium and phosphate containing substances. It is important to note that these other conditions are only manifestations of an underlying systemic disease or condition and are managed according to their underlying pathology.[1] Excluding these conditions is important before establishing a clinical diagnosis of scrotal calcinosis as the approach to the management of this condition is focused only on the scrotal lesions. It is important to emphasize that although scrotal calcinosis may

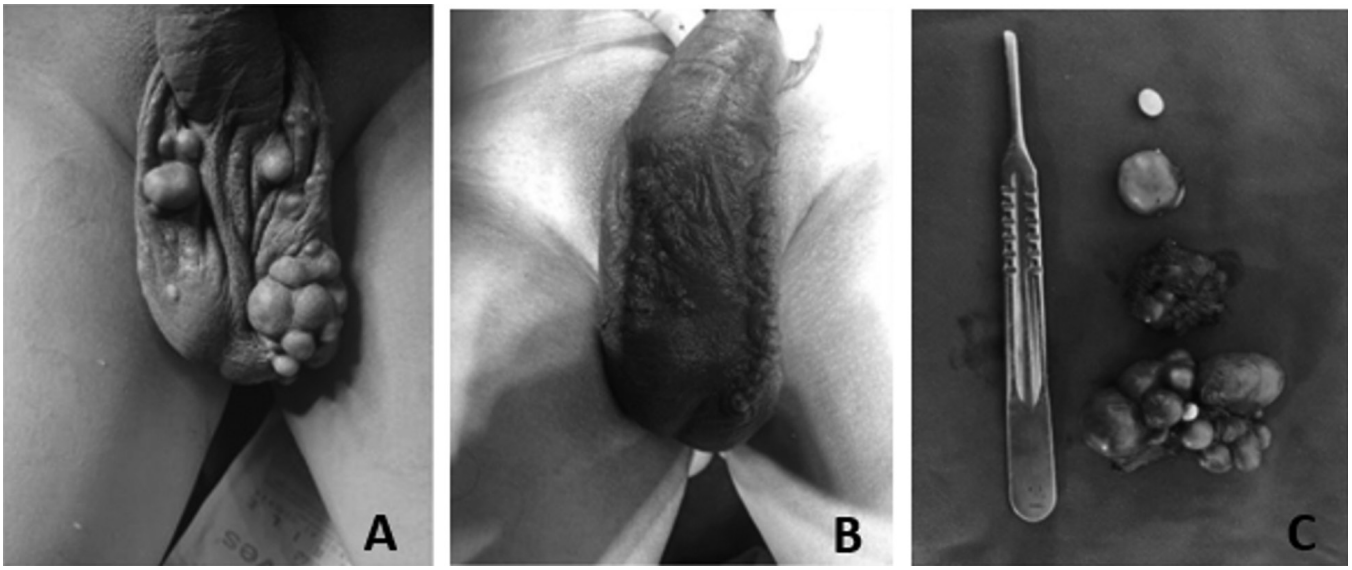

Figure 1. Pre-operative photograph (A), post-operative photograph (B) and photograph of excised lesions (C).
B – Note primary repair done of 2 vertical incisions made over scrotal wall; C - Note gross appearance of opened lesion (white, chalky, rounded).

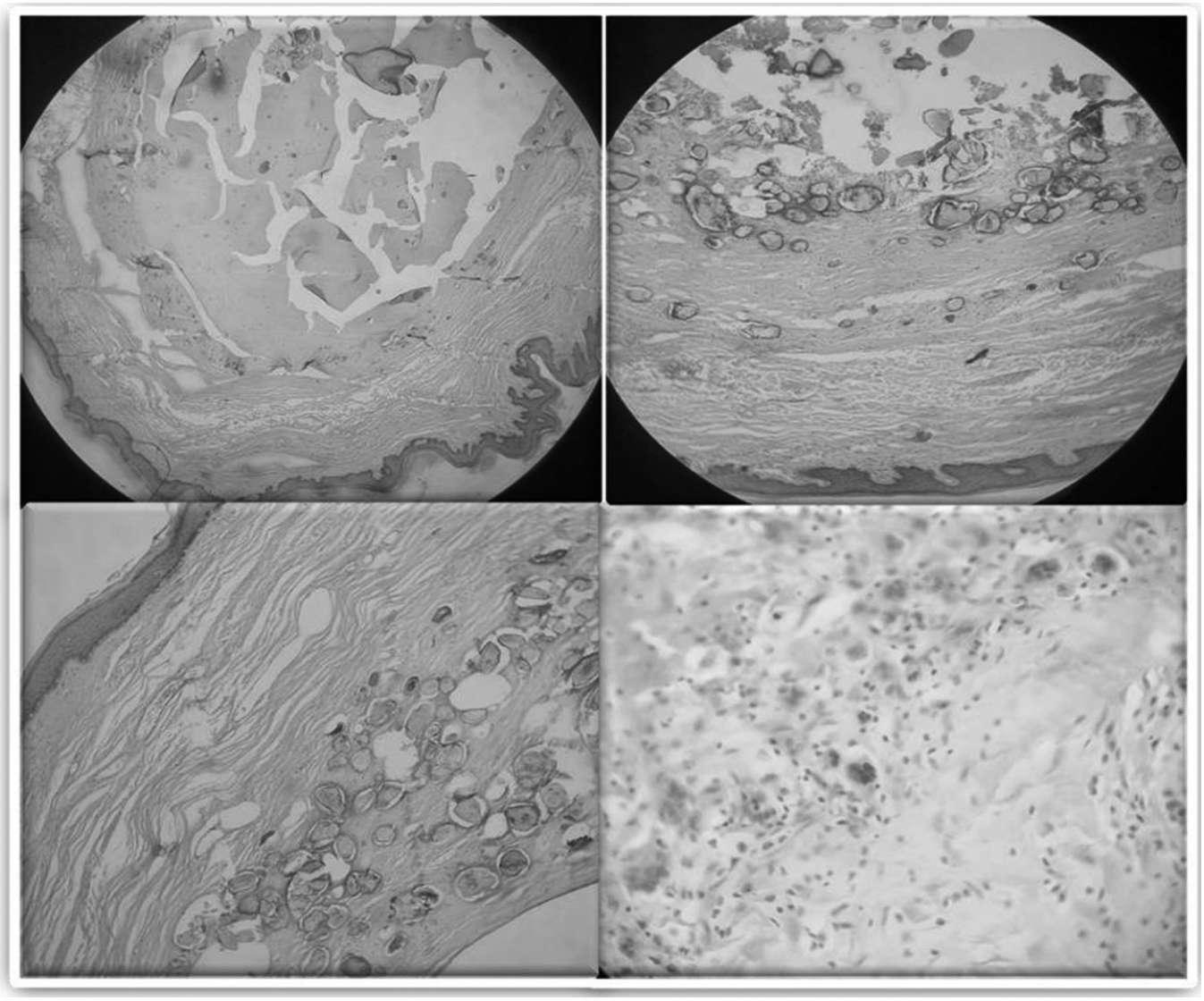

Figure 2. Microscopic sections from the patient's specimen. Note intraluminal basophilic calcific deposit and intact epithelium (upper left); Intradermal calcium deposits and multiple epithelial cysts (upper right and lower left); Further magnification (HPO) shows inflammatory infiltrates, giant body cells and fibrotic areas (lower right).

be secondary to dystrophic changes in either the dartos muscle or a preexisting epidermal inclusion cyst, it is a form of idiopathic calcinosis cutis and that it is a distinct disease entity from a dystrophic calcinosis cutis.

There already have been a number of publications on this disease (102 articles in PubMed database to date) and cases reported are frequently of black patients aged 20-40 years.[8,16,10-13] In the Philippines, there is only one publication reporting this case in a Filipino patient.[2]

Scrotal calcinosis remains to be surrounded by some degree of controversy, particularly surrounding its unclear etiopathogenesis. Several schools of thought have now arisen from recent publications suggesting different etiopathogenetic pathways in its formation. One hypothesis of particular interest is that these lesions are secondary to dystrophic calcification of epidermal inclusion cysts as findings of epithelial cysts and wall remnants are seen on histopathologic review of some cases.[3,7] This is supported by a case series by Shah, et al. 2007 wherein histopathologic findings from 20 different cases of idiopathic scrotal calcinosis were evaluated showing microscopic findings of intact epidermal cysts and remnant structures with variable degrees

inflammation and calcifications.[3] In contrast to this, the histopathologic findings from the publications of Pabuccuoglu, et al. in 2003 and Kelten, et al. in 2005 show calcific deposits within the dartos muscles surrounded by inflammatory cells and hyalinization in the absence of any evidence of epithelial cysts and cyst wall remnants suggesting the cutaneous lesions arising from a degenerative process of the dartos muscle leading to dystrophic calcification.[4,5] Another school of thought is that these deposits are caused by a disequilibrium in eccrine duct secretion leading to accumulation of debris and calcium within the cystic lumen. In a publication by Ito, et al. in 2001, specimen from a patient with scrotal calcinosis turned out to be reactive in special stains differentiating luminal cyst cells and eccrine duct cells, in particular, carcinoembryonic antigen, epithelial membrane antigen and gross cystic disease fluid protein.[6] Electron microscopy done also confirmed these findings confirming that calcified cysts seen on microscopy are of benign eccrine epithelial origin. There are however, publications showing cases devoid of these histologic findings from which a truly idiopathic pathology has been suggested.[10-14] In the case presented in this paper, histologic examination shows presence of multiple intradermal cysts with some calcifications lined by luminal epithelial lining suggesting a scrotal calcinosis secondary to calcific dystrophy of preexisting epithelial/epidermoid cysts in particular. Diagnosis is still confirmed histologically with the common finding of presence of basophilic dermal or luminal calcific deposits with or without inflammatory changes which were also observed in the patient presented in this case.

Surgery has been the only recommended mode of treatment for ISC and is shown to provide excellent outcomes in terms of patient quality of life and self-esteem.[8-10,11,14-16] Patients with this condition consult only due to bothersome symptoms attributed to this disease such as altered self-esteem due to the appearance of the lesions, severe pruritus, which this patient has, and ulcerations in more severe cases.[10,11,13] Management should be planned with consideration to the patient's aesthetic outcome and quality of life. So far, no untoward outcomes have been reported after a simple excision. The case presented, in particular, underwent wide excision of the lesions with primary closure and while histological findings were suggestive of preexisting epidermal cysts that have undergone dystrophic calcification supporting the school of thought that scrotal calcinosis are primarily epidermal inclusion cysts that have undergone calcific dystrophy, the patient on follow up showed similar outcomes from previous publications, regardless of histologic findings, in terms of quality of life and aesthetics.

Scrotal calcinosis is a rare benign condition occurring among young adults with no preexisting conditions or comorbidities. It is however, important to note that although the etiopathology of this disease is still unclear, there are recent publications that disprove traditional thinking that it is simply an idiopathic condition. Case reports are supported by histopathologic findings providing information as to the etiology of each individual case. Whether the pathogenesis is dystrophic or idiopathic, surgery is still the only recommended management for these patients and despite the recent advances in proving etiology, the approach to management and the respective outcomes did not deviate from previous publications. It is acknowledged that the recognition of these different etiologies is beneficial in terms of providing further information on this disease and although some publications describe cases devoid of evidences of these etiologies, careful consideration should still be made in utilizing the term "idiopathic" in describing this condition as to avoid under analysis of future cases. It seems unlikely, however, that these different etiopathogenesis have implications with regards to the management and the overall prognosis of this condition.

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