MONOPHASIC SYNOVIAL SARCOMA OF THE LOWER EXTREMITY: A CASE REPORT AND REVIEW OF THE MEDICAL LITERATURE

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Abstract

Synovial sarcoma is the fourth most common type of soft-tissue sarcoma, accounting for 2.5%-10.5% of all primary soft-tissue neoplasms worldwide. This disease most often affects the extremities, 80%-95% of cases, of adolescents and young adults (15-40 years of age). Despite its name, the lesion does not commonly arise in an intraarticular location but usually occurs near joints. Etiology is still unclear. Synovial sarcoma is an intermediate-to high-grade neoplasm with poor prognosis, and, despite initial aggressive wide surgical resection, local recurrence and metastasis are common. Understanding and recognizing the spectrum of appearances of this malignancy, which reflect the underlying pathologic characteristics, improve radiologic evaluation are essential for optimal patient management.

A twenty year-old girl admitted to our institution with an rapid increasing mass on her left thigh recently. The mass was first noted 5 years ago. Imaging studies including X ray, ultrasound and MRI suggested a soft tissue sarcoma. Definite diagnosis of monophasic synovial sarcoma, fibrous type was made based on histopathological findings.

Key words: *Synovial sarcoma, monophasic.*

1. BACKGROUND

Synovial sarcoma (SS) is the fourth most common type of soft-tissue sarcoma, accounting for 2.5% to 10.5% of all primary soft-tissue malignancies worldwide. This rare and aggressive neoplasm most often affects the extremities (80%-95% of cases), particularly the knee in the popliteal fossa, of adolescents and young adults (15-40 years of age) [1,2,3,4]. Despite its name, SS does not appear to be of synovial origin, but rather from multipotent stem cells that differentiate into mesenchymal and/or epithelial structures. The etiology remains unknown. Almost all SS cells are characterized by the presence of a translocation involving chromosomes X and 18. Complete surgical removal of the primary tumor is the mainstay of treatment [5]. We report a case of monophasic synovial sarcoma admitted to our institution.

2. CASE REPORT

A twenty year-old girl admitted to Hue University Hospital with an increasing left thigh mass. She first noticed the mass 5 years ago with slow progression overtime but recently rapidly increased in size. On clinical examination, a bulging mass locating medially in the left upper thigh was found. The mass was indolent, tough and immobile on palpation. The covering skin was intact with no ulceration, pigmentation or local infection. Neither sensory nor motor deficit was detected. No pelvic lymphadenopathy was noticed. She denied any history of trauma or infection to the extremities. Previous treatment was unremarkable. Laboratory tests reveal normal results.

Ultrasound of the mass showed a heterogeneous mixed echogenicity soft-tissue mass with peripheral calcification and adjacent

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tissue infiltration which is highly suggestive of malignancy. Calcification was then proven on X-ray to be amorphous appearance. Left thigh MRI confirmed a soft-tissue tumor with classical "triple sign", represented by intermixed areas of low, intermediate and high signal intensity on long repetition time images, suggesting a soft-tissue sarcoma. Common femoral arteriography was indicated to study the vasculature pattern of the mass prior to resection. DSA image showed a hypervascular mass with evidence of typical tumor vessels. The definite diagnosis of Fibrous type monophasic synovial sarcoma was confirmed on histopathologic study. The patient denied left thigh amputation and the tumor was left untreated. One year follow up was noticed with advanced stage as evident of distal lymphadenopathy and lung metastasis. She is now undergoing palliative care.

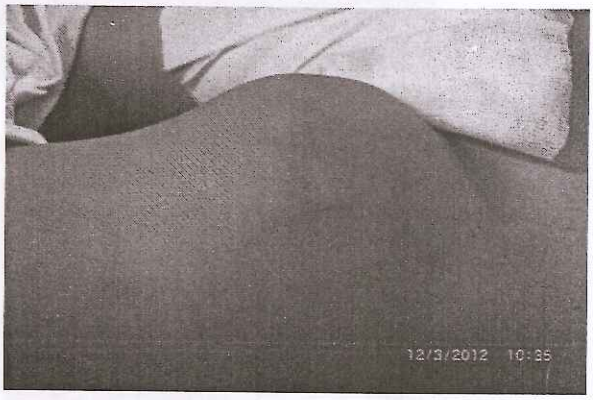

Figure 1. A soft tissue mass in the left upper thigh measuring approximately 9 cm in diameter. The mass is solid, firm, indolent and immobile on palpation. The outer skin appears smooth and intact.

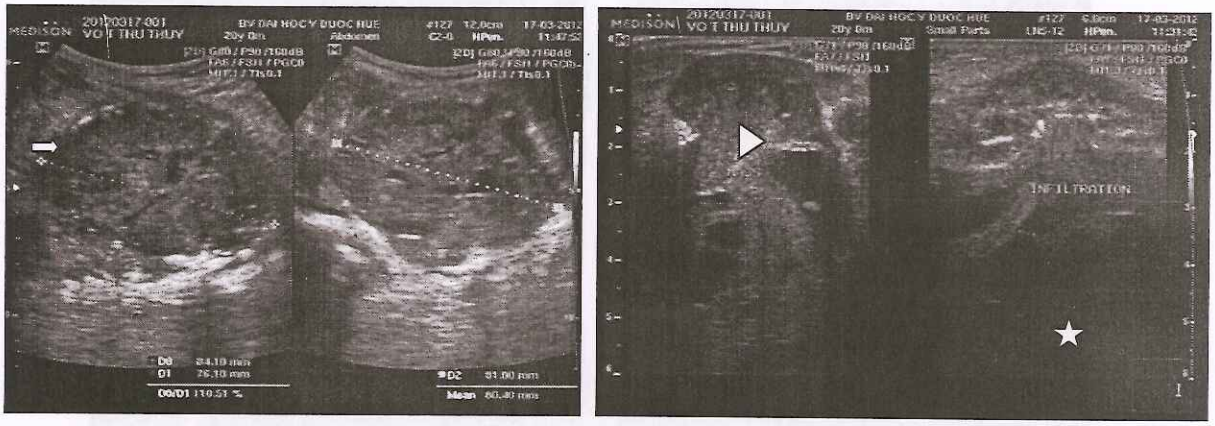

Figure 2. Ultrasound scan showed a large, relatively well-defined hypoechoic mass within muscle layer (caliper). The mass is heterogeneous in nature with irregular border, peripheral calcification (arrow) and areas of necrosis (star). Infiltration to the adjacent subcutaneous tissue was also noted (arrowhead). No evidence of vascular invasion on Color Doppler Ultrasound (not shown).

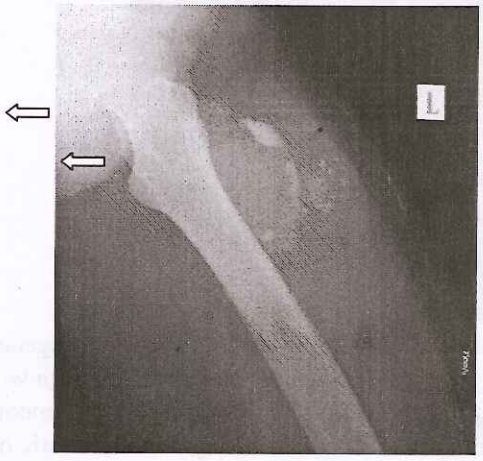

Figure 3. Lateral radiography of the upper left thigh shows a soft-tissue mass with punctuate curvilinear and amorphous calcification (arrow) laterally without obvious underlying bone erosion.

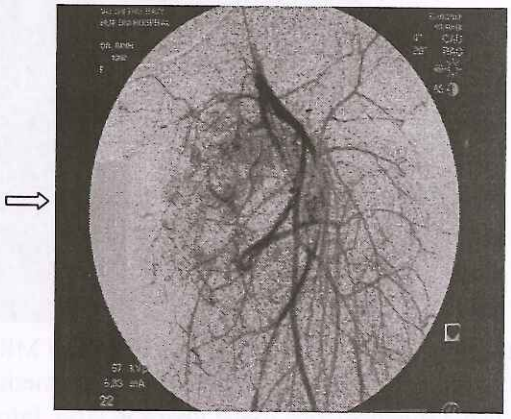

Figure 4. Common femoral angiogram shows hypervascular soft-tissue mass with a fine network of tumor vessels and inhomogeneous capillary blush (arrow) mainly supplied from the profunda femoral artery. Note the compression and spasm of the superficial artery (curve arrow).

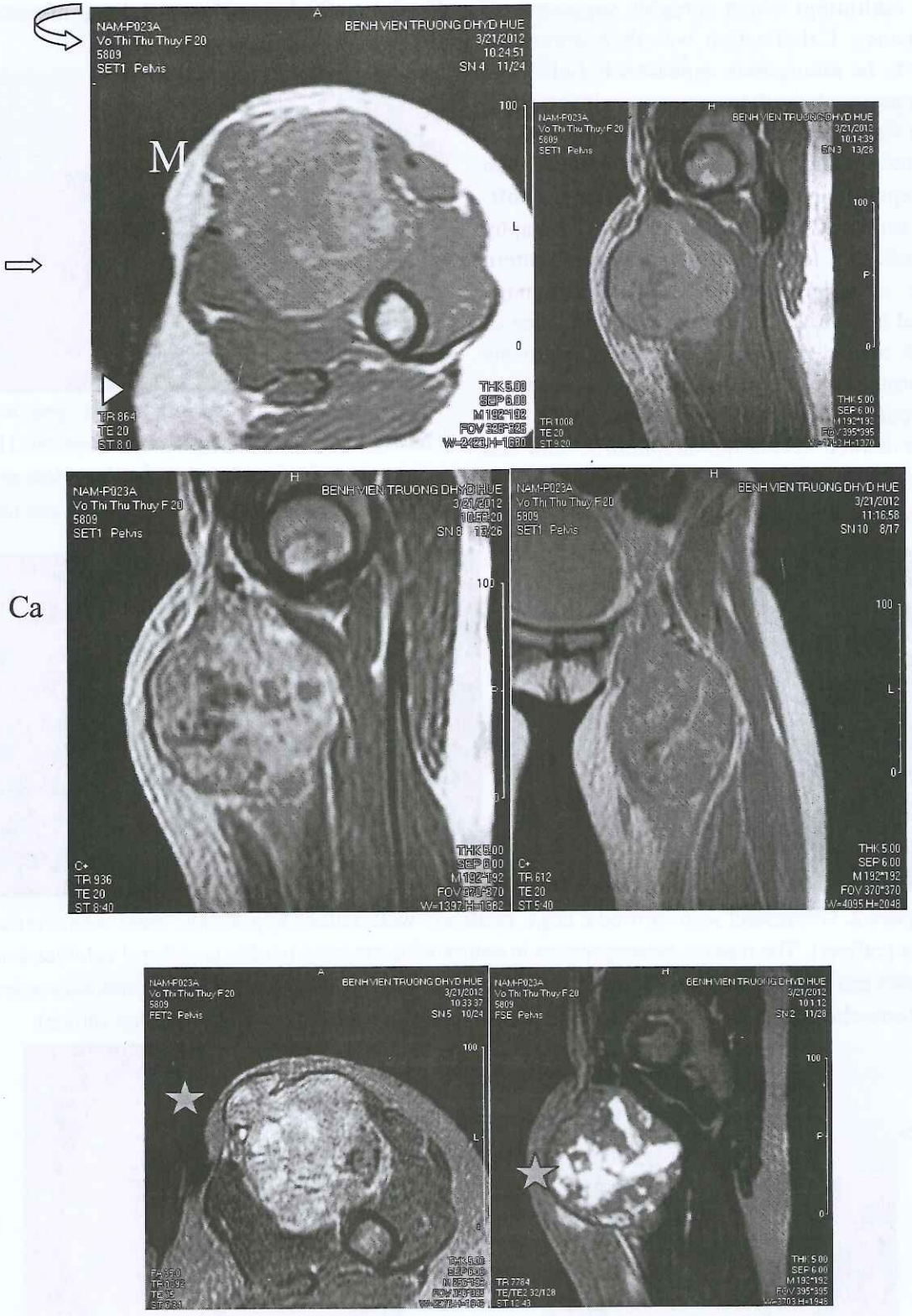

Figure 5. Sagittal and coronal T1 weighted MR images show a 8x9x10 cm well-defined, heterogeneous soft tissue tumor (M) which is hypo-to-intermediate signal intensity in the anterior left upper thigh with extensive subcutaneous tissue invasion (IN). Intravenous contrast-enhanced T1W shows heterogeneous predominant enhancement of the mass, reflecting the intermixture of nonenhancing necrotic, cystic or hemorrhage, and enhancing solid regions. On T2-weighted image, the mass contains strikingly hyperintense cystic areas (star) with irregular septa of intermediate signal intensity. Arrows indicate edema of the adjacent tissue. Note the peripheral rim of hypointensity on both T1 and T2 weighted images represent calcification (Ca) which is proved on X ray.

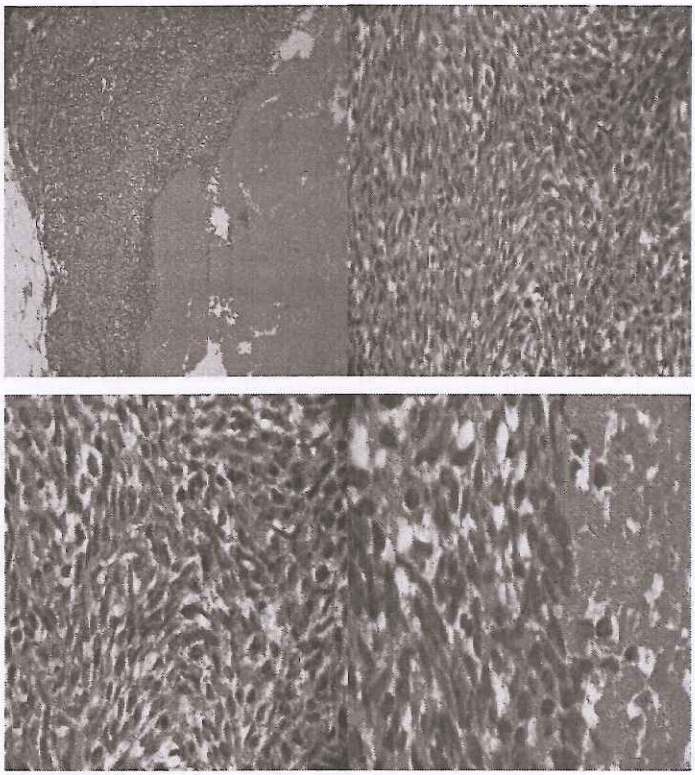

Figure 6. HE staining (x10, x100). Histopathological and Immunohistochemical study (not shown) confirm the diagnosis of monophasic synovial sarcoma, fibrous type.

3. DISCUSSION

Synovial sarcoma (SS) represents only 5% of malignant soft tissue sarcomas, but it is one of the most common soft tissue sarcomas in younger adult patients (15-35 years of age). Although in incident rate of SS peaks in the third decade of life, 30% of SS cases occur in patients younger than 20 years old [6]. Although the name of the lesions suggests an articular process, and although this tumor is often found near a joint, 90% of SS do not originate from a joint. Rather, as with all SS, the tumor is named by the predominant histologic differentiation, in this case tumor cells that resemble synovioblastic cell. There are three main subtypes of synovial sarcoma: biphasic, monophasic and poorly differentiated, among these monophasic is the most common (50-60%). Most SS occur in the lower extremity, especially at or distal to the knee. However, there is a broad spectrum of locations including head, neck, trunk, lung, esophagus, intestine, mediastinum and retroperitoneum [1]. Synovial sarcoma is an intermediate-to high-grade neoplasm with extensive metastatic potential. The major sites of metastatic spread are lungs, less often, lymph node, bone and bone marrow. The clinical course of SS is characterized by a high rate of local recurrence and metastatic disease. Estimated 5-year survival rare ranges from 36% to 76%, at 10 years is from 20% to 63% [7].

Clinically, synovial sarcoma appears as deep-seated, painless, slowly growing masses. Patients with SS usually present with a palpable soft-tissue mass or swelling. The long duration of symptoms and initial slow growth may simulate those of or give a false impression of a benign process. A definitive diagnosis can only be established by an adequate tissue biopsy, but radiological investigations may be useful to characterize this tumor. Radiographs appear normal in approximately 50% of cases of SS, particularly those with small lesions. SS detected at radiography typically appear as nonspecific, round to oval juxtaarticular soft-tissue masses. Calcification is identified in up to 30% of SS at radiography. These calcifications are often eccentric or peripheral within the soft-tissue mass and nonspecific in appearance. Involvement of underlying bone is common seen as extrinsic erosion of bone or periosteal reaction has been

reported in 11-20% of SS. The bone erosion often has an indolent nonaggressive appearance on radiographs, which can lead to misinterpretation of the lesions as representing a benign process. Angiographic descriptions of SS are limited with common findings are hypervascular and displacement of native vessels [1, 7].

The ultrasound appearance of SS has not been extensively reviewed. Common findings are focal, nodular, round or lobulated, solid mass but hypoechoic soft-tissue mass is suggestive of a more indolent, less aggressive process. Doppler US studies would be expected to demonstrate vascularity in the areas of viable tumor. Any large (> 5 cm), solid, non fatty, soft-tissue tumor should be considered sarcoma unless proven otherwise by biopsy [8].

The most common CT appearance of SS is that of a heterogeneous deep-seated soft-tissue mass with attenuation similar to or slightly lower than that of muscle, representing necrosis or hemorrhage areas. SS frequently demonstrates a multinodular morphology on CT scans. Contrast-enhanced CT shows heterogeneous enhancement in 89%-100% of cases. This feature is helpful for distinguishing those SS that initially appear as a cystic lesion or hematoma on precontrast images, as the heterogeneous enhancement pattern excludes these diagnoses. Nodular areas of enhancement may also be seen in these lesions. CT is also useful for detecting calcification and bone involvement [1, 9].

With its superior contrast resolution, MR imaging is the optimal radiologic modality for assessing the extent and intrinsic characteristics of SS for staging and diagnosis. On T1 weighted MRI, SS typically appears as a prominently heterogeneous multilobulated soft-tissue mass with signal intensity similar to or slightly higher than that of muscle. This signal heterogeneity has been described as the triple sign by Jones and co-workers, represented by intermixed areas of low (calcified or fibrotic collagenized regions), intermediate (mixture of cellular elements) and high signal intensity (hemorrhage or necrosis) on long repetition time images. Contrast-enhanced MRI can be particularly important for distinguishing SS with predominantly cystic characteristics with standard T1 and T2 weighted sequences [1].

Synovial sarcoma has three major histological subtypes: biphasic, monophasic and poorly differentiated types. The coexistence of epithelioid cells and spindle cells is a hallmark of the biphasic subtype, while the monophasic subtype is entirely composed of spindle cells. The poorly differentiated subtype is a variant that lacks spindle cells and comprises primitive small round cells. Immunohistochemically, most SS are positive for vimentin, cytokeratin and EMA, but have a lower immunoreactivity for S-100 and CD34 [10].

Approximately 25% of patients have metastases at presentation, frequently to the lungs. Local recurrence at metastatic disease occurs in 80% of patients. Other than the lung, metastases may involve bone, lymph nodes or soft tissues. Lesions with extensive calcification have a better prognosis. Other factors indicating a more favorable prognosis include tumor size < 5cm, those with a low mitotic activity, and tumors arising from the extremities. Other factors associated with a poorer prognosis are extensive hemorrhage (high signal on T1 and T2 images) and the present of a"triple signal" heterogeneous enhancement pattern on T2 images [7].

As with many primary malignant soft-tissue neoplasms, local control of SS is primarily achieved with surgery. The current treatment of choice is wide local excision (removal of the tumor, its pseudocapsule and a normal cuff of surrounding tissue). The surgical margins should be closely evaluated to determine the need for adjuvant therapy. Amputation should be reserved for those cases in which gross resection of the tumor and preservation of a functional limb is not possible. The role of adjuvant therapy remains controversial. Chemotherapy has been used to treat metastatic or residual disease. The agents employed are combinations of adriamycin, cisplatin, vincristine, doxorubicin, ifosfamide. Radiation therapy plays an important role in the treatment of marginally resected tumors. Imaging features associated with a poorer outcome include a large tumor size of >10 cm, absence of calcification and hemorrhage appearance [1,7].

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