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TUMORAL CALCINOSIS: CASE REPORT

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SUMMARY

Tumoral calcinosis (TC) is a rare disease of obscure aetiology. In its classic form, it is characterised by solitary or multiple large foci of mineralisation in the soft tissue adjacent to the bone around large joints in the absence of disorders of calcium metabolism and visceral calcification. A case is presented of TC in a 75-year old Kenyan woman.

INTRODUCTION

Soft tissue calcifications are found in different diseases such as milk alkali syndrome, hypervitaminosis D, primary and secondary hyperparathyroidism and tumoral calcinosis (TC). Among these conditions, TC remains a poorly understood disease in which either solitary or multiple benign calcifications are usually found near large joints, without any involvement of the synovium itself or the adjacent bone in patients with normal serum calcium levels. Although the aetiology of TC is unknown, some authors have associated this disease with metabolic disorders or with trauma(1). TC seems to be more common in the tropics. A case of TC of the right hip in a 75-year-old Kenyan female who had been followed up for 24 months is presented.

Figure 1

Anteroposterior radiograph of the pelvis showing a multilobular calcific mass in the upper region of the right hip. No joint or bone involvement is seen



CASE REPORT

A healthy 75-year old Kenyan woman presented in October 1994 with a firm, but painless mass in the right upper lateral aspect of the hip region. She could not remember the exact onset of her ailment, yet the mass had been there for years and it had progressively increased in size. There was no family history of any similar disease. Whereas it was not possible to recall any cause of the swelling (particularly trauma) the patient affirmed that she tended to lie on her right side while sleeping.

Physical examination revealed a tender, oval, firm, subcutaneous tumour measuring about 5 cm in diameter. The skin over the mass exhibited an orange-peel appearance. Her right hip had a normal range of motion. The anteroposterior radiograph of the right hip revealed a lobulated, calcified soft-tissue mass (Figure 1). A blood analysis, including serum calcium and phosphorus level demonstrated no abnormal findings.

Figure 2

Macroscopic appearance of the completely excised swelling: the lesion consists of an encapsulated, firm, lobulated soft tissue mass (6 x 5 x 1 cm) consisting of fibrous septa surrounding areas containing chalky material



Figure 3

A photomicrograph of the biopsy-specimen showing areas of calcification surrounded by fibrofatty connective tissue. (Haematoxylin-Eosin. Original magnification x 100)



Complete extracapsular excision of the lobulated mass measuring 6x5x1 cm was performed. The macroscopic appearance of the excised mass showed subcutaneous fibrosis of the connective tissue with multiloculated calcification containing a chalky, white friable material (Figure 2). Recovery was uneventful and the histological examination confirmed the diagnosis of TC (Figure 3). At a twenty four-month follow up examination, no signs of recurrence were observed.

DISCUSSION

The term TC defines a condition in which either single or multiple tumour-like calcified masses are present without any associated calcium metabolism disorder. The calcifications normally adhere to the surrounding tissue and frequently appear in the vicinity of joints. TC is a rare disease which has been recognised as a clinical entity since 1899(2). Only 100 acceptable clinical cases were reported in a complete review of the literature by Enzinger and Weiss in 1995(3). More recently Pakasa and Kalengayi described the pathological features in 111 patients registered in Zaire over 30 years(4). The disease seems to affect mainly black adult patients from tropical and sub-tropical regions of Africa(5) and at times is related to a family history(6).

The pathogenesis of TC remains unclear and several theories have been proposed. Although hyperphosphatemia has been described in some patients(7,8). TC in the presence of a normal circulating phosphate concentration is the rule rather than the exception(1). Even though local trauma has been implicated as a possible cause of TC in some patients(9), the suggestion of a post-traumatic origin has been debated on the ground that patients cannot in all cases recall a traumatic incident. No abnormalities of serum phosphate or calcium homeostasis were evident in our patient and she denied a history of trauma at the involved site. The hypothesis that TC is caused by the chronic microtraumatisation of

sleeping on hard surfaces(9), could be supported by the fact that she usually slept on her right side.

The clinical presentation and radiographic features in our patient were quite typical(10) although the diagnosis could only be confirmed by a histological study. Even though some authors have shown that, in addition to radiography, other diagnostic modalities such as computed tomography and magnetic resonance imaging can also be helpful in making a correct preoperative diagnosis of TC(11,12), these examinations could not be performed in our case due to lack of sophisticated facilities at our small rural hospital.

Different forms of treatment have been proposed for TC. Medical treatment such as phosphorus deprivation, calcitonin and diphosphonates, have been tried with only partial success. Patients with TC associated with hyperparathyroidism have also been treated with parathyroidectomy(9). A complete surgical resection of the calcium deposits thus remains the treatment of choice in patients where TC is not associated with any metabolic disorder, yet recurrence is frequent if removal is incomplete(5). Our patient, who demonstrated no metabolic abnormalities, was therefore treated with a simple, but radical, extracapsular resection of the calcified lesion and no recurrence was evident at follow-up.

In conclusion, TC is a benign disease characterised by the presence of a slow growing lump with possible functional limitations secondary to the size and location of the tumour. Therefore, surgical treatment is indicated and the excision has to be complete in order to avoid local recurrences.

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