Case Report

Synovial Sarcoma of Palm Masquerading as a Neural Tumor: A Challenging Differential Diagnosis

Abstract

Synovial sarcoma is one of the common soft-tissue tumors of the body and is usually found on the lower extremity, head-and-neck regions. Reports of monophasic variant of synovial sarcoma in the palm are a rare entity. It can present as a challenge for the surgeons mimicking other conditions. Very few cases of this malignancy are reported in the palm. Painful palmar mass is an unusual presentation for this variant of sarcoma. Preoperative suspicion and planned surgical approach pave the way for the proper management. Neoplasm of the hand is very challenging, as it needs surgical resection balancing with resection margin and functionality of the hand. These cases are notorious for late recurrences and metastasis. Long-term follow-up is of utmost importance. We are reporting a case of palmar monophasic synovial sarcoma with its management and follow-up.

Keywords: *Hand sarcoma, monophasic sarcoma, soft-tissue sarcoma, synovial sarcoma*

Durga Karki, Suvashis Dash, Vamseedharan Muthukumar

Department of Burns, Plastic and Maxillofacial Surgery, VM Medical College and Safdarjung Hospital, New Delhi, India

Introduction

Among soft-tissue tumor of the hand, synovial sarcoma is a rare diagnosis, but it carries a poor prognosis. There are only few cases of palmar synovial sarcoma described in the literature.[1] These present with vague clinical and nonspecific imaging features, which often lead to misdiagnosis.[2] In hand surgery, in view of late recurrences and notorious metastasis, there is a dilemma between choosing the optimum resection versus functionality of the hand, creating a clinical challenge for the surgeon.

Case Report

We are reporting a case of 45-year-old female with the complaints of painful swelling in the right palm for the past 8 months and had difficulty in making a fist. The size of the swelling increased gradually over the duration. On examination, a firm, tender, and nonpulsatile swelling of approximately 2 cm × 3 cm in size was felt in the center of the right palm and skin was free [Figure 1a]. There were paresthesia and weakness in grip strength.

Preoperative X-ray of the right hand showed a soft-tissue shadow without bony involvement. Preoperative magnetic resonance imaging (MRI) showed a well-defined lesion with intermediate-to-hyperintense signals on T1, hypointense signals on T2, and bright signals on the short-tau inversion recovery images. On the postcontrast study, the lesion showed mild peripheral enhancement. The lesion located in the subcutaneous plane of flexor compartment measured about 3 cm × 2 cm × 1.5 cm in size and did not involve extensor compartment [Figure 1b]. The Doppler study was also done to aid in the diagnosis that revealed a well-defined hypoechoic lesion with little to no vascularity over the palmar region. The radiologist gave a provisional diagnosis of infected ganglion cyst or neural tumor.

Excision of the tumor was done under general anesthesia. Tumor was found to be close to the under the surface of the common digital branches of the median nerve. The nerve was stretched over the tumor but was easily separated. Tumor was well-circumscribed and was dissected from the nerve [Figure 2]. The wound was primarily closed.

A 3 cm × 2.5 cm × 1.5 cm soft, grayish tissue was sent to the pathologist, which on microscopy showed well-encapsulated tumor comprising elongated to spindle-shaped cells arranged in intersecting fascicles and bundles. These cells had oval chromatin nuclei with blunt ends, vesicular

How to cite this article: Karki D, Dash S, Muthukumar V. Synovial sarcoma of palm masquerading as a neural tumor: A challenging differential diagnosis. Int J App Basic Med Res 2019;9:176-8.

Received: 06 July, 2018.
Accepted: 05 April, 2019.

Address for correspondence:
Dr. Durga Karki,
Department of Burns, Plastic and Maxillofacial Surgery,
VM Medical College and Safdarjung Hospital,
New Delhi - 110 029, India.
E-mail: dkarki10@yahoo.co.in

Access this article online

Website: www.ijabmr.org

DOI: 10.4103/ijabmr.IJABMR_187_18

Quick Response Code:

chromatin, and inconspicuous nucleoli. Mitotic activity varied from 2 to 4/10 high-power field with few atypical forms. On IHC, tumor cells were diffusely positive for vimentin, focally positive for cytokeratin, and negative for S10, CD 34, and desmin [Figure 3a and b]. These findings were suggestive of monophasic variant of synovial sarcoma.

Postoperatively patient underwent regular physiotherapy and did not have metastatic lesions on further workup. Chemotherapy was planned, but she did not consent for it. She is on follow-up for the past 1 year on outpatient basis.

Discussion

Synovial sarcoma in contrast to its name does not arise from the synovial cells, and rather it arises from the undifferentiated mesenchymal cells. These cells can differentiate into epithelial or mesenchymal components, which are the basis of classification of synovial sarcoma.[3] However, all the cells of synovial sarcoma have the typical molecular marker, i.e., t(X; 18) (p11,2;q11,2) translocation.[4]

Synovial sarcoma is most commonly found in lower extremities, head-and-neck regions. Tendon sheath, bursa, and joint capsule are the areas most commonly associated with synovial sarcoma. The articular surfaces of the joints are less likely involved.[3]

Male predominance is seen in cases of synovial sarcoma; but in synovial sarcoma of the hand, there is no sex predilection. It commonly affects 15–45 years age group of patients.[4] Harjai reported a case in a 6-year-old male child which is the youngest reported case, whereas Casal *et al*. reported a case of synovial sarcoma of the hand in a 63-year-old female which is the oldest age of presentation.[1,5] The hand is a rare site for synovial sarcoma with only 1% of synovial sarcoma occurs in hand. Clinically, they present as a slow-growing tumor. In the reported literature, the average period for the diagnosis is 1–2 years. They present mostly as a painless mass; only a few cases have been reported to be painful.[1,6] Casal *et al*. reported a case of synovial sarcoma of the palm, which presented as painful slow-growing swelling, as it was seen in our case.[1] In most of the cases, there is a delay in clinical diagnosis due to lack of any specific clinical features and their rarity. MRI is the investigation of choice to know the size, extent, preoperative planning, and follow-up. In MRI, T1 images of the lesion show hypo-to-isointense signals, whereas T2 images show hyperintense signals.[4]

The synovial sarcomas have several variants on the basis of cellular components. It may be monophasic, biphasic, or poorly differentiated. The biphasic variant of the tumor is also called the classic variant, has both the epithelial and mesenchymal components. It is necessary to use

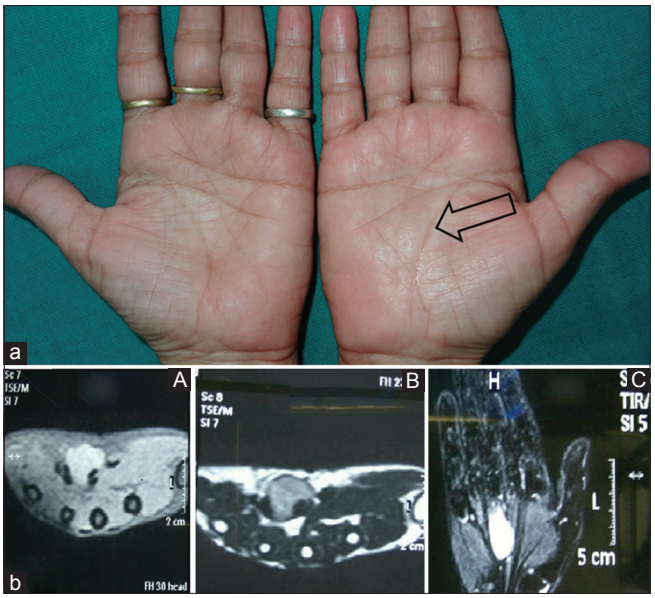

Figure 1: (a) Fullness over palmer surface of the right hand seen as compared to that of the left palm (shown by arrow). (b) Magnetic resonance imaging of the right hand (A) T1 images shows the hyperintense lesion, (B) T2 shows hypointense, and on (C) short-tau inversion recovery images, it is brightly seen. Peritumor planes are free

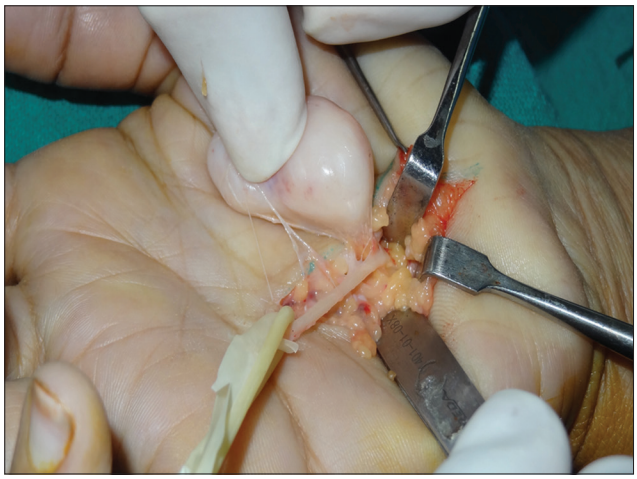

Figure 2: Intraoperative picture showing well-circumscribed tumor and the median nerve is separated from the mass shown looped with rubber band

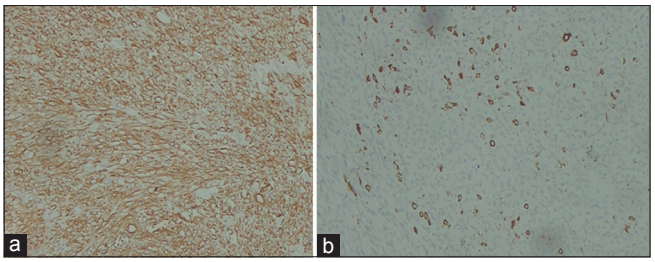

Figure 3: (a) Immunohistochemistry of the tumor showing positive for vimentin. (b) Immunohistochemistry of the tumor showing focal positive for cytokeratin

Immunohistochemistry for the diagnosis of these tissue variants. Monophasic synovial sarcoma stains positively for vimentin, cytokeratin, Epithelial membrane antigen

(EMA), and bcl2. In our case, the tumor was positive for vimentin and cytokeratin and negative for s-100, CD34, and desmin differentiating from the other types of sarcoma.[7] Arumugam *et al*. reported another atypical presentation of monophasic synovial sarcoma which had mucin-filled vacuoles in the epithelioid type of monophasic synovial sarcoma.[8] A new marker for synovial sarcoma is Transducin-like enhancer of split 1 (TLE1) which is now shown to be more sensitive and relatively more specific in single-tissue array.[7]

The management of synovial sarcoma is difficult due to factors such as local recurrences and distant metastasis. In terms of metastasis, the lung is the most common site. Various factors such as size >5 cm, older patients, tumor location over the trunk, bone and neural invasion are associated with poor prognosis. From pathological point of view, high proliferative index, overexpression of p53, and SYT-SSX fusions are high risk for recurrences.[9]

Surgery is the treatment of choice. Due to microinfiltration of the tumor cells in pseudocapsule, it is notorious for recurrences. Although controversy regarding the margin of resection is present, 1–2 cm margin is usually accepted. The important point is preoperative planning, the prognosis of disease deteriorates in unplanned excisions. Puhaindran and Athanasian, in their article, summarized the need for amputation for better control of the disease.[10] The importance of flaps cannot be ignored from the reconstruction point of view. Some cases with incomplete resection margins need adjuvant therapy like radiotherapy or isolated limb perfusion. Multimodality therapy is now followed in a lot of centers for synovial sarcoma, but the efficacy of these therapies is still under debate. Metastasis is an indicator of poor outcome. With its presence, 5-year survival rate decreases to around 30%.

Conclusion

Synovial sarcoma though a rare diagnosis can be deadly. Patients presenting with slow-growing swellings in hand should be approached with a high degree of clinical suspicion keeping this diagnosis in mind.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Casal D, Ribeiro AI, Mafra M, Azeda C, Mavioso C, Mendes MM, *et al.* A 63-year-old woman presenting with a synovial sarcoma of the hand: A case report. J Med Case Rep 2012;6:385.
2. Siegel HJ, Sessions W, Casillas MA Jr., Said-Al-Naief N, Lander PH, Lopez-Ben R, *et al.* Synovial sarcoma: Clinicopathologic features, treatment, and prognosis. Orthopedics 2007;30:1020-5.
3. Labow BI, Rosen H, Greene AK, Lee WP, Upton J. Soft tissue sarcomas of the hand: Functional reconstruction and outcome analysis. Hand (N Y) 2008;3:229-36.
4. Weiss SW, Golgblum JR. Malignant soft tissue tumors of uncertain type. In: Weiss SW, Goldblum JR, editors. Enzinger and Weiss's Soft Tissue Tumors. 5th ed. Philadelphia: Mosby; 2008. p. 1161-220.
5. Harjai MM, Bal RK, Nagpal BM, Sadhotra IP, Maudar KK. Synovial sarcoma of the hand. Indian Pediatr 1999;36:194-7.
6. Siddhi Gaurish SK, Avinash J, Kulkarni M, Patel DS, Zode AB. A rare case of monophasic synovial sarcoma of the hand: Cytological and immunohistopathological study. Oncol Gastroenterol Hepatol Rep 2015;4:116-8.
7. Dreyfuss UY, Boome RS, Kranold DH. Synovial sarcoma of the hand – A literature study. J Hand Surg Br 1986;11:471-4.
8. Arumugam VG, Sundaram S, Ramesh A, Duvuru P, Rajappa S. A tumour in disguise in the right palm-monophasic synovial sarcoma. J Clin Diagn Res 2016;10:ED03-5.
9. Talbot SG, Mehrara BJ, Disa JJ, Wong AK, Pusic A, Cordeiro PG, *et al*. Soft-tissue coverage of the hand following sarcoma resection. Plast Reconstr Surg 2008;121:534-43.
10. Puhaindran ME, Athanasian EA. Double ray amputation for tumors of the hand. Clin Orthop Relat Res 2010;468:2976-9.