Angiomatoid fibrous histiocytoma: case report and review of the literature

Canten Tataroğlu[1], Nil Çulhacı[2], Emre Çeçen[3]

Departments of [1]Pathology and [2]Pediatrics, Adnan Menderes University Faculty of Medicine, Aydın, Turkey. E-mail: cantent@yahoo.com

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Angiomatoid fibrous histiocytoma is a rare soft tissue tumor of uncertain differentiation and low metastatic potential, which occurs predominantly in children and young adults. It occurs mostly within the extremities, trunk, head and neck. It can be associated with systemic manifestations such as anemia, pyrexia and malaise. Its morphology is distinct, with an outer shell of lymphoid tissue, sheets of dendritic-like tumor cells with bland nuclei and blood-filled cystic cavities. Herein, we present a case of angiomatoid fibrous histiocytoma with systemic symptoms before any mass was clinically detectable, arising in the scalp of a 10-year-old girl.

Key words: angiomatoid fibrous histiocytoma, scalp, systemic symptoms.

Angiomatoid fibrous histiocytoma (AFH) is a very rare soft tissue tumor that demonstrates features of both fibrohistiocytic and vascular tumors. It was first described by Enzinger in 1979 as a variant of malignant fibrous histiocytoma[1]. But recent studies have shown that the incidence of metastasis or local recurrence is very low in these tumors[2,3]. For these reasons, the World Health Organization has reclassified AFH as a tumor of uncertain differentiation and low metastatic potential[4,5]. It generally affects children and young adults. It most commonly occurs within the extremities, trunk, head and neck, but can also arise in unusual sites such as the lung, bone, cerebrum and mediastinum[6-9]. A small number of AFH cases recur locally, and rare cases have been known to metastasize[10]. Herein, we describe an AFH with systemic symptoms including abdominal pain and increased sedimentation rate and CRP levels.

Case Report

A 10-year-old girl presented to the pediatric clinic with abdominal pain. The initial examination was unremarkable. The tests showed hemoglobin of 11.2 g/dl, erythrocyte sedimentation rate of 100 mm/h and CRP of 9 mg/dl. Her symptoms initially suggested a genitourinary infection as a primary diagnosis. In addition, an asymptomatic subcutaneous skin mass was palpable on the occipital region of her head, which had first appeared as a small nodule 12 months before and had enlarged gradually. She had noted that the mass had doubled its initial size during this period. Physical examination revealed a firm, painless, nonfluctuant, immobile mass at the retroauricular region measuring 3.5x2 cm. The radiological presumptive diagnosis of the retroauricular mass was lymphadenopathy. A chest radiograph was normal. To confirm or rule out malignant lymphoma, we recommended excisional biopsy. The patient underwent surgery, and total excision of the mass with positive margins was performed. Grossly, the tumor was 3x1x1 cm in diameter and well circumscribed. The cut surface contained cystic, hemorrhagic and solid areas. Microscopic examination of the tissue revealed a fibrous pseudocapsule and a pericapsular lymphocytic infiltrate with a germinal center formation simulating a lymph node. The central portion of the tumor featured blood-filled cystic spaces (Fig. 1.), surrounded by spindle or ovoid cells with bland vesicular nuclei and moderate amounts of eosinophilic cytoplasm. Some of the cells were histiocytoid in appearance (Fig. 2.) Scattered enlarged and mildly pleomorphic

cells were present. Mitosis was infrequent, and necrosis was not found. Following surgery, her symptoms and acute phase reactants were improved. The patient has remained well and recurrence-free for three years.

Immunohistochemistry

The surgical material was fixed in formalin, embedded in paraffin and stained for hematoxylin-eosin. Immunohistochemically, the dendritic-appearing cells expressed desmin; some cells expressed EMA and, weakly, CD99 (Fig. 3.) The lymphoid infiltrate appeared to comprise a mixture of B and T cells (CD3 and CD20 positive). All the other markers were negative, and the ki-67 index was 3%.

Discussion

Angiomatoid fibrous histiocytoma is a rare, slow-growing tumor of the deep dermis and subcutis of children and young adults. It can be associated with systemic symptoms such as fever, anemia, weight loss, nausea, vomiting, fatigue, anorexia, anemia, paraproteinemia or vasculitis, thought to be due to cytokine production by the tumor[4,11-14]. The clinical symptoms disappear following removal of the lesion. Recently, Chen et al. showed that systemic symptoms were observed more frequently in AFH occurring outside the somatic soft tissues[12]. In many cases, systemic symptoms may cause confusion in the differential diagnosis. Therefore, detailed clinical examination is extremely important. After detection of the mass, the clinical symptoms disappear following excision. Histopathologically, the tumor is characterized by a thick fibrous pseudocapsule, a pericapsular lymphoplasmacytic infiltrate, pseudoangiomatoid spaces and a multinodular proliferation of eosinophilic, histiocytoid or myoid cells[4]. It must be examined carefully, because the surrounding lymphocytic infiltrate may closely mimic metastatic disease involving a lymph node[15,16]. Immunohistochemically, the presence of desmin-positive cells with dendritic morphology is characteristic. It is critical to remember that rare AFHs may show muscle actin expression, but they are never positive for myogenin and MyoD1, which are expressed in nearly all rhabdomyosarcomas (RMS)[15]. There is variable expression of EMA, CD68 and CD99[4,12]. Nevertheless, immunohistochemistry has a limited role in the diagnosis of AFH. Cytogenetically, two specific translocations, t(12:16)(q13:p11) and the more recently described t(12:22)

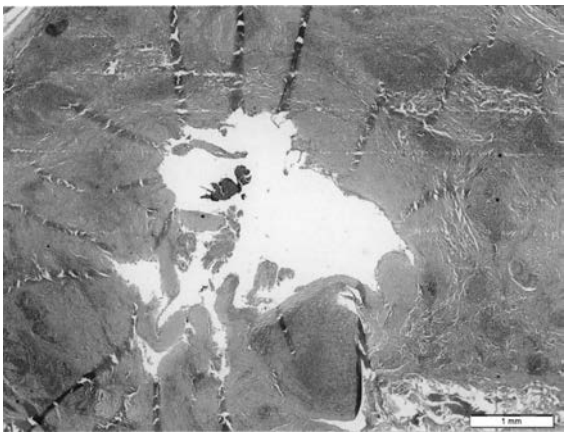

Fig. 1. Bland histiocytoid cells with mild nuclear pleomorphism (HE, x200).

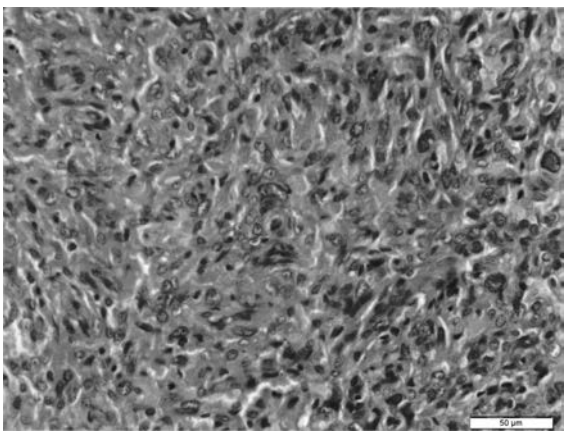

Fig. 2. Lymphocytic infiltrate with germinal center formation around centrally cystic spaces (HE, x40).

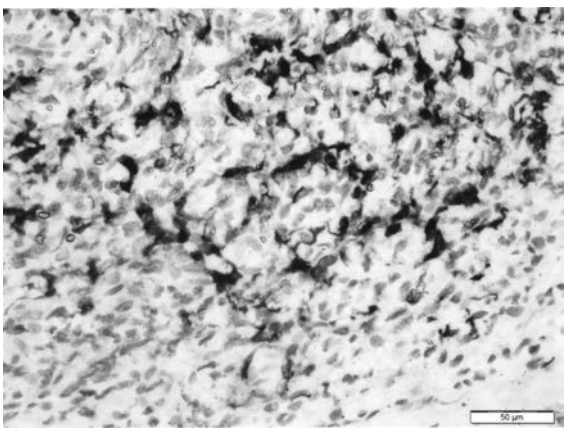

Fig. 3. Desmin expression in some dendritic-appearing cells (Desmin, x400).

(q13:q12), have been characterized[6,8,17]. The differential diagnosis of AFH includes aneurysmal fibrous histiocytoma, metastatic tumor to the lymph node, inflammatory pseudotumor, various vascular neoplasms and RMS. Aneurysmal fibrous histiocytoma arising superficially in young adults typically shows histopathologic features of dermatofibroma. Metastatic tumor to the lymph node has a true nodal architecture, such as subcapsular and medullary sinuses. Inflammatory pseudotumor lacks the pseudovascular spaces of AFH. The vascular tumors contain vascular spaces lined by endothelial cells. Finally, AFH may be misdiagnosed as RMS. Differentiating AFH from RMS is crucial, because the latter is likely to be treated aggressively. RMS is typically deeply localized and larger than AFH. RMS contains atypical cells and expresses desmin and myogenin or MyoD1. Additionally, RMS lacks a surrounding lymphocytic infiltrate or cystic cavities containing blood[6,10,15]. There are no reliable clinical and histologic parameters that predict behavior. Most cases have an excellent prognosis. However, correct diagnosis is important because of the risk of local recurrence and the small risk of metastasis[6]. Wide local excision with follow-up is considered to be the appropriate mode of management. Lymph node and pulmonary metastases are rare (2%-5%).

This case demonstrated that AFH can mimic a spectrum of systemic diseases before any mass is clinically detectable. In conclusion, AFH is a rare disease that is often initially misdiagnosed. AFH should be considered as a differential diagnosis in some cases with increased sedimentation rate and CRP levels. Thus, pathological examination is necessary for diagnosis. Most patients do well with local excision alone. Our patient was well at follow-up, without signs of local recurrence or metastasis.

REFERENCES

1. Enzinger FM. Angiomatoid malignant fibrous histiocytoma: a distinct fibrohistiocytic tumor of children and young adults simulating a vascular neoplasm. Cancer 1979; 44: 2147-2157.
2. Costa MJ, Weiss SW. Angiomatoid malignant fibrous histiocytoma. A follow-up study of 108 cases with evaluation of possible histologic predictors of outcome. Am J Surg Pathol 1990; 14: 1126-1132.
3. Fanburg-Smith JC, Miettinen M. Angiomatoid "malignant" fibrous histiocytoma: a clinicopathologic study of 158 cases and further exploration of the myoid phenotype. Hum Pathol 1999; 30: 1336-1343.
4. Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F (eds). WHO Classification of Tumours of Soft Tissue and Bone (4th ed). Lyon, France: IARC Press; 2013.
5. Petersen I. [The new WHO classification and recent results in soft tissue tumor pathology]. [Article in German] Pathologe 2013; 34: 436-448.
6. Thway K. Angiomatoid fibrous histiocytoma: a review with recent genetic findings. Arch Pathol Lab Med 2008; 132: 273-277.
7. Ren L, Guo SP, Zhou XG, Chan JK. Angiomatoid fibrous histiocytoma: first report of primary pulmonary origin. Am J Surg Pathol 2009; 33: 1570-1574.
8. Dunham C, Hussong J, Seiff M, Pfeifer J, Perry A. Primary intracerebral angiomatoid fibrous histiocytoma: report of a case with a t(12;22)(q13;q12) causing type 1 fusion of the EWS and ATF-1 genes. Am J Surg Pathol 2008; 32: 478-484.
9. Petrey WB, Le Gallo RD, Fox MG, Gaskin CM. Imaging characteristics of angiomatoid fibrous histiocytoma of bone. Skeletal Radiol 2011; 40: 233-237.
10. Thway K, Stefanaki K, Papadakis V, Fisher C. Metastatic angiomatoid fibrous histiocytoma of the scalp, with EWSR1-CREB1 gene fusions in primary tumor and nodal metastasis. Hum Pathol 2013; 44: 289-293.
11. Hothi D, Brogan PA, Davis E, Ramsay A, Dillon MJ. Polyarteritis nodosa as a presenting feature of angiomatoid fibrous histiocytoma. Rheumatology (Oxford) 2004; 43: 245-246.
12. Chen G, Folpe AL, Colby TV, et al. Angiomatoid fibrous histiocytoma: unusual sites and unusual morphology. Mod Pathol 2011; 24: 1560-1570.
13. Fletcher CD. Angiomatoid "malignant fibrous histiocytoma": an immunohistochemical study indicative of myoid differentiation. Hum Pathol 1991; 22: 563-568.
14. Romeo S, Dei Tos AP. Soft tissue tumors associated with EWSR1 translocation. Virchows Arch 2010; 456: 219-234.
15. Billings SD, Folpe AL. Cutaneous and subcutaneous fibrohistiocytic tumors of intermediate malignancy: an update. Am J Dermatopathol 2004; 26: 141-155.
16. Sparreboom E, Wetzels C, Verdijk M, Mulder S, Blokx W. Subcutaneous angiomatoid fibrous histiocytoma mimicking metastatic melanoma. Case Rep Pathol 2012; doi:10.1155/2012/291623.
17. Tanas MR, Rubin BP, Montgomery EA, et al. Utility of FISH in the diagnosis of angiomatoid fibrous histiocytoma: a series of 18 cases. Mod Pathol 2010; 23: 93-97.