Case Report

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Abdominal Wall Synovial Sarcoma, a Rare Presentation

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Citation Nazar E, Shabanzade Z, Ahmadi A. Abdominal Wall Synovial Sarcoma, a Rare Presentation. Case Reports in Clinical Practice. 2022; 7(3): 101-104.

Running Title Abdominal Wall Synovial Sarcoma, a Rare Presentation

Article info:
Received: 21 March 2022
Revised: 09 April 2022
Accepted: 28 April 2022

Keywords:
Synovial sarcoma; Abdominal wall; Surgery

ABSTRACT

Synovial sarcoma is a malignant mesenchymal neoplasm which com- monly arises in the extremities of adults, in close association with joint capsules. Only a few cases of synovial sarcoma occurring in the abdominal wall have been reported. We report an extremely rare case of monophasic synovial sarcoma of abdominal wall in a 58-year-old woman who had presented with painless left ante- rior abdominal wall mass. The patient underwent excisional surgery. Histological and immunohistochemistry examinations revealed monophasic synovial sarcoma. Base on the diagnosis, the patient received chemoradiation.Primary synovial sarcoma is rarely found in the anterior abdominal wall. But, it should be included in the differential diagnosis.

Introduction

Sarcomas are malignant tumors that originate from cells of mesenchymal origin. Synovial sarcoma is one of the subtypes of soft tissue sarcomas [1]. The majority of soft tissue sarcomas are existent in the extremities; however, many other sites can be affected, including the retroperitoneum, abdominal and chest wall, head and neck and subcutaneous tissues [2]. Only, 10–20% of sarcomas are abdominal sarcomas and the overall incidence is 0.3–0.4% per 100 000 of the population. The peak incidence is in the fifth decade of life, although they can happen in any age group [3]. Synovial sarcoma (SS) is a malignant mesenchymal neoplasm with variable epithelial differentiation and with a tendency to occur in young adults, it is usually regarded and treated as a high-grade sarcoma [4]. Herein, we report a monophasic synovial sarcoma originating from the anterior abdominal wall to raise awareness about abdominal wall mass.

Case presentation

A 58-year- old female who presented in the department of surgery in Fajr hospital in Kordestan in Iran with a two months' history of painless

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superficial anterior abdominal mass. At first, outpatient examination finding was unremarkable. The patient's family history and past medical history was unremarkable. General examination was negative except for a movable mass (5cm in greatest diameter) in the left lower anterior abdominal wall. No other masses were detected anywhere else. Routine laboratory investigations were normal. The patient underwent sonography, which showed a solid and hypoechoiec lesion with high vascularity on abdominal oblique muscle. Multisequential multiplanar abdominal MRI (Magnetic Resonance Imaging) with and without contrast showed a fine border lesion with high vascularity but without fat intensity between external and intermediate abdominal wall muscles at left iliac level without enhancement, no invasion to superior or inferior muscle layer. Therefore, clinical diagnosis of sarcoma of the abdominal wall was virtually impossible. Diagnosis required histopathologic examination. The patient was treated by excisional surgery. She well tolerated the procedure without complications. Received specimen for pathology examination consists of multiple pieces of tan soft tissue totally measuring 5x4x2.5cm. Sections show white tan appearance with hemorrhagic areas. Histological examinations revealed a highly cellular neoplastic tissue composed of cells with spindle to oval nuclei and scant cytoplasm which had herring bone pattern. Areas of hemorrhage and necrosis were observed. Mitosis was found easily (Fig. 1). The main differential diagnosis was malignant spindle cell sarcomas including fibrosarcoma and leiomyosarcoma. The specimen sent to Sina hospital pathology laboratory in Tehran for supplementary evaluation. Immunohistochemistry (IHC) staining showed positive reactivity for transducin-like enhancer of split-1 (TLE-1) and BCL-2 and negative reactivity for cytokeratin (CK), smooth muscle actin (SMA) and CD-34. Proliferative activity (Ki-67) was 40% (Fig. 2). As a result, our case was compatible with monophasic synovial sarcoma and diagnosis was confirmed. Base on the diagnosis, the patient received chemoradiation. After 6 months of follow-up examinations, no recurrence was observed and the patient is still asymptomatic.

Discussion

Synovial sarcoma (SS) is an infrequent soft tissue malignant tumor that is common in the extremities of middle aged patients, adjacent to large joints predominantly the knee in the popliteal fossa [5]. SS rarely originates within the abdomen and pelvis [6]. In 1950, Pack and Ariel reported the first two cases of synovial sarcoma originating in the abdominal wall [7]. The anterior abdominal wall is a rare setting for synovial sarcoma growth and, in the English language literature, 44 cases have been published between 1950 and 2005 [8]. Because most explanations from this situation consist of case reports, neither clinical nor prognostic features have been well described [9]. Moreover, the presence of SS in extra-articular locations such as the abdominal wall claims against an origin from arthrogenous mesenchyme [10]. SS in the abdominal wall tend to happen with a much greater frequency in females in contrast to such tumors in the extremities or the neck which tend to happen with a much greater frequency in males [11]. The three histologic subtypes are monophasic, biphasic and the poorly differentiated type. The monophasic subtype mainly consists of spindle cells reminiscent of fibrosarcoma which was similar to our case. Differential diagnosis

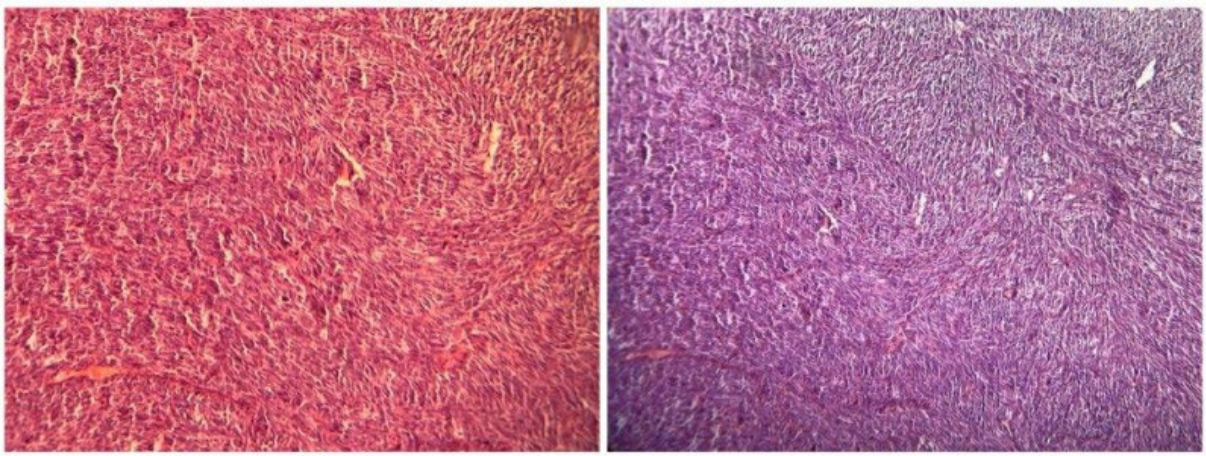

Fig. 1. Section showed a highly cellular neoplastic tissue composed of cells with spindle to oval nuclei and scant cytoplasm which had herring bone pattern (H&E, x200)

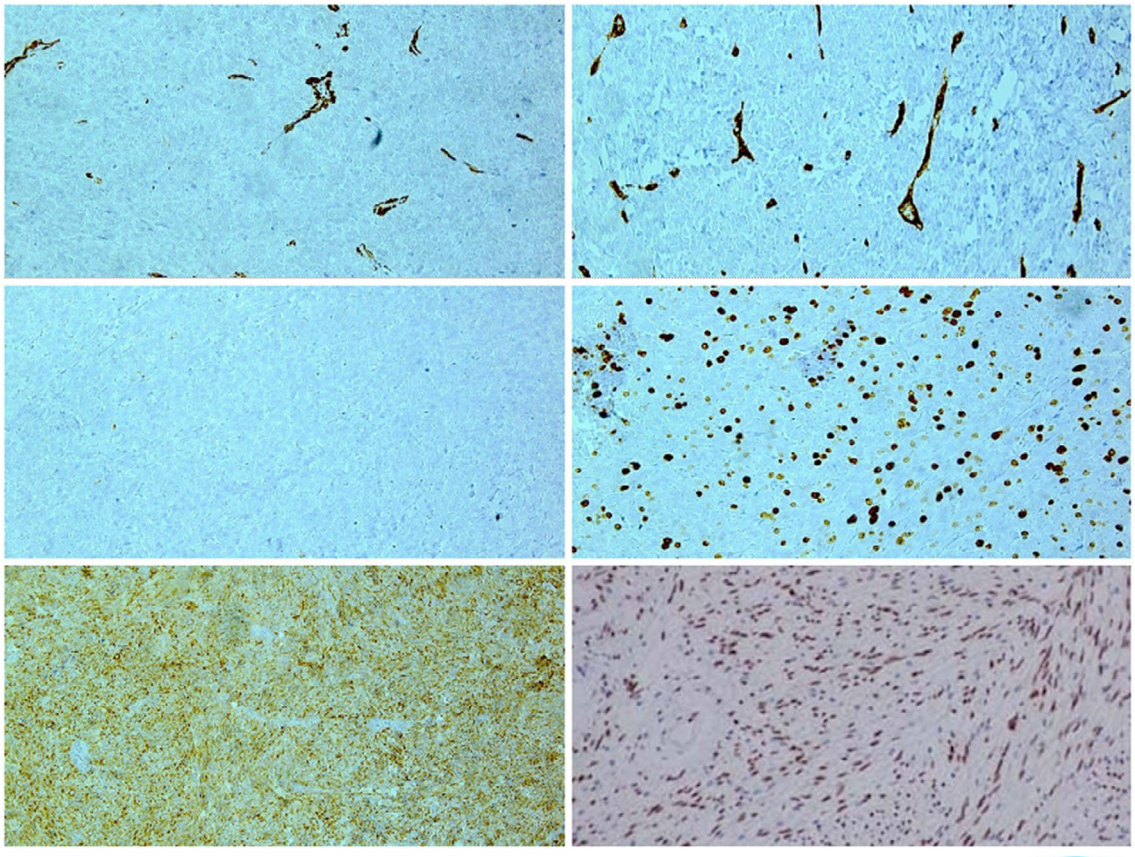

Fig. 2. IHC staining showed (left upper) SMA positive reactivity only blood vessels, (right upper) CD34 positive reactivity only blood vessels, (left middle) CK negative, (right middle) Ki67 about 40%, (left lower) BCL2 positive reactivity and (right lower) TLE-1 positive reactivity (x200).

of abdominal wall tumors in adolescents and young adults includes desmoid tumor (for benign tumors), mesenchymal chondrosarcoma and leiomyosarcoma (for malignant tumors) which in our case excluded by IHC study [12]. Finally, the probability of a metastases to the abdominal wall from a SS of soft tissue should always be ruled out first before making a diagnosis of primary SS at this site [7]. In the present case, the patient did not have a history or evidence of tumor elsewhere (on clinical or radiographic examination), which supported its primary origin at this site. The characteristic t(X;18)(p11.2;q11.2) translocation is a cytogenetic hallmark of SS which presents in nearly all SS and does not occur in other forms of sarcomas [13]. It should be noted that it is an expensive test. In the present case, we confirmed the diagnosis by histology and IHC study, so the identification of a t(X,18) translocation was not performed. Surgical resection is the primary treatment of SS and is supposed to be relatively more chemosensitive than other soft-tissue sarcomas [14]. In the present case, the patient received a complete excision as the tumor had a clear tumor-free margin and underwent chemoradaition. But, it should be included in the differential diagnosis. According to Fisher et al., (https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3041044/ - B1) average survival rates were only 17 months with a high incidence of recurrence [15]. However, it could be understand that SS may be developed in unusual locations (e.g. abdominal wall) even if there is no true synovial cell. Thus, more genetic, pathologic and IHC staining studies are needed for definitive diagnosis in suspicious lesions.

Conclusion

Primary SS is rarely found in the anterior abdominal wall. Nonetheless, it should be included in the differential diagnosis when an adolescent presents with abdominal wall mass.

Ethical Considerations

Consent

The consent was taken from the patient for the case report to be published.

Compliance with ethical guidelines

There were no ethical considerations to be considered in this research.

Funding

This research did not receive any grant from funding agencies in the public, commercial, or non-profit sectors.

Authors' contributions

All authors equally contributed in preparing this article.

Conflict of interest

Authors declare that there is no conflict of interest.

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