

Tumoral Calcinosis of the Scalp: An Unusual Site for a Rare Tumor

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Tumoral calcinosis is a rare calcifying disorder that is associated with deposition of calcium crystals in the periarticular tissues. The mass is most often around the hips, elbows, shoulders, and feet but may be occasionally found elsewhere. We report a case of multiple sporadic tumoral calcinosises in an adult male over the scalp. The scalp as a site of tumoral calcinosis has not been previously reported in adults. Previous surgical excisions done on two occasions had resulted in recurrence of the tumors. This report highlights the need to include tumoral calcinosis in the differential diagnosis of tumors of the scalp. Key Words: Tumoral calcinosis, scalp, calcification.

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INTRODUCTION

Tumoral calcinosis was a term first used by Inclan¹ in 1943 to describe a dense, nodular, calcareous mass in the periarticular subcutaneous tissue. The mass is most often around the hips, elbows, shoulders, and feet but may be occasionally found elsewhere. The calcification usually takes the form of calcium hydroxyapatite crystals surrounded by a foreign body giant cell and histiocytic reaction. We describe a patient with multiple tumoral calcinosises of the scalp. To the best of our knowledge, this is the first report of occurrence of this tumor in the scalp in adults.

CASE REPORT

A 32-year-old healthy man presented to our hospital for cosmetic treatment of multiple progressively enlarging masses over the occipital region of his scalp and upper back. The first lesion had appeared when he was 5 years of age and subsequently a new lesion would crop up every 3 to 4 years. In the last decade, he had developed masses over the upper back, right shoulder, and forehead. He had been operated three times at different centers with recurrence.

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On physical examination, four large swellings in the occipital region were noted. They were mobile with varying consistency (Fig. 1). There were no pressure effects over the bone or skin. Three smaller swellings were noted over the forehead, two swellings were noted in relation to the right shoulder, and four large swellings were present over the upper back. The rest of the physical examination was unremarkable.

Laboratory investigations for calcium, phosphate, and renal functions were normal. Serum alkaline phosphatase and serum intact parathyroid hormone levels were within normal limits. An x-ray of the skull revealed chunky calcification over the area of the mass with no underlying destruction of bone, periosteal resorption or soft tissue swelling (Fig. 2).

An excisional biopsy was performed on one of the tumors in the occipital region. The biopsy showed fibrocollagenous connective tissue exhibiting extensive hyalinization, mineralization, and spicules of bone of various sizes. Based on these histologic findings, the diagnosis of nonfamilial sporadic tumoral calcinosis was made.

DISCUSSION

Tumoral calcinosis is a rare calcifying disorder of unknown origin. There appear to be three distinct varieties of tumoral calcinosis with very different biochemical profiles and pathophysiology. The first type is the sporadic variety in which no biochemical abnormalities or a family history of similar illness is elicited. The second type of tumoral calcinosis is usually secondary to metabolic disturbances associated with an increase in the calcium–phosphate solubility product. The common causes are chronic renal failure, hypervitaminosis D, and very rarely primary hyperparathyroidism.² The third type, which is familial, is associated with two genetic defects, the first of which was identified as biallelic loss-of-function mutations in the GALNT3 gene on chromosome 2q24–q31. This region of the chromosome encodes a glycosyltransferase responsible for initiating mucin-type O-glycosylation.³ Recently, Benet-Pages et al.⁴ reported a homozygous missense mutation (S71G) in the FGF-23 (fibroblast growth factor-23) gene in a boy with hyperphosphatemia, renal phosphate retention, and painful swelling of the elbows. Biochemically, patients with the familial disease have abnormal serum phosphate levels and inappropriately increased 1,25-dihydroxycholecalciferol levels. The scalp is an unusual area for development of these tumors and only one case report has described the development of multiple tumor calcinosises over the occipital region in an infant.⁵



Fig. 1. Right occipital region showing two of the largest swellings on the scalp. The masses were mobile with no pressure effects on the overlying skin.

Surgical excision has been recommended in patients with pain, recurrent infection, ulceration, and functional impairment resulting from tumoral calcinosis of any cause. However, surgical trauma may in itself stimulate further calcification and recurrence is common. The strategies for the medical treatment differ with the pathogenetic mechanism of tumoral calcinosis. In patients with renal failure, the primary goal of therapy is the lowering of plasma phosphate levels. This has been attempted with dietary phosphate restriction, use of phosphate binders, intensification of dialysis therapy, use of low-calcium dialysate, and parathyroidectomy in patients with severe secondary hyperparathyroidism. In familial tumoral calcinosis, also the focus of therapy is to decrease the serum phosphate levels. No definitive medical therapy has been found to be useful in sporadic tumoral calcinosis. Anecdotal reports have suggested that bisphosphonates therapy may be useful in sporadic tumoral calcinosis.⁷

In conclusion, sporadic tumoral calcinosis is a rare disease with no definitive therapy. The scalp is an unusual site for this rare disease. Surgical excision carries with it the risk of recurrence of the tumor.



Fig. 2. X-ray of the skull showing chunky calcification over the occiput in the region of the masses.

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