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석회화 건막 섬유종
- 증례 보고 -

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목적: 석회화 건막 섬유종은 소아 및 청소년층에서 수장부를 가장 많이 침범하는 양성이나 공격적 성향을 띈 섬유성 종양이다. 저자들은 최근에 15세 소년의 수부에 발생한 석회화 건막 섬유종의 치료를 경험하였기에 이를 문헌 고찰과 함께 보고하고자 한다.

증례: 15세 소년이 좌수의 소지구 부위에 재발하는 종물을 주소로 내원하였으며 이 소년은 7년전에 타 병원에서 국소 마취 하에 동일 부위에 발생한 종물을 제거한 기왕력을 가지고 있었다. 종물은 단단하였으며 압통이 있었다. 단순 방사선 촬영상에는 종물 내에 다소성으로 산재된 미세 석회 침착이 보였다. 수술 소견상 종물은 주변과 명확히 구분되지 않았으며 단단하고 옅은 회색빛의 석회화를 동반한 침습성의 결절성 형태를 보였으며 가이언 관내의 척골 신경과 척골 동맥이 바로 인접하고 있었다. 수술 후 제거한 종물을 현미경적으로 관찰한 결과 종물은 많은 석회 침착이 되어 있으면서 이형성 연골양 분화를 보이는 조직이 중간 중간에 섞여 있는 빽빽한 섬유 조직의 증식으로 이루어져 있었고 주변부는 명확히 경계 지어지지 않으며 분엽상을 보이고 있었다.

결론: 이 질환에 있어서 가장 바람직한 치료법은 기능적인 면과 주요 조직을 보존할 수 있는 한도내에서 광범위 절제를 시행하는 것이며 이렇게 치료하더라도 많게는 50%이상의 재발율이 보고되고 있는 실정이다.

색인단어: 석회화 건막 섬유종, 수부, 재발율

INTRODUCTION

Calcifying aponeurotic fibroma is a benign, aggressive fibrous tumor seen in childhood or adolescence that most commonly affects the palm first described by Keasbey[8] in 1953. It is a slow-growing tumor with a predilection for the distal portions of the extremities, and with a tendency to recur at least partially due to its infiltrative nature[1,12]. We hereby present the case of a recurrent calcifying aponeurotic fibroma that occurred in the hand of a fifteen-year-old boy with

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the review of the literature.

CASE REPORT

A fifteen-year-old boy with history of local excision seven years ago and recurrence one year after was presented with a painless, hard and movable mass on the hypothenar area of the left hand. On examination, the tumor was measured approximately at 2 cm in diameter, movable, tender and hard to palpate. Radiographs did not demonstrate any skeletal involvement. At surgery, the mass showed ill-defined, firm, gray-white infiltrative nodular feature with calcifications as small white flecks and ulnar nerve and artery in Guyon's canal were closely adherent to the mass (Fig. 1). In our supposition, similar findings were probably seen in first operation 7 years ago, but they were failed in wide excision due to anxiety about damage to surrounding neurovascular structures. We were dissected the mass widely out with use of 4.5 times loupe magnification, and the ulnar nerve and artery were not compromised. Microscopy showed a fibrous nodular growth that extended with multiple processes into the surrounding tissue with more centrally located foci of calcification and cartilage formation. The tumor

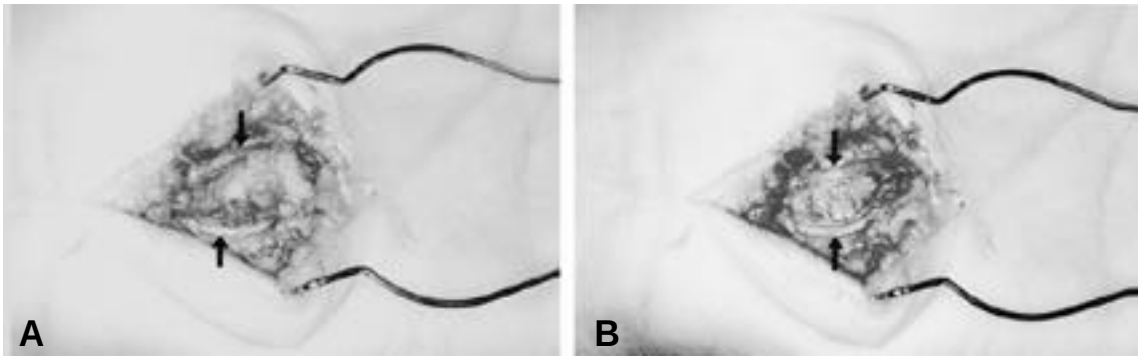

Fig. 1. The mass in hypothenar area of the left hand. (**A**) ulnar nerve and artery in Guyon's canal closely adherent to the mass were seen. Arrow above indicate the ulnar artery and below indicate the ulnar nerve. (**B**) After careful excision, the ulnar nerve and artery visible definitely.

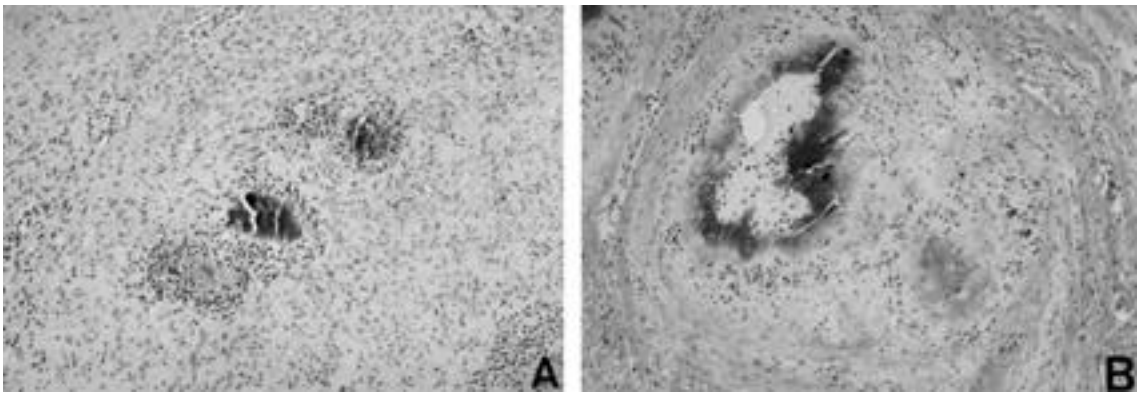

Fig. 2. The histologic sections show poorly defined fascicles of fibroblasts extended from the centrally located calcified areas (**A**) (H-E stain, ×40) and vague nodules of cartilaginous metaplasia with central calcifications (**B**) (×100).

was composed of small regular spindle cells with indistinct cytoplasm embedded within a dense collagenous stroma. Many nodules contained central foci of calcification bordered by areas showing cartilaginous differentiation. The tumor periphery showed infiltrating tongues of fibroblasts between adipocytes, muscles, and tendons (Fig. 2). A pathologic diagnosis of calcifying aponeurotic fibroma was made. Postoperatively, the patient did well, and the lesion had not recurred during one year follow-up period.

DISCUSSION

Calcifying aponeurotic fibroma is a very uncommon fibroblastic lesion that typically occurs in the hands and the feet of children and of young adults[4]. In 1953, Keasbey[8] first described calcifying juvenile aponeurotic fibroma in four children. Keasbey and Fanse-lau[9] changed the term to aponeurotic fibroma in 1961, after it became apparent that the condition was not limited to children. In 1973, Iwasaki and Enjoji[7] used the term calcifying aponeurotic fibroma, which is the current usage endorsed by Enzinger and Weiss[4].

This tumor characteristically presents as a slowly growing, painless, and soft tissue mass. However, Allen and Enzinger[1] noted two cases with mild pain and tenderness, and Keller and Baez-Giangreco[10] reported three cases in which major pain was a presenting complaint. The tumor was initially described in the palms, fingers, and soles of the feet of cjildren. Enzinger and Weiss[4] reported that 77% of patients had a tumor in the hand and 13% had a tumor in the foot. The age at presentation has been reported to range from birth to 67 years of age, with a median age of 12[1,5,6,12]. It is more common in male, with male to female ratio of approximately 2:1, and tends to recur locally with the time interval for recurrent disease ranging from 6 months to 23 years[5,12,14]. There is no evidence of any increased familial prevalence.

The etiology and pathogenesis are unresolved, but it is thought to represent certain type of a reactive process of connective tissue[13]. Fetsch and Miettinen[5] suggested an origin from fibroblastic cells attempting to differentiate into dense regular connective tissue and also into fibrocartilage and hyaline cartilage.

Most lesions are ill-defined, firm or rubbery, and gray-white; most are less than 3 cm in diameter at most. Portions of the surrounding fat, skeletal muscle, and fibrous tissue frequently merge with the tumor. Calcifications are occasionally evident as small white flecks. Radiogrphs usually demonstrate the calcification[4,11].

The microscopy reveals a fibrous growth that extends with multiple processes into the surrounding tissue. Fibrous tumor tissue extended from the main lesion may infiltrate muscles, tendons, nerves, or adipose tissues. Calcifying apneurotic fibroma presents multiple cellular nodules composed of fibroblasts surrounding the centrally located areas of calcification and the nests of cartilage. The cellularity varies from region to region. The tumor cells have plump, oval nuclei with vesicular chromatin and indistinctly outlined cytoplasm separated by a densely collagenous stroma. Calcification and cartilage formation are much more pronounced in lesions removed from older children and young adults. The calcifications are usually small and vary from fine granules or string-like deposits to large amorphous masses. These calcified foci are sur-

rounded by radiating columns of cells that resemble chondrocytes, with rounded nuclei lying in lacunae. Multinucleated giant cells resembling osteoclasts are occasionally present adjacent to the calcific foci[4,11].

The differential diagnosis for calcifying aponeurotic fibroma includes the following: infantile and juvenile forms of fibromatosis, fibrous hamartoma of infancy, the monophasic fibrous subtype of synovial sarcoma, and chondroma of soft parts[5].

Herein we report a case of typical calcifying aponeurotic fibroma with recurrence in a fifteen-year-old boy. The recommended treatment for capcifying aponeurotic fibroma is wide excision allowing for sparing of functional or vital structures, but as this case if the mass is closely adherent to surrounding neurovascular structures, wide excision may be impossible. So, the recurrence rate has been reported to be more than 50%[3] Because of its infiltrative nature as this case. Also, there are two case reports of malignant transformation of a calcifying aponeurotic fibroma in the literature and radical surgery may be required in those very rare cases that display evidence of malignant transformation[2]. So, closed follow-up should be recommended after surgical management due to its high recurrence rate and possibility of malignant transformation.

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Abstract

Calcifying Aponeurotic Fibroma
- A Case Report -

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Calcifying aponeurotic fibroma is a benign, aggressive fibrous tumor seen in childhood or adolescence that most commonly affects the palm. We report our experience in treating calcifying aponeurotic fibroma in the hand of fifteen-year-old boy. He was presented with a recurrent mass in hypothenar area of his left hand. He had a history of local excision of the mass 7 years ago in local clinic. The palpable mass was hard, tender and movable. The simple X-ray revealed multifocally scattered microcalcifications in the mass. The excised mass showed the features of dense fibrotic soft tissue tumor with multiple small whitish calcifications. The microscopic sections showed a lobulated and a poorly circumscribed proliferation of dense fibrous tissue, containing islands of metaplastic chondroid differentiation with prominent calcification. The recommended treatment of this lesion is wide excision allowing for sparing of functional or vital structures, but high recurrence rate as this case has been reported to be more than 50 percent.

Key Words: Calcifying aponeurotic fibroma, Hand, Recurrence

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