

## Gigantic idiopathic scrotal calcinosis: A rare case report and review

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### Abstract

Idiopathic scrotal calcinosis is a rare skin condition characterized by multiple asymptomatic calcified nodules of unknown aetio-pathogenesis. It is a benign condition that may be complicated with ulcerations or secondary infections occasionally. Diagnosis depends on histology and treatment is needed for mostly cosmetic reasons. We report a 40-year-old male with extensive idiopathic scrotal calcinosis and review the literature.

**Keywords:** Idiopathic scrotal calcinosis, Calcinosis cutis.

### Report

A forty-year-old male patient, a farmer by occupation, presented with multiple asymptomatic nodular scrotal growths of five years duration. They were gradually progressing in size and number. He did not give any history of diabetes mellitus, immunological diseases, sexually transmitted diseases, neoplastic diseases or intake of medications. There was no past history of acute or chronic scrotal trauma, infection or surgery.

Examination revealed multiple firm cutaneous nodules on the scrotum measuring three to thirty millimeters in size. Most of them were coalescing with each other and specks of chalky white matter were visible on the surface. They were non-tender on palpation. There was an associated left sided direct inguinal hernia. Underlying testis appeared normal with no associated signs of inflammation. Based on the clinical presentation, a provisional diagnosis of steatocystoma multiplex with calcification was made.

Routine biochemical tests including serum calcium and phosphorous were normal. He did not have any similar lesions elsewhere on his body. Excisional biopsy was done and histopathology revealed calcified nodules with amorphous homogeneous material staining positive for calcium, surrounded by thick fibrous walls and intact epidermis. Based on the clinical picture and histopathology, diagnosis of idiopathic scrotal calcinosis was made. (Fig. 1 & 2)



**Fig. 1: Gigantic idiopathic scrotal calcinosis in a 40-year-old male.**



**Fig. 2: The scrotal calcinosis was surgically excised.**

### Discussion and Review

Calcium and phosphate deposition in tissues is called calcification or calcinosis. The three main types of calcinosis are:

- 1) Dystrophic calcification that occurs in tissues damaged by an underlying pathology eg: dermatomyositis, pseudo xanthoma elasticum, Ehlers Danlos syndrome.<sup>1</sup>
- 2) Idiopathic calcification not associated with any tissue damage or metabolic derangement eg: calcinosis universalis, calcinosis circumscripta or tumoral calcinosis.<sup>1</sup>
- 3) Metastatic calcinosis secondary to abnormal calcium and phosphorus metabolism as in hypercalcemia or hyperphosphatemia. In this type visceral organs are more affected than the skin.<sup>1</sup>

Idiopathic scrotal calcinosis is a rare cutaneous disease first reported by Lewinski in 1883.<sup>2</sup> Shapiro et al later described this condition as a separate entity.<sup>3</sup> It is commonly seen in 2<sup>nd</sup> and 3<sup>rd</sup> decade of life.<sup>4</sup> This condition is more common in the dark coloured races and does not run in families.<sup>5</sup>

Exact aetio – pathogenesis of idiopathic scrotal calcinosis is not known but various theories have been proposed. Song et al suggested that it was actually not idiopathic but dystrophic calcification of preexisting epidermal cysts. The calcification of epidermal cysts initiates an inflammatory response that results in resorption of the cyst walls and hence the remnants of epithelial lining of the walls are not evident on histopathology.<sup>6</sup>

Dare and Axelsen showed involvement of eccrine ducts using immunohistochemistry.<sup>7</sup> Carson proposed infection by nanobacteria through repeated microtrauma. Few consider it as dystrophic calcifications of degenerated dartos muscle.<sup>9</sup> Since none of the above theories could be well substantiated, it is believed to be of idiopathic origin.

Clinically they present as firm, non-tender, yellowish nodules that occur isolated or in groups on the scrotal skin. They vary in size from pinhead to several centimetres and in number from solitary to more than 100. The lesions slowly progress in size and patient often presents several years after onset.<sup>10</sup> Rare cases present like pedunculated tumors - "polypoidal scrotal calcinosis" that result in gross deformity of the scrotum.<sup>11</sup>

Most cases are asymptomatic and patient presents only for cosmetic concerns.<sup>13</sup> Occasionally patients complain of heaviness and pruritus. Complications include ulceration, extrusion of chalky yellow material and secondary infection.<sup>13</sup> Similar idiopathic calcinosis are described in females on vulva and around areola. Rare cases of idiopathic penile calcinosis have been described.<sup>14</sup>

Calcified onchocercoma, solitary neurofibroma, schwannoma, steatoma, lipoma and fibromas need to be differentiated. Angiokeratoma and lymphangioma circumscriptum need to be excluded.<sup>15</sup>

X-ray of scrotum is usually not advised but if done shows numerous radiopaque shadows corresponding to the calcifications.<sup>16</sup> Biopsy with histologic examination is necessary for definitive diagnosis. The lesions are hard with calcific deposit and thin rim of fibrous tissue on cut section. Under microscopy, the epidermis is intact and amorphous basophilic deposits are noted in the dermis surrounded by a foreign body type of granulomatous inflammation.<sup>16</sup>

The calcific deposits stain blue on Grunwald-Giemsa stain, dark blue to black on Von Kossa staining and red colour with Alizarin Red S.<sup>15</sup>

Shivkumar et al described fine needle aspiration as a diagnostic modality in calcinosis cutis wherein the aspirate reveals amorphous chalky material that stains positive for calcium.<sup>18</sup> Transmission electron microscopy shows crystal and mineral deposits on degenerated collagen fibrils. Chemical analysis shows deposits of calcium, carbonates and phosphates. Infrared spectroscopy in experimental studies has shown equal proportion of calcium phosphate and magnesium ammonium phosphate.<sup>19</sup>

Serum and urine biochemical examination for calcium, phosphate, uric acid, alkaline phosphatase, parathyroid hormone and calcitonin and total vitamin D3 are normal.<sup>17</sup>

Patient seeks attention mainly for cosmetic concerns.<sup>13</sup> Surgical excision is the treatment of choice. In extensive lesions skin grafting may be required. Punch excision can be used as therapeutic modality in

smaller lesions.<sup>13</sup> Recurrences are usually not seen since it is a benign condition. However, few cases of recurrences have been reported when small foci are left behind during surgery.<sup>13</sup>

## Conclusion

Idiopathic scrotal calcinosis is a skin condition characterized by asymptomatic, gradually progressive nodules of varying sizes on scrotal skin in young men. It is not associated with any derangement of calcium metabolism. The condition is benign and definitive diagnosis hinges on histopathology. Surgical excision in the treatment of choice and recurrences are rare.

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