Case Report

Angiomatoid Fibrous Histiocytoma with Pain in a Child

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Angiomatoid fibrous histiocytoma (AFH) is a rare soft tissue tumor with low-grade malignancy which occurs chiefly in children and young adults. Mostly, the tumor develops in the extremities or the trunk. The tumor has a relatively rare metastasis and its overall clinical outcome is excellent. The authors present a case of AFH of the subcutis on a left elbow of an 11-year-old boy. Clinically, the tumor appears as a cutaneous nodule with slow growth. Pain and adjacent lymphadenopathy are leading symptoms of the patient. Histological examination shows the classical morphological features of AFH with focal cellular atypia. The tumor has immunoreactivity to vimentin and CD68 but negative to CD34, desmin, CD117 and S-100 protein. Because of its rarity, we report a case with an uncommon clinical presentation in a Thai patient.

Keywords: *Angiomatoid fibrous histiocytoma, Fibrous histiocytoma, Angiomatoid tumor*

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Angiomatoid fibrous histiocytoma (AFH) is primarily a tumor in children or young adults, with the median age of manifestation of 13 years[1-3]. In most cases, it arises in the extremities (65%) as a slowly growing cystic mass of soft tissue. Other preferential sites of the tumor are the trunk (28%) and head and neck (7%)[4]. It normally manifests as a nodular subcutaneous growth that hardly causes any tenderness or pain. The exact pathogenesis of the tumor is still obscure and uncertain[3]. Enzinger presumed that AFH was a tumor of fibrohistiocytic origin judging from its spindle to round-shaped cells, histiocytes, its phagocytosis activity and positivity to CD68. However, a recent electron microscopic study found a mixture of fibrohistiocytic, myofibroblastic, and undifferentiated cells that contained cytoplasmic process and dense core granules. Immunohistochemical study was concordant with the electron microscopic finding and exhibits mixed histiocytic, myofibroblastic, epithelial, and neural reactivity[5]. The distinctive clinical feature, prognosis, and various expression of immunophenotype are totally in contrast with conventional malignant fibrous histiocytoma (MFH) which separate AFH into the intermediate malignancy group of fibrohistiocytic tumor[6,7]. The authors present a case of angiomatoid fibrous histiocytoma presenting with an unusual clinical presentation.

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Case Report

An 11- year-old boy presented with a painful mass on the medial aspect of his left elbow for approximately 4-5 months. The tumor progressively enlarged with pain. Physical examination revealed a well-circumscribed, mobile and soft mass of about 3.0x4.0 cm^2 with tenderness on the medial aspect of the left elbow. In the adjacent area an enlarged lymph node with smooth surface about 1.2 x 1.2 x 1.5 cm^3 was found. Magnetic resonance imaging showed a soft tissue mass of 2.0 x 3.0 cm^2 at the subcutis and two lymphadenopathies (Fig. 1). Other laboratory findings were unremarkable. The patient had no other associated systemic manifestation. Operative finding found a well-circumscribed soft tissue mass, approximately 3.0 cm at its widest diameter. The wide excision of the mass and its adjacent lymph nodes was done.

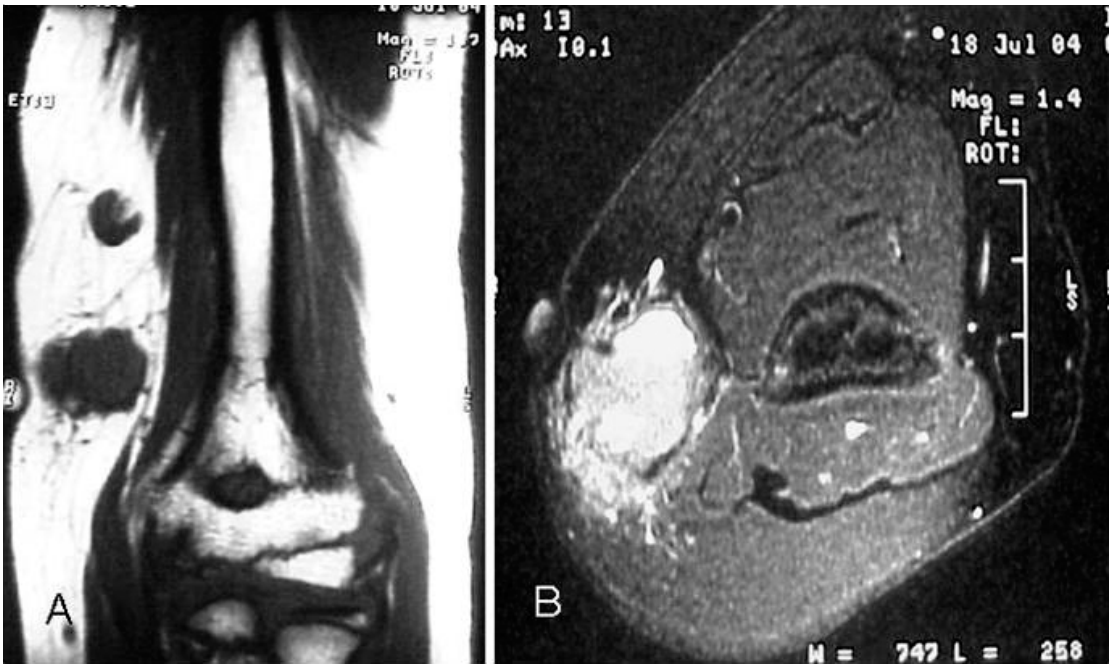

Fig. 1 A) Coronal T1WSE of elbow showed the mass (hypo-intense) located at medial aspect and one reactive hypo-intense lymph node enlarging above the mass. B) Axial T2WSE of elbow demonstrated high signal in the central portion and intermediate signal in peripheral irregular solid portion

Pathologic examination revealed a piece of yellow-brown tissue measuring 3.0x4.0x4.0 cm^3. Serial sections showed irregular bloody spaces, separated by solid grey white and yellow areas (Fig. 2). There was no evidence of infiltrative foci into the surrounding tissue. Adjacent lymph nodes were also received and measured 1.2 x 1.2 x 1.5 cm^3.

Histological examination of the tumor revealed a cystic mass surrounded by dense fibrous tissue and multiple irregular foci of lymphoid tissue. The central areas exhibited blood filled-spaces. The lining septae were composed of solid packed of histiocyte-like cells with multiple foci of hemorrhage (Fig. 3A). Most of the tumor cells had round to oval nuclei, faintly eosinophilic cytoplasm, and indistinct cell border as epithelioid and spindle cells (Fig. 3B). The more cellular areas showed large oval to polygonal-shaped cells with atypia (Fig. 3C). Another characteristic feature of the lesion was diffuse intra- or extra- cellular deposition of hemosiderin granules. The surrounding lymphocytic cuffs were composed of lymphocytes with occasional germinal centers formation. Mitotic figures were not common. Focal areas of large atypical cells with mitotic rate 1/10 HPF were detected. The submitted adjacent lymph node revealed reactive lymphoid hyperplasia.

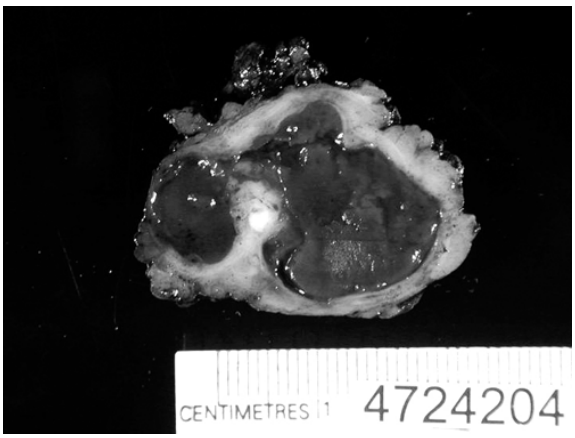

Fig. 2 Gross specimen of AFH showed irregular bloody spaces, separated by solid grey white and yellow areas

The immunohistochemical study exhibited reactivity to vimentin and CD68 (Fig. 3D) and negative to CD34, desmin, CD 117 and S-100 protein. The assembly of the clinical features, histological findings and immunohistochemical study, the final diagnosis is angiomatoid fibrous histiocytoma. At the time of writing (six months postoperatively), the patient remains well with no sign of recurrence.

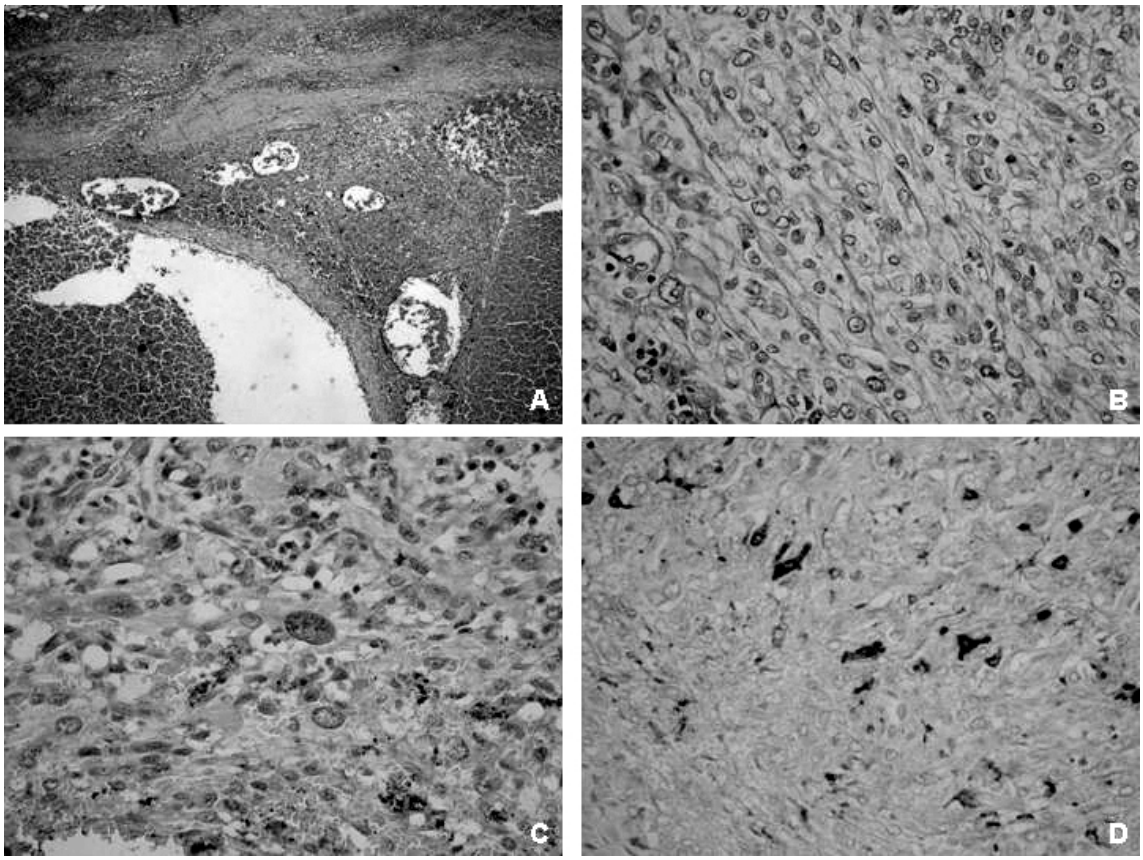

Fig. 3 A) The tumor revealed cystic spaces surrounded by dense fibrous tissue and multiple irregular foci of lymphoid tissue. Histiocyte-like cells were arranged in solid sheets. B) Spindle cell area in angiomatoid fibrous histiocytoma. C) Focal area of cellular atypia in AFH. D) CD68 immunoreactivity within AFH

Discussion

Soft tissue mass previously termed angiomatoid malignant fibrous histiocytoma was recently renamed by a committee of the World Health Organization as angiomatoid fibrous histiocytoma (AFH). Its common histological features include solid sheets of histiocyte-like cells, central hemorrhagic cyst-like spaces and surrounded by irregular aggregate of chronic inflammatory cells. The expression of central space is similar to vascular spaces, but they are not really lined by endothelial cells which are confirmed by its negative reaction to factor VIII and CD34[8]. However, the patient presented with both recent and old hemorrhagic areas that may explain its uncommon presentation such as pain and tenderness. The periphery of the mass is composed of pseudofibrous capsule admixed with chronic inflammatory cells including plasma cells and lymphoid tissue with numerous germinal centers formation giving this tumor a resemblance to a lymph node. This finding including the adjacent lymphadenopathy supports the possibility of a lesion which may arise from an area of normal lymphoid tissue or myoid cells of lymphoid tissue.

Immunohistochemical study is helpful in differential diagnosis[9]. The tumor is commonly positive for vimentin (100%), desmin (50% to 100%), and CD68 (47.4 to 100%)[5,6,10]. Hasegawa found other immunophenotypes expressions such as synaptophysin, alpha-smooth muscle actin, epithelial membrane antigen, neuron-specific enolase and CD99 (0-13)[5]. Most of the studies found that the tumor is uniformly negative to S-100 protein[11,12]. The result of immunohistochemical study in the presented case was consistent with its histiocytic differentiation and vascular tumor was definitely excluded.

The preoperative clinical impression from symptomatic subcutis mass is usually mistaken for lymphadenitis, hematoma or hemangioma. AFH seldom occurs in differential diagnosis. The authors report a case of a rare form of soft tissue sarcoma in children

which has distinctive clinical course and prognosis. The tumor typically occurs in young adults with the mean age of manifestation ranging from 10.5 to 17 years[5,13]. Pettinato et al reported patients with AFH ranged in age from 3 months to 42 years[14]. Most of the tumors arose in the extremities (65%), trunk (28%) and head and neck (7%). Tumor mass tended to have recurrence if it was incompletely excised (12%) and developed into local nodal metastasis (1%), and the patients could die from the disease (1%)[4]. Because of the rarity of its metastasis, this tumor is, therefore classified as low-grade sarcoma[2]. The histological feature in this case also showed cellular atypia. Nevertheless, the histological parameters such as mitotic figure rate and pleomorphism did not influence the final outcome. Costa et al found that the behavior of the tumor with local and distant metastasis correlated with its invasion into deep fascia or muscle. Complete surgical excision without adjuvant therapy was considered appropriate treatment[4]. However, radiotherapy after postsurgical recurrences was successful[14].

In the present case, tumor mass was a hemorrhagic cyst with focal cellular atypia combined with adjacent lymph node enlargement. The symptom of pain might be caused by hemorrhage or from the lymphoid reaction within and around the tumor mass.

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อาการปวดของเนื้องอก Angiomatoid fibrous histiocytoma ในเด็ก

พิริยา สุทธิเรืองวงศ์, วรนุช ธนากิจ, อภิชาต อัศวมงคลกุล

Angiomatoid fibrous histiocytoma เป็นมะเร็งของเนื้อเยื่อเกี่ยวพันที่มีความรุนแรงต่ำ และพบได้น้อย พบส่วนใหญ่ในเด็กและวัยรุ่น เนื้องอกชนิดนี้มักเกิดที่บริเวณแขนขา หรือลำตัว พบการแพร่กระจายไปที่อวัยวะอื่นได้น้อย และมีผลการรักษาที่ดีมาก ผู้เขียนได้รายงานผู้ป่วยโรคนี้ 1 ราย เป็นเด็กชายอายุ 11 ปีมีรอยโรคที่ข้อศอกซ้าย โดยเนื้องอกมีลักษณะเป็นก้อนที่บริเวณใต้ผิวหนัง โตค่อนข้างช้า อาการปวดและต่อมน้ำเหลืองข้างเคียงโตพบเป็นอาการแสดงของผู้ป่วย ผลตรวจทางพยาธิวิทยา พบลักษณะจำเพาะเจาะจงกับเนื้องอกชนิดนี้ และมีบางบริเวณที่มีเซลล์รูปร่างผิดปกติร่วมด้วย ผลการย้อมทางอิมมิวโนฮีสโตเค็มของเนื้องอก พบว่าเนื้อเยื่อย้อมติด vimentin, CD68 และย้อมไม่ติด CD34, desmin, CD117 และ S-100 เนื่องจากเนื้องอกชนิดนี้พบได้น้อย ทางผู้เขียนจึงได้รายงานผู้ป่วยที่มีอาการแสดงที่แตกต่างจากปกติในผู้ป่วยไทย