Pediatric hand masses are often seen in outpatient department of the hospital. There are kinds of masses that can occur and it is important to discriminate between malignant and benign, since the treatment may be different. Calcifying aponeurotic fibroma (CAF) is a rare benign soft tissue in children and adolescents. It primarily occurs on the distal portion of the extremities, typically fingers, palms of the hands and plantar aspects of the feet. The recommended treatment of CAF is conservative local excision. Considering its benign characteristics and the rare metastasis, a strong and enough efforts should be made, whenever possible, to preserve the function of involved extremities, especially in child. In our report, we present a case of a 4-year-old girl with a CAF found near the course of motor branch of ulnar nerve at the palm and being performed surgical removal successfully with preserving the nerves.

**Key Words:** Hand, Pediatrics, Neoplasms

Calcifying aponeurotic fibroma (CAF) is a rare benign soft tissue in children and adolescents. It primarily occurs on the distal portion of the extremities, typically fingers, palms of the hands and plantar aspects of the feet. This mass appears slow-growing and ill-defined lesions related to tendons and aponeuroses in the body. It also tends to infiltrate the surrounding tissues such as surrounding fat, muscle tissue and rarely the bone. CAF has a high rate of local recurrence after surgical...
treatment with well-characterized histopathological features. These features include the presence of proliferative fibroblasts, chondrocytes and foci of calcification.

Malignant transformation of the tumor is very rare but not at all. Lafferty et al. reported a case of a CAF with metastatic fibrosarcoma of the lungs and the bones. In consideration of the high local recurrence and malignant transformation of residual tumor, surgical excision of CAF is recommended. However, considering its benign characteristics and the rare metastasis, a strong and enough efforts should be made, whenever possible, to preserve the function of involved extremities, especially in child. In our report, we present a case of a 4-year-old girl with a CAF found near the course of motor branch of ulnar nerve at the palm and being performed surgical removal with no recurrence 2 years after the operation.

CASE REPORT

A 4-year-old girl presented with a palpable mass on palm of the right hand. The patient hadn’t any medical and traumatic history. The family had recently noticed the mass. The physical examination showed an indolent, slightly movable and poorly circumscribed mass in the palm of her right hand (Fig. 1). The overlying skin didn’t show a dimpling sign or fixation to mass. The motor and sensory functions of the hand were normal. For more accurate diagnosis, ultrasonography and X-ray was performed. The ultrasonography finding showed a 0.5 cm sized isoechoic nodule with dark signal intensity in peripheral portion and poor vascularity, but not calcification (Fig. 2). There was no specific findings and bony abnormality in X-ray (Fig. 3). Although, there was no pain, surgical excision was performed because of the discomfort of daily life and the appearance of slow growth. Because mass was located near the course of branch of ulnar nerve at the palm, we considered that a fine surgical approach under microscope was needed.

Under general anesthesia, a transverse incision was
performed. Meticulous surgical approach was done with microscope. Mass was located and attached at the palmaris brevis muscle fascia and palmar aponeurosis. We excised the tumor carefully with no injury of surrounding structure. Macroscopically, it was a yellowish 15×10 mm mass with areas of speckled calcification (Fig. 4).

The microscopic examination with Hematoxylin and Eosin stain (Fig. 5) revealed an ill-demarcated tumor in dermis and subcutaneous tissue, which infiltrates surrounding fat, muscle and fibrous tissue. Variable-sized calcification were multifocally found within the lesion. The tumor was composed of bland-looking fibroblasts with round to oval nuclei and intervening collagen lay-down.

Two years after the operation, the patient remains asymptomatic and no signs of recurrence have been noted (Fig. 6).

**DISCUSSION**

Pediatric hand masses are often seen in outpatient department of the hospital. There are kinds of masses that can occur and it is important to discriminate between malignant and benign, since the treatment may be different. Among them, CAF which is benign fibrosing tumors in pediatric population consists of a tumor of differentiated fibroblasts, which follow an infiltrative pattern of growth. Since the first report by Keasbey in 1953, less...
than 100 cases of CAF have been reported in the literature. It usually occurs on the distal portion of the extremities, typically fingers, palms of the hands and plantar aspects of the feet. This mass appears slow-growing and ill-defined lesions related to tendons and aponeuroses in the body.

Histologic examination with surgical biopsy is necessary for diagnosis. Microscopical features of CAF are characteristic. Those features include foci of calcification and chondroid areas surrounding by fibroblast. Radiologic examination including X-ray and computer tomography may be helpful in diagnosis of CAF, but are not necessary. The presence of a soft tissue mass with presence of foci of calcification due to mineralization of the lesion can be found at the X-ray. Magnetic resonance imaging (MRI) can be useful at preoperative differential diagnosis of CAF from other fibrous tumors. Unlike other recent reports about CAF, our case does not show any suspicious radiographic findings of CAF. So we also initially suspected benign soft tissue tumor such as dermatofibroma.

If the physicians can’t rule out non-calcified CAF as in our case or the case reported by Kwak et al., they can take MRI. MRI findings of the mass show the lower signal intensity than that of muscle on T1WI and T2WI with intense heterogeneous enhancement after gadolinium administration. The possibilities of local recurrence are relatively high and these may raise concern. Some studies reported that more than 50% of tumors are recurred after the surgical removal. According to some reports, these CAFs can be classified into two types with a histologic pattern. Among them, the type, which is more infiltrative and lacks calcification, which typically occurs in younger children has a higher recurrence rate.

In usual case of benign tumors of the hand, if there are no symptoms affecting the daily life, they will be monitored and resected if necessary. CAF is a rare benign tumor that has never exceeded 100 cases since it was first reported. Thus, It is difficult to consider CAF when the patient visited the hospital with mass of the palm and it is easy to be mistaken for general benign tumor such as dermatofibroma. Because CAF tends to infiltrate the surrounding tissues such as surrounding fat, muscle tissue and rarely the bone and can be located deep layer of the hand, there may have possibility of the injury of the neurovascular structures during the operation without concerns.

The recommended treatment of CAF is conservative local excision. Lafferty et al. reported a case of a CAF with metastatic fibrosarcoma of the lungs and the bones. So, in consideration of the high local recurrence and malignant transformation of residual tumor, radical excision of CAF can be considered. However, several different studies have reported that radical excision should be avoided as the natural course could be benign. Therefore, considering its benign characteristics and the rare metastasis, a strong and enough efforts should be made, whenever possible, to preserve the function of involved extremities, especially in child. An important point is that the neurovascular bundles should be preserved even if they are found to be invaded by the tumor. If CAF occurs at the fingers or toes, mass should be resected incompletely that do not cause functional problem, not amputation. A report in the past shows extreme cases of misdiagnosis of CAF have led to amputation.

Considering CAF when child with hand mass visited hospital and the awareness of the natural history of mass can provide clues for approach and avoid the need for aggressive treatment.

CONFLICTS OF INTEREST
The authors have nothing to disclose.

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소아 수부 위험 부위의 석회화 건막 섬유종

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소아 수부의 종양은 병원 외래에서 흔하게 볼 수 있는 환자군이다. 발생할 수 있는 소아 수부의 종양 종류는 다양하며, 이 종류에 따라 치료 방법이 달라질 수 있기 때문에 양성 또는 악성 가능성을 감별하는 것이 중요하다. 석회화 건막 섬유종은 소아 수부에 드물게 발생하는 종양으로 일반적으로 사지 끝에 주로 발생한다. 석회화 건막 섬유종의 치료는 절제 및 재발률이 높고 일반적으로 양성의 성향을 보여 근치 절제술보다는 국소 보존적인 절제술이 필요하다. 특히 이 종양은 근막, 힘줄 또는 뼈까지 침윤되는 경우가 있어 수술 시행 시 주변 구조물에 손상을 입히지 않도록 하는 것이 중요하다. 본 증례에서는 4살의 어린 환아가 손바닥의 위험 부위에 종양이 있어 내원하였고 국소 재발 및 신경 등의 주변 구조물의 손상 없이 성공적으로 수술을 시행하였으며 발생률이 매우 낮은 석회화 건막 섬유종이 나와 이를 보고하고자 한다.

색인단어: 수부, 소아, 종양

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