



Cervical Angiomatoid Fibrous Histiocytoma

*Mohammad Al Felasi, *Joao Barosa, *Sebastian Arnaud, *Marc Makeieff

*Head and Neck Surgery Department
University Hospital of Montpellier
Montpellier
France

Abstract

Background: Angiomatoid fibrous histiocytoma (AFH) is a rare type of sarcoma with low-grade malignancy that usually occurs in young subjects. AFH is uncommon in the head and neck region.

Methods: We describe an exceptional case of localization in the neck. This is the first report of a rare variant of AFH presenting in a 42-year-old woman. The tumor was situated posterior to the carotid artery adhering to the prevertebral plane and invading the sympathetic cervical chain. In this setting, we decided to perform a surgical exploration with frozen section biopsy. The frozen section revealed a fusiform tumor proliferation resembling a sarcoma. A FISH study concluded on the presence of a EWSR1 22q (22) gene rearrangement. Furthermore, the immunohistochemical study revealed anti-EMA positive cells. The final pathological description concluded on the presence of an AFH, which was excised by surgery alone.

Results: After a 2 year follow-up period, the patient is free of disease. Angiomatoid fibrous histiocytoma (AFH) is a rare sarcoma subtype, and mis diagnosis can lead to its over treatment. A precise description of the pathological response and a multidisciplinary discussion can lead to a correct decision.

Conclusion: AFH is a tumor with local evolution, and surgery is the mainstay of its management. Complete local excision is recommended to hamper local recurrences.

Introduction

Angiomatoid fibrous histiocytoma (AFH) is a rare low-grade malignancy sarcoma type that usually occurs in young subjects ¹. Enzinger et.al. first described AFH and its features in 1979 due to its slow growth rate and low risk of metastasis¹. Few cases have been described in the literature and most of the AFH tumors were in the extremities and trunk, while only 5-7% of them were found in the head and neck region ². Genetic studies can help in the definitive diagnosis of AFH ³. We present the first case located in the prevertebral space.

Clinical Presentation

A 42-year-old woman presenting with a left cervical mass was referred to our outpatient clinic. The clinical examination revealed an anterior left cervical mass, pushing the piriform sinus with no mucosal lesions in the upper airways.

Echography and CT scan showed a left cervical 30 mm mixed cystic mass with a necrotic center situated in the prevertebral space, in contact with the fifth cervical vertebra with no bone erosion.

The MRI examination revealed a clearly defined multilobed and heterogeneous prevertebral lesion situated posteriorly to the great vessels, with T2 hypersignal and isosignal areas and T1 hyposignal. These findings suggested a neurogenic tumor. (Fig. 1)



Figure 1

A cervical exploration was programmed. An external cervical surgical approach with a lateral transverse incision is

carried. The tumor was situated posterior to the carotid artery, adhering to the prevertebral plane and invading the sympathetic cervical chain. In this setting, we decided to perform a frozen section biopsy.

The frozen section diagnosis was a fusiform tumor proliferation suggesting a sarcoma. Based on these findings, we stopped the surgery while awaiting the final pathological results. The pathological specimen was sent to a specialized sarcoma center. A FISH study concluded on the presence of anEWSR1 22q (22) gene rearrangement. Furthermore, the immunohistochemical study revealed anti-EMA positive cells, while the final pathological conclusion was AFH. (Fig. 2)

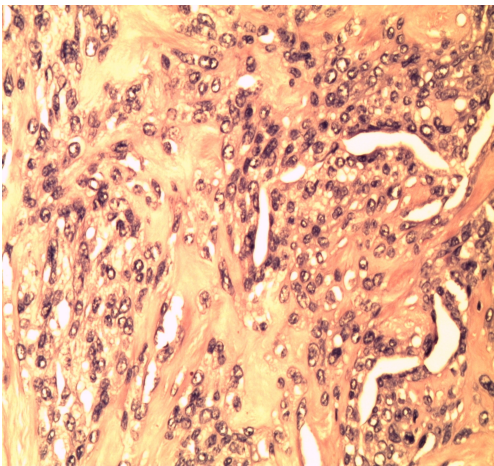


Figure 2

Total surgical excision was decided on the basis of a multidisciplinary oncological team discussion.

Surgery was programmed after informing the patient of all the risks. The tumor was excised through a lateral cervical approach with no complications. The pathological study confirmed the AFH diagnosis after a genetic study that revealed the presence of the EWS-ATF1 EWSR1 22q (22) fusion gene. The patient is free of recurrence for the past two years.

Discussion

AFH is typically found on the extremities or trunk, very seldom in the head and neck region ². Most reported cases are located within the subcutis or deep dermis ⁴. The AFH age range is between 6 months to 43 years with slight male predominance ¹. MRI and CT scan findings are nonspecific, with fluid levels and intralesional hemorrhage, which cannot be differentiated from severe malignant fibrous histiocytoma, hematoma or soft tissue haemangioma. Diagnosis is thus based on histopathological findings ^{1,4}. The four main distinctive features include: (i) a fibrous pseudo capsule; (ii) a dense lymphoplasmic response; (iii) blood-filled focal areas of cystic change; and (iv) proliferation of spindle or round cells.

Differential diagnosis of AFH is difficult. AFH can be mistaken for hematoma. However, compact sheets of histiocyte-like cells are never observed in an organized hematoma ⁴. Moreover, vascular neoplasms are excluded based on the fact that dilated vascular spaces are rare in AFH, as compared to hemangiopericytoma or hemangioendothelioma. Microhaemorrhages and extravasated erythrocytes occasionally suggest a Kaposi disease, but our case did not display the typical spindle cell or slit-like pattern of this entity. Moreover, the absence of pleomorphism and the presence of histiocyte-like cells excluded angiosarcoma³.

Waters reported the first cytogenetic characterization of AFH, noting the presence of the translocation t (12; 16)(q13; p11) and the production of a FUS-ATF1 fusion gene. This was soon followed by reports of EWSR1-ATF1 fusion transcripts, resulting from the translocation t (12; 22) (q13; q12)⁵.

Most authors recommend complete surgical excision, although Costa treated three recurrence cases by post surgical irradiation therapy². AFH has a low recurrence rate, with less than 2% metastasis risk and 1% mortality rate².

References:

1. Enzinger FM. Angiomatoid malignant fibrous histiocytoma: a distinct fibrohistiocytic tumor of children and young adults simulating a vascular neoplasm. *Cancer* 1979; 44: 2147-57.
2. Costa MJ, Weiss SW. Angiomatoid malignant fibrous histiocytomas. A follow-up study of 108 cases with evaluation of possible histologic predictor outcome. *Am. J. Surg. Pathol.* 1990; 14: 1126-32.
3. Fanburg-Smith J.C. Angiomatoid "Malignant" Fibrous Histiocytoma: A clinicopathologic Study of 158 Cases and Further Exploration of the Myoid Phenotype. *Human Pathology* 1999; 30: 1336-1343.
4. Pratibha R., Ahmed S..Angiomatoid variant of fibrous histiocytoma: a case report and review of literature. *Int J Paediatr Dent.*2006; 16: 363-369.

5. Waters BL, Panagopoulos I, Allen EF. Genetic Characterization of Angiomatoid Fibrous Histiocytoma Identifies Fusion of the *FUS* and *ATF-1* Genes Induced by a Chromosomal Translocation Involving Bands 12q13 and 16p11. [*Cancer Genetics and Cytogenetics*](#) 2000; 121: 109-116.