Orthopaedic Case of the Month

A 14-Year-Old Boy With a Medial Thigh Soft Tissue Mass

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History and Physical Examination

A 14-year-old boy presented to the outpatient clinic for a 7-month history of a slow-growing mass in the right upper thigh and an approximate 30-pound (14-kg) weight loss. He had no history of trauma, constitutional symptoms such as fevers or chills, or substantial pain. The patient could walk without any problem but experienced fatigue and discomfort with strenuous physical activity requiring him to withdraw from football. His medical and surgical

histories were noncontributory. There was no family history of medical illnesses, including history of cancer.

On physical examination, he was a pale-looking adolescent in no acute distress. Palpation of his right lower extremity revealed a firm, fixed, 9- × 8-cm, nontender, nonpulsatile soft tissue mass in the medial proximal thigh. There were no skin changes, increased local temperature, or fluctuance associated with the mass. The patient had full ROM of the right hip and knee, and there were no neurovascular deficits distally. Enlarged right inguinal lymphadenopathy was noted.

His initial workup showed elevated serum alkaline phosphatase (539 IU/L, normal 38–126 IU/L) and erythrocyte sedimentation rate (> 130 mm/hour, normal 0–10 mm/hour), with microcytic, hypochromic anemia (hemoglobin, 10.7 g/dL; mean corpuscular volume, 75 fL; mean corpuscular hemoglobin, 21.8 pg) and thrombocytosis (platelet count, 642 k/mm³). Given the thigh mass, laboratory abnormalities, and history of weight loss, malignancy was a concern. Workup included MRI of the primary site (Fig. 1), CT of the chest, abdomen, and pelvis, positron emission tomography (PET) scan, nuclear bone scan, molecular analysis, and needle biopsy 3 weeks before evaluation at our center.

Based on the history, physical examination, laboratory studies, and imaging studies, what is the differential diagnosis at this point?

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Imaging Interpretation

MRI showed a large (7- × 5- × 4.2-cm), deep, heterogeneous soft tissue mass with fluid-fluid levels at the medial proximal thigh in the adductor musculature (Fig. 1). The mass was abutting the femoral vessels medially, extending

Fig. 1A–C (A) An axial T2 postcontrast MR image shows the proximity to the femoral bundle, which is visible at the superomedial aspect of the mass. Hyperintensity, the presence of fluid-fluid level, a possible enhancing thickened pseudocapsule superiorly, and lack of

complete enhancement are evident. (B) A coronal T1 postcontrast MR image shows solid and cystic areas. (C) An axial T1 precontrast MR image shows fluid-fluid level.

to, but not involving, the medial femoral cortex. There was no intraosseous component or signal changes. There was high T2 signal extending from the mass to the inguinal area, where multiple enlarged inguinal lymph nodes were identified.

CT of the chest, abdomen, and pelvis revealed no evidence of metastatic disease, except for the presence of right iliac and inguinal lymphadenopathy.

The PET scan showed a cystic and solid right thigh mass with multiple active inguinal and external iliac nodes. The maximum standard uptake value of the right thigh mass was 2.3. A metabolically active node in the right inguinal area had a maximum standard uptake value of 4.1 and measured 1.6×1.83 cm. A smaller right inguinal node had a maximum standard uptake value of 2.2. Three external right iliac nodes with a maximum standard uptake value of 3.5, 2.9, and 2.6 also were present.

Nuclear bone scan revealed no concern for skeletal metastases, but radioactive uptake was noted in the primary tumor in the medial aspect of the right thigh (Fig. 2).

Differential Diagnosis

- Rhabdomyosarcoma
- Synovial sarcoma
- Malignant peripheral nerve sheath tumor
- Extraosseous Ewing's sarcoma
- Metastatic tumor to lymph node
- Angiomatoid fibrous histiocytoma

The patient initially underwent needle biopsy of the lesion, followed by a primary resection and ipsilateral inguinal lymph node sampling for staging (as part of the

Children's Oncology Group protocol for resection of soft tissue sarcomas). The biopsy and resected specimens were evaluated histologically (Fig. 3).

Based on the history, physical examination, laboratory studies, imaging studies, and histologic picture, what is the diagnosis and how should the patient be treated?

Histology Interpretation

The resected specimen (with clean margins) was a well-circumscribed oval mass of red-tan soft tissue measuring $9.0 \times 6.5 \times 6.0$ cm (Fig. 3A). The cut surface showed an encapsulated lesion with solid and cystic areas. The largest cystic lesion (approximately 5 cm) contained approximately 25 mL of blood. Surgical margins appeared grossly free of tumor.

Microscopically, the biopsy and resected specimens showed the same features. The tumor had areas with decreased cellularity and myxoid change and other densely cellular sheets of tumor (Fig. 3B–C). The cells had round to oval to spindle-shaped nuclei with fine open chromatin and inconspicuous nucleoli. Mild nuclear pleomorphism and hyperchromasia were observed. Scant to moderate eosinophilic and basophilic cytoplasm was present with indistinct cell borders. The inked surface of the resection and the neurovascular margin were free of tumor.

The immunohistochemical profile performed on the initial biopsy and definitive surgery specimens showed expression of vimentin, CD99 (Fig. 3D), epithelial membrane antigen, and nuclear INI-1. The tumor did not express keratin AE1/3, CAM 5.2, CD30, CD31, CD34, CD45, fascin, smooth muscle actin, CD68, factor XIIIa, S-100 protein, CD1a, desmin, myogenin, synaptophysin,

Fig. 2A–B (A) The patient's nuclear medicine bone scan shows the right medial thigh uptake (arrow). (B) A magnified view again shows the medial thigh mass (arrow).

and ALK-1. Ki67 showed a proliferative index of approximately 5% to 10% in tumor cells.

Cytogenetic studies showed normal male karyotype. Fluorescence in situ hybridization (FISH) analysis showed no rearrangement of the 18q11.2 SYT region. Reverse-transcriptase (RT) PCR on samples of tumor mass was negative for EWS/FLI1, EWS/ERG, PAX/FKHR, SYT/SSX, EWS/WT1x9, and ETV6/NTKR3 translocations.

Excisional biopsies of the proximal and distal superficial femoral nodes showed no metastatic involvement.

Diagnosis

Angiomatoid fibrous histiocytoma (AFH)

Discussion and Treatment

Certain elements of the patient's history, physical examination, and imaging suggested AFH. The patient's age was consistent with that in which AFH is most commonly found (< 30 years), and he reported a 14-kg weight loss—a common paraneoplastic effect. On physical examination, the characteristics of the mass were more suggestive of tumor (nontender, nonpulsatile, local lymphadenopathy), and its location in the right lower extremity was consistent with the common location of AFH. In addition, laboratory evaluation showed a microcytic anemia, a common paraneoplastic effect of AFH, and an elevated erythrocyte sedimentation rate, occasionally seen in patients with AFH. Imaging showed a primarily soft tissue mass, lack of osseous involvement, lack of metastatic foci, and heterogeneity with cystic involvement and fluid-fluid levels, all features suggestive of AFH. Histologic findings that helped confirm the diagnosis included round to oval to spindle-shaped nuclei, myogenin and S100-negative, and vimentin and CD99-positive immunohistochemistry.

Rhabdomyosarcoma is the most common type of soft tissue sarcoma in the pediatric population [35, 43], although adult-type soft tissue sarcomas become more predominant in the adolescent age group [39]. As in most other soft tissue tumors, MRI features of rhabdomyosarcoma can be nonspecific [28, 29]. If intratumoral necrosis is present, rhabdomyosarcoma can exhibit cystic changes, as seen in our patient. Histologic analysis reveals characteristic rhabdomyoblasts, which exhibit eccentric round nuclei and eosinophilic cytoplasm [37]. The lymphoplasmacytic infiltrate present in AFH [20] also is not characteristic of rhabdomyosarcoma. On immunohistochemistry, specific skeletal muscle markers (myogenin, MyoD, myoglobin, muscle-specific actin) [8, 38] are seen in rhabdomyosarcoma that are absent in AFH. Desmin often is positive in rhabdomyosarcoma [37, 38] and it also is seen in AFH [20]. Synovial sarcoma is the most common nonrhabdomyosarcoma soft tissue malignancy seen in children, comprising 7% to 8% of all pediatric malignant soft tissue tumors [1]. Similar to AFHs, synovial sarcomas primarily occur in the extremities [22, 42] and are deep-seated [22]. Radiographically, approximately 30% of the time they exhibit calcifications [33] that are not typically seen in AFH. Grossly, both tumors appear multicystic, with fluid-fluid levels [22], although this pattern is more commonly observed with AFH. Histologically, synovial sarcomas contain spindle-shaped cells with round to oval nuclei, indistinct cell borders, and rare mitoses [22]. Often, they exhibit a biphasic histologic picture with epithelial and spindle cells [21]. Immunohistochemistry also may reveal vimentin, epithelial membrane antigen, and cyto-keratin positivity [22]. The absence of the characteristic

Fig. 3A–D (A) A cross section shows solid yellow and cystic spaces, some of which contain blood. (B) Microscopically, the tumor is composed of myxoid (star) and densely cellular areas (arrow) (Stain, hematoxylin and eosin; original magnification, $\times 2$). (C) Higher magnification of myxoid areas shows round to oval cells with open

chromatin, occasional nuclear grooves, and inconspicuous nucleoli (Stain, hematoxylin and eosin; original magnification, $\times 40$). Abundant eosinophilic and basophilic cytoplasm is present. (D) Immunohistochemistry for CD99 shows focal membranous expression in tumor cells.

chromosomal translocation (X;18) [45] and SSX1 and SSX2 fusion genes [12] of synovial sarcoma was critical to differentiating these two entities. Malignant peripheral nerve sheath tumors usually present as enlarging soft tissue masses in the extremities [49]. Histologically, malignant peripheral nerve sheath tumors are characterized by spindle-shaped cells but show considerable mitotic activity [49]. Although MRI does not reveal any specific features, close proximity of the tumor to nerves is suggestive of a malignant peripheral nerve sheath tumor [29]. These tumors may arise *de novo* or be secondary to degeneration of neurofibromas. Approximately $\frac{1}{2}$ of patients with malignant peripheral nerve sheath tumors have neurofibromatosis, Type I (NF-1) [16, 17, 27, 46, 51]; however, the cumulative risk of having malignant peripheral nerve sheath tumors develop among patients with NF-1 varies from 2% to 29% [17, 34, 47]. On immunohistochemistry, S-100 positivity frequently is seen [13, 34]. Extrasosseous Ewing's sarcoma is a malignant small, round, blue-cell tumor that occurs mostly during or after adolescence, with

a slight male predominance [2, 31, 44, 52]. The extremities are the most common sites of involvement [41, 48]. Histologically, this tumor is characterized by spherical to polygonal cells with round to oval nuclei and glycogen content [26, 31, 52]. The described immunohistochemistry varies, although immunoreactivity to CD99, S-100, and vimentin, with absence of cytokeratin and desmin, has been noted [9, 25, 30]. FISH and RT-PCR analyses reveal the fusion gene EWS-FL1 [t(11;22)(q24;q 12)] is present in greater than 80% of cases and an EWS-ETS fusion gene in most others. AFH may be misdiagnosed as metastatic disease to lymph nodes, although a diagnosis of the latter is favored by tumor presentation in a typical nodal location. Histologically, lymph node metastasis is suggested by the presence of typical nodal architecture, tumor invasion of germinal centers, cytologic atypia, and a paucity of blood-filled cystic spaces [20, 50].

Originally described by Enzinger in 1979 [19], AFH is a soft tissue tumor of low to intermediate grade most often found in the subcutis and deep dermis [50]. Anatomically,

the tumor occurs primarily in the extremities (most commonly in the upper extremities) and torso and less frequently in the head and neck region [11, 19, 24]. AFH predominantly affects individuals younger than 30 years, with a median age of 13 years [19]. There is no sex predilection [11, 19]. Paraneoplastic effects, notably, anemia, weight loss, fever, and chills, secondary to cytokine expression can be the primary presenting symptoms [11, 14, 19]. AFH is primarily a slow-growing tumor with an indolent course [11, 19, 20], although more aggressive behaviors have been described [10].

AFH characteristically appears as a multinodular mass, with cystic spaces and hemorrhage [11, 19]. Although fluid-fluid levels seen in AFH are nonspecific, they are seen more commonly with AFH. Histologically, four features have been described: proliferation of round or spindle cells, dense lymphoplasmacytic infiltrate, fibrous pseudocapsule, and areas of cyst formation with hemorrhage and hemosiderin [11, 19, 20, 50]. However, only the multinodularity and round or spindle cells are consistent findings [50]. Mitotic figures are rare [5, 10, 11, 19, 50].

The immunohistochemical profile often varies, although some consistencies have been noted. Desmin, vimentin, calponin, CD99, and CD68 are usually positive [20, 50]. Smooth muscle (caldesmon, smooth muscle actin) and vascular endothelial (CD31, CD34) markers are sometimes positive [7, 20, 36]. Skeletal muscle markers (MyoD1, myoglobin, myogenin [MYF4], CD21, CD35, S-100 protein, cytokeratin 8/18) are always negative [20].

AFHs reportedly express EWSR1-CREB1, EWSR1-ATF1 (also present in histologically distinct clear cell sarcoma, where it is associated with MTF-M transcript expression), and FUS-ATF1 fusion genes, all of which can assist with the diagnosis [3, 4, 40].

Wide resection is the recommended definitive treatment for AFH [11, 24]. In patients with an unresectable or recurrent tumor, adjuvant radiotherapy or chemotherapy may be indicated [15]. Chemotherapy is not indicated for resectable cases [6, 15]. Local recurrence rates vary from 0% to 63% [10, 11, 18–20, 23]. Additionally, paraneoplastic effects generally resolve within weeks to months after surgical resection of the tumor [32]. Ultimately, the low-grade histologic features, rare distant metastases, and low recurrence rates (mainly local) have resulted in the renaming of this rare entity that previously was known as angiomatoid malignant fibrous histiocytoma to AFH.

In our patient, wide resection was performed, and despite the proximity to the neurovascular bundle and need to sacrifice the femoral vein, free surgical margins were successfully achieved. The femoral artery could be dissected free from the tumor and the femoral vein was sacrificed since there was sufficient collateral flow. The patient did not receive any adjuvant treatment and has been

continuously free of disease at 24 months' followup, having returned to normal physical activity and regained his weight within 3 months. He is being followed serially with physical examination, MRI of the primary site, and chest radiography.

Our experience suggests, although rare, AFH should be included in the differential diagnosis of extremity soft tissue masses that are multinodular and cystic and exhibit round or spindle-shaped cell histologic features. Early identification allows prompt excision, which leads to resolution of the tumor and any paraneoplastic effects.

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