

Case Report

Response of bisphosphonate therapy in recurrent tumoral calcinosis

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ABSTRACT

Tumoral calcinosis is a rare condition characterized by solitary or multiple, periarticular masses. Surgical excision of the tumoral calcinosis lesion is a well-documented treatment, but recurrences are not uncommon. A case of 42 year old male patient of tumoral calcinosis presented to us with history of repeated surgical excision twice in past 8 years for recurrent swellings. He was started on IV zoledronic acid and the patient has shown improvement after two years with no recurrence in follow up.

Keywords: Calcinosis, Di(bis)phosphonate, Hyperphosphatemia, Parathyroid hormone

INTRODUCTION

Tumoral calcinosis has been studied time to time but has remained a controversial clinico-pathological entity. Teutschländer used the term progressive lipocalcinogranulomatosis for a case of TC in 1935.¹ This entity is divided into two types -primary (with no associated disease). Primary (Familial) type is of two subtypes hyperphosphatemic or normophosphatemic tumoral calcinosis., which are due to inactivating mutation of FGF-23 and SAMD9 gene mutation respectively. Secondary type (particularly chronic renal failure, secondary and tertiary hyperparathyroidism).

Both surgical and medical modalities are being tried in TC with complete excision being the ultimate goal. Although surgery is considered to be definitive treatment modality, recurrence do occur due to incomplete excision of tumoral mass. Medical management of tumoral calcinosis comprises of dietary phosphate restriction, phosphate binders and rarely acetazolamide. Only a few articles have been published which has shown some role of bisphosphonate therapy in such cases. A case is presented of recurrent tumoral calcinosis who was diagnosed clinically, radiologically and biochemically

and later treated with bisphosphonate (IV zoledronic acid) at the centre with no recurrence in 2 years follow up.

CASE REPORT

A 42 years old male presented in the OPD with complains of swelling and discharging sinus over left gluteal region and small sinuses over scalp. There was gradual increase of pain which brought the patient to attention. There was no history of any fever or chronic illness or any kind of physical trauma. His past treatment history revealed that he was misdiagnosed by some local practitioners as a case of cold abscess gluteal region and was started on antitubercular therapy for 2 years but no response to the therapy was found. Thereafter he was diagnosed outside as tumoral calcinosis and was treated with surgical excision along with phosphate binding agents like calcium carbonate and sevelamer.

Now the patient presented with recurrence after one year in the OPD with swelling of left gluteal region (Figure 1) and this time with multiple discharging sinuses from which chalky white material was oozing. He also had chalky discharge from his scalp (Figure 2) and gums

along with loss of premolar tooth. His routine biochemical investigations came to be normal.



Figure 1: Discharging sinus which was operated earlier with excision of tumoral mass.



Figure 2: Scalp showing sinus with chalky white discharge.



Figure 3: Inhomogeneous mass around left hip joint.

Serum total calcium was 8.8 mg/dL and inorganic phosphorus 6.2 mg/dL. Serum 25(OH)D was 46 (30-50 ng/ml). Serum parathyroid hormone was 38 (10-65 ng/L).

Plain radiograph of left hip revealed inhomogeneous soft calcification over medial aspect of left thigh (Figure 3). MRI left hip showing periarticular mass (Figure 4). Patient was treated with IV zoledronic acid, dose 2 mg along with dietary restriction of phosphate, which showed great response in terms of resolution of present lesions and no recurrence of tumoral mass in follow up period of two years.



Figure 4: Amorphous deposition of calcium phosphate crystals around the joint and soft tissue.

DISCUSSION

Tumoral calcinosis is rare disorder of phosphorous metabolism which is characterized by periarticular soft tissue deposition of the calcium around large joints of the body. It is a kind of dystrophic calcification which primarily has normal calcium metabolism. It manifests as painless, tumor-like growth around the joints that may cause limitation of joint movement. Masses are usually found around large joints more than small ones, such as the hips, shoulder, elbow, knee.² Small joints like metacarpal and metatarsal are also involved.³ Plain radiograph of large joints are diagnostic tool for tumoral calcinosis which shows the typical appearance of cauliflower like peri-articular soft tissue calcification.⁴ Biopsy is usually avoided due to risk of infection, it can be considered in difficult cases to settle the diagnosis. Although genetic mutation analysis is confirmatory for the diagnosis of tumoral calcinosis.

Treatment of tumoral calcinosis is usually categorized as surgical and medical. Surgery still remains definitive treatment for tumoral calcinosis. Treatment for primary type has been classically described as complete surgical excision of tumoral deposits.⁵ Although there are chances of high rate of recurrence even after complete surgical excision, medical treatment prior to surgical approach is reasonable especially in primary variety. Treatment with dietary restriction of phosphates and phosphate binders such as oral aluminum hydroxide, sevelamer, lanthanum carbonate has been used so far with variable success rates in both normophosphatemic and hyperphosphatemic subtypes.⁶ There are limited literatures which favors the

use of bisphosphonates as treatment modality with variable success.⁷ Successfully zolendronate have been used in this case which has shown decrease in recurrence rate after 2 years follow up with regression in tumor size.

CONCLUSION

Tumoral calcinosis though a rare entity needs to be diagnosed correctly in earlier stages as this case was earlier misdiagnosed and treated as cold abscess of gluteal region with multiple discharging sinuses. Surgical excision is still considered as treatment of choice for many but authors should also be aware of the fact of high recurrences even after complete removal of mass as the defect lies biochemically in phosphate metabolism. Therefore, medical treatment in form of IV zolendronic acid may be warranted for this type of recurrent case to achieve better results.

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