Childhood soft tissue chondroma. Two cases report.

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Chondromes des tissus mous: A propos de deux cas pédiatriques.

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RÉSUMÉ

Prérequis : Le chondrome des tissus mous est une tumeur cartilagineuse bénigne rare, qui se voit généralement à l'âge adulte. Il s'agit d'une métaplasie hétérotopique qui se localise le plus souvent au niveau des mains et des pieds. L'implication de microtraumatisme dans la genèse de cette tumeur reste à vérifier.

But : Rapporter deux cas pédiatriques de chondrome des tissus mous.

Cas cliniques : Le premier est un chondrome axillaire gauche chez un garçon de 3 ans dont le scanner a montré une masse sphérique attachée au muscle infra épineux, de densité graisseuse sans calcifications évidentes. Le deuxième cas est un chondrome du 5ème doigt de la main droite chez une fille de 9 ans dont les radiographies standard ont montrés une opacité en regard de l'inter phalangienne proximale du 5ème rayon, siège de quelques calcifications ponctuées avec scalopping de l'os adjacent. L'IRM a objectivé une masse des tissus mous lobulée hyperintense en T2 et attachée au tendon fléchisseurs. Les deux patients ont été opérés avec exérèse complète de la tumeur. L'étude anatomopathologique a montré du tissue cartilagineux dans les deux cas. Les suites opératoires ont été simples avec une bonne récupération fonctionnelle et sans récidive.

Conclusion : Le traitement chirurgical des chondromes des tissus mous est souvent simple et l'exérèse doit être complète pour éviter les récidives.

Mots-clés

Chondrome, tissus mous, enfant.

SUMMARY

Background: Soft tissue chondroma is a rare benign tumour, which is generally seen in adult. It consists of islands of heterotopic cartilaginous tissue and most localised on the hands and the feet. The hypothesis that microtrauma is involved in the aetiology of this condition has yet to find any factual support.

Aim: To report two paediatric cases of soft tissue chondroma.

Cases report: The first is a soft tissue chondroma of the posterior aspect of the left axilla in a 3-year-old boy. CT scans showed a spherical fatty density soft tissue mass without evident calcifications, attaching the infraspinous muscle. The second patient is a 9-year-old girl presented with a right auricular finger soft tissue chondroma. Radiographs showed several punctuated calcifications with adjacent bone scalloping. MRI revealed a lobulated soft tissue mass attaching the flexor tendons. The tumours were entirely removed. Histological examination showed cartilaginous tissue in both cases. At follow-up, the patients had good functions without evidence of recurrence.

Conclusion: Simple excision should suffice to treat soft tissue chondroma but care should be taken to make the excision complete if recurrence are to be avoided.

Key-words

Chondroma, Soft tissue, Child

Soft tissue chondroma is a rare benign slowly growing tumour. It predominantly occurs in the third and fourth decades with a predilection for the hands and the feet. We report here two rare cases of soft tissue chondroma in children. At follow-up, the patients were disease-free with no evidence of recurrence.

CASES REPORT

Case 1

A 3-year-old right-handed boy presented with one year history of a slowly enlarging painless mass arising in the left retro-axillar region. There was no history of trauma or any medical problems. Physical examination revealed a deep soft tissue tumour of the posterior aspect of the left axilla. It was firm, with a size of an apricot, slightly painful when pressed and not infiltrative with good-defined margins. The patient was able to fully move the limb and there were no neurovascular problems. Radiograph showed a dense soft tissue without evident calcifications or bone lesion (Figure 1). CT scans showed a soft tissue mass with spherical shape and fatty density, about 4 by 4 by 3.5cm in size attaching the infraspinous muscle (Figure 2). Biopsy revealed a white encapsulated tumour reminding cartilaginous tumours. Histological examination showed a neoplasm composed predominantly of mature hyaline cartilage with variable cellularity and plump nuclei arranged in a lobular pattern at the periphery of the tumour. The histopathology report was benign chondroma of the soft tissue. The lesion was then treated with marginal excision. At six years follow-up, the patient was asymptomatic without evidence of recurrence.

Figure 1 : Plain-X ray: Para-scapular dense soft tissue without calcifications

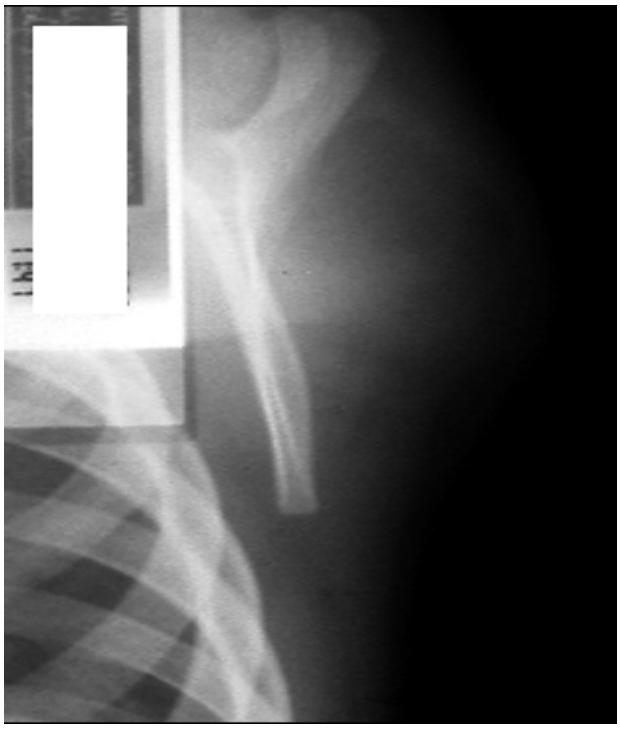
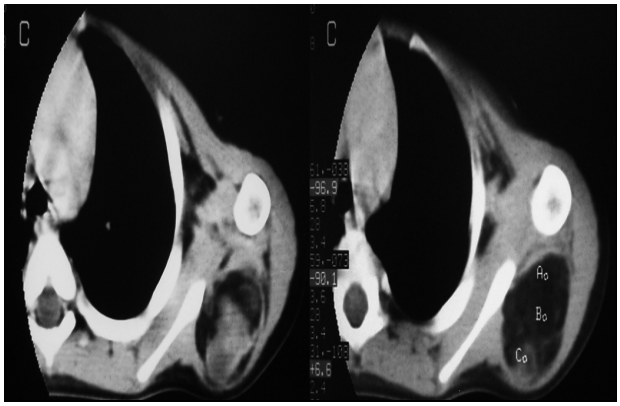


Figure 2 : Axial contrast-enhancement CT scans show a para-scapular spherical soft tissue tumour with fatty density



Case 2

A 9-year-old right-handed girl presented with four years history of right auricular finger swelling arising in the region of the proximal phalanx without any history of trauma. Although asymptomatic, it became gradually bigger and gave rise to concern. Physical examination revealed a nodular mass of the proximal phalanx near the inter-phalangeal joint laterally. It was firm, with a size of an olive, slightly painful when pressed. The overlying skin was reddish. The patient was able to fully extend the finger. However, flexion of the proximal inter-phalangeal joint was not full because of the intervening mass. There were no neurovascular problems of the finger. Radiographs showed several peripheral punctuated calcifications within a dense soft tissue and adjacent bone scalloping (Figure 3).

Figure 3 : Plain-X ray: Punctuated calcifications (arrow) in a dense palmer soft tissue with bone scalloping.

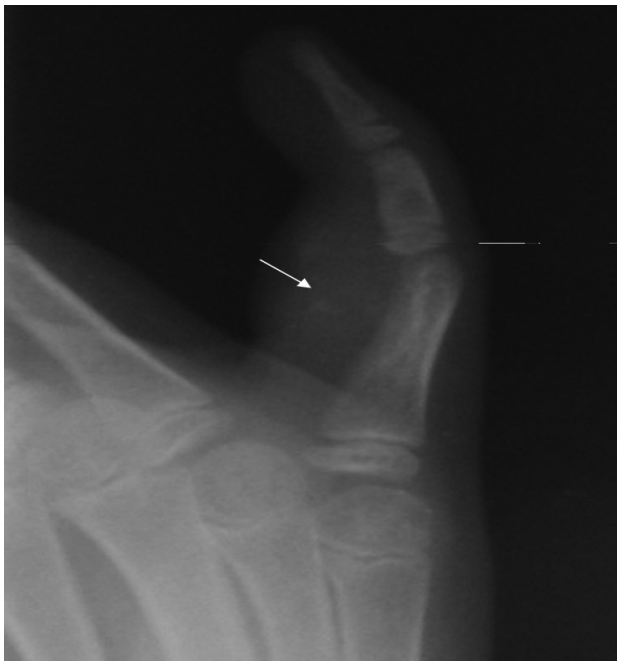
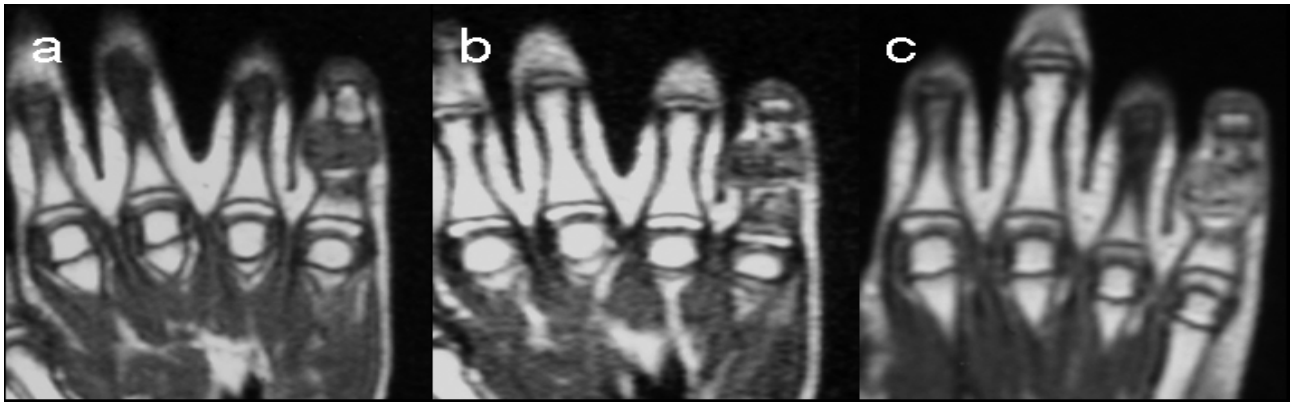


Figure 4 : MRI images: a) coronal T1-weighted image, b) coronal T2-weighted image, c) coronal T1-weighted enhancement MRI image: show a lobulated soft tissue tumour with a heterogeneous signal and enhancement after Gadolinium injection.



phalangeal joint capsule. Histological examination concluded to benign chondroma. At seven years follow-up, there was no recurrence and the finger's function was good.

DISCUSSION

Extraskeletal chondroma or soft tissue chondroma are defined by the presence of benign solitary cartilaginous masses in soft tissues [1]. It constitutes only 1.5% of benign soft-tissue tumours [2] and has a predilection for the distal part of the extremities [3, 4]. Short series or sporadic cases of the tumour are the most reported and only Dahlin and Salvador [3] and Chung and Enzinger [4] presented series of respectively 70 and 104 patients. The tumour, common in adulthood and occurring mainly between the fourth and sixth decades, had been also but rarely reported in young children [5, 6]. Miki et al [7] reported a case of congenital chondroma in a newborn, located between the spinous processes of T12 and L1 vertebras affecting equally both genders [8, 9, 10]. Soft tissue chondroma arises in majority cases with a solitary localization [1] but multiple soft tissue chondromas have been also reported [11, 12]. Malignant transformation occurs infrequently [13].

For many authors, chondromas of the soft tissues are believed to be of teno-synovial origin. In fact, their tendency to be near bones and joints, their sometimes multicentric occurrence, and their association with multicentric intra-synovial cartilaginous masses or solitary loose bodies are highly suggestive of this derivation. Furthermore, their histological similarity to major articular synovial chondromatosis, as well as the demonstration in some of these lesions of areas resembling giant cell tumours of tendon sheaths, lends further evidence that these lesions are most probably of teno-synovial origin [3, 14, 15, 16, 17, 18].

These tumours are asymptomatic and undetected unless a mass appears and so the clinical history consists often on a painless slowly enlarging soft tissue mass [1]. The lesion may be painful when located in the plantar aspect of the foot [19, 20, 21]. On palpation, the tumour is a painless, firm, well delimitated nodular mass with a variable size.

The imaging features are variable. Focal calcifications are often present in the center of the lesions [3, 4, 13, 22] but as seen in our second case, these calcifications may be noted in the periphery aspect of the tumour. Late diffuse calcification of the entire tumour was reported [13]. Chung and Enzinger [4] reported that the tumour never involved the underlying bones. However compression deformity, bone remodelling, bone erosion or bone sclerosis due to the soft-tissue mass have also been reported [4, 24, 28, 30]. In our second case, a cortical scalloping was originated by the tumour without bone expanding. Descriptions of MRI of soft-tissue chondromas are rare in the literature. They have been reported to be of intermediate signal intensity on T1 weighted images and of high signal on T2 weighted images [1, 20, 23, 24, 25, 26]. The high signal is due to a high water content of the mucopolysaccharide component or myxoid changes [24, 27]. In two reports, it exhibited low signal on both T1 and T2 weighted images which is due to a dense calcified matrix [28, 29].

Grossly, chondromas of soft tissues are well-encapsulated with lobular architecture [22]. Histologically, lobules of mature hyaline cartilage with varying cellularity are seen [13, 22]. The external border of the tumour is usually well delineated from the adjacent tissues. The chondrocytes are located in rounded spaces and have usually single nucleus. The tumour can demonstrate marked cellularity, binucleated cells and mitoses. These findings may lead to the misdiagnosis of malignancy [12, 13, 22, 31] but in children, they suggest an active chondroma because chondrosarcomas and malignant transformation are extremely rare. The chondrocytes are positive for vimentin and S-100 protein [22]. The molecular biology had revealed two types of chromosomal aberrations: 12ql3-ql5 translocation and monosomy of chromosome 6 with rearrangement of chromosome 11 [27].

It is necessary to differentiate other tumours from chondroma, especially when calcifications are present [8, 30]. In children, some others tumours can be discussed. Periosteal chondroma typically arises from the surface of the bone and beneath the periosteum, and has saucer-like erosion with a well defined rim of reactive bone [8, 30]. Myositis ossificans has a characteristic zonal pattern and occurs less frequently in the extremities [30].

Malignant tumours such as extraskeletal osteosarcoma tend to be larger with a preference for the thighs [30]. Synovial sarcomas have more irregular contours and a larger size and when calcifications are present, they are often at the periphery [8, 24]. Tumoral calcinosis, arising commonly near elbows and hips, tends to have amorphous, nodular or multilobulated calcifications with relatively poorly defined margins [8, 30]. Synovial chondromatosis, Hoffa's disease and extraskeletal chondrosarcoma occur rarely in children.

Treatment usually consists of complete marginal excision. Local recurrence had been reported up to 18% [11, 29, 31, 33] and can be treated with repeat excision [1, 3, 4].

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