



Case Report

Tumoral Calcinosis (TC) of iliac Crest in a 70 year-Old Female: A Case Report

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Abstract

A seventy-year-old lady had a hard tumor mass in left sacroiliac region. She was operated. Biopsy was taken from the left iliac crest and histopathological examination was done.

Tumor measured 3×2×2 cms approximately. It was chalky-white ball-like. Its surface was lobulated. Microscopically, it showed small and large areas of necrosis and calcification. Areas of mild-to-moderate lymphocytic infiltration were seen. Fibrosis and hyalinization were seen. Bony trabeculae and osteoid formation were also seen. The patient was finally diagnosed as a case of primary tumoral calcinosis of sacroiliac region.

Keywords: *Soft tissue calcification necrosis osteoid formation chronic inflammation.*

Introduction

Tumoral calcinosis (TC) is characterized by periarticular deposition of calcium salts in hip, shoulder and elbow. Deposition of calcium salts may occur gradually in soft tissues^[1]. Tumor mass may gradually increase in size for several years. Tumor mass may be hard and the patient may

have intermittent drainage of white chalk-like material. Aspirated fluid may be rich in calcium, phosphates and proteins. Further, the disease may be autosomal dominant^[2]. Hyperphosphatemia and trauma may be important etiological factors^[3]. Rarely, TC might develop fistula in the skin. Moreover, neural and vascular compression may

occur^[3]. TC is produced by pathogenic genes encoding for FGF 23, GALNT3 and KLOTHO^[4]. Herewith, we report a case of primary tumoral calcinosis of sacroiliac region.

Case Report

A seventy-year-old lady had a painless hard swelling in soft tissues involving left sacroiliac joint. Swelling measured 3×2×2 cms approximately (figure 1).



Figure 1: X-ray of pelvis showing the lesion of tumoral calcinosis. Chalky white lobulated appearance of the lesion is seen (red arrow). Size of tumor mass was 3×2×2 cms.

Biopsy was taken from left iliac crest. It's surface was lobulated. It consisted of irregular deposition of calcified amorphous phosphate material. Small and large-sized areas of calcification and osteoid formation were seen. Focal mild and moderate

perivascular lymphocytic infiltration was also seen (figure 2). Lesion was finally diagnosed as primary tumoral calcinosis (Active phase).

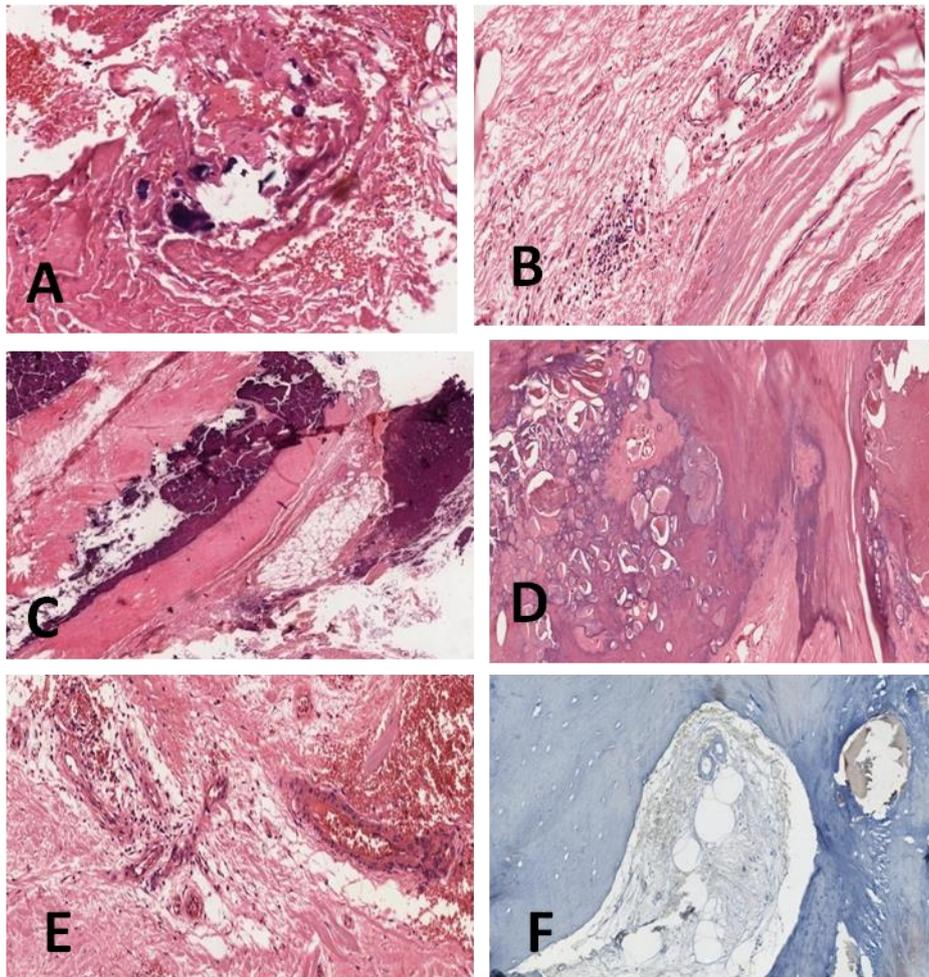


Figure 2: (A) Photomicrograph shows a small area of coagulative necrosis superadded with calcification. (B) Photomicrograph shows perivascular lymphocytic infiltration. (C) Photomicrograph shows a large area of coagulative necrosis and calcification. (D) Photomicrograph shows small cystic areas. Bony trabeculae formation is seen. (E) Shows hemorrhage in soft tissue of tumoral calcinosis. (F) IHC using anti-CD 46 antibody failed to stain the tumoral calcinosis cells.

Discussion

Primary tumoral calcinosis (TC) is a rare crippling metabolic genetic disorder which may manifest as a hard tumor-like structure. Sex ratio is nearly equal. Secondary TC may develop in a patient with hyperparathyroidism^[4]. Differentials may be tophaceous gout, myositis ossificans and calcific myonecrosis. Most significant feature of current report was perivascular lymphocytic infiltrations suggesting chronic inflammation. In addition, fibrosis was also seen suggesting repair of necrosed tissue by fibrous tissue. Large areas of necrosis and calcification were also seen in the

current patient. Moreover, osteoid tissue with woven bone formation was also seen. Exact etiopathogenesis of tumoral calcinosis is not known. Current patient was aged 70 years. Late onset of disease may favour gene-environmental interactions and effect of ageing to manifest the disease. Another report of two cases of TC in Sudanese siblings have been reported which further suggested the role of genetic factors in pathogenesis of tumoral calcinosis. Another case of TC has been reported from Yemen, suggesting presence of pathogenic gene in middle eastern countries^[4]. Another case of primary TC has been

reported from Morocco^[5]. Nodular lesions of TC have been described in larynx^[6]. Rarely, TC may develop around wrist in extensor indicis proprius tendon^[7]. The disease was described earlier by Giard and Duret^[8,9]. Later, the term 'Tumoral calcinosis' was coined by Inclan et al^[10]. Primary calcinosis may be of two types, a hyperphosphatemic type and a normophosphatemic subtype. Secondary tumoral calcinosis may be associated with chronic kidney failure^[11]. The disease primarily develops in pediatric age-group. Conversely, our patient was an adult, aged 70 years. Further, firm tumor-like structures may develop around the joints which may lead to restriction of their movement. Cystic spaces may form with different fluid-levels, resulting in a positive sedimentation sign^[12]. Surgical excision of large tumor masses may be done. However, recurrence and ulceration may develop later.

Conclusion (S)

Periarticular inflammation of soft tissue associated with calcification in pediatric age-group may be diagnosed as tumoral calcinosis. On the contrary, our patient was an adult. Restriction of excess of phosphates containing diet and aluminum hydroxide may prevent recurrence of tumor after primary excision.

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