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A Rare Calsinosis Soft Tissue Tumor on Hip Region

I Wayan Juli Sumadi[1], Ni LuhGde Sagita Dewi[1], Vika Indriani[1], Elysanti Dwi Martadiani[2], IGede Eka Wiratnaya[3]

[1]Pathology Anatomy Department, Medical Faculty, Udayana University, Denpasar, Bali, Indonesia

[2]Radiology Department, Medical Faculty, Udayana University, Denpasar, Bali, Indonesia

[3]Orthopedic Surgery Department, Medical Faculty, Udayana University, Denpasar, Bali, Indonesia

Abstract: ***A 14 years old male came with soft tissue tumor on right hip region. The confusing diagnosis finally confirmed by histopathology examination. The diagnosis was confirmed byhistopathological examination following surgical excision. Histopathology examination revealedislands of amorphous psammomatous calcified material surrounded by bands of fibrous tissue showing histiocytic and multinucleated giant cell reaction. The prognosis is good due to no metabolic disorders accompanying in this patient.***

Keywords: tumoralcalcinosis, soft tissue mass, hip region, histopathology

1. Introduction

Tumoralcalcinosis (TC) is a rare clinical and histopathologic syndrome characterized by calcium salt deposition in different periarticular soft tissue regions.It often lead to diagnostic confusion.The pathogenesis and genetic background gradually have been unraveled since its first description in 1943. Most common region of TC are soft tissues of peri-articular upper limb (shoulder and elbow) and hip regions. Since its first description by Inclanet al, the term tumoralcalcinosis has been widely used in the literatures and sometimes broadened to other conditions with similar clinico-pathologic features, or even imprecisely used to describe any massive collection of peri-articular calcifications. We report one case of tumoralcalcinosis on hip regions. [1]

2. Case

A 14 years old malecame with lump on the right hip since 6 months prior to admission. It started small and getting bigger after receiving physiotherapy treatment. He also complained intermittent pain on the lump,there was no difficulty to do his dailyactivities. There was ni history of trauma and chronic cough but he lost his body weight due to loose of appetite.No previous medical problem nor family historyof the same medical condition.

Figure 1: Lump on the right hip, size 10x10 cm, weel defined, fixed, and firm consistency.

On physical examination, there was a lump on the right lateral pelvic, with a shiny skin, no visible venectasis or deformity. Size 10x10 cm, defined border, smooth surface, hard consistency, attached to deeper structure but not attached to the skin. On the palpation was warm, dorsalis pedis artery can be palpable. There is also disruption of pelvic joint movement.

Laboratory results show normal blood count, normal serum liver and serum creatinine tests. Calcium serum calcium levels within normal limits, phosphate levels increased (8.4 mg / dl).

Radiological examination revealed subtle multilobular calcifications, no apparent erosions or fractures on the bone. MRI examination showed well defined soft tissue mass, dimension 14.44 cmx 7.02 cm x 6.26 cm, on T1 phase seen predominantly hipointense mass in gluteus maximus muscle, there is also the focus of the hypointens component. In phase T2 the mass represents a cystic dominant lesion with fluid-fluid levels with persistent hypointens component lesions (calcification). In the fat set some lesions appear hyperintense, indicating the presence of fluid components. While on contrast showed heterogenous contrast enhancement with capsule enhancement, resulting in a suspicious impression of an intra muscular abscess of gluteus maximus with the calcification component with tumoral calcinosis as a differential diagnoses.

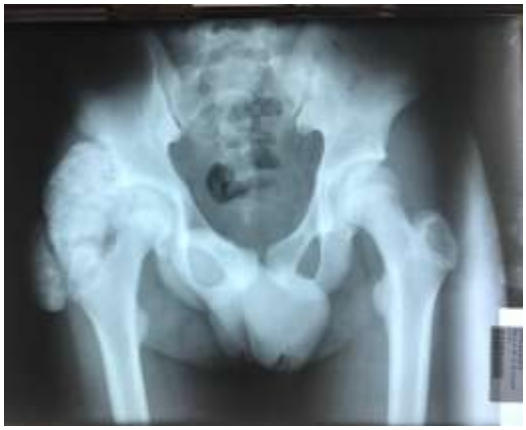


Figure 2: Pelvic X-Ray AP view.Subtle multilobular calcifications, no apparent erosions or fractures on the bone.

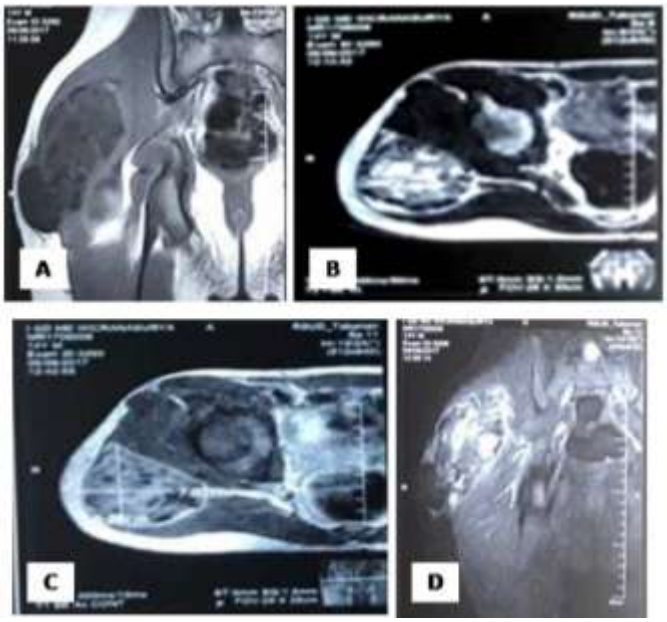


Figure 3: A. T1 phase, predominantly hipointense mass in gluteus maximus B. In T2 phase the mass represents a cystic dominant lesion with fluid-fluid levels with persistent hypointens component lesions (calcification). C. The fat set show some hyperintense lesions, indicating the presence of fluid components. D. On contrast showed heterogenous contrast enhancement with capsule enhancement.

Orthopedic department conducted debridement and excisional biopsy, at the time of the debridement there was a milky paste material came out from the tumor. The tissue sent to anatomic pathology laboratory for histopathological examination.

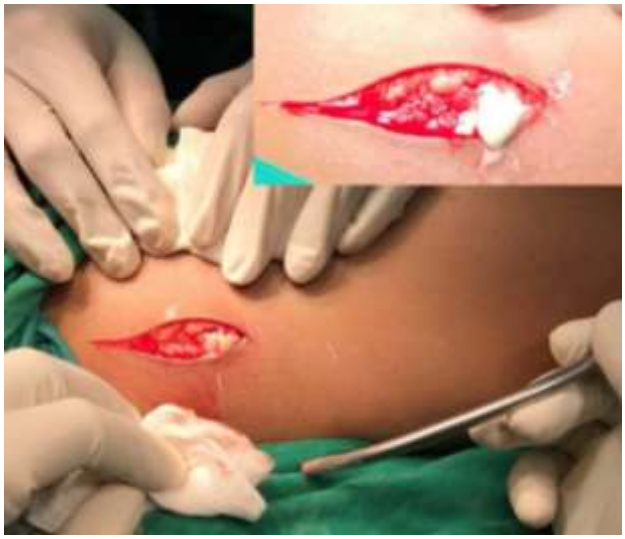


Figure 4: Excision and debridement of tumor. On sectioning, there was a milky paste material.

On macroscopic examination the tissue fragments was whittish gray, irregular shaped, chewy consistency. The largest size was 2.5x0,8x0,7 cm, the smallest was 0.7x0,5x0,3 cm. Histopathologic examination revealed wide area containing eosinofiilic amorphous material partially constrained fibrous connective tissue with lymphocytes, plasma cell, histiocyte and multi nucleated giant cells. There were foci of calcification both in amorphous materials and among fibrous connective tissues. Some calcification was spherical and concentric resembling psammoma bodies (calcospherites). Pathology reports confirmed the diagnosis as tumoralcalcinosis.

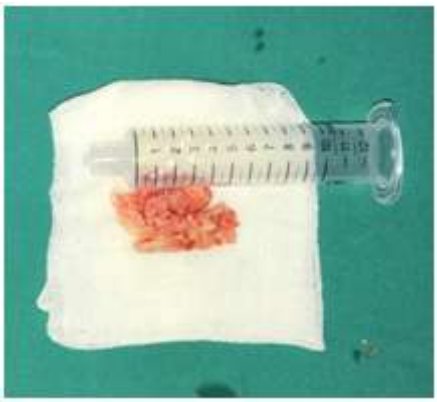
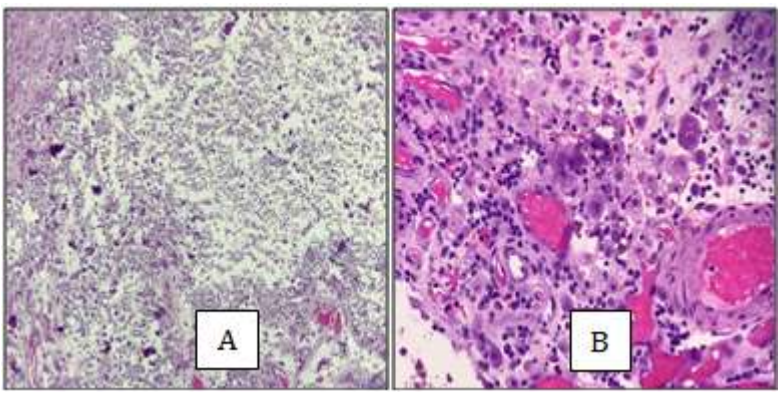


Figure 5: Biopsy specimen : pieces of tumor excision, whittish gray, irregular shaped, chewy consistency.



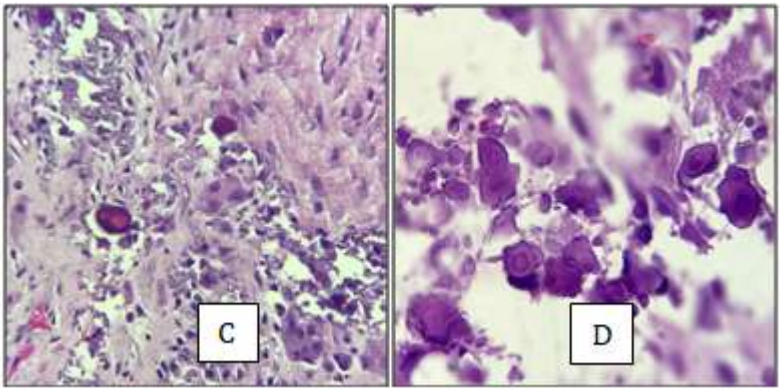


Figure 6: Histopathology examination. A. Wide area containing eosinofiilic amorphous material B. Fibrous connective tissue with lymphocytes, plasma cell, histiocyte and multi nucleated giant cells. C. Foci of calcification both in amorphous materials and among fibrous connective tissues. D. Spheric and concentric calcification, resembling psammoma bodies

3. Discussion

Tumoralcalcinosis is a rare condition described in the literature as deposition of calcium salts in soft tissues. In most cases, TC occurs in young patients as in this patients. TC is often confusing with osteosarcoma, chondrosarcoma, and myositis ossificans. TC can be described as a calcium deposit syndrome that occurs mainly in the juxta-articular region, so far only about 250 cases have been reported worldwide. The term 'tumoralcalcinosis' was first used by Inclan et al., who developed a standard for diagnosing the disorder although previously described by Duret in 1889 and Teutschlaender did the same in 1930, calling it "Teutschlaender Disease".[1,2,3]

Tumoral calcinosiscan mimic several conditions and therefore should be differentiated from calcinosis of chronic renal failure, calcium circumscripta, soft tissue chondroma, calcinosisuniversalis, pseudogout, milkalkali syndrome, hypervitaminosis D, calcareous tendinitis. The diagnosis of TC can be made by carefulhistory taking and good imaging; if there is any doubt, a biopsy will confirm the diagnosis. Tumoralcalcinosis has a typical appearance on radiographs, showing amorphous, cystic and multilobulated calcification located in a periarticularregion. In addition to this, absence of erosion or osseous destruction by adjacent soft-tissue masses is a distinguishing finding in tumoralcalcinosis.[4,5,6]

Etiology of TC remains uncertain despite several theories that have been proposed.The underlying cause of the idiopathic tumoralcalcinosis also has still to be proven. Slavin*et al*. suggested three theories of the pathogenesis of tumoralcalcinosis, as repetitive trauma leading to reparative dysfunction, periarticular forces dissecting histiocytic aggregates that initiate osteoclastic activity, and haemorrhage from microtrauma causing an exaggerated reparative response.[3]

Smack *et al* proposed a pathogenesis based on classification into three subtypes. The first subtype is primary normophosphataemictumoralcalcinosis in which patients have normal serum phosphate, normal serum calcium, and no evidence of disorders previously associated with soft tissue calcification. the second subtyoe is primaryhyperphosphataemictumoralcalcinosis, in this TC subtype patients have elevated serum phosphate, normal serum calcium, and no evidence of disorders previously associated with soft tissue calcification. The last subtype is secondary tumoralcalcinosis, patientshaveaconcurrent disease capable of causing soft tissue calcification.[4]

Inprimary hyperphosphatemictypemulti factorial calcification is initiated by elevated calcium phosphorus product with hyperphosphatemia as the overwhelming component. This hyperphosphatemia can be explained by genetic mutations in *FGF23*, *GALNT3* or *KLOTHO* gene resulting in inactivation of the phosphaturic protein *FGF23* . The secondary hyper-phosphatemictypeis associate with secondary hyperparathyroidism resulting from chronic renal failure. On the other hand, in the primary normophosphatemictype, transient hyperphosphatemia is the proposed mechanism. This transient hyperphosphatemia is either produced locally due to tissue injury leading to release of phosphate from injured cells into extracellular space specially when injury involves muscles (main phosphate store in soft tissue), or induced by excessive oral or rectal use of a phosphate-saline laxatives.In this case phosphate serum was elevate with no other medical condition causing secondary calcification, indicating this case was the primary hyperphosphataemictumoralcalcinosis.

The gold standard treatmentfor TC is considered to be surgical excision, but several studies claim that an additional medical treatment would be helpful. In this case, tumoralcalcinosis was successfully treated by debridement and surgicalexcision. Indications for surgery include: pain, recurrent infection, ulceration and functional impairment.[7-10]Surgical excision of tumoralcalcinosis is the primary treatment described in the literature for masses that cause discomfort, function limitation, or cosmetics. However, complications and recurrence frequency limited to patients with significant disability or deformity. In patients without metabolic abnormalities, reported recurrence rates range from 0% to 33%. Some authors reported that the recurrence rate is lower after complete excision than after incomplete excision. In cases with metabolic abnormalities, the lesions tend to recur, especially after incomplete excision. TC can also lead to postoperative complications, such as prolonged drainage. Prolonged drainage can cause delayed wound healing and even sinus tract formation.[10-13]

4. Conclusion

TC is a rare clinical and histopathologiccondition characterized by calcium salt deposition in different periarticular soft tissue regions. It may lead a diagnostic confusion. The good histopathologic examination confirmed the diagnosis. Excision and debridement is the gold standar treatment.

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