A rare case of monophasic synovial sarcoma of the hand: Cytological and immunohistopathological study

Sir,

Soft tissue sarcomas of the hand are rare and challenging neoplasms. Hand surgeon usually comes across one or two undiagnosed cases of synovial sarcoma during their career. The combination of an indolent presentation and the overwhelming predominance of benign hand tumors often lead to a lack of urgency in obtaining a diagnosis and hence delay in treatment. Synovial sarcoma of the hand is notorious for recurrence and metastasis often with devastating results despite apparently complete resection. The decision about the extent of resection requires a balance between the requirement for negative resection margins, which is associated with reduced local recurrence and improved overall survival, and the functional requirement for greater preservation of native tissue, which improves functional outcome. Prompt diagnosis is vital in treating a potentially malignant soft tissue mass. Diagnosis of synovial sarcoma of the hand is based on histopathological and immunohistochemical (IHC) evaluation, which is absolutely necessary to determine the tumor type and for predicting biological behavior.

A 30-year-old female presented with swelling over her right palm which was increasing in size over the last 1-year. The swelling showed a sudden increase in size and became painful over the past 3 months. Local examination showed 10 cm × 6 cm, tender, ill-defined, firm swelling on the radial aspect of the right-hand thenar eminence. Overlying skin showed presence of scar of incisional biopsy. Systemic examination findings were noncontributory. Ultrasonography of the right hand done using a 5 MHz probe showed 5 cm × 4.4 cm ill-defined heterogenous mixed echogenic lesion at right thenar space. There was no evidence of calcification, and the bones appeared grossly normal with no evidence of erosion. Pre- and post-contrast multiplanar multiecho magnetic resonance imaging (MRI) of the right hand showed a large, well-defined heterogenous signal intensity lesion in the region of thenar eminence and extending in between the first and second metacarpals measuring 7.4 cm × 6.9 cm × 6.2 cm. The lesion was hyperintense on T2-weighted images and hypointense on T1-weighted images. Few areas of fat intensity were seen on the posterior aspect of the lesion. On postcontrast study, the lesion showed heterogenous enhancement. On palmar aspect, the lesion displaced the flexor tendons toward the medial side. Dorsally, it extended up to the skin. However, the skin was not breached. Medially, the lesion extended between the second and third metacarpal and along palmer aspect of third metacarpal, the latter’s inferior margins were scalloped. Laterally, the lesion extended adjacent to the first metacarpal and proximal phalynx of the thumb but did not cross the thumb onto its lateral aspect. Minimal fluid was noted in the carpometacarpal joint. Proximally, the lesion was seen extending to the level of distal carpal row trapezium and trapezoid. Distally, it was seen extending at the level of second and third metacarpophalangeal joint. Small soft tissue component of the lesion was seen extending into second metacarpal head region on the palmar aspect. Altered marrow signal intensity was visualized in proximal shaft of the second metacarpal with the postcontrast enhancement. Cortical breach was noted at multiple places along the lateral and volar aspects of the second metacarpal. Diffuse subcutaneous soft tissue edema was seen in the hand region. A radiological diagnosis of primary soft tissue neoplasm possibly liposarcoma was offered with lesion showing cortical involvement and marrow infiltration.

Fine-needle aspiration cytology (FNAC) was advised. FNAC revealed moderately cellular smears composed of clusters of spindle cells with nuclear atypia, hyperchromasia, and moderate amount of cytoplasm against a hemorrhagic background. Cytological report of spindle cell sarcoma was offered. Incisional biopsy was then done which showed evidence of spindle cell sarcoma.

In view of radiological and cytohistological findings, patient was taken up for below elbow right-hand amputation. Fish mouth incision was taken 8–10 cm below the right elbow. It was deepened in layers with muscles cut, arteries, nerves, and veins sutured and ligated. Radius and ulna were cut. Myodesis was done. Bones were covered with muscles. Drain was kept and suturing done. The specimen was sent for histopathological confirmation.

We received right hand below elbow amputation specimen. It showed an ill-circumscribed tumor on the thenar aspect of the right hand measuring 9 cm × 7.5 cm × 7.5 cm [Figure 1]. On the extensor surface, scar of incisional biopsy was noted. Cut section of the tumor showed ill-circumscribed fleshy, grayish white tumor. Representative sections were studied. Multiple sections examined from the swelling showed an ill-circumscribed tumor composed of monotonous spindle-shaped cells with elongated hyperchromatic nuclei and scanty cytoplasm [Figure 2a]. Mitosis was 1–2/high power field. No areas of necrosis were noted. Diagnosis of grade 3 spindle cell sarcoma was offered according to French Federation of Cancer Centers Sarcoma Group grading system and representative immunohistopathological study.
sections for IHC studies were submitted. The surgical resection margins were free of tumor.

Immunohistochemical study was performed with the following panel of antibodies viz., vimentin (clone V-9, Dako), cytokeratin-7 (CK-7) (clone OV-TL12/30, Dako), CK-19 (clone RCK108, Dako), epithelial membrane antigen (EMA) (clone E29, Dako), CD99 (clone 12E7, Dako), bcl2 (clone bcl2/100/D5), Ki 67 (clone MIB-1, Dako), desmin (clone 33, Dako), and S-100 (Leica) for diagnosing the lesion. The tumor cells showed strong cytoplasmic immunoreactivity for vimentin [Figure 2c] and focal immunoreactivity for EMA [Figure 2e]. They showed strong nuclear immunoreactivity for CD 99 and Bcl2 [Figure 2b]. They were nonreactive for CK-7 [Figure 2d], CK-19, desmin, and S-100. Ki 67 labeling index was found to be 60% confirming its malignant nature. Histopathological and IHC findings confirmed the diagnosis of monophasic synovial sarcoma. In view of free surgical resection margin, patient was not given radiotherapy or chemotherapy. Postoperatively, the patient is disease free and on follow-up, there is no evidence of recurrence for a period of 6 months.

Soft tissue sarcomas of the hand are rare and comprise 1% of the adult malignancies.[5] They have a higher recurrence rate and worse survival than sarcomas located in other upper extremity sites.[6]

The reported incidence for synovial sarcoma of the hand is only 8.5%.[7] Further, its location in the palm of the hand is exceedingly rare. It is located less commonly in the digits than carpus of the hand.[8] Synovial sarcoma has a wide histopathological spectrum and is defined as a distinct translocation positive sarcoma. The gold standard for diagnosis is molecular analysis method that identifies the characteristic t (X; 18) (p11.2, q11.2). Radiological investigations are important to know the extent of the tumor in planning surgery and the detection of mainly metastatic disease. MRI is the imaging modality of choice for diagnosis and staging of synovial sarcoma.[9]

The histologic spectrum of tumors of the hand is vast and uncommon subtypes tend to predominate. Histomorphological examination along with IHC studies helps to finally clinch the diagnosis and is mandatory for better patient management.[9] Monophasic and biphasic synovial sarcomas are the two major histological subtypes of synovial sarcoma depending on presence and absence of epithelial components. Apart from these, it has calcifying variant and poorly differentiated subtypes. Monophasic synovial sarcoma as in our case has to be differentiated from other spindle cell sarcomas such as fibrosarcoma, leiomyosarcoma and malignant peripheral nerve sheath tumor (MPNST). Monophasic synovial sarcomas show vimentin, EMA, CD99, bcl2 immunoreactivity and
absence of CK, desmin, and S100 immunoreactivity. A high ki-67 labeling index helps in confirming the malignant nature of synovial sarcoma. Transducin-like enhancer of split 1 a transcriptional repressor essential in hematopoiesis, neuronal differentiation, and terminal epithelial differentiation, has recently been shown in a single tissue microarray study to be a highly sensitive and relatively specific marker of synovial sarcomas.

Fibrosarcoma characteristically does not show EMA immunoreactivity. Leiomyosarcomas show immunoreactivity for desmin and bcl2. MPNST shows focal S100 immunoreactivity.

Presence of metastasis, tumor size > 5 cm, invasiveness, high histologic grade (based on mitosis and tumor necrosis), positive surgical margins, and poor histological differentiation are factors known to be associated with adverse clinical outcome. Only tumor size (>5 cm) is found to be consistently associated with a negative outcome.

The management of soft tissue sarcomas of the hand is a matter of controversy and presents challenges to both the surgeon and oncologist. It is one of the few adult chemosensitive spindle cell sarcomas. The current standard treatment is wide resection followed by polychemotherapy with or without irradiation. This approach is difficult to apply to the hand because of its anatomy and the lack of readily expendable soft tissue. The loss of important nerves, blood vessels, and tendons to gain an adequate margin may result in significant functional loss. Alternatively, aiming to preserve function by just removing the tumor itself may compromise the outcome. Furthermore, it has been reported that radiotherapy to the hand is poorly tolerated and associated with an increased risk of long-term toxicity and poor outcome.

Our experience with the present case highlights the significance of prompt detailed clinical, radiological, and immunohistopathological analysis in the diagnosis of soft tissue lesions of the hand in young patients. Synovial sarcoma should be considered in patients with a slowly growing, painless swelling in the hand.

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