Scrotal calcinosis: pathogenesis and case report

J. Hicheri, T. Badri, B. Fazaa, R. Zermani, N. Kourda, S. Ben Jilani and M. R. Kamoun

SUMMARY

Scrotal calcinosis (SC) is a rare and benign condition defined as the existence of multiple calcified nodules within scrotal skin. We report the case of a 39-year-old male patient with a three-month history of scrotal tumours that increased rapidly in number and size. Histopathological and immunohistochemical investigations showed no evidence of epithelial structure. Whether SC is idiopathic or the result of the calcification of pre-existing cysts is still a controversial issue. In some cases, as well as in our case, no evidence of cystic structure was found around calcified material, despite minutely careful studies. This suggests that SC might be truly idiopathic.

KEY WORDS

calcinosis, scrotal, case report

Introduction

Scrotal calcinosis (SC) is a rare and benign condition defined as the existence of multiple calcified and asymptomatic nodules within scrotal skin that occur without any anomaly of the phosphor/calcium metabolism. These nodules usually occur during childhood or early adulthood (1). Histologically, SC 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 (2).

The pathogenesis of scrotal calcinosis is still controversial. Some authors think that scrotal calcinosis is the result of dystrophic calcification of preexisting structures such as epidermal, pilar, hybrid, indeterminate and ductal cysts. It may also be due to the degeneration of the dartoic muscles. Some authors did not find any evidence of preexisting cystic structures and think this condition is idiopathic (3).

We report the case of a patient with scrotal calcinosis and we discuss whether this disease is idiopathic or not.

Case report

A 39-year-old male patient presented with a three-month history of scrotal tumours that increased rapidly in number and size. His past medical history was unremarkable. Cutaneous examination revealed multiple, firm, painless, yellowish, subcutaneous nodules beneath the scrotal skin, 1 to 3 cm in size (Figure 1). Further

physical examination did not reveal any changes. Serum and urinary levels of calcium and phosphates were normal.

The excision of a nodule was performed. Histological examination showed under a normal epidermis a dermal globular bluish nodule containing an amorphous and homogenous substance, corresponding to deposits of calcium. The nodule was surrounded by a fibrous capsule, without any identifiable epithelial structure on several sections (Figure 2). There was also a foreign body-type granulomatous reaction. Immunohistochemistry using broad spectrum cytokeratin (DAKO) was negative. Surgery based on subtotal excision of scrotal skin was performed successfully with a good aesthetic result. No recurrence was observed after a 6-month follow-up period.

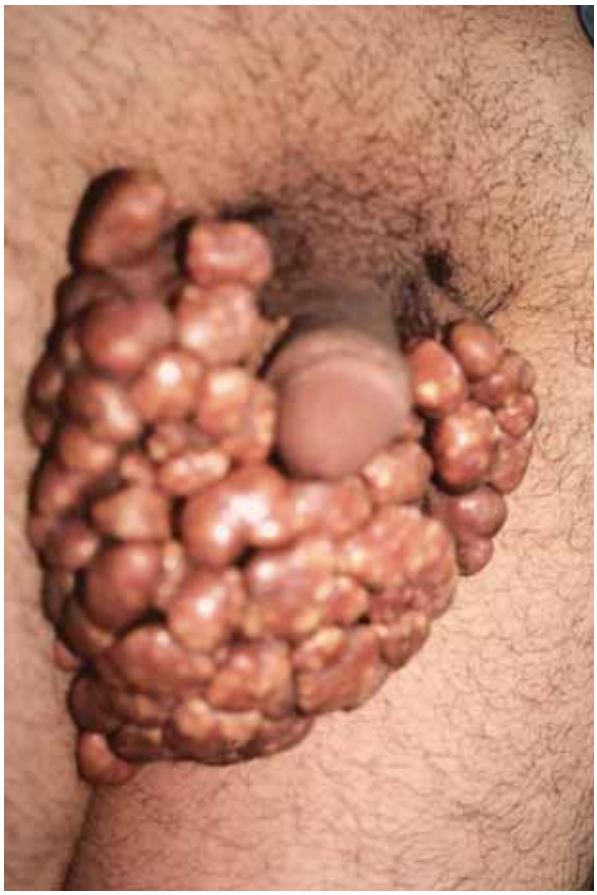

Figure 1. Multiple scrotal nodules.

Discussion

SC is a rare and benign condition described initially by Lewinski in 1883. It occurs mainly in men, 20 to 40 years old. The youngest and the oldest reported patients were a 9-year-old and an 85-year-old respectively (4).

Clinically, there are hard yellowish nodules that vary in size: from 1 mm to several centimetres. Lesions can be solitary or multiple, located within scrotal skin and can share the characteristics of epidermoid cysts. Although these nodules are usually asymptomatic, they can cause itching or may discharge a chalky material. A case of SC presenting with prostatitis-like symptoms (chronic pelvic and perineal pain) was reported by Tsai et al. (5). The interval between the occurrence of the disease and therapy is often several years (4). Usually, scrotal nodules develop slowly over many years. However, in our patient, the onset and the spreading of nodules was rapid and lasted three months. In most cases, there are no associated calcinosis elsewhere (1). Cecchi et al. reported an unusual association of calcinosis circumscripta cutis of the face with scrotal calcinosis in a 54-year-old man which had been present since youth (6).

Whether SC is idiopathic or may be the result of calcification of preexisting cysts remains a controversial issue.

Swinhart and Golitz (7) observed epidermal cysts in three cases of SC; some were calcified with partial or total disintegration of the epithelial walls, associated with an inflammatory reaction. The conclusion of the authors was that SC results from calcification of epidermal cysts in the following sequence: inflammation of epidermal cysts, dystrophic calcification of their content and subsequent rupture of the cyst wall, and a granulomatous proliferation. The cyst wall is destroyed and only calcium deposits remain in the dermis.

Song et al. (2) studied 51 scrotal nodules in the same patient and identified 37 cysts with intact cyst walls. The calcification of the keratin content was observed in about half of the cases. Among the 37 cysts, the authors

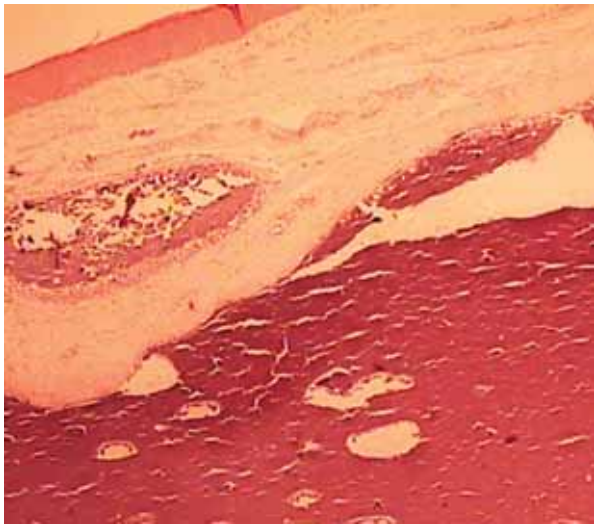

Figure 2. Calcified nodule surrounded by a fibrous capsule (HE 250x).

identified 3 epidermal cysts, a calcified pilar cyst and a calcified hybrid cyst. The majority of the remaining cysts were labeled as indeterminate cysts because they had flattened walls that made the classification difficult. Half of the indeterminate cysts were calcified. Furthermore, the authors also observed, within the dermis, an accumulation of a calcified keratinous material associated with an inflammatory reaction and simple granular depositions of calcium.

On the base of these histopathological findings, Song et al. established the following pathogenic pattern for formation of SC: cysts (epidermal, pilar or hybrid) are formed, implying calcification of the intracystic keratinous content with enlargement of the cyst and a subsequent attenuation of the wall. The structure of the cysts becomes indeterminate, along with their wall ruptures and the exposure of their content. This triggers a mononuclear cell inflammation or foreign body granuloma with resorption of the cyst walls and of the keratinous material. Finally only calcified deposits remain. Among the 51 studied nodules, only one showed both inflammation and the remnant cyst wall. This means that after the cyst ruptures, the resorption of its wall is rapid and is prone to inflammation. The keratin content might be more resistant to inflammation than the attenuated cell wall. Calcified keratinous material surrounded by inflammatory cells was present even when the cyst wall could not be observed any more. Song et al. consider that histopathological findings vary with the age of the cysts, the oldest lesions no longer containing epithelial cells.

The role played by epidermal cysts in the pathogenesis of SC is not accepted by all authors. Epidermal cysts are usually not prone to calcification. If epidermal cysts develop in the scrotal skin according to Song et al. (2), they tend to be multiple and to calcify easily, since various conditions are known to cause calcification: inflammation, varicosities, tumours and trauma (8).

King et al. stress the possible role of dystrophic calcification of the dartoic muscle in the genesis of SC (9). Some authors suggested that degeneration and necrosis of the dartoic muscle are the initial events in the pathogenesis of the disease, followed by coalescence of the necrotic bundles, thus creating an acidophilic amorphous necrotic mass that eventually triggers a dystrophic calcification (3). A number of other theories link SC to the calcification of lymphangiomas, xanthomas, fibromas, teratomas or gonadoblastomas (4, 10). Calcification of the scrotum may occur in infants, secondary to meconium peritonitis, with leakage of meconium through the processus vaginalis during foetal life (4, 10).

Ito et al (11) reported a case of SC originating from eccrine epithelial cysts. This eccrine origin was discovered via an immunohistochemical study using antibodies against carcinoembryonic antigen (CEA), epithelial membrane antigen (EMA) and gross cystic disease fluid protein-15 (GCDPF-15). There was a positive reaction surrounding the lamina and in the contents of both cyst and ductal structures. The pathogenic mechanism seemed to be due to the excessive discharge and accumulation of matrical debris and sulphated mucopolysaccharides in the lumina.

In certain reports, as well as in our case, no evidence of cystic structure was found around calcified material, despite minute studies.

Wright and al (12) who examined 63 lesions in nine patients, failed to find any epithelial material around the calcified nodules even after staining with the antikeratin monoclonal antibodies LP34 and PKK1. Füzesi et al (13), who did not find any cellular structure in the calcified nodules, concluded that SC is a degenerative phenomenon resulting from alterations in the chemical microenvironment leading to the deposit of calcium and phosphates.

This suggests that SC may be truly idiopathic. Our observations support the view that SC is not a secondary phenomenon. First is the the rapid onset of the calcified scrotal nodules within a three-month period, without pre-existing lesions. To our knowledge, such a rapid evolution of SC has not been reported. Secondly, no remnant epithelial structures were identified in histopathological or immunohistochemical studies.

The condition is benign. Treatment is only recommended for aesthetic reasons. Surgical excision must be limited to the scrotal skin since calcified nodules are localized within the dermis (14). The risk of recurrence is controversial. Some authors think that surgery is a solution while others insist on the high probability of recurrence of SC (14).

In our patient, the result of the surgical treatment was satisfactory while the period of follow-up has been too short to confirm the adequacy of the treatment.

REFERENCES

1. Özgenel GY, Kahveci R, Filiz G, Özcan M. Idiopathic scrotal calcinosis. Ann Plast Surg 2002; 48(4): 453–4.

2. Song DH, Lee KH, Kang WH. Idiopathic calcinosis of the scrotum: Histopathologic observations of fifty-one nodules. J Am Acad Dermatol 1988; 19: 1095–101.

3. Pabuççuöðlü U, Canda MS, Güray M, Kefi A, Canda E. The possible role of dartoic muscle degeneration in the pathogenesis of idiopathic scrotal calcinosis. Br J Dermatol 2003; 148: 827–9.

4. Saad AG, Zaatari GS. Scrotal calcinosis: is it idiopathic? Urology 2001; 57:365.

5. Tsai YS, Tzai TS, Lin JS, Tong YC. Scrotal calcinosis presenting with prostatitis-like symptoms. Urology 2002; 59: 138.

6. Cecchi R, Giomi A, Rapicano V, Innocenti S. Idiopathic scrotal calcinosis associated with idiopathic calcinosis circumscripta cutis. Les Nouvelles Dermatologiques 1998; 17:11–3.

7. Swinehart JM, Golitz LE. Scrotal calcinosis: dystrophic calcifications of epidermoid cysts. Arch Dermatol 1982; 118: 985–8.

8. Martin B. Les calcifications en échographie scrotale. Ann Radiol (Paris) 1985 ;28: 637–44.

9. King DT, Brosman S, Hirose FM, et al. Idiopathic calcinosis of the scrotum. Urology 1979; 14: 92–4.

10. Fritz H. Verkalkte Fibrome der Haut Des Hodensackes. Roentgenpraxis 1941; 13: 417–8.

11. Ito A, Sakamoto F, Ito M. Dystrophic scrotal calcinosis originating from benign eccrine epithelial cysts. Br J Dermatol 2001; 144: 146.

12. Wright S, Navsaria H, Leigh M. Idiopathic scrotal calcinosis is idiopathic. J Am Acad Dermatol 1991; 24: 727–30.

13. Füzesi L, Hollweg G, Lagrange W, et al. Idiopathic calcinosis of the scrotum: scanning electron microscopic study with X-ray micro-analysis. Ultrastruct Pathol 1991; 15: 167–73.

14. Ruiz-Genao DP, Rios-Buceta L, Herrero L, Fraga J, Aragüés M, García-Díez A. Massive scrotal Calcinosis. Dermatol Surg 2002; 28: 745.

A U T H O R S ' A D D R E S S E S

Jihene Hicheri MD, dermatologist, Department of Dermatology, Charles Nicolle Hospital, 1006, Tunis, Tunisia, e-mail: hicherijihene@yahoo.fr
Talel Badri MD, dermatologist, same address
Becima Fazaa MD, Professor of Dermatology, same address
Rachida Zermani MD, Professor of Histopathology, Department of Histopathology, Charles Nicolle Hospital, 1006, Tunis, Tunisia
Nadia Kourda MD, Assistant Professor of Histopathology, same address
Sarra Ben Jilani MD, Chief of the Department of Histopathology, same address
Mohamed Ridha Kamoun MD, Chief of the Department of Dermatology, Charles Nicolle Hospital, 1006, Tunis, Tunisia