Aggressive tumoral calcinosis in an infant thoracotomy scar

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Abstract

The authors report a case of tumoral calcinosis (TC) in a six-month-old infant, which developed within the thoracotomy scar from previous aortic coarctation repair. After initial resection of the lesion, the child returned with a large mass of TC restricting movement of the left shoulder. Repeated total resection was successful with no recurrence in 12 months’ follow-up. This is the first report of TC that developed in a postoperative scar and is unusual in its recurrence and aggressive growth. Pathogenesis, diagnosis and treatment of this rare event is discussed.

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1. Introduction

Tumoral calcinosis (TC) is a problem of unknown etiology, usually occurring as calcification of periarticular soft tissues. The condition is exceptionally rare in infancy and has not been described in a postoperative scar.

2. Case report

The female infant presented with coarctation of aorta, restrictive ventricular septal defect and a bicuspid aortic valve. She underwent end-to-end repair of coarctation via posterolateral thoracotomy at nine weeks of age. The wound was closed using absorbable sutures; Dexon 3-0 (Ethicon, Johnson and Johnson, PA, USA) and Vicryl rapide 4-0 (Ethicon, Johnson and Johnson, PA, USA). Recovery was uneventful and she was discharged on the 12th postoperative day.

At the age of six months, four months after coarctation repair, a 50×20×20-mm irregular, painless, firm, non-fluctuant mass was found within the thoracotomy scar. She was afebrile and inflammatory markers (C-reactive protein and white cell count) were repeatedly within the normal range. Ultrasound suggested an encapsulated hematoma. This was excised completely and there were no signs of infection. Histology showed multiple cavities with calcium phosphate depositions and foreign body giant cells in the stroma, in-keeping with TC. Screening for metabolic and genetic diseases proved negative. Three months later, the wound had healed and the ventricular septal defect closed spontaneously. She was discharged from surgical follow-up.

At 22 months of age, the child returned with a 100×70×50-mm tumor in the thoracotomy scar, now restricting movement of the left shoulder. Unbeknownst to us, partial resection of recurrent tumor had taken place in another hospital 12 months previously. Chest X-ray and CT-scan showed a radiopaque mass extending into the neck beneath the left clavicle and scapula (Fig. 1).

The tumor and scar were completely resected using dual approach with a new supraclavicular incision. Histology confirmed the diagnosis of TC (Fig. 2). The patient was discharged after 34 days and at 34 months of age did not have any signs of recurrence.

3. Discussion

TC was first described by Inclan in 1943. The condition usually presents around large joints as a mass of calcium phosphate and carbonate, and is more frequent in dark-skinned populations. Extra articular forms are rare as is manifestation in infancy [1].

The problem occurs in three clinical settings. Firstly, as a complication of renal failure and dialysis due to disordered calcium and phosphate homeostasis. Secondly, as a genetic disorder with two subtypes: hyperphosphatemic familial TC with N-acetylgalactosaminyltransferase 3 (GALNT3) and normophosphatemic familial TC with sterile alpha motif domain containing protein 9 (SAMD9) genes abnormalities. Thirdly, as a sporadic condition of unknown etiology with trauma as possible cause in some cases [1–4].

Review of TC in infants by Hammoud et al. illustrates differences when compared to adults. There was usually no predisposing condition, trauma or family history of disease were not identified in any infant. Partial or complete resection was curative with no local recurrence [2].
forms of the disease need to be differentiated from soft tissue tumors [2, 5, 6].

Surgical resection is the treatment of choice. Dietary restriction of calcium and phosphate has been suggested but is not feasible in a growing child. Outcomes with biphosphonate, non-steroidal anti-inflammatory agents, steroids, radiotherapy and calcitonin are not encouraging [7–9]. Spontaneous regression of a TC lesion in an infant has also been reported [10]. We did not use medical therapy or dietary restrictions as phosphate and calcium levels were within the normal range in our patient. Aggressive growth with movement restriction of the scapula, possible future involvement of vital neck structures and risk of ulceration and infection of the lesion guided us to repeated resections. Given that surgical trauma is the most plausible cause of TC in this patient, we cannot satisfactorily explain why repeated complete resection of the lesion did not lead to further recurrence. Avoidance of suture materials used for soft tissue closure in the original surgery and initial total resection of TC, normalization of possible local abnormalities in calcium–phosphate metabolism and immunological maturation of the growing child might be possible explanations.

In summary, TC is a rare problem which poses both diagnostic and therapeutic dilemmas. It can develop in a postoperative scar and radical resection remains the treatment of choice.

References