

CASE REPORT

TUMOR CALCINOSIS: A RARE CASE REPORT

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ABSTRACT: A 15 year old male Arun Suresh Kammar patient presented with the chief complaints of right elbow swelling with discharging sinus since 5 years. Left elbow swelling and ulcer over left gluteal region since 3 years. Patient noticed gradually progressive, painless swelling over right elbow 5 years back and left gluteal region 3 years back for which he underwent surgery 4 and 3 years back respectively. One year latter patient developed gradually progressive diffuse swelling at left elbow with painful left elbow joint movement. Blood investigations were normal except raised phosphorus-6.8mg/dl(N-2.5-4.5mg/dl) and Alkaline phosphatase-52IU/L(N-20-40IU/L).Total excision was done for the left elbow and left gluteal region (periarticular) swelling and right elbow swelling subsided by itself.

KEYWORDS: Tumour calcinosis, Rare tumoural calcinosis, Elevated alkaline phosphatase.

INTRODUCTION:

- Idiopathic tumoral calcinosis is an unusual benign condition characterized by the presence of calcified soft tissue masses of varying size around the joints.^{1,2,3} It was previously described by Giard in 1898,¹ Duret in 1899 and Teutschländer in 1935 but the term "tumoral calcinosis" was used for the very first time by Inclan in 1943.¹
- Idiopathic tumoral calcinosis should be diagnosed by eliminating other conditions in which similar calcified masses are seen, such as chronic renal failure, hypervitaminosis D, milk-alkali syndrome, sarcoidosis, primary hyperparathyroidism, calcinosis universalis, calcinosis circumscripta, collagen vascular diseases and paraneoplastic Syndromes.¹
- These latter diseases are associated with a high serum calcium level while normocalcaemia is seen in idiopathic tumoralcalcinosis.¹ Hence, the diagnosis of tumoral calcinosis has to be refuted if an elevation of the blood levels of urea, calcium or non-protein nitrogen is found.¹ There are very few studies presenting the magnetic resonance (MR) imaging characteristics of this disorder.⁴

CLINICAL PRESENTATION: A Patient Named Arun Suresh Kammar 15 years old male, student, residing at po: Akkatangerhalli TQ: Gokak District: Belgaum; admitted in orthopedics C unit KIMS HUBLI, Presented with Right elbow swelling gradually increased in size with discharging sinus since 5 years, Left elbow swelling since 3 years, Ulcer over left gluteal region since 3 years.

Physical examination revealed a painless diffuse swelling of left elbow, a plain x-ray of left elbow showed a multiloculated calcified, soft tissue collection around left elbow joint.

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X-ray of right and left elbow shows evidence of calcified soft tissue mass around both elbow suggestive of tumor calcinosis (Right>>left)

HISTORY OF PRESENTING ILLNESS:

- Patient was apparently alright 5 years back. Then his mother noticed a swelling over right elbow which was almost an apple size, painless, gradually progressive for which patient underwent surgery for this swelling 4 years back at ESI hospital Belgaum,
- Again he developed swelling with discharging sinuses at same region for which he had got operated 3 years back, at ESI hospital Belgaum. Patient developed swelling at the left gluteal region 3 years back which was about a lemon size, gradually progressive non tender for which he underwent surgery 3 years back ESI hospital at Belgaum.
- One year later patient developed diffuse swelling at left elbow region which was gradually progressive, painful on movements at elbow joint. An ulcerated mass over left gluteal region.

NEGATIVE HISTORY:

- No h/o sudden weight loss.
- No h/o chronic cough with expectoration.
- No h/o any contact with tuberculosis patient.
- No h/o evening rise fever.
- No h/o night cries.

PAST HISTORY:

- No h/o dm, hypertension, tuberculosis, asthma, convulsions, or any past trauma
- H/o surgical interventions 3times at ESI hospital Belgaum before 4 years, before 3 years - 3 years respectively.

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RIGHT ELBOW

LEFT ELBOW:



Pre-op

Post-op

LEFT GLUTEAL REGION:



Pre-Op

Post-Op

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Excision of periarticular tissue from left elbow



Intra-operative excision of periarticular tissue from left elbow

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HISTOPATHOLOGY REPORT

Name: *Aman* Age: *14y* Sex: *male* #
Hospital I. P. No. / O. P. No. *20983* Work *202* Date of Receipt: *7/7/14*

Anatomical Source of Specimen: *Specimens sent in two bottles*
Gross - Bottle 1 -> specimen consists of multiple bits of grey
clinical diagnosis: brown to grey black tissue largest measuring 4x8x1 cm
Bottle 2 is specimen consists of 3 bits of grey white to grey
Microscopic appearance: brown tissue, largest measuring 1x1x0.4 cm.

Microscopic appearance: *Section studied shows features of tendral*
calcinosis.

Dr. Prof. & Head of the Dept. of Pathology
K. I. M. S., Hubli. *[Signature]*

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BLOOD INVESTIGATIONS:

- Hb = 10.9 gms %.
- RBC = 4.75 M/cumm.
- PCV = 34%.
- Total count = 10,600/cumm.
- MCV = 71.6 cumm.
- MCH = 22.9 mmg.
- MCHC = 32.1 %.
- DC = N65% E1% L34% B0%.
- Platelets = 7.99 lacs/cumm.
- Ionized Calcium = 1.38 mmol/L.
- Phosphorous = 6.8 mg/dl (2.5 – 4.5 mg/dl).
- Alkaline phosphatase = 52IU/L.

DISCUSSION:

- Idiopathic tumoral calcinosis is an unusual benign condition characterized by the presence of calcified soft tissue masses of varying size around the joints. It was previously described by Giard in 1898, Duret in 1899 and Teutschländer in 1935 but the term "tumoral calcinosis" was used for the very first time by Inclan in 1943.¹
- Radiographs showed well demarcated, lobulated and calcified soft tissue masses, located in the periarticular areas, unattached to bone in all cases. The masses consisted of conglomerations of multiple small, round opacities with different size and density separated by radiolucent septa. Fluid calcium levels were noted in 2 cases.⁴
- Radiographs also showed periosteal reaction associated to ill-defined patchy areas of osteocondensation within the medullary cavity of the tibiae in one patient. On scintigrams, increased radionuclide uptake was seen in the tibial diaphysis. These lesions resolved completely on follow up. As patient could not afford for expensive investigations. We had to operate with only radiograph as available investigation.^{4,3}
- Idiopathic tumoral calcinosis should be diagnosed by eliminating other conditions in which similar calcified masses are seen, such as chronic renal failure, hypervitaminosis D, milk-alkali syndrome, sarcoidosis, primary hyperparathyroidism, calcinosis universalis, calcinosis circumscripta collagen vascular diseases and paraneoplastic syndromes.¹ These latter diseases are associated with a high serum calcium level while normocalcaemia is seen in idiopathic tumoral calcinosis.¹
- Hence, the diagnosis of tumoral calcinosis has to be refuted if an elevation of the blood levels of urea, calcium or non-protein nitrogen is found.¹
- There are very few studies presenting the magnetic resonance (MR) imaging characteristics of this disorder.⁴

Idiopathic tumoral calcinosis is characterised by soft-tissue calcified masses around the large joints.^{2,4} Statistically hip joint is the most common site being involved. It is not uncommon for the calcific masses to become secondarily infected.²

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Pathogenesis of the disease is unclear, but an inherited error of inorganic phosphate and vitamin D metabolism is the most probable hypothesis. Hyperphosphataemia seen in some of the patients supports this theory.⁵ In this group, the disease develops earlier than 20 years of age. Such cases are familial with multiple lesions during disease course.^{2,3}

The exact pathogenesis of tumoral calcinosis is not defined; so the disease treatment is mainly symptomatic. Diets restricted in calcium and phosphate along with phosphate binding antacids have been used in some cases. This modality may be successful in hyperphosphataemic patients.²

Complete surgical excision of the mass is the optimal treatment.²

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