ACTA
ORTHOPAEDICA
et
TRAUMATOLOGICA
TURCICA

Author's translation
Acta Orthop Traumatol Turc 2008;42(2):145-147

Calcifying aponeurotic fibroma: a case report

Kalsifiye aponörotik fibrom: Olgu sunumu

Nilüfer ONAK-KANDEMIR, Aylin EGE-GUL, Nimet KARADAYI

Zonguldak Karaelmas Üniversitesi Tıp Fakültesi Patoloji Anabilim Dalı;
[1]Dr. Lütfi Kırdar Eğitim ve Araştırma Hastanesi, Patoloji Kliniği

Kalsifiye aponörotik fibrom nadir rastlanan, histopatolojik özellikleri iyi tanımlanmış, benign fibröz bir tümördür. Genellikle çocuk ve gençlerde distal ekstremitelerde görülür. Yirmi üç yaşında erkek hastada sol el palmar yüzde yaklaşık beş yıldır var olan ağrısız serbest yumuşak doku kitlesi cerrahi olarak çıkarıldı. Histopatolojik incelemede kalsifikasyon ve kondroid metaplazi alanlarını çevreleyen fibroblastik proliferasyon alanları ve yoğun kollajenöz stroma izlendi. Lezyona kalsifiye aponörotik fibrom tanısı kondu. Hastanın bir yıllık izleminde nüks görülmedi.

Anahtar sözcükler: Kalsinozis; fibrom/patoloji; el; yumuşak doku neoplazileri.

Calcified aponeurotic fibroma is a rare benign fibrous tumor with well-characterized histopathologic features. It is usually seen in distal extremities of children and youngsters. A 23-year-old male patient underwent surgical excision of a free, painless soft tissue mass that developed in the palmar aspect of the left hand. Histopathologic examination showed fibroblastic proliferation areas and dense collagenous stroma surrounding calcifications and chondroid metaplasia. The diagnosis was made as calcified aponeurotic fibroma. No recurrences were detected within a year follow-up.

Key words: Calcinosis; fibroma/pathology; hand; soft tissue neoplasms.

Calcifying aponeurotic fibroma (KAF) is a rare benign fibrous tumour with well-characterized histopathological features. They are generally slowly growing lesions related to tendons and aponeuroses in the distal extremities of children and youngsters. During the stage following rapid growth phase, an increase of calcification and a pause of growth are seen. Lesion often shows local recurrence. Due to functional efficiency total excision of the lesion is suggested. In differential diagnosis, infantile or juvenile fibromatosis, fibrous hamartoma, monophasic fibrous type synovial sarcoma and romatoid nodule should be taken into account.[1-3]

Our case has been presented for being a rare lesion with characteristic histopathological features, its occurence in youngsters and its presentation as a free soft tissue mass.

Case report

Painless free soft tissue mass existing for a long time and localised in left hand palmar side of 23-year-old male was totally excised. The lesion was 10x10x7 cm, yellow-cream and elastic with calcification areas in cut surface. Histopathologically, there were fibroplastic proliferation areas of spindle cells with round-oval nucleus, eosinophilic cytoplasm and unclear borders which were sorrounding areas of calcification and chondroid metaplasia, besides dense collagenous stroma (Figure 1). The cells forming the lesion did not show pleomorphism, atypia or mitotic activity. Lesion was clearly separated from the adjacent tissue and was surrounded with a thin, fibrous capsule. It was diagnosed as "Calsifying Aponeurotic Fibroma (CAF)" as a result of histopathological examination.

Correspondence / *Yazışma adresi:* Dr. Nilüfer Onak Kandemir. Zonguldak Karaelmas Üniversitesi Tıp Fakültesi, Patoloji Anabilim Dalı, 67600 Kozlu, Zonguldak. Phone:+9 0372 - 261 01 83 Fax:+9 0372 - 261 01 55 e-mail: niluferkandemir@yahoo.com

Submitted / *Başburu tarihi:* 23.11.2007 **Accepted / *Kabul tarihi:*** 05.02.2008

Discussion

CAF is first described by Keasbey at 1953 as "Juvenile Aponeutoic Fibroma". Today, since it is recognised in a wider age group, the preferred names are "aponeurotic fibroma" and "calsifying aponeurotic fibroma". Approximately 100 case reports has taken place in the literature, till now.[1-5]

CAF, unlike juvenile fibromatosis, though seen in a wider age group, is mostly observed in 0-16-year-aged children and adolescents. It generally is presented as a mass in palm of the hand and fingers. There are few cases localised in the feet.[1,2] Our patient is 23-year-old and this supports the view that it can also be found in young adulthood. Lesion in our case is localised in palm which is the localization CAF mostly prefers.

Clinically, it is presented as a painless soft tissue mass mostly related to aponeurosis, tendon and fascia. It can rarely also be seen as a free soft tissue mass, if the relation of the mass with adjacent tissue disappears in the course of time.[1-6] In our case, the mass existed for a long time as a free soft tissue mass not causing pain or function loss.

Radiological findings are not diagnostic. In direct radiography, a mass with unclear borders with calsific loci is seen. The mass has to be differentiated from ganglion in ultrasonographic examination and panniculitis ossificans, extra-skeletal or parosteal chondroma in tomographic examination.[1,4]

The lesion, macroscopically, is mostly smaller than 3 cm, poorly demarcated, elastic, gray-white nodule.

Definite diagnosis is always given with histopathological examination and in some cases with immunohistochemical and ultrastructural studies. Microscopical features of the lesion are characteristic. Generally the-

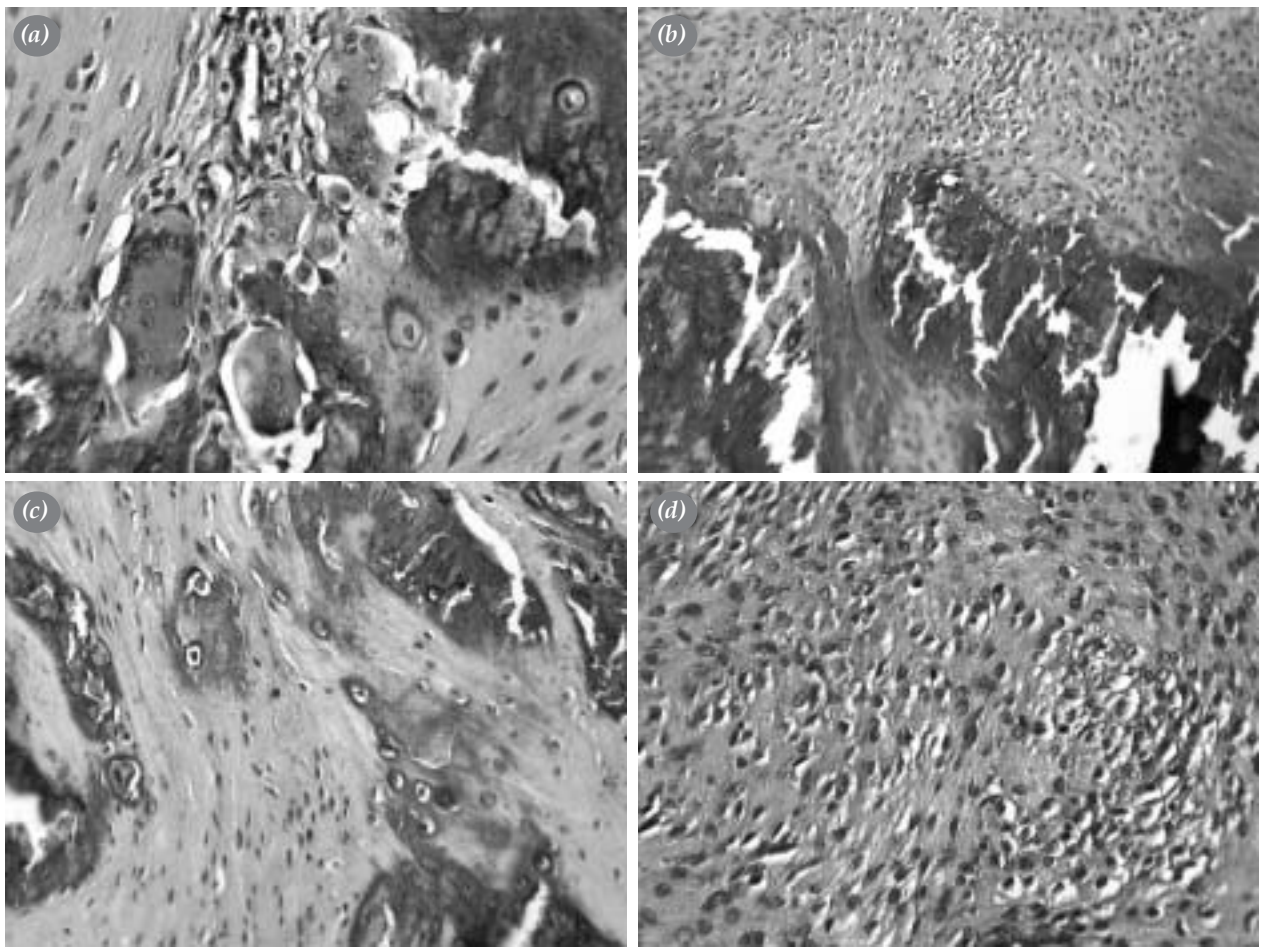

Figure 1. Calcifying aponeurotic fibroma. **(a)** Multinuclear giant cells surrounding focal calcification areas (H&E, X100). **(b)** Fibroblastic proliferation areas adjacent to calcification areas. (H&E, X60). **(c)** Areas showing chondroid metaplasia (H&E, X100). **(d)** Myofibroblast-like cellular proliferation areas (H&E, X100).

re are calcified and chondroid areas in the center and fibroblastic proliferation surrounds these areas. Plump fibroblasts with round and oval nucleus are separated with dense collagenous stroma. Mitotic figures are rare. Fibrous growth is mostly related to tendon and aponeurosis surrounding blood vessels and nevre fibers. In 1/3 of the cases, calcification is related to cartilage. In some cases, there are multinuclear giant cells around calcified areas. Ossification is rare. Borders are more clear and cellularity is less in older cases.[1] Our case shows all the features of CAF. The lesion, existing for a long time, is separated from the adjacent tissues with a clear border and has dense calsification and chondroid metaplasia areas.

Histopathological differential diagnosis is associated with lesion age. In infants and young children, since there is no or few calcification, it is difficult to differentiate from infantile or juvenile fibromatosis. Plump fibroblasts, their relation to dense and hyalinised collagen and localization of the lesion help to differentiate. Palmar and plantar fibromatosis are rare in children. They do not have calcification and chondroid metaplasia areas. Malign spindle cell tumors should be taken into account in CAFs with dominant spindle cell pattern. In cases belonging to elderly patients, to differentiate from soft part chondroma can be a problem. Since both lesions are localised in the same area they can clinically be confused. Chondromas have less fibrous tissue microscopically, are lobulated and calcification is not focal or linear as in CAF but shows a homogenous distribution. Panniculitis ossificans, nodular fasciitis, calcifying soft tissue leiomyoma and romatoid nodule are the others lesions that should be considered in differential diagnosis .[1, 2, 7, 8]

In more than half of the cases local reccurence occurs. Reccurence is common especially in infants and children. Malign transformation is very seldom. Although total excision of the mass is suggested, functional efficiency should be considered.[1, 5, 9] In our case, reccurence is not seen in 1 year follow-up.

In conclusion, calsifying aponeurotic fibroma which is a rare soft tissue tumor, as in our case, can present as a free soft tissue mass. These lesions mostly are observed in childhood, however can also be seen in young adults as the case presented here. In the differential diagnosis of distal localised fibrous lesions, calsifying aponeurotic fibroma should be taken into account and attention should be payed to the age of the lesion while diagnostic histopathological features are being evaluated.

References

1. Enzinger FM, Weiss SW, editors. Fibrous tumors of infancy and childhood. In: Soft tissue tumors. 3rd ed. St. Louis: Mosby; 1995. p. 256-61.
2. Fetsch JF, Miettinen M. Calcifying aponeurotic fibroma: a clinicopathologic study of 22 cases arising in uncommon sites. Hum Pathol 1998;29:1504-10.
3. Iwasaki H, Kikuchi M, Eimoto T, Enjoji M, Yoh S, Sakurai H. Juvenile aponeurotic fibroma: an ultrastructural study. Ultrastruct Pathol 1983;4:75-83.
4. Hasegawa HK, Park S, Hamazaki M. Calcifying aponeurotic fibroma of the knee: A case report with radiological findings. J Dermatol 2006;33:169-73.
5. Sethi S, Mishra K, Rajni. Calcifying aponeurotic fibroma: a case report and review of literature. Indian J Pathol Microbiol 2003;46:223-6.
6. Sferopoulos NK, Kotakidou R. Calcifying aponeurotic fibroma: a report of three cases. Acta Orthop Belg 2001;67:412-6.
7. McCurdie I, Jawad SS. Unusual and memorable. Calcifying aponeurotic fibromata. Ann Rheum Dis 1998;57:78.
8. DeSimone RS, Zielinski CJ. Calcifying aponeurotic fibroma of the hand. A case report. J Bone Joint Surg [Am] 2001;83:586-8.
9. Amaravati R. Rare malignant transformation of a calcifying aponeurotic fibroma. J Bone Joint Surg [Am] 2002;84:1889.