Journal of Pediatric Endocrinology

Open Access

Austin Publishing Group

Case Report

Calcifying Aponeurotic Fibroma in Children: Case Report

Kessab A[1,3]*, Chalouah B[2,3], Azzakhmam M[1,3], Benchakroun M[2,3], Bouaabid AS[2,3] and Rouas L[1,3]

[1]Department of Anatomy Pathological, Laboratory Children's Hospital Ibn Sina CHU, Morocco

[2]Department of Orthopedic Surgery, Military Hospital, Morocco

[3]Faculty of Medicine and Pharmacy of Rabat (FMPR), Morocco

***Corresponding author:** Amine Kessab, Pathological Anatomy Laboratory Children's Hospital Ibn Sina CHU, Faculty of Medicine and Pharmacy of Rabat (FMPR), Rabat-Morocco

Received: December 28, 2020; **Accepted:** January 21, 2021; **Published:** January 28, 2021

Abstract

Calcifying aponeurotic fibroma is a rare benign fibrous tumour with characteristic histopathological features commonly observed in children and adolescents. It usually involves the most distal part of the extremities. Despite being a benign tumour, it has a high risk of local recurrence. Our case is a 14-year-old child, attending school, followed for chronic renal failure with small kidneys who presented with a pathological fracture in his left big toe with a lytic X-ray image. Calcifying aponeurotic fibroma is a rare tumor, usually located in the subcutaneous layer, but it can manifest as a diffuse intra-articular calcified mass with numerous bone erosions. Calcification is absent at the early stage, hence the importance of careful radiological examination and complete histological analysis in order to rule out a probable malignant tumor. The rate of recurrence is important, hence the importance of surgical excision with healthy margins.

Keywords: Aponevrotic; Children; Antibodies; Fibroma; Toe

Introduction

Calcifying aponeurotic fibroma is a rare benign tumor (represents less than 1% of benign soft tissue tumors) and is most often found in children or young adults with a peak incidence between 8 and 14 years of age and sex. Ratio of 2 men to 1 woman (predominantly male). It develops, in 70% of cases, in contact with aponeuroses, fasciae and tendons of the palmar face of the hands. The second location in terms of frequency is the foot, especially the plantar surface. This tumor grows slowly, it remains painless, does not limit the course of the joint. Radiologically, the mass is poorly limited, with a heterogeneous signal on MRI. Calcifications are sometimes absent; in their presence they remain fine, punctate and of have mainly a central distribution. A biopsy is essential before accessing the excision in order to rule out the main differential diagnosis: synovialosarcoma.

Case Presentation

This is a 14-year-old child, in school, followed for chronic kidney disease with small kidneys. Currently he has microcytic hypochromic anemia, which has resulted in hair loss with significant inflammatory syndrome, all in a context of deterioration of his general condition. A pathologic fracture in the left toe revealed by swelling showed on the standard radiograph a well-defined lytic image with a periosteal reaction (Figure 1). A surgical biopsy was taken from the outgrowth and the fragments were sent to our laboratory. Two fragments were examined measuring respectively 0.3x0.3 and 0.3x0.2cm, they are completely included in a block. Histological study showed tumor proliferation made of elongated fibroblasts and epithelioids with oval or elongated nuclei with vesicular chromatin, the mitotic index is low estimated at mitosis by ten fields with Gx40. These fibroblasts are dispersed within a collagen-rich stroma, forming palisades around collagen nodules, which are sometimes calcified. PAS staining was performed with positivity in the collagen nodules. Inflammatory cells such as foamy histiocytes as well as bone sequesters are observed (Figure 2).

An immunohistochemical study was carried out which showed:

- A positivity of fibroblast tumor cells for the anti vimentin antibody.

- Focal positivity of fibroblast tumor cells for anti PS100 and anti AML antibodies (Figure 3).

- Negativity for the anti CD34 and CD68 antibodies.

A diagnosis of calcifying aponeurotic fibroma is made.

Discussion

Calcifying aponeurotic fibroma is a rare benign fibroblastic tumor

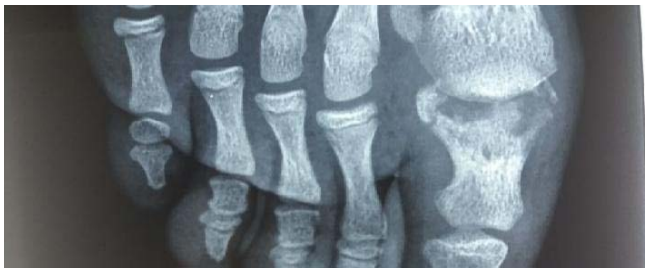

Figure 1: X-ray image showing lytic lesion of the left big toe.

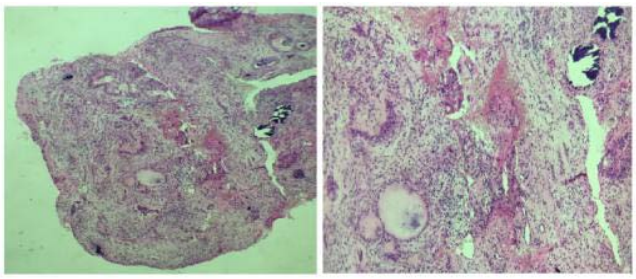

Figure 2: Histological Images [HI] showing tumor proliferation made up of elongated fibroblasts and epithelioids with oval or elongated nuclei with vesicular chromatin dispersed within a collagen-rich stroma forming palisades around the collagen nodules which are sentîmes calcified.

Citation: Kessab A, Chalouah B, Azzakhmam M, Benchakroun M, Bouaabid AS and Rouas L. Calcifying Aponeurotic Fibroma in Children: Case Report. J Pediatri Endocrinol. 2021; 6(1): 1039.

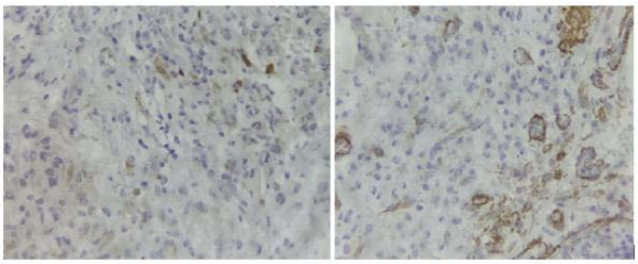

Figure 3: Images showing focal immunohistochemical staining of fibroblast cells by PS100 (right) and AML (left) at the tumor level.

first described in 1953 by Keasbey as juvenile aponeurotic fibroma [1]. Selon une etude anglaise faite par Shim et al., intitulee: Fibroma aponeurotic calcifying upper and lower limbs [2], According to an English study made by Shim and all, titled: Fibroma aponeurotic calcifying upper and lower limbs, the duration of discovery of the tumor mass varies between 4 days and 20 years and generally it is symptomatic after one year with no decrease in mobility in 71% of cases [2]. The standard radiograph shows a poor limitation of the tumor, with fine, dotted calcifications in the center of the lesion. Extrinsic erosion of adjacent bone is rarely seen [3].

The MRI shows in T1 hyperostosis and calcifications with signs of the presence of adipose tissue. This examination remains insufficient to make the diagnosis, hence the importance of the histological examination [4,5]. Macroscopic examination shows a flesh-colored, pseudo-encapsulated mass with lesional limits. Microscopic examination remains the key to diagnosis with a characteristic appearance: the presence of foci of calcification and collagen deposits surrounded by a proliferation of fibroblasts with round and oval nuclei. Mitotic figures are rare. In some cases, there are also multi-nucleated giant cells around the calcified areas. Ossification is rare. Scattered foci of chondroid differentiation are also noted. Recently, cytologic examination can help diagnose benignity, especially if the radiology is uncertain [6]. Immunohistochemical data remain of minimal importance given the non-specificity of positive markers such as PS100 and AML and vimentin [7]. The transformation towards malignancy is extremely rare, Lafferty et al., reported a case of calcifying aponeurotic fibroma transformed into metastatic pulmonary and bone fibrosarcoma [8].

The tumor is locally invasive, usually affecting the distal extremities, most often the fingers, palms of the hands and the soles of the feet [9]. The recurrence rate after surgical excision is generally reported in 50% of cases, with an interval varying between 6 months and 23 years, hence the need for an accurate and complete diagnosis before excision [7]. Local recurrence is seen more frequently in children under 5 years of age [10], especially during the first three years postoperatively [11].

The reasons which determine the recurrence remain unknown. Some authors [11-13] have made the link between the histological characteristics and the rate of recurrence, and they have distinguished two types: the first generally affects young children, more infiltrative, with little calcification [11,7] with a high recurrence rate, the second type is nodular in structure and usually contains significant calcification and occurs in older children, with a lower tendency to relapse [14,5]. These two forms could explain the correlation between the age and the risk of recurrence.

The differential diagnoses of calcifying aponeurotic fibroma are essentially malignant soft tissue tumors encountered in the hand and wrist, especially in the presence of intratumoral calcifications, including epithelioid sarcoma, synovialosarcoma, and undifferentiated pleomorphic sarcoma [15,16].

Conclusion

Calcifying aponeurotic fibroma is a rare tumor, usually located in the subcutaneous layer, but it can manifest as a diffuse intra-articular calcified mass with numerous bone erosions. Calcification is absent at the early stage, hence the importance of careful radiological examination and complete histological analysis in order to eliminate a probable malignant tumor. The rate of recurrence is important, hence the importance of surgical excision with healthy margins.

References

1. Murphey MD, Ruble CM, Tyszko SM, Zbojniewicz AM, Potter BK and Miettinen M. Musculoskeletal fibromatoses: radiologic- pathologic correlation 1. Radiographics. 2009; 29: 2143-2173.
2. Shim SW, Kang BS, Lee CC, Suh JH and Shim HS. MRI features of calcifying aponeurotic fibroma in the upper arm: a case report and review of the literature. Skelet Radiol. 2016; 45: 1139-1143.
3. Morii T, Yoshiyama A, Morioka H, Anazawa U, Mochizuki K and Yabe H. Clinical of magnetic resonance imaging in the preoperative differential diagnosis of calcifying aponeurotic fibroma. J Orthop Sci. 2008; 13: 180-186.
4. Nishio J, Inamitsu H, Iwasaki H, Hayashi H and Naito M. Calcifying aponeurotic fibroma of the finger in an elderly patient: CT and MRI findings with pathologic correlation. Exp Ther Med. 2014; 8: 841-843.
5. Takaku M, Hashimoto I, Nakanishi H and Kurashiki T. Calcifying aponeurotic fibroma of the elbow: a case report. J Med Invest. 2011; 58: 159-162.
6. Keasbey LE and Fanselau HA. The aponeurotic fibroma. Clin Ortho. 1961; 19: 115-131.
7. DeSimone RS and Zielinski CJ. Calcifying aponeurotic fibroma of the hand. A case report. J Bone Joint Surg Am. 2001; 83: 586-588.
8. Allen PW and Enzinger FM. Juvenile aponeurotic fibroma. Cancer. 1970; 26: 8570-867.
9. Goldman RL. The cartilage analogue of fibromatosis (aponeurotic fibroma). Further observations based on 7 new cases. Cancer. 1970; 26: 1325-1331.
10. Fletcher CDM, Unni KK and Mertens F. Pathology and genetics of tumours of soft tissue and bone. World Health Organization, classification of tumors. Lyon: IARC Press. 2002; 69-71.
11. Fetsch JF and Miettinen M. Calcifying aponeurotic fibroma: a clinicopathologic study of 22 cases arising in uncommon sites. Hum Pathol. 1998; 29: 1504-1510.
12. Keasbey LE. Juvenile aponeurotic fibroma (calcifying fibroma); a distinctive tumor arising in the palms and soles of young children. Cancer. 1953; 6: 338-346.
13. Keasbey LE and Fanselau HA. The aponeurotic fibroma. Clin Orthop 1961; 19: 115-131.
14. Lichtenstein L and Goldman RL. The cartilage analogue of fibromatosis. A reinterpretation of the condition called 'juvenile aponeurotic fibroma'. Cancer. 1964; 17: 810-816.
15. Drape JL and Le Viet D. MR imaging of the fingers. In: Stoller DW, Li AE, Bredella MA, Potter HG, Rosenberg ZS, Bencardino JT, eds. Magnetic resonance imaging in orthopedics and sports medicine, 3rd ed. Philadelphia, PA: LW & W. 2007: 1847-1932.

16. van Vliet M and Kliffen M, Krestin GP, van Dijke CF. Soft tissue sarcomas at a glance: clinical, histological, and MR imaging features of malignant extremity soft tissue tumors. Eur Radiol. 2009; 19: 1499-1511.