Angiomatoid fibrous histiocytoma, previously called angiomatoid malignant fibrous histiocytoma, is now considered to be an intermediate malignancy. This tumor is categorized as a tumor of uncertain differentiation by the 2002 WHO classification. Four cases of angiomatoid fibrous histiocytoma that have been reported in Korea were all less than 3 cm at the greatest dimension, and they were located on the scalp and the upper arm. Here, we describe another case of angiomatoid fibrous histiocytoma in an 11-year-old boy.

**CASE REPORT**

An 11-year-old boy was referred to our hospital with an ovoid mass in his left shoulder. His mother said that the mass had first been noted five months before, and that it had been slowly increasing in size. Neither pain nor tenderness was present. The magnetic resonance (MR) images showed a well capsulated multilobulating deep subcutaneous mass (8 cm at the greatest dimension) with hemorrhage and fluid-fluid level. There was no evidence of muscle invasion, and the lesion was clinically regarded as a sort of benign hemorrhagic cystic mass. An excision was performed.

Grossly, the tumor was an encapsulated, ovoid, multilocular cystic mass, and it measured 7.5 × 7.5 × 4.0 cm (Fig. 1). The external surface was brownish tan and smooth. The cut surface revealed several locules that were filled with hemorrhagic fluid. The internal surface of the locules was purplish brown, irregular and trabecular. The thickness of the wall ranged from 0.1 cm to 0.8 cm.

Histologically, the tumor consisted of fairly cellular solid areas with variable-sized blood-filled cystic spaces (Fig. 2A). The tumor was surrounded by a fibrous capsule; a dense lymphoplasmacytic infiltration was present, especially in the peripheral areas, and those two features mimicked metastatic tumor in a lymph node (Fig. 2B). The blood-filled clefts or cystic spaces were lined by flattened tumor cells rather than endothelium proper. A diffuse, intra- and extracellular deposition of hemosiderin pigments was another characteristic feature of this tumor (Fig. 2A). The solid areas consisted of spindle histiocytoid cells with a storiform or short fascicular pattern. The individual cells had bland looking ovoid nuclei and elongated cytoplasm (Fig. 2C). Mitotic figures

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were rare, but a focal area with mitotic counts up to 3/10 HPF was present in the periphery of the tumor. On the immunohistochemical staining, the tumor cells showed diffuse positivity for CD99 and CD68 and focal positivity for desmin (Fig. 3). The Ki-67 labeling index was up to 10%. The patient has shown no evidence of persistence or recurrence of the tumor for the postoperative 5 months.

**DISCUSSION**

In 1979, Enzinger described 41 cases of unusual sarcoma of a fibrohistiocytic lineage, which were characterized by their primary occurrence as nodular subcutaneous growth in the extremities or, more rarely, in the head and neck of young individuals and children. The author named those cases "angiomatoid malignant fibrous histiocytoma" (AMFH), and they were regarded as a variant of malignant fibrous histiocytoma, based on the presence of metastasis. However, according to a report by Fanburg-Smith et al., the metastatic rate was only 1% in a large study.
of angiomatoid fibrous histiocytoma. Moreover, Costa and Weiss documented that local recurrences were noted only in 12% of patients. All of the tumors had initially positive resection margins, and they were subsequently cured by re-excision. Based on these reports, the World Health Organization has reclassified angiomatoid (malignant) fibrous histiocytoma as tumor of uncertain differentiation and intermediate malignancy rather than as fully malignant sarcoma. This tumor typically affects children and young adults, and it presents as a painless, slowly growing subcutaneous soft tissue mass that is usually located in the extremities and less commonly in the trunk, head and neck. Clinically, it may resemble hematoma, hemangioma or a benign cyst, as it was in our case.

Histologically, thick fibrous pseudocapsule around the tumor, variable-sized pseudovascular spaces between the tumor cells and a prominent cuff of lymphoplasmacytic infiltration are characteristic features of angiomatoid fibrous histiocytoma. The tumor cells are arranged in a variety of growth patterns, including sheets, whorls and short fascicles. Individual tumor cells of angiomatoid fibrous histiocytoma look cytologically bland, but in occasional cases, they may show a striking pleomorphism, which does not appear to have clinical significance. The mitotic rate is low.

Partial immunopositivity for desmin, epithelial membrane antigen, CD68 or CD99, which was also be seen in our case, have been reported in about half of the cases of angiomatoid fibrous histiocytoma. This rather nonspecific multiphenotype is attributed to uncertain differentiation of angiomatoid fibrous histiocytoma. Despite its name, this tumor does not express endothelial markers such as CD31, CD34 or factor VIII-related antigen.

The differential diagnosis of angiomatoid fibrous histiocytoma includes dermatofibroma with aneurysmal/hemosiderotic changes (aneurysmal benign fibrous histiocytoma), metastatic tumor in a lymph node, various vascular neoplasms and Bednar tumor. We can discriminate angiomatoid fibrous histiocytoma from vascular neoplasms by the lack of endothelium. Bednar tumor contains melanin pigments, but not hemosiderin pigments. Aneurysmal fibrous histiocytoma, a variant of dermatofibroma by the 2002 WHO classification, has histologic features similar to angiomatoid fibrous histiocytoma, except the lack of pseudocapsule and a dense cuff of lymphoplasmacytic cells. Although they are classified into different groups, some authors have used these two terms interchangeably.

Fusion of FUS and ATF1 genes has been reported in two cases of angiomatoid fibrous histiocytoma. Recently, a case with fusion of the EWSR1 and ATF1 genes, which was previously identified in clear cell sarcoma, has been reported. Further cytogenetic analysis of more cases will be needed to clarify whether or not the presence of these fusion genes is pathognomonic for angiomatoid fibrous histiocytoma.

Death due to the disease is extremely rare in patients with angiomatoid fibrous histiocytoma. Local recurrence has been reported in 12% of cases in one large study, and it is common in cases with infiltrating margins, involvement of the skeletal muscle and the head and neck region. A wide surgical excision
and a careful follow-up are recommended since there is no evidence that adjuvant therapy may play a role in the primary management of this tumor.  

The present case is different from the four previously reported Korea cases in respect of the size, which was 7.5 cm in the greatest dimension, and the location in the shoulder. In spite of the uncommon location, angiomatoid fibrous histiocytoma should be considered as part of the differential diagnosis of hemorrhagic cystic mass of soft tissue in young individuals, and the diagnosis can, to a certain extent, be confirmed by immunohistochemical staining.

REFERENCES