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ULTRASOUND IMAGING AND PATHOLOGIC FEATURES OF IDIOPATHIC SCROTAL CALCINOSIS IN A YOUNG ADULT NIGERIAN AND REVIEW OF THE LITERATURE

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ABSTRACT

Background: **Idiopathic scrotal calcinosis is a rare and benign disease of the scrotal skin that presents as solitary or multiple painless calcified nodules or papules in the absence of systemic disorders of calcium or phosphorus metabolism. Although some theories have been proposed as to the cause of this rare disease, the exact cause remains unknown. In a resource-poor medical setting like Nigeria, a confident diagnosis of this condition can be made with ultrasonography.**

Objective: **The objective of this report is to emphasize the role of ultrasound in the imaging diagnosis of idiopathic scrotal calcinosis.**

Case Presentation: **This is a case report of a 38-year-old man who presented with recently discharging but longstanding multiple painless scrotal nodules of 22-years duration.**

Conclusion: **This case illustrates the prompt and accurate diagnosis of idiopathic scrotal calcinosis using an ultrasound, a readily available imaging modality in a low-resource setting.**

Although histology remains the gold-standard for diagnosing idiopathic scrotal calcinosis following surgical excision, this benign disorder has unique sonographic characteristics that could aid the radiologist in making a confident diagnosis.

Keywords: Idiopathic scrotal calcinosis, Calcification, Ultrasound, Computed tomography, Magnetic resonance imaging.

INTRODUCTION

Idiopathic scrotal calcinosis (ISC), sometimes called dystrophic scrotal calcinosis, is a rare benign disease of the scrotal skin that presents with solitary or multiple, painless calcified nodules or papules of varying sizes on the scrotal wall. [1,2] Although, few cases have been reported in infants and the elderly,[1,3,4] this condition usually presents in early adulthood in the absence of systemic disorder of the calcium or phosphorus metabolism.

Historically, this condition was first reported by Lewinski in 1883[2] and about a century afterwards Shapiro named it as 'idiopathic scrotal calcinosis'.[1] Several theories have been proposed as to the cause of ISC; Shapiro *et al* [5] suggested that it could be idiopathic while Karabulut *et al*[6] proposed dystrophic calcification following calcium deposition on epidermal cysts. However, the exact cause and nature of ISC have remained elusive.[2,4] Although histology remains the gold standard of diagnostic confirmation[7] after surgical excision,[1] the diagnosis of ISC can be confidently made radiologically.

This is a case of ISC seen in our institution. The summary of sonographic findings is presented, and we reviewed the literature.

CASE REPORT

A 38-year-old man presented in August 2021 with 22-year history of multiple slow-growing nodules on the scrotum. The nodules had progressively increased in size and number with occasional spontaneous thick chalky white discharge from the nodules. There was no history of trauma or previous infection of the scrotum.

On physical examination, the scrotum had multiple, non-tender, hypopigmented, firm skin nodules of different sizes (Fig 1). There is no nodule seen in other parts of the body.

Patient was referred for a scrotal ultrasound which revealed multiple, circumscribed, oval to round hypoechoic masses of varying sizes in the dermis of the scrotal skin bilaterally (Fig 2). Multiple foci of high-

Figure 1: Photograph showing multiple skin-coloured scrotal nodules of varying sizes.

level echoes showing posterior acoustic shadowing were also seen within these masses. The masses showed no colour flow on Doppler interrogation (Fig 3) and the largest of the masses measured 12.6mm in its widest dimension with approximate volume of 0.7ml. The two testicles are however normal in size, outline, parenchyma pattern, position, and echogenicity. There was no inguinal lymphadenopathy seen. A radiologic diagnosis of idiopathic scrotal calcinosis was suggested.

Laboratory evaluation revealed normal serum phosphorus and calcium. The patient consented for surgical excision biopsy of the scrotal nodules which was performed under local anaesthesia.

Histology of the excised specimen showed nodular aggregates of deeply basophilic calcified materials in the dermis with moderate infiltration of the papillary dermis by lymphocytes and plasma cells (Fig 4). The

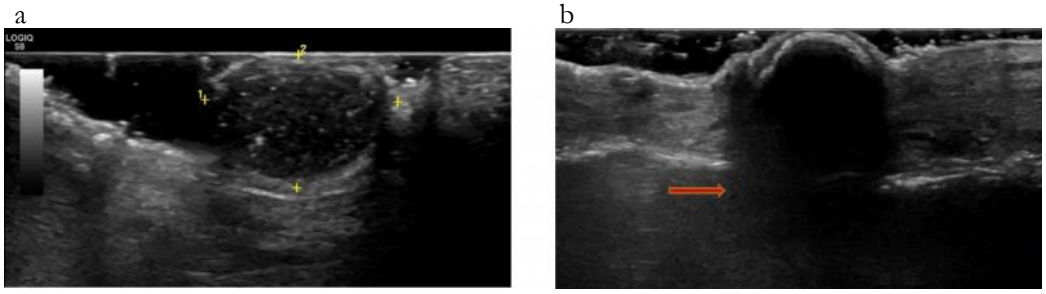

Figure 2: B-mode ultrasound images of the scrotum. (a) Circumscribed hypoechoic nodule (calipers) within the dermis of the scrotum with echogenic foci seen within it (arrow heads). (b) Posterior shadowing from one of the nodules (arrow)

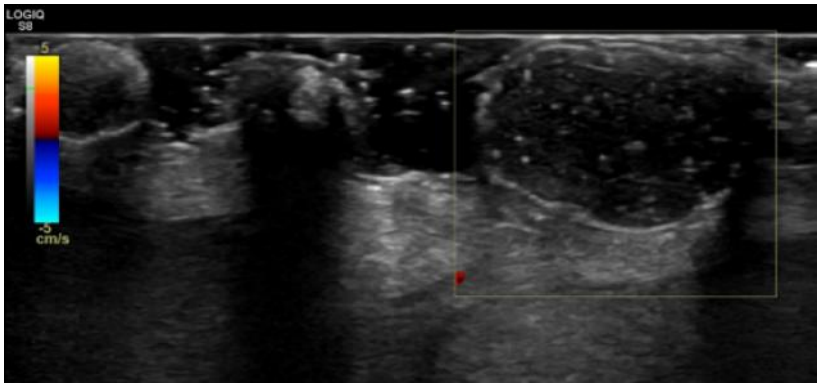

Figure 3: Duplex ultrasound image of the scrotum showing the nodule as non-vascular (arrow).

overlying epidermis showed hyperkeratosis and focal areas of acanthosis. Histopathologic diagnosis of idiopathic scrotal calcinosis was made.

The usual presentation of ISC was seen in our index case, (firm papules and nodules that appeared in childhood and gradually increased in number and size). Also, there was a history of intermittent spontaneous white chalky discharge from the nodules as previously described.[1]

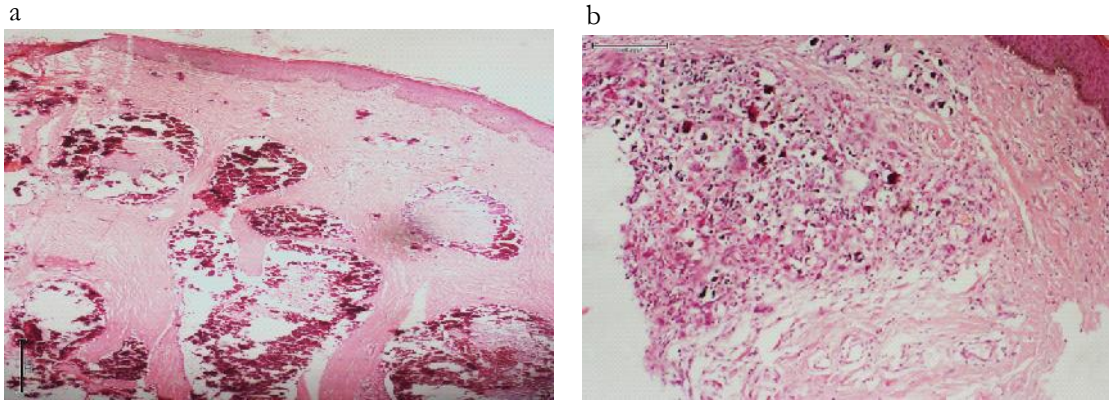

Figure 4: Photomicrographs showing various nodules of calcified amorphous materials within the dermis (a) and areas of dystrophic calcification with moderate chronic inflammation of the dermis by lymphocytes and plasma cells (b)

DISCUSSION

ISC is a rare benign disease of the scrotal skin that usually presents as painless, firm papules and nodules that appear in childhood and adolescence and gradually increase in number and size. Although occurrence in infants and elderly have been reported [1,4,8] however, most patients present between the ages of 20 and 40.[2] ISC is common in blacks suggesting an ethnic susceptibility however; no predisposing factors have been documented.[9] Although a female counterpart of ISC, "idiopathic vulvar calcinosis", has been reported,[10] ISC is seen in males. Idiopathic scrotal calcinosis initially takes the colour of the skin, but as they grow larger, they become yellowish and lobulated breaking down spontaneously or when compressed they produce a chalky white material.[11] The nodules are usually asymptomatic but could be itchy or painful.[1]

This rare condition was first described by Lewinski in 1883 but the name "idiopathic scrotal calcinosis" was given by Shapiro in 1970 after exclusion of the presence of epithelial lining, residual cysts, lipid, or organism on histology.[1,5,11] Several theories have been proposed as to the cause of ISC however, the exact etiology remains obscure as controversy exists as to whether the disease is idiopathic or the result of dystrophic calcification of preexisting structures including epidermal cyst, eccrine epithelial cyst, and degenerated dartos muscle.[12] Imaging plays a significant role in making a diagnosis and for follow-up, but histopathology remains the gold standard of diagnostic confirmation after surgical excision.

Literature review indicated that a plain pelvic radiograph would show solitary or multiple homogeneously dense, superficial calcifications of varying size projected over the scrotal shadow and may distort the contour of the scrotum. Extra-testicular calcifications like scrotal pearl, foreign body, calcified scrotal onchocercoma etc., can give a similar radiographic appearance. Ultrasound remains a safe, readily available, and sensitive imaging modality which in trained hands, can confidently confirm the diagnosis of ISC. The scrotal nodules appear as smooth oval circumscribed mostly hyperechoic avascular nodules within the dermis of the scrotum (Figs 2). The advantage of ultrasound over plain x-ray is that the testicles can also be examined concomitantly to rule out testicular lesions.

Computed tomography and magnetic resonance are advanced imaging modalities that can also be employed to further characterize the scrotal lesions if diagnostic doubt still exits. Computed tomography is the gold standard imaging technique for demonstrating calcifications, and it shows ISC as solitary or multiple homogeneously hyperdense structures within the scrotal wall on non-enhanced images. Magnetic resonance shows the lesions as T1 and T2 hypointense nodules within the scrotal dermis with no enhancement post gadolinium administration.

Cosmetic concern remains the main reason why affected patients seek medical consultation[4] and till date, surgery is the only recommended mode of treatment for ISC even though recurrence is common.[1,2] Smaller lesions are amenable to the novel 'pinch-punch' excision which involves pinching the scrotal skin with the fingers and punching the scrotal lesions with a disposable biopsy punch to remove them.[12] Larger lesions will usually require wide excision and direct skin closure.[2,4] However, in cases of massive calcinosis or when most of the scrotum is involved, a subtotal excision or total scrotal skin removal and grafting may be necessary.[2]

CONCLUSION

ISC is a rare, benign, and nodular disorder of unknown aetiology affecting the scrotum. Although histopathology remains the gold-standard of diagnosis following surgical excision, ISC has specific sonographic and radiographic characteristics, the knowledge of which will help the radiologist to make confident diagnosis.

ETHICAL STATEMENT

Written informed consent for the case to be published (including images, case history and data) was obtained from the patient.

CONFLICT OF INTEREST

The authors have no conflicts of interest to declare.

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