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Case Report

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Multifocal Tumoral Calcinosis: Address of a Huge Hip Tumor with Vastus Lateralis Muscle Reconstruction: A Case Report and Literature Review

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Abstract

Background: Tumoral calcinosis is a rare diagnosis characterized by periarticular calcific deposits, mainly affecting the periarticular tissue of the hips, shoulders and elbows. It's usually progressive, and can lead to painful lesions. Incomplete surgical resection is usually associated with a high recurrence rate.

Case report: Here, we report a case of a 39-year-old man with multifocal tumoral calcinosis, affecting both shoulders and his left hip. He presented with a variant of the GALNT3 gene of unknown significance. Due to intense pain in his hip, the tumoral lesion was resected along with the part of the vastus lateralis involved by the tumor. The muscle was reconstructed by filling the gap with a polypropylene mesh, normally used for hernia repair. One year after surgery, the patient referred no restrictions or inabilities in his daily life. The imaging study showed no recurrence of the tumor.

Conclusion: Here we describe a possible new association between a specific mutation in the GALNT3 gene and tumoral calcinosis. We believe we're also the first to report the usage of polypropylene mesh as a substitute for a muscular gap. Considering the good clinical outcome, this method may constitute a good solution for future similar cases.

Keywords

Tumoral calcinosis, Muscle reconstruction, Calcium deposits, Polypropylene mesh

Background

Calcifications of the musculoskeletal soft tissue are frequently found in routine radiographs [1]. There are multiple differential diagnoses to consider. Tumoral calcinosis, a rare diagnosis characterized by periarticular calcific deposits, is among them.

Tumoral calcinosis, firstly named by Inclan, et al. [2] in 1943, mainly affects the periarticular tissue of the hips, shoulders and elbows, and the joint is normally spared [2-5]. These tumors are often asymptomatic and grow slowly [3], and sometimes they can be multifocal [3]. It usually appears among adolescents and young adults [2,4,5] with African ascendance [1,5].

Olsen, et al. [6] state the term should be used to describe only a rare familial disease of autosomal recessive transmission, which can be either hyperphosphatemic or normophosphatemic [3,4,7]. It's usually progressive, and can lead to painful ulcerative lesions and skin and bone infections [3,8]. Several genes (such as fibroblast growth factor-23 (FGF23), an FGF23-glycosylating enzyme (GALNT3), and the FGF23 co-receptor α-Klotho (αKL)) have already been implied and their role described [7]. Other authors include in the term the sporadic cases (where calcium and phosphate are usually normal; in spite of the fact that, in some patients, the serum phosphate may be elevated [1,9]) and the complications of renal dialysis with deregulation of the calcium and phosphate homeostasis [3,10], and so currently any calcium deposit around a joint is called "tumoral calcinosis" [3].

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In clinical practice, we can divide these calcific periarticular masses termed "tumoral calcinosis" into primary or idiopathic (with no associated diseases and normal levels of serum calcium) and secondary (when they result from disorders with high serum calcium levels, such as chronic renal failure, hyperparathyroidism, pseudoxanthoma elasticum, sarcoidosis, scleroderma, hypervitaminosis D, milk-alkali syndrome and massive osteolysis) [3]. The primary type is the most frequent and 30% of the cases are hereditary [3,11].

Evaluation starts with a good clinical history. A progressive growth of a calcified mass can be associated with pain, functional impairment, and nerve compression [3]. Common clinical manifestations are joint tenderness, limited range of motion of joints, and skin ulcerations [12]. Plain radiographs, ultrasound, computed tomography, and magnetic resonance imaging can all play a role in the diagnostic workup. Definitive diagnosis requires histological examination [1,13]. Differential diagnoses include connective tissue diseases, neoplastic diseases (such as osteosarcoma or chondrosarcoma), metaplasia (synovial osteochondromatosis), and degenerative diseases [3,4].

Treatment can be either conservative or surgical. Medical treatment includes dietary restrictions; intake of calcitonin, bisphosphonates, steroids, phenylbutazone; or radiation therapy [3,12]. If calcium and/or phosphate levels are altered, medical management includes using low-calcium dialysate, and giving non-calcium-containing phosphate binders [12]. It's usually unsuccessful and recurrence is rather frequent [3,14,15].

Surgical treatment allows the eradication of the lesion. Incomplete resection is usually associated with a high recurrence rate, and mass growth is commonly faster than the initial lesions [14]. Hyperphosphatemia and familial predisposition negatively impact the outcome of surgery [9,11]. For patients with huge masses that interfere with joint function, mobility, or neurovascular structures, medical treatment alone is only justifiable for patients with complicated comorbidities [3], for whom a surgical resection implies an important life risk.

Here, we report a case of a 39-year-old man with multifocal tumoral calcinosis, affecting both shoulders and his left hip, and we explain a unique method for muscle reconstruction with a polypropylene mesh, normally used for hernia repair.

Case Report

A 39-year-old man was referred to our hospital due to multifocal tumoral calcinosis affecting both shoulders and his left hip, with intense pain for the last 3 months resulting from his left hip mass. He was being managed by an Endocrinologist at another institution. He had no family history of tumoral calcinosis and no relevant past medical history. Calcium serum levels were normal, as were the levels of vitamin D and parathyroid hormone, but he had hyperphosphatemia. Mutations normally associated with tumoral calcinosis weren't present, but a variant of unknown clinical significance was detected in the GALNT3 gene (c.1765T>C-p. (Trp589Arg)).

Computed tomography (CT) for the chest, abdomen, and pelvis was performed and no metastases were found. The scan showed a large calcified lesion in the inner border of the left iliac bone (Figure 1). Magnetic resonance imaging (MRI) of the hip lesion was ordered (Figure 2). A biopsy was performed and it was compatible with tumoral calcinosis, with no signs of malignancy.

The hip tumoral lesion was then resected in the operating room, but considering its large volume, a huge gap remained between the proximal and distal stumps of the vastus lateralis

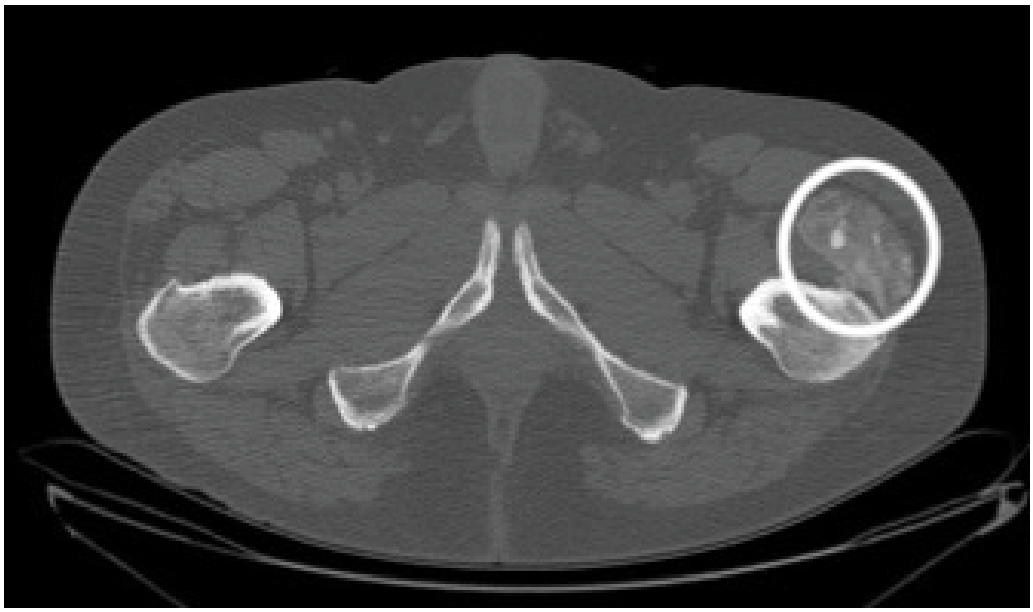

Figure 1: CT scan (axial view) showing a calcified lesion (41 mm × 80 mm × 108 mm) in the inner border of the left iliac bone extended laterally to the left lower abdominal wall with infiltration of the lateral abdominal wall. The left iliac bone was intact with no evidence of bone destruction or periosteal reaction.

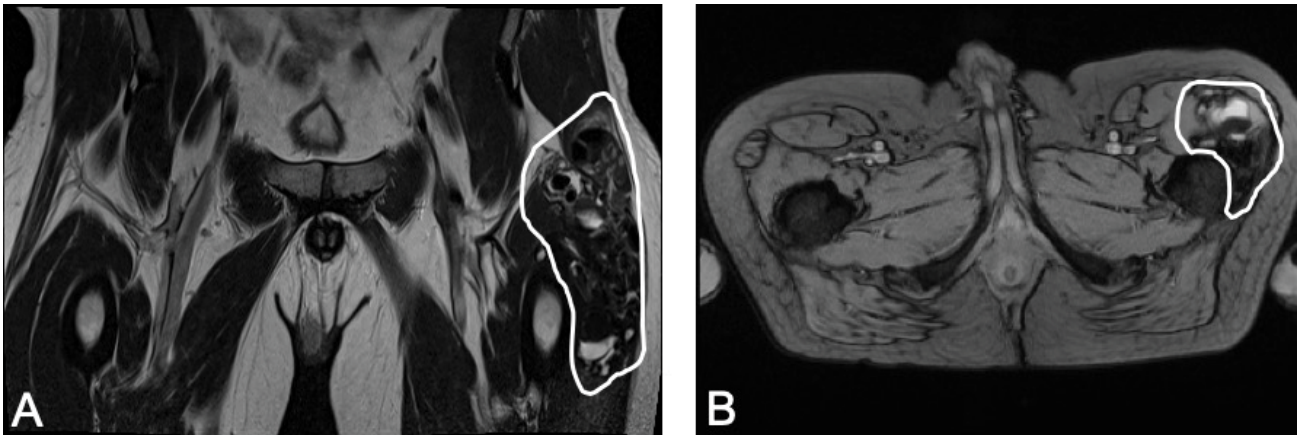

Figure 2: MRI showing a fairly defined cauliflower-like multilocular lesion, mainly a single void sclerotic lesion with thick internal septations, intimately related to an inner surface of the left iliac bone with no definite intraosseous origin. The mass is seen fungating laterally through the lateral pelvic muscles until the subcutaneous tissue and causes splaying of the lateral abdominal wall muscles and stretching of the overlying skin. The lesion displays mixed low and intermediate signal at T1 and mixed signals at T2.

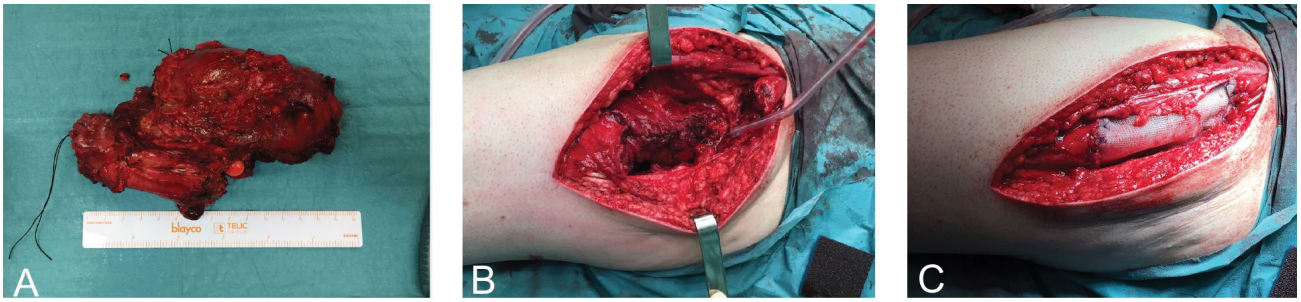

Figure 3: (A) Resection specimen; (B) Gap between the muscular stumps; (C) Reconstruction of the gap with a polypropylene mesh.

(Figure 3). The surgical team decided to reconstruct the muscle by filling the gap with a polypropylene mesh (Figure 3), normally used for hernia repair.

In the immediate post-operative period, no atrophy of the hip was noted (diameter of around 56 cm bilaterally), and the Musculoskeletal Tumor Society (MSTS) score was 14 (pain 4, function 1, emotional 5, supports 0, walking 2, gait 2). He was discharged 1 day after the surgery walking with two crutches. He initiated physical rehabilitation immediately after discharge.

Three months after surgery, the surgical wound was closed, without any signs of complication. A residual pain at the wound remained, though, when flexing the hip counter-resistance. The diameter of the hip was still similar on both sides (around 56 cm). The MSTS score improved and was then 18 (pain 3, function 4, emotional 1, supports 5, walking 3, gait 2). The patient maintained his program of physical rehabilitation, in order to regain his basal level of ability.

One year after surgery, the patient referred no limitations in his daily activity. The residual pain referred when flexing the hip counter-resistance was still present. The diameter remained similar on both sides (56 cm) with muscle strength similar bilaterally. The MSTS score was 28 (pain 4, function 5, emotional 4, supports 5, walking 5, gait 5). So, the patient was back to his normal daily life with no restrictions and with no inabilities. The MRI showed no relapse of the calcinosis, with fibrosis around the mesh.

Discussion

Tumoral calcinosis is a rare benign tumor, mainly seen in adolescents and young adults [2,4,5], that can be treated either medically or surgically. Hips, shoulders and elbows are the most affected locations [2-5]. As previously stated here, medical treatment has poor efficacy for big masses with functional impact and, when pain or functional limitation arise, the tumor should be surgically extirpated, if possible.

We present here a case of a young patient, with masses in both shoulder and left hip and with important pain and limitation related to his hip lesion. After carefully studying his various lesions and histological confirmation of the diagnosis, we opted to remove his hip tumor. In order to remove all the tumor and prevent recurrences, we had to excise part of his vastus lateralis. In an attempt to preserve the muscle function, we used a polypropylene mesh, normally used for hernia repair by general surgeons. We opted for this solution considering Nag, et al. [16] had successfully used it in the recent past to perform adelayed repair of the quadriceps muscle. There are no other reports of this usage for muscle repair. However, what we did here was a bit different and riskier.

Nag, et al. [17] used the mesh as a means of reinforcement of the underlying muscle and not as a connection between two stumps without any muscle in between, as in our case. Here, with the mesh guiding the way, we hoped fibrosis between the stumps would allow the recovery of some function. After 1 year, the patient did indeed recover his normal gait function, with no atrophy or strength deficits of the limb. Early mobilization decreases muscle atrophy and promotes healing [16] and we believe the enrolment of the patient in an early rehabilitation program played an important role in his recovery. The presence of other agonists that remained intact also helped the favorable outcome. In terms of imaging, there weren't any signs of recurrence as well.

Managing calcium and phosphate homeostasis is also an important key in the treatment cascade. The patient in this case had hyperphosphatemia being managed by the Endocrinologist, but the tumoral masses were not controlled. The role of the Endocrinologist is essential to rule out medically treatable causes in order to prevent further tumors. As mentioned, the variant c.1765T>C-p. (Trp589Arg) in the GALNT3 gene was detected. This variant affects a highly conserved residue, but its clinical significance is unknown. Some pathogenic variants of the GALNT3 gene cause familial hyperphosphatemic tumor calcinosis with autosomal recessive inheritance [7], though, so further studies on this new variation and its possible relation to tumoral calcinosis are needed. Unfortunately, with the information currently available, this should be classified as a variant of unknown significance and we cannot state for sure it is responsible for this case of tumoral calcinosis.

Conclusion

Tumoral calcinosis is a rare diagnosis. The disease is benign, but it can cause pain, functional impairment, and compression of neurovascular structures. The diagnostic workup includes a good clinical examination, imaging, and histopathological examination.

Here, we report a case of a young male successfully treated with surgical excision. Resecting the whole mass is essential for a good outcome and for preventing recurrences.

For the first time, the c.1765T>C-p.(Trp589Arg) variant in the GALNT3 gene was described in a patient with tumoral calcinosis. The real significance of this finding is unknown, but we believe it could possibly be the cause behind this pathology in our patient, since other variants of the same gene were associated with this disease in the past.

We believe we're also the first to report the usage of polypropylene mesh as a substitute in a muscular gap. The fibrosis around the mesh and between the two stumps, the early rehabilitation program, as well as the presence of other agonists allowed a good clinical outcome, with the patient returning to preoperative activity and functional levels. This method may constitute a good solution for future similar cases.

Disclosure

The authors declare no conflicts of interest.

Ethical Disclosures

The authors declare that no patient data appear in this article. The patient, however, agreed with this publication and gave informed consent.

References

1. Freire V, Moser TP, Lepage-Saucier M (2018) Radiological identification and analysis of soft tissue musculoskeletal calcifications. Insights Imaging 9: 477-492.
2. Inclan A LP, Camejo MG (1943) Tumoral calcinosis. JAMA 121: 490-495.
3. Del Bravo V, Liuzza F, Perisano C, et al. (2012) Gluteal tumoral calcinosis. Hip Int 22: 585-591.
4. Al-Rumaih MH, Al-Otaibi MA, Alshammari AN (2020) Tumoral calcinosis presentation in operated developmental dysplasia of the hip: A case report and review of literature. Cureus 12: e9948.
5. Chebbi W, Belhaj G, Belhaj Messaoud A (2021) Tumoral calcinosis: A rare disease. Tunis Med 99: 306-310.
6. Olsen KM, Chew FS (2006) Tumoral calcinosis: Pearls, polemics, and alternative possibilities. Radiographics 26: 871-885.
7. Farrow EG, Imel EA, White KE (2011) Miscellaneous non-inflammatory musculoskeletal conditions. Hyperphosphatemic familial tumoral calcinosis (FGF23, GALNT3 and alphaKlotho). Best Pract Res Clin Rheumatol 25: 735-747.
8. Topaz O, Indelman M, Chefetz I, et al. (2006) A deleterious mutation in SAMD9 causes normophosphatemic familial tumoral calcinosis. Am J Hum Genet 79: 759-764.
9. Hammoud S, McCarthy EF, Weber K (2005) Tumoral calcinosis in infants: A report of three cases and review of the literature. Clin Orthop Relat Res 2005: 261-264.
10. McGregor DH, Mowry M, Cherian R, et al. (1995) Nonfamilial tumoral calcinosis associated with chronic renal failure and secondary hyperparathyroidism: report of two cases with clinicopathological, immunohistochemical, and electron microscopic findings. Hum Pathol 26: 607-613.
11. Polykandriotis EP, Beutel FK, Horch RE, et al. (2004) A case of familial tumoral calcinosis in a neonate and review of the literature. Arch Orthop Trauma Surg 124: 563-567.
12. Yano H, Kinjo M (2021) Tumoral calcinosis. Cleve Clin J Med 88: 208-209.
13. Chaabane S, Chelli-Bouaziz M, Jelassi H, et al. (2008) Idiopathic tumoral calcinosis. Acta Orthop Belg 74: 837-845.
14. Savaci N, Avunduk MC, Tosun Z, et al. (2000) Hyperphosphatemic tumoral calcinosis. Plast Reconstr Surg 105: 162-165.
15. Bittmann S, Gunther M, Ulus H (2003) Tumoral calcinosis of the gluteal region in a child: Case report with overview of different soft-tissue calcifications. J Pediatr Surg 38: E4-E7.
16. Nag HL, Jain G, Nayak M, et al. (2021) Result of delayed repair of quadriceps muscle following a sharp cut injury. BMJ Case Rep 14: e239863.
17. Tomori Y, Nanno M, Takai S (2020) Acute calcific periarthritis of the proximal phalangeal joint on the fifth finger: A case report and literature review. Medicine (Baltimore) 99: e21477.