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Case Report

Tumoral calcinosis, a diagnostic dilemma: a case report

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ABSTRACT

Tumoral calcinosis is a rare diagnosis characterized by deposition of calcium salts in peri-articular soft tissue regions. It is divided into primary and secondary varieties. The primary tumoral calcinosis is further divided into two types; primary hyperphosphatemic type and primary normophosphatemic type. The secondary variety occurs in association with chronic renal failure. Biochemical assessment and typical radiographic features help in diagnosis. Mainstay of treatment for primary variety is surgical. Secondary variety is mainly treated by medical measures. Surgical intervention is reserved for patients who do not respond to medical therapy.

Keywords: Chronic renal failure, Hyperphosphatemia, Normophosphatemia, Tumoral calcinosis

INTRODUCTION

The term 'tumoral calcinosis' was first coined by Inclan in 1943.[1] The usual presentation is, swelling around the joints (solitary or multiple) associated with discomfort, pain and/or limitation of joint movements.[2,3] Joints most commonly affected include the hip, elbow, shoulder, foot, and wrist.[4] These lesions are slowly growing and progress over years. Lesions may be associated with ulceration of the overlying skin.[5]

Etiology of tumoral calcinosis is evolving. Till date many theories have been proposed, however all of them are inconclusive. It is divided into primary and secondary types. The primary tumoral calcinosis is further divided into two types; primary hypophosphatemic type and primary normophosphatemic type.

Primary type is characterized by normal serum calcium levels and normal or raised serum phosphate levels. Secondary type is most commonly found in association with chronic renal failure. Differential diagnosis of tumoral calcinosis include synovial sarcoma, osteosarcoma and myositis ossificans. A certain preoperative diagnosis is rarely made. A high degree of suspicion on radiological and biochemical profile leads to diagnosis.[6] Biopsy is usually indicated to rule out other causes.

Mainstay of treatment for primary variety is surgical. Secondary variety is mainly treated by medical measures. Surgical intervention is reserved for patients who do not respond to medical therapy.

CASE REPORT

Authors present a case of 55-year-old gentleman, with no comorbidities. He presented with history of progressive slowly growing soft tissue mass over left buttock area of 1-year duration. Patient was having difficulty in walking. The mass was painless with no skin involvement with normal joint movements. On clinical examination, a large approximately 30x15cm smooth, round, oval, mobile, painless mass was observed in the left gluteal region (Figure 1).

Preoperative MRI scan was done which was showing large soft tissue mass of about 30x20 cm in dimension arising from left gluteus maximus muscle with no extension or involvement of retro gluteal structures or

pelvis. No bony involvement was reported on MRI (Figure 2). Core biopsy was done was suggestive of low-grade spindle cell tumour.

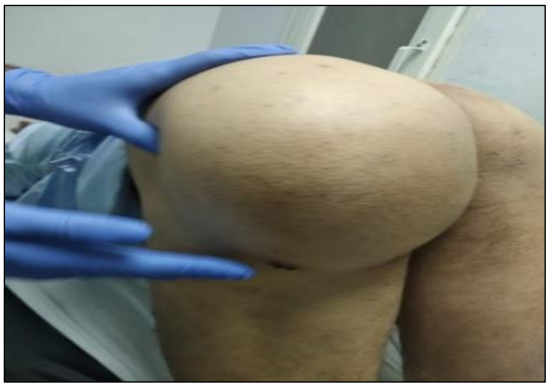

Figure 1: Mass in the left gluteal region.

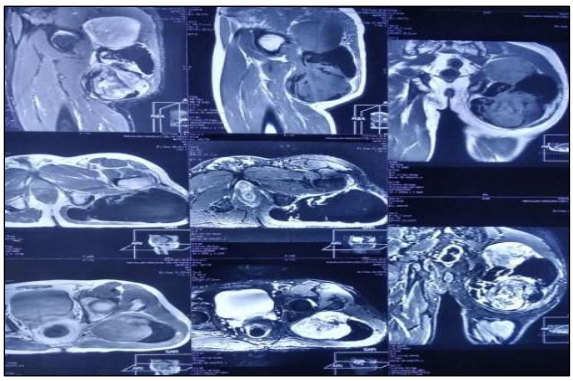

Figure 2: Mass arising from left gluteus maximus.

Patient was planned for wide local excision. A longitudinal elliptical incision was made in the left gluteal region and the mass was excised with more than 1 cm margins (Figure 3).

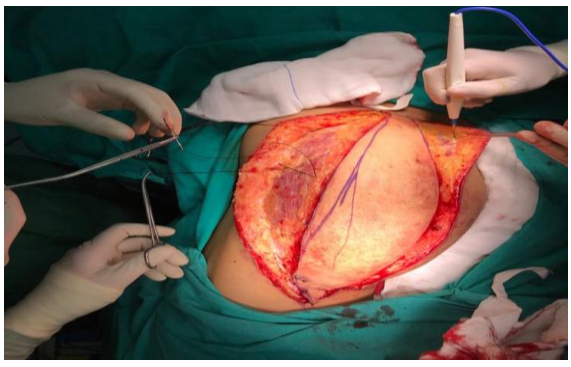

Figure 3: Elliptical incision.

The mass was excised en-mass without any spillage. The mass was abutting left sciatic nerve which was preserved with careful dissection (Figure 4). The specimen was sent for histopathological examination (Figure 5). On gross examination, a 32x17x10 cm, bosselated mass was seen on cut section, chalky white milky fluid was seen coming from multiple cystic cavities (Figure 6). On microscopy, multiple cystic cavities were seen separated by fibrocollagenous stroma cavities were filled with amorphous calcified deposits without lining epithelium. Spindle cell stroma with myxoid degeneration was noted.

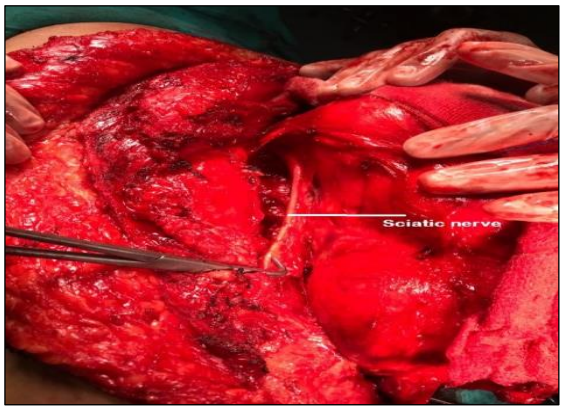

Figure 4: Sciatic nerve being saved.

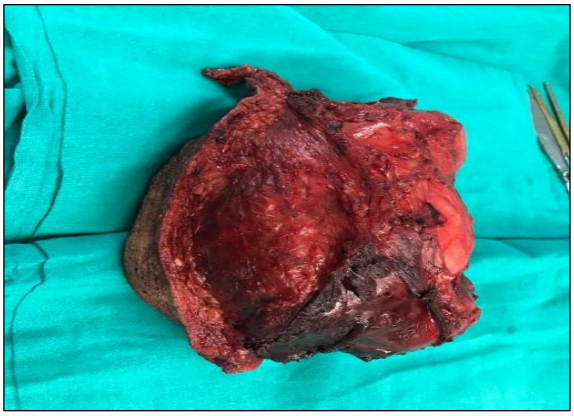

Figure 5: En bloc specimen.

Figure 6: Tumoral calcinosis.

No cellular atypia or mitosis was seen. All these features were in favour of tumoral calcinosis. Patient was discharged on day 3 and followed up after one week and is on regular follow up. Postoperatively, serum calcium, serum phosphorus and parathormone levels were done

and were within normal limits. At 3 months of follow up our patient had no recurrence.

DISCUSSION

Tumoral calcinosis is a very rare diagnosis. It is divided into primary and secondary varieties. In primary normophosphatemic type, the calcium and phosphorous levels are typically normal. This entity usually presents before the 2nd decade of life. This variety is thought to be familial, involving gene encoding for SAMD9 protein.[6,7] Primary hyperphosphatemic type is characterized by normal calcium levels and high phosphorous levels. Patients with this entity usually present in the first and second decades of life.[3,4] This variety has a genetic predisposition. Various genes that have been found to be associated with primary hyperphosphatemic variety include GalNAc transferase 3 gene, GALNT3, and KLOTHO.[4] Secondary tumoral calcinosis is most commonly associated with chronic renal failure.

The usual presentation is, swelling around the joints (solitary or multiple) associated with discomfort, pain and/or limitation of joint movements.[2,3] Joints most commonly affected include the hip, elbow, shoulder, foot, and wrist.[4] These lesions are slowly growing and progress over years. Lesions may be associated with ulceration of the overlying skin.[5]

Preoperative diagnosis of tumoral calcinosis is usually by various radiological techniques. Findings of amorphous, multiloculated and cystic calcifications in a periarticular location on plain radiographs are suggestive of tumoral calcinosis4. Computed tomography serves as a guide for surgical planning as it determines the extent and relations of the lesion. Sedimentation sign (cystic loculi with fluid-fluid levels caused by calcium layering) on computed tomography is typical of tumoral calcinosis.[8] T2-weighted MRI sequences show inhomogeneous high signal intensity with nodular pattern having alternating areas of high signal intensity and signal void. T1-weighted MRI sequences inhomogeneous lesions with low-signal intensity.[9] Ultrasonography has got a limited role and is usually of some importance in loculated fluid collections.[4,9]

Differential diagnosis of tumoral include; calcinosis universalis, calcinosis circumscripta, calcific tendonitis, synovial osteochondromatosis, synovial sarcoma, osteosarcoma, myositis ossificans, tophaceous gout, and calcific myonecrosis. This is settled by combination of biochemical and radiological features.[4] Biopsy is indicated in cases where there is a suspicion of malignancy like soft tissue sarcomas.[6]

Treatment mainly depends on type of the lesion, site, size and relations of the lesion along with symptom. Mainstay of treatment for primary tumoral calcinosis surgical excision.[10] Recurrence is managed by repeat excisions5. In case of very large lesions which cannot be excised completely, partial excision is recommended for symptomatic relief.[5] Recurrent ulceration with infection and functional impairment warrant excision. Variable success rates in both primary types have been achieved by dietary phosphate depletion.[5] Recurrence after surgical excision are high therefore, medical treatment could be considered before the surgical approach, especially in the hyperphosphatemic entity.

Secondary tumoral calcinosis is managed by medical measures. Failure of medical treatment is an indication of biopsy/surgical intervention.[11] Calcium and phosphorus depleted diets, dialysates, and phosphate binders (except aluminium containing binders) form the mainstay of treatment. Various other medications that have used with variable success rates include Vinpocetine, Sodium thiosulfate and Pamidronate. Secondary tumoral calcinosis may be associated with secondary or tertiary hyperparathyroidism, therefore in such cases if the medical management fails, subtotal or total parathyroidectomy is warranted.

CONCLUSION

Tumoral calcinosis is a rare condition with poorly explained aetiology. Accurate preoperative diagnosis may be difficult in view of similarities between various other benign and malignant conditions. Biochemical profile combined with radiological features forms the basis of preoperative diagnosis. Biopsy is not always indicated and is reserved for cases where the diagnosis is not certain. Surgery constitutes the mainstay of treatment in primary tumoral calcinosis, however in some cases medical treatment may be instituted prior to surgical treatment. Secondary tumoral calcinosis is managed by medical measures primarily and surgery is in the form of subtotal or total parathyroidectomy reserved for patients resistant to medical treatment or failure of medical treatment.

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