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Idiopathic Scrotal Calcinosis Masquerading as Epidermal Inclusion Cyst

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Abstract: ***Idiopathic scrotal calcinosis is an uncommon benign disorder involving the scrotal skin characterized by multiple calcified intradermal nodules. It was first described by Lewinskey in 1883. Deposition of calcium in the skin, subcutaneous tissue, muscles, and visceral organs is known as calcinosis. Commonly it involves the skin and it is called calcinosis cutis. Pathogenesis is still under debate as to whether the calcification is dystrophic or idiopathic. In this article we report a 45 year oldasymptomatic male with multiple, painless, hard nodular scrotal skin nodules. There was no history of trauma or any systemic illness. A provisional diagnosis of sebaceous cyst of scrotum was made. His haematological and biochemical parameters were normal The patient was treated by primary excision at our hospital and diagnosed as a case of idiopathic scrotal calcinosis cutis on histopathology.***

Keywords: Calcinosis, Epidermal Cyst, Idiopathic, Scrotum, Calcium

1. Introduction

Idiopathic scrotal calcinosis was first described by Lewinsky in 1883 [1-3], as a subtype of calcinosis cutis [4]. It is an uncommon benign disorder of the scrotal skin characterized by multiple calcified intradermal nodules that occur in the presence of normal calcium and phosphate metabolism. The nodules may vary in size and number. The deposition of calcium in the skin, subcutaneous tissue, muscles, and visceral organs is known as calcinosis, and when it involves the skin, it is called as calcinosis cutis. As per the etiology, there are four types of calcinosis cutis; dystrophic, metastatic, iatrogenic, and idiopathic. Commonly seen in the age group of 20- 40 years [5-7].Various theories on pathogenesis have been proposed by authors favoring idiopathic and dystrophic calcification. The exact pathogenesis of the condition is still controversial [8]. In dystrophic calcification, calcification occurs as a consequence of pre-existing condition such as an epidermal cyst, etc. and when there is no evidence of pre-existing pathology it is termed as idiopathic scrotal calcinosis. Metastatic calcifications are usually generalized and due to metabolic changes such as hypercalcemia and hyperphosphatemia as in end-stage renal diseases and hyperparathyroidism and dermatomysositis [9]. Histopathologically, scrotal calcinosis is characterized by the presence of calcium deposits within the dermis. Our aim is to report this rare disease of the scrotum commonly misdiagnosed as epidermal inclusion cyst and highlight the importance of follow up of such patients for timely detection of recurrence.

2. Case Report

A 45-year-old patient with no known co-morbidities presented in the outpatient department with multiple painless swellings in the scrotal region of two years duration. The swelling gradually increased in size over the years. He denied history of scrotal trauma, history suggestive of any metabolic, systemic, neoplastic or autoimmune disease. No history of sexually transmitted diseases. On physical examination, the patient was healthy and systemic examination was within normal limits. Local examination revealed multiple yellowish to white nodular lesions involving the scrotal skin. The nodules were of varying sizes, hard, non-tender, with no ulceration and discharge. Penis was not involved. Clinical diagnosis of multiple sebaceous cysts of scrotum was made. All hematological and biochemical parameters including calcium and phosphorus were within normal limits. Diabetes and retroviral screening were negative. Excision of the nodules from the scrotal skin was done.

Histopathologic examination:

On gross examination, multiple excised nodules were received. The nodules had a chalky-white cut surface and were gritty on cutting. The nodules varied in size and colour, largest measuring 2 x 2 cm. The nodules were fully submitted for further processing.The routine hematoxylin and eosin (H & E) stained sections examined from the tissue showed epidermis and dermis with lobules of amorphous calcium deposits in thein dermis and subcutaneous tissue. The borders of the lobules were fibrotic along with foreign body reaction (Figure 1).

Based on the above findings, a final diagnosis of idiopathic calcinosis cutis of scrotum was made. At the time of this writing the patient had been discharged from the hospital and is on a close follow upwith no recurrence.

3. Discussion

Idiopathic calcinosis of scrotum was first described by H.M. Lewinsky in 1883 [8] and then in 1888 by Hutchinson [10]. However, Shapiro *et al*. must be given credit for establishing idiopathic scrotal calcinosis as a distinct entity in 1970 [11]. It is a rare and benign condition, the exact incidence of

which is not known. Deposition of calcium in the skin, subcutaneous tissue, muscles, and visceral organs is known as calcinosis, and it more commonly involves skin and it is called calcinosis cutis. Calcinosis cutis is classified into four major types according to etiology: Dystrophic, Metastatic, Iatrogenic and idiopathic.

Scrotal calcinosis is usually asymptomatic but occasionally causes heaviness, itching, ulceration, and chalky white exudative discharge [2,5-7,9]. This benign scrotal lesion, though commonly occurs between third and fourth decades of life, can affect both adult and paediatric age groups with age range between 9 to 85 years reported in the literature [12]. Initially, it resembles the color of scrotal skin later it changes into yellow, and duration is about 10 years ranging from 3 months to 46 years [9].

Many authors proposed that dystrophic calcification of preexisting lesion like epidermal cyst [13], eccrine duct milia, degenerated dartos muscle as the underlying aetiopathogenesis of this disease. Dubey et al. suggest that inflammation of epidermal cyst leads to calcification of the cyst wall; with subsequent degeneration of cyst wall living only the calcific deposits in older lesions [13]. Dare and Axelsen using immunohistochemistry and CEA antibodies demonstrated the involvement of eccrine duct milia in scrotal calcinosis [14]. He proposed the term hydra calcinosis of the scrotal skin. In the dystrophic form, the serum calcium and phosphorus levels are normal. It may also be observed in connective tissue diseases like scleroderma, dermatomyositis, SLE and secondary to trauma and in ammation [3,13].

The main reason patient seek intervention is because of cosmetic concern. Patient with intense pruritus or ulceration will require surgical intervention. If swelling is <4 mm, pinch and punch excision is advised. Surgery is the treatment of choice.If it is massive, subtotal excision of the scrotal wall is preferred. If it is extensively involved, excision followed by complex scrotal reconstruction using meshed split thickness skin graft as the scrotal skin is rugged.Recurrence is very low mainly due to microscopic foci of calcification left over [9]. The diagnosis of scrotal calcinosis is established by histopathological examination.

Microscopic picture shows amorphous basophilic calcium deposits within dermis surrounded by lymphocytic infiltration, histiocytes, and hyalinization [7,9]. Histological picture shows muscle, epithelial cells, and foreign body granuloma during early stage and it shows only calcification in the advanced stage [2].

Song *et al*. described spectrum of changes takes place in scrotal calcinosis as mild to moderate inflammation of epidermal cyst is followed by mononuclear cell infiltration and foreign body granuloma formation and lastly resorption of cyst wall and keratin remnants leaving calcium deposits only [9] which is supported by Swinhart *et al*., Akosa *et al*., Saad *et al*., Dubey *et al*., Parlakgumus *et al*., and Dini and Colatraneschi *et al*. [2,7].

Differential diagnosis are teratoma, gonadoblastomas, leydig cell tumors, calcified onchocercoma, neurofibroma, ancient schwannomas, steatomas, lipomas, fibromas, and scrotal calcinosis may also be due to chronic epididymitis, calcified appendix testis, appendix epididymis, and sperm granuloma due to sperm extravasation and hematoma [9].

Even though scrotal calcinosis is a benign condition, it is important to let the patient know about the possibility of recurrence [15]. Recurrence may be due to left over microscopic foci of calcification.

In our case, there was no history of connective tissue diseases, trauma and pre-existing scrotal cysts. The biochemical parameters were also normal. Hence, the diagnosis of Idiopathic calcinosis cutis of scrotum was rendered. The excision of the nodules was done and the patient has been under follow up with no recurrence in a period of 3 months.

4. Conclusion

To conclude, the authors report a case of idiopathic scrotal calcinosis cutis in an asymptomatic individual. Idiopathic scrotal calcinosis cutis is a rare benign lesion and commonly misdiagnosed as epidermal inclusion cyst. Metabolic and hormonal work-up is required to rule out other causes. Irrespective of the etiology, surgical excision is required both for confirming the diagnosis as well as for treatment. Scrotal calcinosis must be included in the differential diagnosis of cutaneous swellings in the scrotal region. Even though scrotal calcinosis is a benign condition, precise diagnosis is crucial for a prompt and appropriate surgical procedure. At the same time the patient should be educated regarding the possibility of recurrence and therefore a regular follow up must be ensured.

5. Conflicts of interest

The authors declare no conflicts of interest.

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Figure 1

Legends;
Fig.1;(A-D),H&E stained photomicrographs of the paraffin sections of the scrotal nodules. A, 40X view of H&E stained biopsy tissue showing epidermis and dermis along with basophilic calcific deposits;B,100X view of the same tissue showing lobules of basophilic calcific deposits;C, 400X view of calcific deposits; D, 400X sections showingforeign body giant cells surrounding the calcific deposits.