CASE REPORT

Idiopathic tumoral calcinosis presenting in early childhood

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SUMMARY

Idiopathic calcinosis (IC) is deposition of calcium hydroxyapatite crystals or amorphous calcium with no predisposing cause. IC presents in juxta-articular tissue in a tumour-like fashion is known as idiopathic tumoral calcinosis (ITC). Recurrent trauma and chronic pressure are thought to be the triggering factors. It usually presents in adolescents and adults. Presentation in early childhood is a rarity. We are reporting ITC in an 8-year-old child with the onset at 4 years of age.

BACKGROUND

Calcinosis cutis is an uncommon disorder characterised by the deposition of insoluble calcium salts such as hydroxyapatite crystals or amorphous calcium phosphate in the skin and subcutaneous tissue and rarely deeper tissues such as muscle and tendons.¹ Depending on the cause calcinosis cutis is of five subtypes, that is, dystrophic calcification, metastatic calcification, idiopathic calcification (IC), iatrogenic calcification and calciphylaxis.² Calcinosis is labelled as idiopathic when no cause can be determined (no local tissue insult or metabolic disorder). IC is of different types depending on depth of calcification. We present a case of idiopathic tumoral calcinosis (ITC) with an onset of disease at 4 years of age.

CASE PRESENTATION

An 8-year-old female child presented to dermatology outpatient department with multiple skin coloured to erythematous pea-sized nodules over lateral aspects of both feet for 6 months and ulceration over the right foot for the last 15 days. She also had a skin coloured swelling over left elbow, which had been gradually progressing in size over a period of 6 months. Lesions were otherwise asymptomatic. There was a history of similar lesions over right foot 4 years ago which was diagnosed as calcinosis cutis and surgical excision was done. There was no evidence of any connective tissue disorder or trauma. On examination, there was an ill-defined skin coloured swelling 8×4 cm approximately with no surface changes present over the left elbow (figure 1). On both knees, lateral aspects of both feet and left lateral malleoli, there were multiple ill-defined swellings ranging in size from 0.5×0.5 cm to 2×2 cm approximately with lesion over the lateral aspect of a right foot showing central ulceration and discharge of chalky white material (figure 2). The patient was diagnosed as

a case of multicentric calcinosis cutis. There was no evidence of any systemic or connective tissue disease on examination.

INVESTIGATIONS

Her complete haemogram, liver functions, renal functions, muscle enzymes, serum electrolytes were within normal limits. Erythrocyte sedimentation rate was 5 mm and C reactive protein was not raised. Rheumatoid factor, Antinuclear antibody (ANA) by Hep2, anticentromere and anti-Scl-70 antibodies were negative. There was no evidence of hepatitis or HIV. Radiographs of left elbow revealed the presence of a lobulated calcified mass of size 10×3 cm seen at the elbow joint posteromedially within the soft-tissue with normal underlying bones, suggestive of calcinosis cutis (figure 3). Radiography also revealed 0.5×0.5 cm foci of calcification on the lateral border of left foot (figure 4). Serum uric acid 4.4 mg/dL, serum calcium 8.3 mg/dL, serum phosphorus 4.1 mg/dL, serum PTH 22 ng/L, vitamin D 26 ng/dL were within normal limits.

DIFFERENTIAL DIAGNOSIS

ITC needs to be differentiated from, calcific tendinitis, other types of TC and synovial osteochondromatosis. Calcific tendinitis is localised within a tendon. Synovial osteochondromatosis shows calcification in rings and arcs pattern within the joint. In soft-tissue osteochondroma, osteosarcoma no calcification occurs.

TREATMENT

She was advised surgical excision followed by medical therapy in the form of tablet diltiazem.

OUTCOME AND FOLLOW-UP

She was referred to orthopaedic department. She instead visited a private practitioner, where the lesion on the right foot was excised. As rests of the lesions were asymptomatic, she was not interested in getting them excised. She did not come for follow-up.

DISCUSSION

TC is tumour-like deposits of calcium in the juxta-articular tissue. TC was first described by Duret³ and was termed as 'endotheliome calcifié'. Teutschlaender described it as lipocalcinogranulomatosis.⁴ Inclán *et al* coined the term TC and gave diagnostic criteria and radiological features of the disease.⁵ It has been synonymously called as calcifying bursitis,

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Figure 1 Swelling around right elbow.

calcifying collagenolysis and kikuyu.⁶ Smack *et al* classified TC into three groups: familial, primary and secondary.⁷ Secondary TC can be due to endocrinal and metabolic disorders like chronic renal failure, hyperparathyroidism, vitamin D intoxication, milk-alkali syndrome, connective tissue disease or malignancy induced bony destruction. Familial form is inherited in an autosomal recessive pattern, is associated with elevated serum phosphate and vitamin D due to defective phosphate excretion and decreased vitamin D synthesis. Primary ITC is sporadic and is not associated with any systemic, metabolic or endocrinal disorder and has normal calcium, phosphate, vitamin D levels.

Depending on the site and morphology, IC can be idiopathic scrotal calcinosis, subepidermal calcified nodules, TC and miliary calcinosis. Sporadic form of these deposits is usually seen in adolescents and young adults. Very few cases have been reported in early childhood in literature.⁸ Our patient of ITC presented to us at 8 years of age with an onset of lesions before 4 years of age, which is very rare in this age group.

ITC presents as painless slowly progressive deposits of calcium resembling neoplasms commonly seen around major joints such as hip, elbow, shoulder, foot and wrist in decreasing order. As seen in our case, lesions are quite often multicentric. Trauma and chronic pressure act as a triggering factor.⁹ Trauma leads

Figure 2 Ulceration on lateral aspect of right foot.

Figure 3 Plain X-ray elbow showing extra-articular calcinosis in nodulocystic pattern.

to collection of the blood, provoking fibrosis and fat necrosis, ultimately leading to calcification. But early age of presentation in our case puts a question mark as this factor to be implicated in the causation.

In ITC periarticular amorphous, cystic and multilobulated calcification gives a cobblestone appearance in plain X-rays. Cystic lesions separated by fibrous septa give a chicken wire type of lucent appearance. CT though not essential may additionally show the fluid level which is termed 'sedimentation sign'. There is usually no involvement of adjacent bones or joint.¹⁰ Harkess and Peters suggested diagnostic criteria for ITC.¹¹

ITC needs to be excised. The lesions are usually asymptomatic. Surgical excision is required in painful, ulcerated lesions impairing movements.¹² If excised completely it does not recur at the same site. Medical treatments like steroids, calcitonin, phenylbutazone, bisphosphonates have been tried but not found to be helpful.¹⁰

Figure 4 Plain X-ray showing calcium deposits on feet (red arrows).

Learning points

- ▶ Calcinosis cutis may be familial, primary or secondary.
- ▶ Idiopathic tumoral calcinosis is deposition of calcium in periarticular tissue without any systemic, metabolic or endocrinal disorder and has normal calcium, phosphate, vitamin D levels.
- ▶ Chronic trauma and friction are thought to be the triggering factors. Our patient was very young for these factors to be causative.
- ▶ Radiologically multilobulated and cystic deposits of amorphous calcium are seen.
- ▶ Symptomatic lesions can be excised.

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