Large Tumoral Calcinosis in The Gluteal Region:  
A Case Report


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SUMMARY

Background. Tumoral calcinosis is a poorly understood phenomenon. It can be described as a syndrome of calcium deposits principally affecting the juxta-articular areas. It is a rare entity that has been poorly understood. Our aim is to highlight a special and unusual case of an 11-year-old with a large, relatively painless lump in her buttock.

Clinical case. An 11-year-old girl of African descent presented to our Bone Tumour Unit after being referred by her local hospital. The girl presented with a large lump on the posterolateral aspect of the right buttock, measuring 15cm in diameter. Due to the delay in referral/diagnosis, tethering of the skin had progressed to necrosis, with a sinus discharging milky-white fluid. A MRI scan further characterised the lump as a densely calcified area within the gluteus maximus, extending to the subcutaneous tissue. The characteristic features of the calcified mass on the images led to the diagnosis of tumoral calcinosis. Laboratory test did not demonstrate any metabolic disturbances. Pathology reports further confirmed the diagnosis and the lump was successfully resected. There were no recurrences on follow-up.

Conclusion. Several cases of tumoral calcinosis have been described in the literature; however, it remains a rare entity. Being aware of the possibility and having knowledge of tumoral calcinosis is paramount in preventing confusion and delay in diagnosis for patients and clinicians.

Key words: tumoral, calcinosis, gluteal, hip
BACKGROUND

Tumoral calcinosis can be described as a syndrome of calcium deposits mainly affecting the juxta-articular areas, of which only approximately 250 cases have been reported so far worldwide. The term ‘tumoral calcinosis’ was first used by Inclan et al, who developed a standard by which to diagnose the disorder, although it had been previously described [1]. Duret described it back in 1889 and Teutschlaender did the same in 1930, attaching his name to this condition, naming it “Teutschlaender’s Disease” [2,3].

Tumoral calcinosis tends to occur more often in people of African descent and is commonest in the second decade of life, but can affect any age [4,5,6]. It is commonly seen as a complication of renal dialysis, but occasionally the idiopathic form occurs. The aim of this article is to highlight and share an interesting case.

CASE HISTORY

An 11-year-old girl of African descent, presented to our Bone Tumour Unit after being referred by her local hospital. The girl presented with a large lump on the posterolateral aspect of the right buttock. Her mother had noticed this a few weeks previously. The patient had previously been complaining of some pain around her right thigh, but this gradually subsided. The patient was otherwise fit and well; past medical history and family history were unremarkable.

On examination, the lesion was approximately 15cm in diameter and was hard and smooth within the right gluteal region. Tethering of the skin was originally noted by the referring hospital; however, by the time that the patient was examined at our hospital, the tethering had progressed to skin necrosis, with a sinus discharging milky-white fluid. There was a full range of movement in the hip and no pain on movement. Note was made of a benign-feeling cervical lymphadenopathy, although admittedly the patient was also recovering from a recent cold.

Laboratory tests, performed to exclude inflammatory processes and metabolic disturbances, yielded the following results:

- CRP <2 (<5 mg/L)
- ESR 11 (<20 mm/hour)
- Hb 13.5
- WCC 5.6 (4-10 x109/L)
- Platelets 303 (150-350 x109/L)
- AF 125 (13-120 U/L)
- Ca 2.30 (2.25-2.75 mmol/L)
- Mg 1.10 (0.74-1.48 mmol/L)
- P 1.35 (0.65-1.30 mmol/L)

The patient also tested negative for sickle cell disease and thalassaemia. A large heterogeneous calcified para-articular mass was demonstrated in the right hip on plain radiographs (Fig. 1).

Subsequently, a CT scan of the pelvis was performed to determine the extent of the development of the mass. The scan confirmed a large mass of calcification at the lateral aspect of the right hip (Fig. 2). The site of origin was difficult to determine due to its size. It appeared to be sited partially within the gluteus maximus, displacing muscles at the anterior aspect of the hip and separated from the greater trochanter by only a small strip of soft tissue. It extend-
ed into subcutaneous fat and appeared to be ulcerating the skin at the level of the lesser trochanter. Its medial aspect was closely related to the sciatic nerve. A MRI scan further characterised this as a densely calcified area, with dimensions of 11.2 x 7.3 x 6.5 cm within the right gluteus maximus muscle, extending to the subcutaneous tissue. The characteristic features of the calcified mass on the images led to the diagnosis of tumoral calcinosis, a rare benign condition. Pathology reports further confirmed the diagnosis, describing ‘islands of amorphous calcified material surrounded by bands of soft tissue showing histiocytic and multinucleated giant cell reaction’.

The lesion was well localised and could be resected completely, thereby reducing the risk of recurrence. The tumour was successfully and completely excised (Fig. 3,4). No complications occurred post-operatively.

**DISCUSSION**

Tumoral calcinosis is a rare disorder characterised by calcified masses mainly in the juxta-articular areas. The condition usually presents as a painless lump, but can be complicated with compression of peripheral nerves and erosion of the skin and drainage of chalky, milk-like material through sinus tracts. The most commonly affected locations are large joints, such as the hip, shoulder and elbow [7]; however, cases have been described where tumoral calcinosis has been found on rarer locations, such as hands, scalp and feet [8].

Tumoral calcinosis can mimic several conditions and should therefore be differentiated from calcinosis of chronic renal failure, calcium circumscripita, soft tissue chondroma, calcinosis universalis, pseudo-gout, milk-alkali syndrome, hypervitaminosis D, calcareous tendinitis. The diagnosis of tumoral calci-
nosis can be made by careful, focused history-taking and imaging; if there is any doubt, a biopsy will confirm the diagnosis.

Tumoral calcinosis has a typical appearance on radiographs, being an amorphous, cystic and multilobulated calcification located in a periarticular distribution. In addition to this, absence of erosion or osseous destruction by adjacent soft-tissue masses is a distinguishing finding in tumoral calcinosis [9].

Primary tumoral calcinosis can be divided into a familial form and an idiopathic form. Recent advances give us a clearer picture in the cause of the familial form. These recent studies show involvement of several genes. In the hyperphosphatemic familial tumoral calcinosis (FTC), mutations in GALNT3 and FGF23 have been observed [4,10-15], whereas in the normophosphatemic FTC, the SAMD9 gene has been suggested to be involved [16]. In this patient, there was no family history or trauma to suggest or imply an aetiological factor.

The underlying cause of the idiopathic tumoral calcinosis also has still to be proven. Slavin et al. suggested three theories of the pathogenesis of tumoral calcinosis, these are [17]:
1. repetitive trauma leading to reparative dysfunction
2. periarticular forces dissecting histiocytic aggregates that initiate osteoclastic activity, and
3. haemorrhage from microtrauma causing an exaggerated reparative response.

Over the past decades, a consensus has not yet been reached regarding the classification of tumoral calcinosis. Smack et al proposed a pathogenesis-based classification into three subtypes [18]:

1. Primary normophosphataemic tumoral calcinosis: patients have normal serum phosphate, normal serum calcium, and no evidence of disorders previously associated with soft tissue calcification;
2. Primary hyperphosphataemic tumoral calcinosis: patients have elevated serum phosphate, normal serum calcium, and no evidence of disorders previously associated with soft tissue calcification;
3. Secondary tumoral calcinosis: patients have a concurrent disease capable of causing soft tissue calcification.

However, Olsen et al. have claimed that this classification is not practical for the clinician. They propose only to use the historical definition of tumoral calcinosis and to use it strictly in reference to a disease caused by hereditary metabolic dysfunction of phosphate regulation associated with massive periarticular calcinosis [9].

The gold standard treatment is considered to be surgical excision, but several studies claim that an additional medical treatment to this would be helpful [19-22]. In our case, tumoral calcinosis was successfully treated by surgical excision alone. Indications for surgery include: pain, recurrent infection, ulceration and functional impairment. Disadvantages of surgical excision include post-operative complications and recurrences, the latter being common.

Several cases of tumoral calcinosis have been described in the literature, however, it remains a rare entity. Being aware of the possibility and having knowledge of tumoral calcinosis is paramount in preventing confusion in diagnosing the condition for patients and clinicians.

REFERENCES


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