

Calcifying Aponeurotic Fibroma of the Elbow – A Case Report –

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Calcifying aponeurotic fibroma is a rare soft tissue tumor that mostly occurs in the distal extremities of children and adolescents. We report here on a case of calcifying aponeurotic fibroma of the right elbow in an 8-year-old boy, and the tumor was diagnosed by surgical excision. The patient complained of painless swelling and mild limitation of the range of motion of the elbow joint. Radiologically, the mass was ill-defined and showed stippled calcification with shallow bony erosion. Microscopically, the tumor was composed of spindle cells with nodular deposits of hyalination and calcification, and these deposits were surrounded by palisading polygonal plump cells. Immunohistochemically, the tumor showed a diffuse positive expression for CD99 and negativity for smooth muscle actin, S-100 protein and CD34. The patient has been well with no signs of recurrence during the 42 months after surgery.

Key Words : Fibroma; Soft tissue neoplasm

Calcifying aponeurotic fibroma is a rare soft tissue tumor that was first described by Keasbey in 1953 as juvenile aponeurotic fibroma.¹ This tumor is a slow growing, painless palpable mass and generally detected on the palms and the soles of young children and adolescents under 20 years of age. This tumor usually infiltrates into the surrounding fascia or muscle, and has a tendency to recur after surgical resection. To the best of our knowledge, six cases of calcifying aponeurotic fibroma have been reported in the Korean medical literature²⁻⁷ and there has been no reported case in which the tumor occurred at the elbow. We report here on a case of calcifying aponeurotic fibroma that occurred at the right elbow of an 8-year-old boy and we review the relevant medical literature.

CASE REPORT

An 8-year-old boy without a history of any trauma presented with a palpable mass on the right elbow, and this mass had been

incidentally detected the previous month. The mass was painless except for the mild discomfort when putting direct pressure on it. The patient denied any local neurological symptoms and he was otherwise systemically well: he had no fever or other constitutional features. The physical examination showed a firm mass with equivocal swelling and tenderness, and the mass measured 2 × 2 cm on the anteromedial aspect of the right elbow. The overlying skin was unremarkable and nonadherent without dimpling. Mild limitation of movement in the right elbow joint was noted. The plain radiographs of the elbow showed a punctated calcified soft tissue mass in front of the medial epicondyle in the right elbow with probable smooth margined bony erosion (Fig. 1). The CT scan and three dimensional reformed images confirmed a calcified mass with bony erosion in the right elbow (Fig. 2). MRI revealed that the mass showed intermediate signal intensity on the T1-weighted images and high signal intensity on the T2-weighted fat-suppressed images, and a heterogeneously well-enhanced mass was seen on the gadolinium enhanced fat-suppressed T1-weighted coronal MRI (Fig. 3). The patient was



Fig. 1. Plain radiograph shows a punctated calcified mass (black arrows) with smooth marginated bony erosion (white arrow) in front of the medial epicondyle in right elbow on lateral view.



Fig. 2. Three dimensional reformed CT image shows a punctated calcified mass with bony erosion (arrows).

operated on under general anesthesia and the mass was near-totally excised except for an intramuscular portion. At surgery, the mass was poorly demarcated and it had infiltrated into the surrounding muscle. The microscopic sections revealed a proliferation of spindle cells with nodular deposits of hyalination, calcification and chondroid differentiation that were surrounded by palisading of polygonal plump cells (Fig. 4) and the tumor had infiltrated into the adjacent fatty tissue and skeletal muscle (Fig.

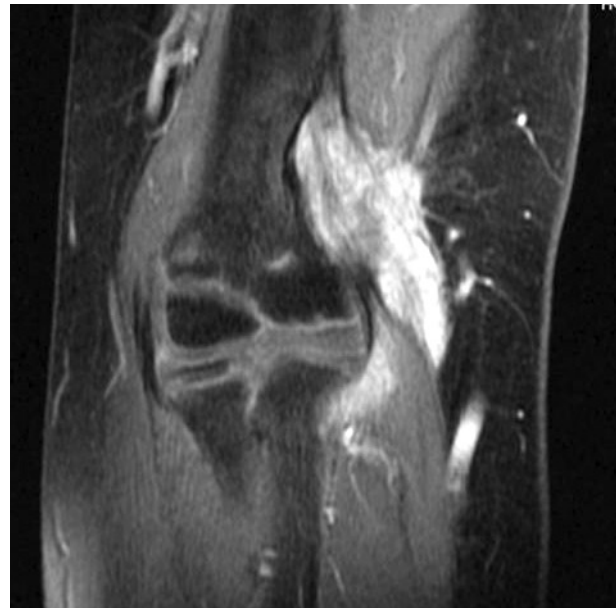


Fig. 3. Gadolinium enhanced fat-suppressed T1-weighted coronal MR image shows a soft tissue mass with heterogeneous well enhancement and cortical bony erosion in the anteromedial aspect of the right elbow.

5). Any mitotic figures were not seen. Immunohistochemically, the mass showed a diffuse positive expression for CD99 (1:1,000, Zymed, San Francisco, CA, USA), and negativity for smooth muscle actin (1:1,000, Neomarker, Fremont, CA, USA), S-100 protein (1:1,000, Neomarker, Fremont, CA, USA) and CD34 (1:1,000, Neomarker, Fremont, CA, USA). The final pathologic diagnosis was calcifying aponeurotic fibroma. The patient has been well with no signs of recurrence during the 42 months after surgery.

DISCUSSION

Calcifying aponeurotic fibroma was first defined by Keasbey¹ in 1953 as a neoplasm that's characterized by a proliferation of fibroblasts, areas of calcification and chondroid differentiation. Because most of the patients were children and young adolescents, it was named juvenile aponeurotic fibroma. Then in 1961, Keasbey and Fanselau found that the tumor presented not only in children, but in adults too, so they called the tumor aponeurotic fibroma. After that, Lichtenstein and Goldman in 1964 defined a cartilage analogue of fibromatosis. At last, Iwasaki and Enjoji in 1973 named these tumors as calcifying aponeurotic fibroma.^{8,9}

Although calcifying aponeurotic fibroma usually appears in

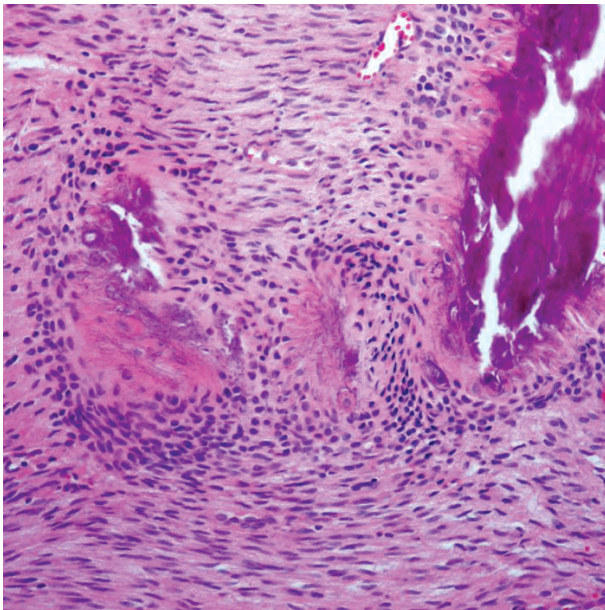


Fig. 4. The tumor is composed of fibroblastic cells with nodular deposits of calcification and chondroid differentiation that surrounded by palisading of polygonal plump cells.

Table 1. Clinicopathologic characteristics of Korean cases

Case	Sex/age	Site	Location	Size	No. of reference
1	F/36	Finger (volar side)	Superficial	1.5 × 1.0 cm	2
2	M/14	Chin	Superficial	2.5 × 2.5 cm	3
3	F/22	Toe (plantar side)	Superficial	2.0 × 1.5 cm	4
4	M/14	thumb (ulnar side)	Superficial	2.0 × 1.5 cm	5
5	F/24	Finger (tip)	Superficial	0.5 × 0.5 cm	6
6	M/11	Foot (plantar side)	Superficial	1.5 × 1.5 cm	7
7	M/8	Elbow (anteromedial side)	Deep	2.0 × 2.0 cm	Our case

the first decade of life, it can be detected from the time of birth to 62 years of age.^{10,11} The peak incidence is between 8 and 14 years of age.¹⁰ Most of the small published series have not reported a distinct gender predilection, but 70% of the patients in the series by Allen and Enzinger were male.¹² There is no evidence of a familial or racial prevalence.¹² The pathogenesis of calcifying aponeurotic fibroma is still unknown, but it is thought to be a tumor with a fibroblastic origin.¹⁰ This tumor was first reported on the palms, fingers and plantar region of children,¹ but isolated tumors have been reported at other sites, including the neck, forearm, elbow, upper arm, thigh, popliteal fossa, knee, lumbosacral region, scalp, chin, and abdominal wall.^{3,9,12-15} Local recurrence is common because of its infiltrative growth pattern and it often

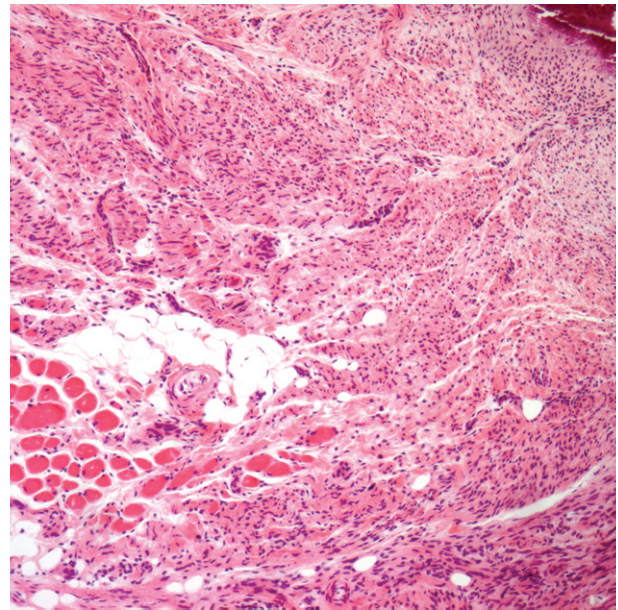


Fig. 5. The tumor cells infiltrate to adjacent fatty tissue and skeletal muscle.

has a sensitive location that mandates performing conservative surgery. Very few cases of malignant transformation have been reported.¹⁶ On physical examination, calcifying aponeurotic fibroma presents as a firm, palpable mass that has shown slow growth for a few months to a few years.^{10,11} There is frequently no history of trauma or local irritant factors. Although most of these tumors have presented as a painless nodule, a few cases have been reported with the patients complaining of significant pain.⁹

The cases in the Korean literature and our case are summarized in Table 1 and these cases have shown no gender predilection. The mean patient age (range: 8 to 36 years) of these Korean cases was 18.4 years, which was somewhat higher than that of the other previous reports.¹⁰ Most of the Korean cases occurred on fingers and toes like that of the other previous papers. The size of the masses was always under 2.5 cm at the greatest dimension and the masses were almost all painless palpable masses except for case 5. All the Korean cases were located in superficial soft tissue except for our case.

Radiologically, calcifying aponeurotic fibroma may show a soft tissue mass with fine stippling calcification. Most of the cases are not associated with osseous lesion. However, in extremely rare cases, occasional scalloping of the cortex and thickening of the bone have been reported. Shallow bony erosion was noted in our case, and this required us to differentiate this mass from other tumors including malignant tumors.

Histologically, there has been some reports about the phases of calcifying aponeurotic fibroma. Keasbey and Fanslau have

defined that there are two phases during the growth of this tumor.⁸⁻¹⁰ In the first phase, the lesion is a destructive infiltrative tumor and calcification is usually absent. This phase is named the diffuse or florid phase. In the other phase, the lesion forms a nodular mass with a well-defined border. Therefore, this phase is named the compact or demarcated phase. This phase shows calcification and cartilage formation. The diffuse phase is mostly seen in infants and young children, and the compact phase tends to be seen in adolescents. For this reason, Keasbey has proposed that the compact phase is formed by fusion and growth of the diffuse phase, and the histologic differential diagnosis differs from case to case depending on the age of the patient at the time the lesion is excised. In infants and small children, infantile fibromatosis, palmar and plantar fibromatoses and monophasic fibrous synovial sarcoma should be considered when making the differential diagnosis. In older patients, soft part chondroma may rarely be mistaken for calcifying aponeurotic fibroma. In our case, the fibroblastic proliferation was predominant, but calcification and cartilage formation were relatively scant. From these findings, we think that our case falls into the diffuse phase. We also reviewed the microscopic findings of four cases (cases 2 to 5) that were reported in the Korean literature. Case 3 and 5 showed scant fibroblastic proliferation and abundant cartilage formation and calcification. However, cases 2 and 4 revealed relatively abundant fibroblastic proliferation and scant cartilage formation and calcification. These findings seem to corroborate that this tumor shows different morphology according to age.

To eliminate a possible misdiagnosis such as sarcoma which might require amputation, it is important to make an accurate diagnosis of calcifying aponeurotic fibroma, and especially for the case with equivocal radiologic findings showing osseous involvement like our case.

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