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Angiofibroma of soft tissue in the subcutis: A radiologic-pathologic study of three cases

Ikuo Kudawara

ABSTRACT

Introduction: Angiofibroma of soft tissue (AFST) in the subcutis is a rare condition with few cases reported in the literature.

Case Series: Three cases of angiofibroma of soft tissue in the subcutis of extremities are presented. Two cases which received surgery have no recurrent tumor for three years and one year, respectively. The remaining case underwent only excisional biopsy and was lost to follow-up.

Conclusion: Radiological features of AFST were commonly well-circumscribed, round or oval configuration with homogeneous intermediate signal intensity on T1-weighted image on magnetic resonance imaging (MRI). The tumors were located in the subcutis of extremities and attached to the fascia or aponeurosis, but histologically no evidence of infiltration. The histopathological features were composed of oval or spindle cells with various degree of the cellularity, small vessels, and collagen fibers or myxoid matrix.

Keywords: Angiofibroma of soft tissue, Magnetic resonance imaging, Pathology, Radiology

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INTRODUCTION

Angiofibroma of soft tissue (AFST) is rare and benign fibrovascular tumor that occurs in the extremities [1–3]. It displays the karyotype with a recurrent translocation t (5;8) (p15;q13), resulting in the expression of *AHRR-NCOA2* fusion transcript [4]. Some authors reported cases of angiofibroma of soft tissue [5–15]. However, most articles primarily discuss histopathological findings of the tumor. Therefore, the clinical, radiological, and pathological characteristics of angiofibroma of soft tissue have not been well recognized. Herein, we report the features of radiological, pathological findings, and clinical course in three cases of angiofibroma of soft tissue in the subcutis of extremities.

CASE SERIES

Case 1

A 57-year-old man had three-year of a slowly growing painless mass in the right thigh. Magnetic resonance imaging (MRI) showed a mass in the subcutis and attached to the femoral fascia that had homogeneous intermediate intensity (slightly high intensity than that of the muscle) on T1-weighted image and homogeneous high intensity on T2-weighted image (Figure 1A and B). Axial diffusion-weighted imaging (DWI) showed hyperintense mass, and apparent diffusion coefficient (ADC) map also showed high signal intensity (Figure 1C and D). This case was initially diagnosed epithelioid hemangioendothelioma. We received a consultation regarding the definite diagnosis of the patient who received surgery for the tumor at the requesting hospital. Histologically, the tumor composed of oval shaped cells without nuclear atypia and abundant small vessels and fibrosis and myxoid change (Figure 2A). Immunohistochemically, the tumor cells

and vessels were positive for CD31 and CD34 (Figure 2B). Mib-1 (Ki-67) index was less than 1%. There was no tumor necrosis. From these findings, definite diagnosis of AFST was made. He was lost to follow-up after definitive histological diagnosis.

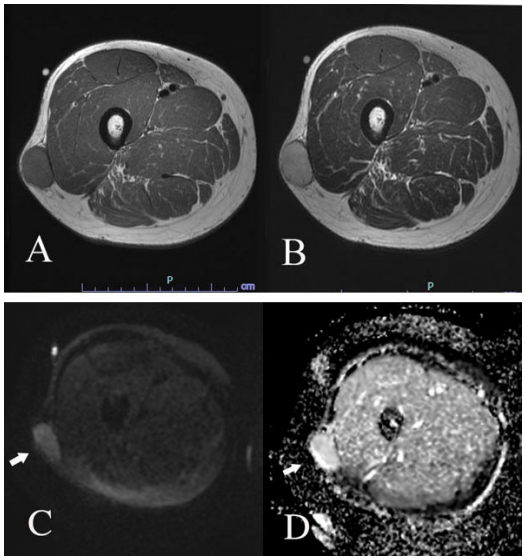

Figure 1: Case 1. Angiofibroma of soft tissue in the lateral thigh. Axial T1-weighted MRI (TR/TE=825/12.42) demonstrates a well-circumscribed subcutaneous mass adjacent to the femoral fascia with homogeneously slightly high signal intensity compared to the muscle (A). Corresponding axial T2-weighted MRI (TR/TE=4000/82.46) reveals a mass to be homogeneously high signal intensity (B). Axial diffusion-weighted imaging (DWI) (TR/TE=4000/61) shows hyper intense mass (C), and apparent diffusion coefficient (ADC) map also shows high signal intensity (D).

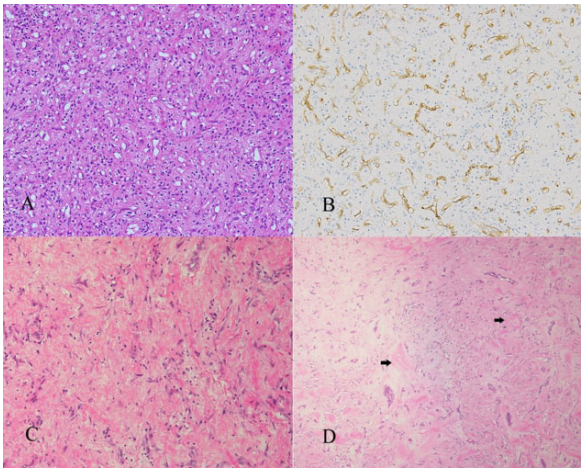

Figure 2: Microphotograph of three cases. Case 1. The tumor composed of oval cells with relatively high cellularity and abundant small vessels. H.E. 100 (A). Case 1. Immmunohistochemically, tumor cells and endothelial cells are positive for CD34 (B). Case 2. The tumor composed of oval shaped cells without nuclear atypia and abundant small vessels and fibrosis and myxoid change. The cellularity is low. H.E. × 100 (C). Case 3. The tumor composed of spindle cell without nuclear atypia and abundant various sized vessels with around hyalinization the cellularity of the tumor is medium. Note the hyalinization around the vessels (arrows). H.E. × 100 (D).

Case 2

A 52-year-old man had dysesthesia of left ring and little fingers of three-year duration and a soft tissue mass in the left palm of six months duration.

On physical examination, a rubbery mass with skin bulging in the left palm was palpable, which measured a size of 22 mm × 25 mm using vernier caliper. There were no changes of the skin including heat, red flare, ulcer, and pigmentation. Spontaneous local pain was not identified, but tenderness on the tumor was observed. MRI showed a well-circumscribed mass in the subcutis and attached to the palmar aponeurosis that had homogeneous intermediate intensity on T1-weighted image and inhomogeneous high intensity on T2-weighted image (Figure 3A and B). Clinical and radiological differential diagnoses include hemangioma, neurogenic tumor, synovial sarcoma, and clear cell sarcoma. Excisional biopsy was performed using air tourniquet under general anesthesia. The tumor was located within the subcutis. The tumor capsule was not identified and the border was unclear in place. Macroscopically, the tumor surface was whitish and fibrous appearance. The palmar branch of ulnar nerve was identified the bottom of the tumor. The nerve was gently detached from the tumor using blunt dissection.

Dysesthesia of the left ring was disappeared soon after surgery. Microscopically, the tumor had thin fibrous capsule but partially absent. The tumor composed of oval shaped cells without nuclear atypia and abundant small vessels and fibrosis and myxoid change (Figure 2C). The cellularity was medium. Immunohistochemically, the tumor cells and vessels were negative for CD31, CD34, S100, SMA, and ALK. Mib-1 (Ki-67) index was less than 5%. From these findings, histological diagnosis was AFST. There was no recurrent tumor three years after the surgery.

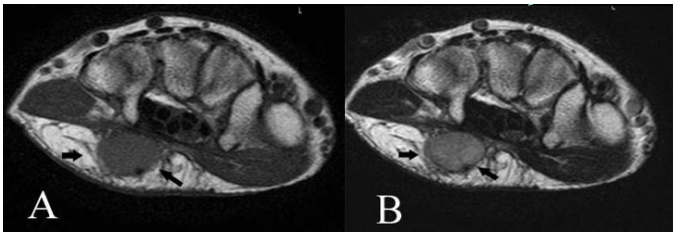

Figure 3: Case 2. Angiofibroma of soft tissue in the palm. Axial T1-weighted MRI (TR/TE=529.63/23) demonstrates a well-circumscribed subcutaneous mass adjacent to the palmer aponeurosis with homogeneously slightly high signal intensity (arrows) (A). Corresponding axial T2-weighted image (TR/TE=2652.90/83.85) reveals a mass to be inhomogeneously high signal intensity (arrows) (B).

Case 3

A 65-year-old woman had six-month of left buttock painless soft tissue mass. She underwent polypectomy for colon adenoma one year ago. On physical examination, a

rubbery and movable mass in the buttock was palpable, which measured a size of 17 mm × 17 mm using vernier caliper. Coronal and axial MRI showed a mass in the subcutis of buttock and attached to the gluteal fascia that had homogeneous intermediate intensity on T1-weighted image (Figure 4A and B). Corresponding axial T2-weighted image revealed a mass to be inhomogeneously high signal intensity (Figure 4C).

A percutaneous needle biopsy was performed under local anesthesia using 1% lidocaine. Histologically, the tumor is composed of spindle cells without nuclear atypia and abundant vessels with perivascular hyalinization (Figure 2D). Immunohistochemically, the tumor cells were negative for CD34, S100, SMA, and β-catenin. Mib-1 (Ki-67) index was less than 2%. The histological diagnosis was suggested AFST. At surgery, the tumor was resected with the cuff of surrounding fatty tissue and also with gluteal fascia since the bottom of tumor firmly attached to the underlying fascia. The definite diagnosis of AFST was made from surgical specimen. There was no recurrent tumor one year after the surgery.

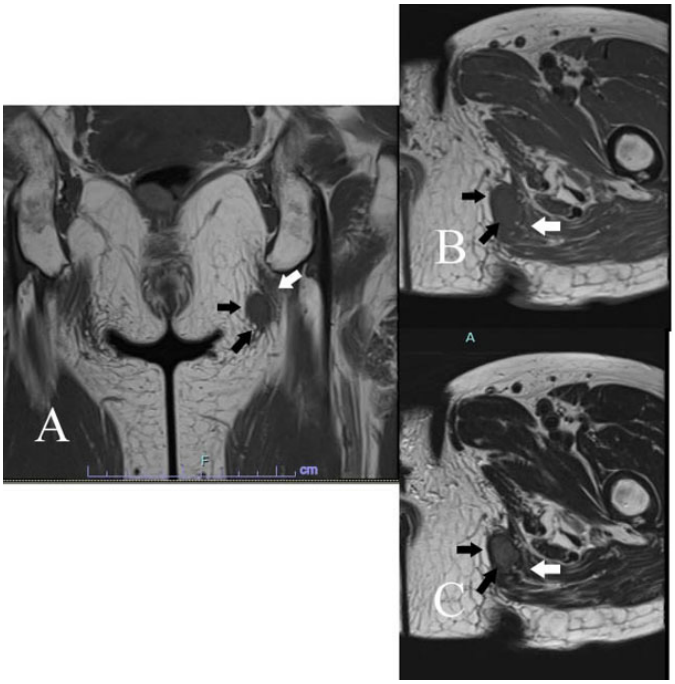

Figure 4: Case 3. Angiofibroma of soft tissue in the buttock coronal (A) and axial (B) MRI shows a mass in the subcutis of buttock (black arrows) and attached to the gluteal fascia (white arrow) that had homogeneous intermediate intensity on T1-weighted image (TR/TE=476/9.90). Corresponding axial T2-weighted image (TR/TE=3110/105) reveals a mass to be inhomogeneously high signal intensity (C).

DISCUSSION

Mariño-Enríquez and Fletcher first described 37 cases of AFST in 2012, as a benign fibroblastic tumor composed of spindle cells, vessels, and fibromyxoid matrix [3]. The tumors present most commonly as a slowly growing painless mass located in mainly lower extremity, often close to a joint, tendon, or fascia [1, 2, 16]. There are a limited number of published reports of AFST, the most of their authors were describing concern histopathology [5, 11–15]. Only five reports briefly discussed the MRI findings of this tumor [6–10]. Moreover, locations of all extremity AFST were deep-seated. There is little information available on radiological findings of AFST in the subcutis.

The common MRI features of the presented cases are round or oval shaped and well-circumscribed tumor located in the subcutis had homogeneous intermediate signal intensities on T1-weighted image and high signal intensities on T2-weighted image. Flow voids were not observed on MRI despite the presence of abundant vessels in the tumor. Although these findings are almost similar to the previous reports [6–10], we would designate that the tumors of small size (less than 5 cm of maximum diameter) in the subcutis and attached to the fascia or aponeurosis had slightly high signal intensity compared to the surrounding muscle on T1-weighted image. Radiological differential diagnosis of the superficial soft tissue tumors less than 5 mm in a diameter such as present cases were suggesting benign tumor includes vascular, neurogenic, myofibroblastic tumor, nodular fasciitis, and clear cell sarcoma [17, 18].

To determine histological diagnosis, a percutaneous needle biopsy possible or excisional biopsy is needed.

Case 2 was not always clinically typical this tumor compared to the previously reported cases. The tumor in palm was painful and the border was unclear at surgery. One possible explanation for above findings was continuous pressure to the palm in the daily life or his occupation of mechanics. Dysesthesia of the ring that may be due to neuropathy of palmar digital nerves from superficial branch of ulnar nerve was disappeared soon after tumor resection.

Most of fibroblastic or myofibroblastic tumors such as desmoid fibromatosis, desmoplastic fibroma, nodular fasciitis, proliferating myositis, proliferating fasciitis are not remarkably vascularized [2]. On the contrary, the characteristichistologicalfindingsofAFSTareproliferation of tumor spindle cells and distinct and complex vascular tissue. The complex vascular pattern showed small, thin-walled, branching vessels distributed throughout the lesion. Immunohistochemical study indicated tumor cells were positive for epithelial membrane antigen (EMA) in 44% cases and endothelial cell were positive for CD34 [1]. Yamada et al. discussed that estrogen receptor (ER) and CD 163 were useful markers to diagnose of AFST from detailed immunohistochemical study [13]. The histological differential diagnosis of AFST in the extremities includes solitary fibrous tumor (SFT), low-grade fibromyxosarcoma, low-grade myxofibrosarcoma, and myxoid liposarcoma. The most common misdiagnosis was SFT, which occurred in 5 of 13 cases [11]. STAT6 is relatively specific immunohistochemical marker for SFT, but one case of AFST had weakly positive for STAT6 [11].

Solitary fibrous tumor typically shows strong and diffuse nuclear staining of STAT 6 [19].

Eight of 13 cases in RT-PCR had *AHRR-NCOA2* fusion genes and 9 of 13 cases were NCOA2 rearrangement-positive in FISH analysis [11]. If *AHRR-NCOA2* fusion is identified by RT-PCR or FISH, it will be exclusively diagnosis of AFST [15, 16].

A standard surgical procedure for this tumor is unknown because of small size of reported cases [1, 11]. The local recurrence rate was reported of 14% after the marginal excision [1]. It seems to be relatively higher than those of other benign soft tissue tumor such as lipoma or neurogenic tumor. Recent reviews indicated that AFST sometimes shows infiltration growth pattern to the surrounding soft tissue [20, 21]. Wang et al. reported that minimal infiltration of tumor cells into adjacent soft tissue was observed in 3 of 8 cases [15]. The infiltrative growth pattern was not confirmed in our cases. Clinically, it would be safer to resect tumor with the cuff of the surrounding tissue in case of the tumor adhesion.

Further investigation of future reports of cases with AFST would clarify the more detailed clinical, pathological, and radiological features.

CONCLUSION

Radiological features of angiofibroma of soft tissue in the subcutis were commonly well-circumscribed, round, or oval configuration with homogeneous intermediate signal intensity (slightly high intensity than that of the muscle) on T1-weighted image, high signal intensity on T2-weighted image on MRI. The tumors were located in the subcutis of extremities and attached to the fascia or aponeurosis, but histologically no evidence of invasion of their structures by macroscopically and microscopically. The histopathological features were composed of oval or spindle cells with various degree of the cellularity, abundant small vessels, and interstitial collagen fibers or myxoid matrix. Two cases that received surgery have no recurrent tumor for three years and seven months, respectively.

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Author Contributions
Ikuo Kudawara – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest
Author declares no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.