Case Report

Calcifying aponeurotic fibroma in a girl: MRI findings and their chronological changes

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Abstract

Calcifying aponeurotic fibroma (CAF) is a rare benign tumor that typically develops in the fascia and tendons. On MRI, CAF usually shows isointensity on T1WI and heterogeneous high intensity on T2WI. We report a case of 17-month-old girl with CAF on her right ankle. MRI showed an unusual signal intensity for CAF. At 3 years and 4 months of age, MRI revealed hyperintense nodules on T1WI and T2WI inside of the CAF. These fatty areas corresponded to coarse calcified nodules on CT. Radiologists should be aware of the fact that fat-like signals can appear in CAF on MRI.

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Introduction

Calcifying aponeurotic fibroma (CAF) is a rare benign tumor that typically develops in the fascia and tendons of young individuals. Pathologically, the condition is characterized by fibroblast proliferation with an indistinct border and calcification [1]. The tumors show invasive growth; therefore, they are often difficult to distinguish from malignant tumors [2]. Although calcification is a characteristic imaging finding, it is not easily identified at early stages in some cases [3,4]. Previous studies revealed that MRI is useful to evaluate CAF for differentiating from malignant tumors [3,5]. Here, we report a case of CAF that occurred on the dorsum of the right foot of a 17-month-old girl, whom we were able to follow-up with imaging for 2 years. We discuss chronological changes of image findings of CAF and novel MRI findings which have never been described previously.

Case report

A 17-month-old girl was referred to our hospital for examination of a mass on her right ankle. There was no remarkable past medical or family history. The swelling on the medial...
right ankle had been observed since the age of approximately 10 months. Physical examination showed a soft tumor with a solid portion. Ultrasonography revealed a heterogeneous echogenic lesion of 2 cm with an indistinct border. Radiographs of the right ankle revealed the tumor with a slightly high density area (Fig. 1). MRI revealed a mass lesion measuring 22 × 10 mm with an indistinct and irregular border in front of the right ankle joint (Fig. 2). T1WI showed a lesion which was isointense to muscle. T2WI showed a heterogeneously high intensity. A needle biopsy was performed. Histologically, proliferation of fibroblast-like spindle cells was observed. There was no calcification; however, some cartilage-like tissue was found. Pathologic diagnosis was consistent with CAF. Since the tumor was benign and her parents did not wish for surgical resection, it was told to the parents that they should visit our hospital again when the tumor enlarged or any symptom appeared. At 3 years and 4 months of age, the tumor had grown significantly and hardened, increasing the limitation to the range of motion of the right ankle joint. Therefore, the patients visited our orthopedic surgery department again. Radiograph and unenhanced CT revealed the right foot lesion containing calcification (Fig. 3). On CT and MRI, tumor size was enlarged to 25 × 16 mm. The signal intensities of the background in the tumor were not changed on T1WI and T2WI. However, hyperintense nodules newly appeared on T1WI and T2WI in the tumors (Fig. 3). Because these hyperintensities of the nodule decreased on short tau inversion recovery imaging, we considered that these nodules contained fat. These fatty areas existed in the calcified nodules on CT (Fig. 4).

The tumor was resected. Microscopically, specimens were contained cartilage tissue and fibroblasts (Fig. 5), and pathologically, diagnosis was confirmed as CAF. Moreover, tumor had coarse calcified nodules which contained fatty component. We considered that these fatty component of calcification was identical to high intensity area of T1WI and T2WI. Follow-up observation was performed, during which no recurrence was observed.

Discussion

CAF is a rare invasive fibroblastic tumor that was first reported by Keasbey [1] in 1953 as juvenile aponeurotic fibroma. Although its prognosis is good, the recurrence rate is high following resection [6], and there have been 2 reported cases of malignant transformation [2]. Onset typically occurs in young infants, particularly boys, and often arises in the distal extremities and on the palm, fingers, and wrists [2]. Primary treatment is surgical resection, and histological evaluation is required for diagnosis. The tumor is histologically characterized by proliferation of fibroblasts with calcification [1].

Imaging findings include subcutaneous neoplastic tumors with indistinct borders, showing a tendency of invasion into the surrounding tissues and edematous changes. The tumor is located next to the fascia and tendon sheath. T1WI shows...
hypointense to isointense signals, and T2WI shows heterogeneous signals. Heterogeneous contrast enhancement is also seen [3,7,8].

Benign lesions that should be differentiated from CAF include giant cell tumors of the tendon sheath (GCTTS) and fibromatosis. On MRI, GCTTS shows lobulated margins with indistinct borders and heterogeneous hypointense signals associated with hemosiderin deposition [3,9]. These MRI findings can occasionally be found in CAF. However, GCTTS can be distinguished from CAF because calcifications in GCTTS are atypical and GCTTS rarely occur in children. Fibromatosis resembles CAF in the clinical course and image findings. Since fibromatosis also shows hypointensity on T2WI and has a distinct border and invasion to the surrounding tissue [4,9], it is sometimes difficult to distinguish this tumor from CAF. Malignant tumors arising on the ankles are rare, but synovial sarcoma and clear cell carcinoma should be differentiated from CAF. These tumors grow slowly and are therefore occasionally diagnosed as benign tumors. Synovial sarcoma is also associated with intratumoral bleeding [9,10].

Clear cell carcinoma dedifferentiates into melanocytes and therefore appears as a hyperintense signal on T1WI [3]. These findings are useful for the differential diagnosis of CAF.

CAF growth process is divided into two phases as follows. In the early phase, CAF is often not accompanied by calcification. In the latter phase, calcification and chondrocyte differentiation become clear. Although calcification is a characteristic imaging finding of CAF, it is often unnoticeable at the initial stage. Therefore, it is important to know this fact when radiologists interpret images [3,4]. In the present case, we observed a hyperintense area on T1WI and T2WI, suggesting the presence of fat within the calcified lesion. Pathologically, fat in the calcification was demonstrated, and it looked like the yellow bone marrow in the tumor. We considered that these fatty tissues represent the hyperintense area on T1WI and T2WI. To the best of our knowledge, it has not been reported previously that CAF contains these fatty signals in calcifications. In other entities such as myositis ossificans, it has been reported that fatty areas resembling the yellow bone marrow, appeared in calcified areas [11].

Fig. 3 — (A) Radiographs at 3 years and 4 months old revealed the tumor had more prominent calcification than those at 10 months old (arrow). (B and C) Axial and sagittal CT images at 3 years and 4 months old showed coarse calcification in the tumor on the anterior ankle.

Fig. 4 — MRI at 3 years and 4 months old. High intensity foci on both axial T1WI (A) and T2WI (B) appeared in the tumor (arrows). These fatty signals were suppressed in STIR image (C). Background intensities of the tumor were the same as those on MRI at 10 months old. STIR, short tau inversion recovery.
diffuse idiopathic skeletal hyperostosis and ossification of the posterior longitudinal ligament, fatty signals can be seen in calcifying lesions on MRI [12]. In CAF, we first reported this phenomenon. However, we considered this fatty area in the calcification can appear in a later phase of CAF. To clarify this fact, a further study is needed.

Because calcification is difficult to detect with MRI alone, CAF at a later phase containing calcification with fatty tissue can be misdiagnosed as other fatty tumors. Therefore, it is important to recognize existence of calcification using radiography or CT.

In conclusion, calcification of CAF does not appear during the early stage. Radiologists should be aware of this fact in differentiating CAF from other soft tissue tumors. On MRI in the later stage, fat-like signals can appear in the site of calcification.

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Fig. 5 – (A) Hematoxylin and eosin staining demonstrates cartilaginous metaplasia observed in the calcified areas (asterisk). The spindled fibroblastic component was around there (arrow). (B) Multinucleated osteoclast-type giant cells were seen around the calcified areas (arrow). (C) A fatty area (asterisk) which resembled yellow bone marrow was observed in the coarse calcified lesions (arrow).