Our Dermatology Online

Case Report

Subungual exostosis and subungual osteochondromas: diagnostic challenge in children

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ABSTRACT

Subungual exostosis (SE) and subungual osteochondroma (SO) are an uncommon osteocartilaginous benign, tumor that affects the distal phalanx. Both lesions are the prerogative of children. Controversy surrounds whether SE and a SO are the same clinical entity. We report the case of two little girls aged respectively 11 and 7 years old, which presents a similar clinical picture with the final diagnosis of exostosis for the first and osteochondroma for the second, which leads us to discuss the differences and similarities of these two entities as well as their management in children.

Key words: Subungual; Exostosis; Osteochondroma; Diagnostic; Children

INTRODUCTION

SE was described in 1847 by Dupuytren who, through a series of 30 cases, specified that it was a phalangeal lesion and not a nail lesion [1]. SE can be found at any age, but it predominates in children and young adults; it affects both sexes, with male dominance [2]. The most common clinical presentation of this tumour is a firm subungual nodule, which lifts the nail plate, resulting in nail dystrophy and pain when wearing shoes [3]. It is difficult to diagnose ES on the basis of clinical presentation alone and misdiagnosis or delays in diagnosis of this lesion are common. Indeed, X-ray examination and histopathology remain mandatory for the diagnosis; moreover some few dermoscopic descriptions have recently been published. Local excision of the mass with the overlying fbrocartilaginous cap is the preferred treatment [4]. Controversy surrounds whether SE and a subungual osteochondroma (SO) are the same clinical entity. There is a paucity of case reports on SO [5–7]. Some previous reports suggested that cases diagnosed as SE were in fact SO [8]. We report the case of an eleven year-old girl with an SE of the left big toe. Clinical, radiological and histological description of the case were given, as well as a new dermoscopic description. The patient was treated effectively by surgical excision with a two-year follow-up without recurrence. We also report a second case of a seven-year-old girl who presents a similar case but the final diagnosis was an osteochondroma, which will allow us to compare these two entities.

CASE REPORT

Case 1

This is a 7-year-old girl, with the notion of a trauma to the right foot dating back 3 months, then the appearance of a mass on the big toe of the same foot gradually increasing in size, painful when putting on shoes and sometimes when walking.

Dermatological examination found laterodistal onycholysis of the right hallux with elevation of the nail plate by an ovoid nodule of approximately 1.5 cm

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in diameter, pinkish-yellow, fixed and of firm with a hyperkeratotic smooth surface (Fig. 1).

Dermoscopy showed a yellowish-white central area corresponding to hyperkeratosis onycholysis and vascular ectasia (Fig. 2).

Radiographs show a pedunculated radio-opaque mass on the medial surface of the distal phalanx, homogeneous, well circumscribed, without continuity or remodeling of the underlying bone (Fig. 3). The diagnosis of exostosis was based on a series of anamnestic, clinical and radiological arguments. Our approach was to perform a complete removal of the lesion with a careful curettage of the bone plane to minimize the risk of recurrence. Microscopic findings include trabecular bone formation at the basement layer, a proliferating fbrocartilaginous cap, and some cyto-nuclear atypia which are very suggestive of the diagnosis (Fig. 4). The evolution was favorable with a resolution of the pain, good healing, complete regrowth of the nail and a standard X-ray of normal control, with a 2-year follow-up without recurrence.

Case 2

We report for comparison the case of an eight-year-old girl with no notable pathological antecedents, without notion of trauma, who presented for one year, a painful nodule of the right big toe, of hard consistency with distal onycholysis (Fig. 5). The clinical-radiological picture was similar to the first case. Surgical excision with the same technical was performed. The anatomopathological result showed some differences, namely a hyaline cartilage cap, less cellularity and the absence of cyto-nuclear atypia that would lead to the diagnosis of osteochondroma. The evolution was made by a normal regrowth of the nail.

DISCUSSION

Osteochondroma is the most common benign skeletal neoplasm and accounts for 10% to 15% of all bone tumors and 20% to 50% of benign bone tumors and its usually solitary (90%). Subungually located osteochondromas are rare and more common in toes than in fingers [9].

Osteochondromas are more frequently observed in adolescents and young adults, with a male to female ratio of 2:1, whereas subungual exostosis is frequently identified in patients 20 to 40 years of age, with a female to male ratio of 2:1 [10].

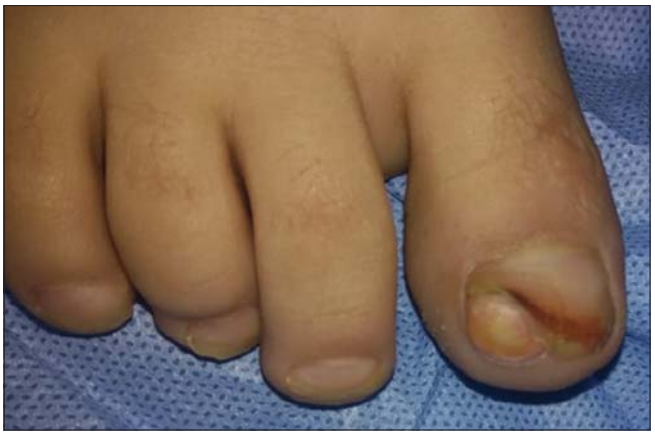

Figure 1: Clinical appearance of subungual exostosis with elevation of the nail plate.

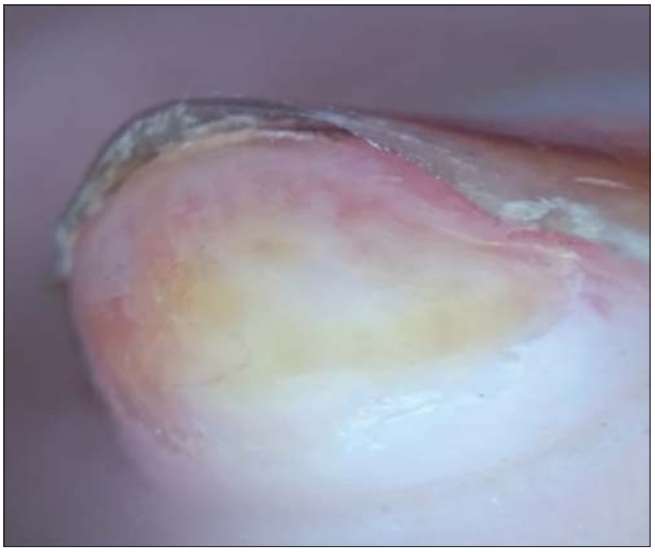

Figure 2: Dermoscopy of nail plate showing a yellowish-white central area, hyperkeratosis, onycholysis and peripheral vascular ectasia.

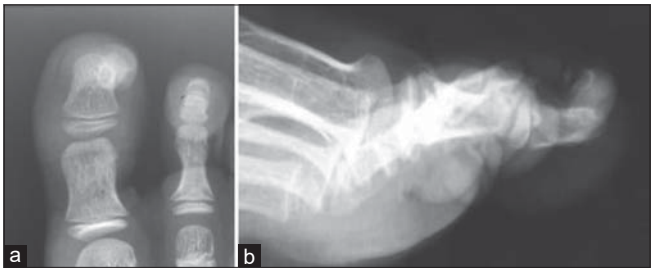

Figure 3: (a-b) X-ray revealing bony proliferation originating from the distal phalanx of the first toe in two different projections

In a recent retrospective study [8], who compared twenty-five cases histopathologically confirmed as SE or SO, there were 14 patients in the SE group and 11 patients in the SO group. There were no statistically

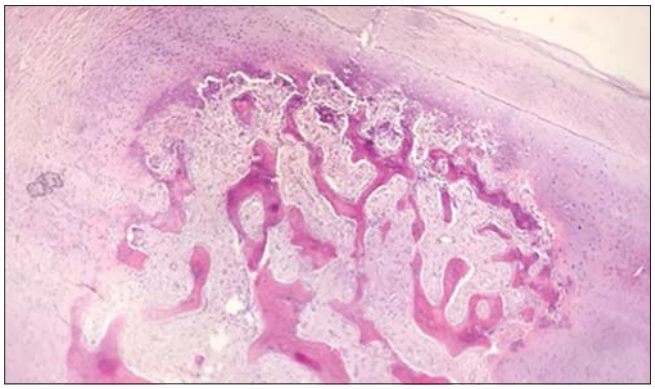

Figure 4: Light microscopic image of a histological specimen taken intraoperatively. It shows dermal tissue with trabecular bone and fibrocartilaginous overgrowth, consistent with subungual exostosis.

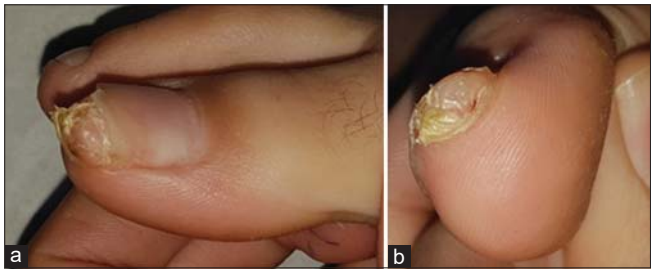

Figure 5: (a-b) Histologically confirmed sub-ungual osteochondroma in an eight-year-old girl.

significant between-group differences in age, gender, duration of illness, or presence of pain.

In both conditions, subungual masses develop progressively over several months and they enlarge may produce nail deformation and pain.

The pathogenesis of subungual exostosis remains uncertain, although trauma, infection, tumor, hereditary abnormality, or activation of a cartilage cyst have all been advanced as possible etiologies of subungual exostosis [2]. The majority consider it to be a reactive metaplasia resulting from microtrauma (1); More recently The translocation t(X;6)(q22;q13-14) has been reproducibly linked to subungual exostosis, implying it is a true neoplasm instead of being a reactive process in response to trauma [11]. However, at this time, there is no strong evidence to confirm a specific pathogenesis.

There is debate whether subungual osteochondroma is the same clinical entity as subungual exostosis. Some authors differentiate between ESU with a distal base of implantation and osteochondromas with a proximal base of implantation. Osteochondromas are considered to be congenital tumours of very slow growth, observed mainly in young males. Deformation of the nail shelf is secondary to hyperpressure [12].

Standard radiography provides several arguments to differentiate between these two entities, as the ES has a distal phalangeal implant base, whereas in the osteochondrome it is proximal. Moreover, osteochondromes appear as pedunculated or sessile exophytic growths and are composed of cortical and medullary bone with continuity of these two elements with the underlying bone defining this lesion, whereas ES appears as a homogeneous opacity without distinction between cortical and medullary and without continuity with the underlying bone, which makes it possible to distinguish them. However, other authors have pointed out that these radiological differences are not always obvious and that some cases remain very difficult to differentiate on standard radiography [8,9].

The major histologic difference between subungual exostosis and subungual osteochondroma is the composition of the cartilaginous cap. In subungual exostosis, this is composed of fibrocartilage, and the distal phalangeal tuft normally develops through enchondral ossification of fibrocartilaginous anlage [2,8,10].

Complete operative excision is the treatment of choice for both subungual exostosis and subungual osteochondromas. In our review of the literature, we found no difference in the recommended management or recurrence rate. Surgical treatment remains the gold standard in pediatric populations with very low recurrence with minor adverse events.

CONCLUSION

SE and SO is a relatively rare tumor, which affects many children, a good knowledge of its clinical and radiological aspects allow an early diagnosis to minimize secondary nail dystrophy. The surgical treatment allows a good healing.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be

published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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