Case Reports in Oncology

Case Rep Oncol 2024;17:217–224
DOI: 10.1159/000536125
Received: December 8, 2023
Accepted: December 28, 2023
Published online: February 8, 2024

Case Report

A Rare Case of High-Grade Spindle Cell Sarcoma of the Breast: A Case Report

Zoaib Habib Tharwani[a] Zehra Habib[b] Yumna Ahmed[b] Samreena Ishrat[c] Abdulqadir J. Nashwan[d]

[a]Faculty of Medicine, Dow Medical College, Dow University of Health Sciences, Karachi, Pakistan; [b]Usman Memorial Hospital, Karachi, Pakistan; [c]Shaheed Mohtarma Benazir Bhutto Medical College Lyari, Karachi, Pakistan; [d]Hamad Medical Corporation, Doha, Qatar

Keywords
Spindle cell carcinoma · Breast cancer · Case report · Phyllodes tumor

Abstract
Introduction: Spindle cell sarcomas are rare breast lesions which are difficult to diagnose due to resemblance with other breast lesions. Histopathological examination and immunohistochemical staining are essential for diagnosis. ***Case Presentation:*** We present a rare case of a 15-year-old female presenting with high-grade rapidly progressive spindle cell sarcoma of the breast, differentiated as phyllodes tumor, with axillary lymph node involvement. Her lesion, on the left breast, measured 16.9 × 10.1 × 13.7 cm. Histology revealed malignant neoplasm arranged in sheets and individual neoplastic cells with an epithelioid to spindled morphology with scant cytoplasm and irregular nuclear membranes. Immunohistochemistry showed weakly positive focal CD-99, and negative WT-1, Myogenin, Desmin, p63, Cytokeratin, Synaptophysin, and CD-34 markers. She was successfully managed with modified radical mastectomy and discharged with regular follow-up advised. ***Conclusion:*** Spindle cell sarcomas have a very aggressive course and prompt diagnosis, and management is mandatory for better patient outcomes. Modified radical mastectomy is the mainstay of treatment.

Introduction

Spindle cell sarcomas are rare lesions of the breast which range from benign to high-grade malignant carcinomas, with a prevalence of less than 0.5% per year [1–3]. Differential diagnosis of these lesions includes sarcomatoid/metaplastic carcinoma,

Correspondence to:
Abdulqadir J. Nashwan, anashwan@hamad.qa

Case Rep Oncol 2024;17:217–224
DOI: 10.1159/000536125

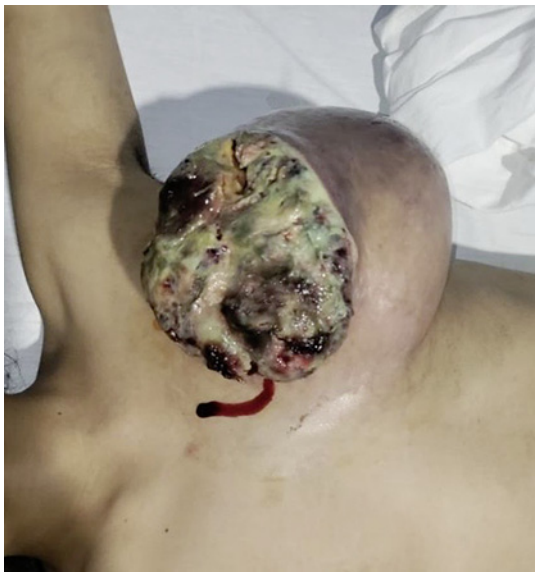

Fig. 1. Gross image of the lesion on previous visit (10 days before admission).

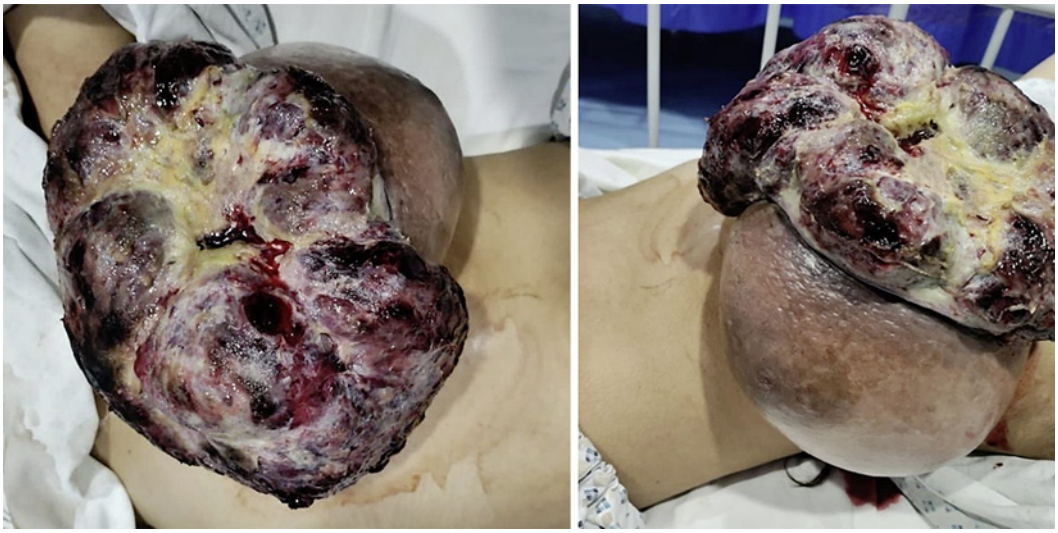

Fig. 2. Gross image of the lesion on current visit.

primary breast sarcoma, and phyllodes tumor (PT) [4]. Diagnosing this lesion can be extremely challenging due to similarity with other breast lesions and mixed morphological patterns on histology; however, histological examination of trucut biopsies along with immunohistochemical staining has made it easier to differentiate spindle cell sarcoma from other breast lesions [1, 2]. This article presents a rare case of a 15-year-old female with a high-grade spindle cell sarcoma with the aim of enhancing clinical awareness to promote early detection and better management plans for such rare cases. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material 1 (for all online suppl. material, see https://doi.org/10.1159/000536125).

Case Rep Oncol 2024;17:217–224
DOI: 10.1159/000536125

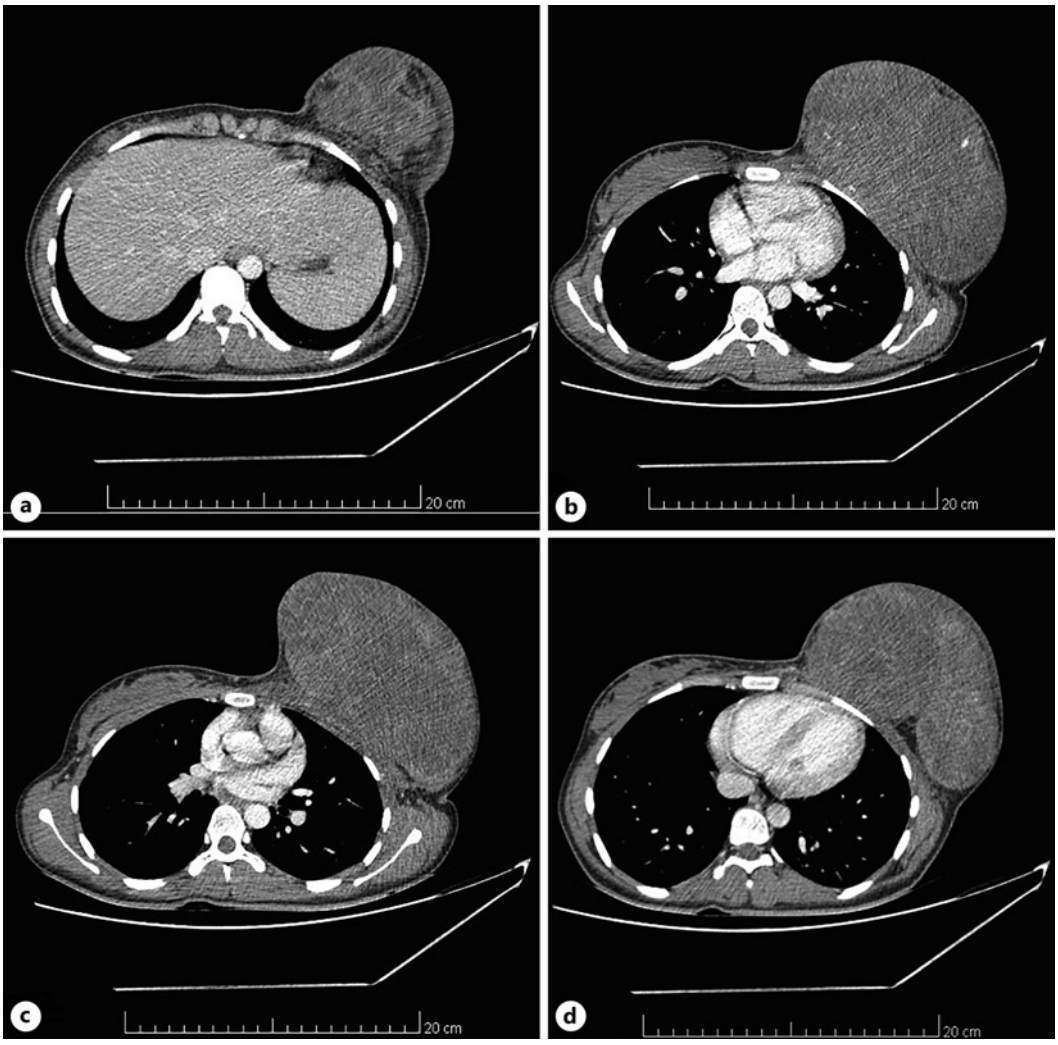

Fig. 3. a–d CT scan with contrast.

Case Presentation

A 15-year-old female with no known comorbidities presented with the complaint of a gradually progressive lesion on her upper left breast. The patient previously visited our hospital 10 days back but opted out of further intervention, however, the lesions rapidly progressed (Fig. 1, 2) and she currently visited the ER with the complain of purulent discharge and bleeding from the lesion for the past few days. She also complained of fever for 2 days which was undocumented, intermittent, with no rigors or chills and no other associations.

Examination revealed a coconut-shaped erythematous mass on the left breast with no dimpling or nipple retraction (Fig. 2). The mass was warm and tender on touch and there was no nipple discharge. Imaging involved CT with contrast and HRCT which were performed 22 days apart, HRCT being the recent one. On CT scan with contrast (Fig. 3), a large heterogeneously enhancing soft tissue density multilobulated mass was seen, which occupied nearly all left breast parenchyma. Anteriorly, the mass was inseparable from the adjacent skin and posteriorly from the anterior chest wall muscles. The mass approximately measured 16.9 × 10.1 × 13.7 cm. Left axillary lymph nodes were found out to be enlarged, with the largest lymph node measuring approximately 1.8 × 1.3 cm. Few sub-centimeter lymph nodes

Case Rep Oncol 2024;17:217–224
DOI: 10.1159/000536125

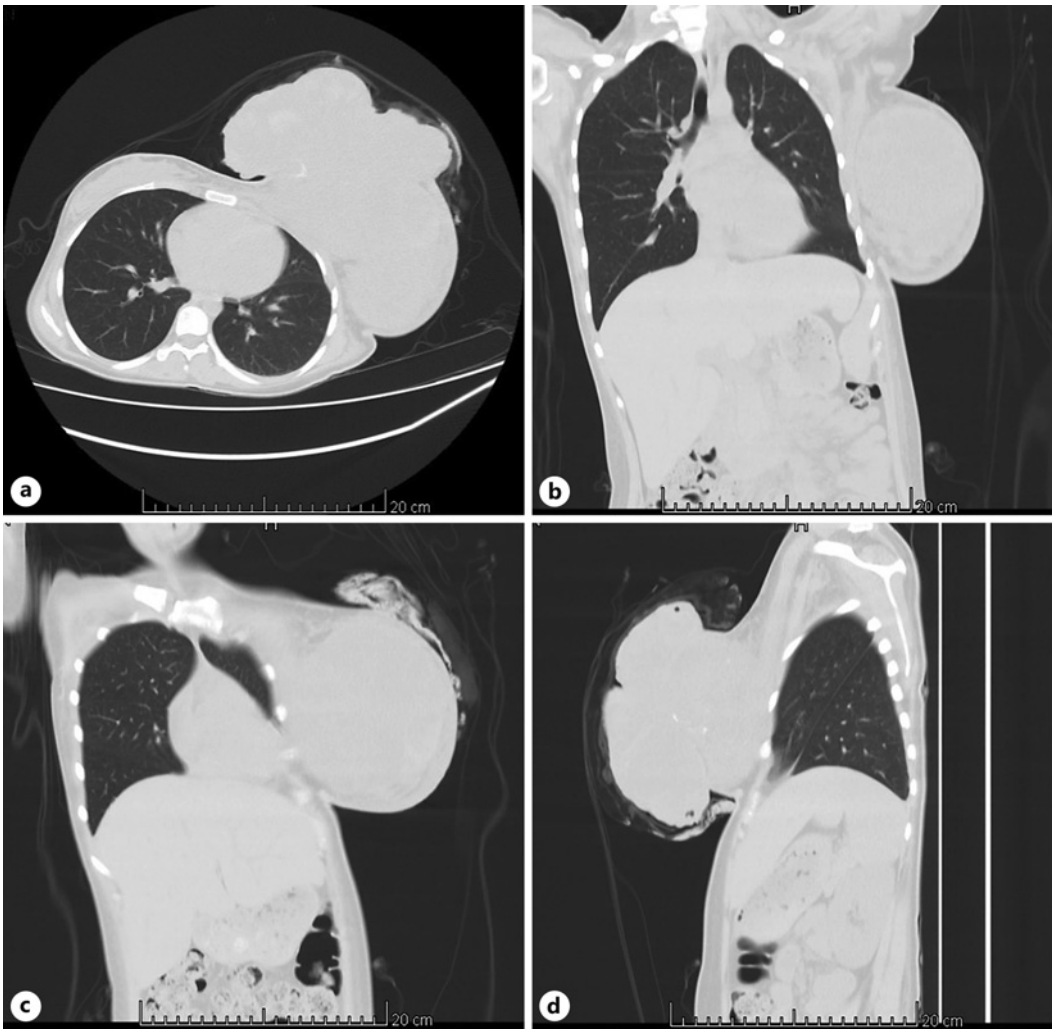

Fig. 4. HRCT scan: (**a**) axial view; (**b** + **c**) coronal view; (**d**) sagittal view.

were also identified in the right axilla with cortical thickness measuring approximately 0.4 cm. No evidence of bony or pulmonary metastasis was seen on non-enhancing images. HRCT showed grossly enlarged heterogeneous lobulated exophytic mass with overlying skin ulceration and multiple coarse calcifications (Fig. 4). The mass measured approximately 14.0 × 22.6 cm on axial images, showing rapid progression when compared with the dimensions on CT with contrast 22 days back. The mass was posteriorly inseparable from the underlying pectoralis muscle and enlarged left axillary lymph nodes were also observed with the largest lymph node measuring approximately 1.5 × 1.2 cm. Trucut needle biopsy was performed to confirm the diagnosis of high-grade spindle cell sarcoma.

Histopathological specimen was transferred in formalin with intact core in multiple fragments with separately present piece of tissue. The cores ranged in size from 1.3 × 0.1 cm to 0.1 × 0, 1 cm. Separately present fragment of tissue measured 2.0 × 1.0 × 0.5 cm. Sections were taken in five blocks as A1-5. Histological examination of sections from the cores revealed a malignant neoplasm arranged in fascicles with intervening epithelial elements. Few areas showed stromal overgrowth with increased cellularity. The individual neoplastic cells in these areas displayed an epithelioid to spindled morphology, moderate to severe nuclear pleomorphism, and hyperchromasia with scant cytoplasm and irregular nuclear membranes,

Case Rep Oncol 2024;17:217–224
DOI: 10.1159/000536125

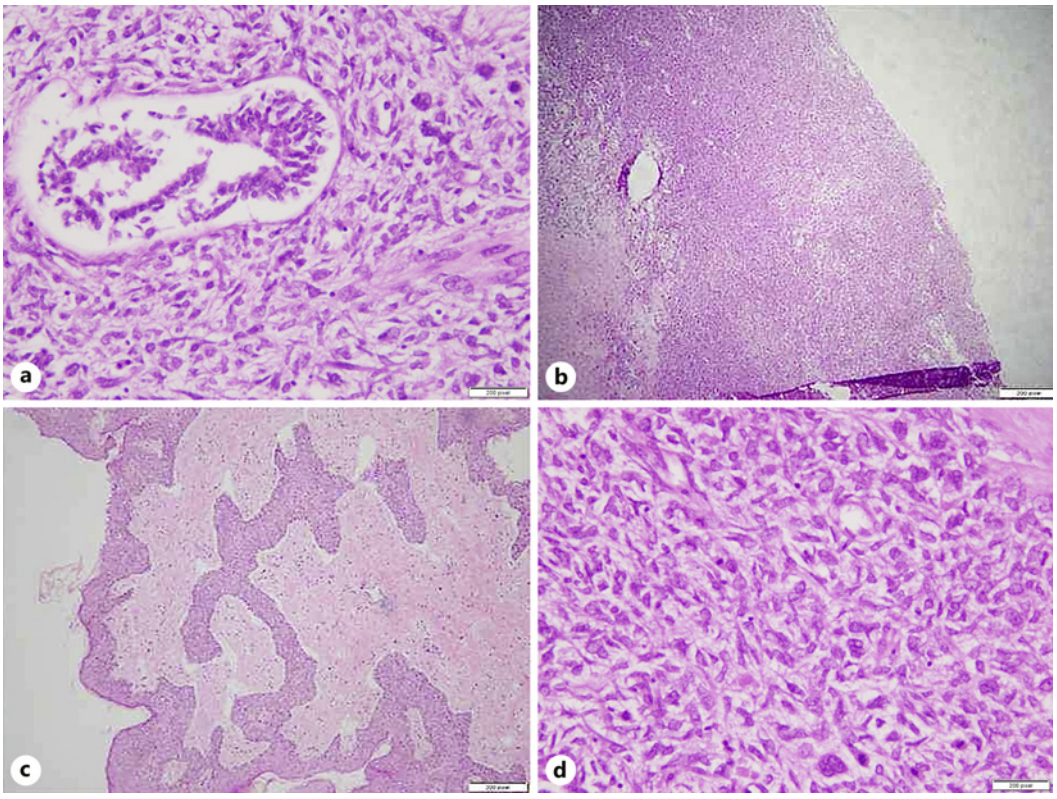

Fig. 5. a–d Histopathological images of the mass.

Figure 5 shows the histological images of the biopsy. Brisk atypical mitosis (up to 12/10 H.P.F) was also observed. Areas of necrosis are seen. No heterologous elements are noted. Sections from the separately present fragment revealed fibroconnective tissue with acute necrotizing inflammation along with areas of infarction. Scant areas showed atypical cellular proliferation of the abovementioned morphology. Overlying skin was ulcerated. Benign ducts entrapped in malignant neoplasm were noted. Nipple areola complex and all resection margins are free of tumor.

Immunohistochemical Staining Markers of the Mass Are Shown

The patient underwent modified radical mastectomy (MRM) of the left breast and was monitored for recovery and any complications, Figure 6 shows the postoperative image(s). During her stay, the patient also received two blood transfusions and was monitored for fever or other complications associated with transfusions. An echocardiography was also ordered after transfusion which came out normal. Medications plan for the patient included ceftriaxone, ciprofloxacin, and tranexamic acid. The patient was discharged and advised regular follow-up for reconstruction and recurrence management Table 1.

Discussion

Most common type of cancer among women is breast cancer, with 2.3 million cases diagnosed annually and 684,996 deaths [5, 6]. Spindle cell sarcomas of the breast are a type of mesenchymal tumors of the breast which are extremely rare, with an incidence of 4.6 cases per million women per year [4]. These lesions can be arduous to diagnose due to similarity

Case Rep Oncol 2024;17:217–224
DOI: 10.1159/000536125

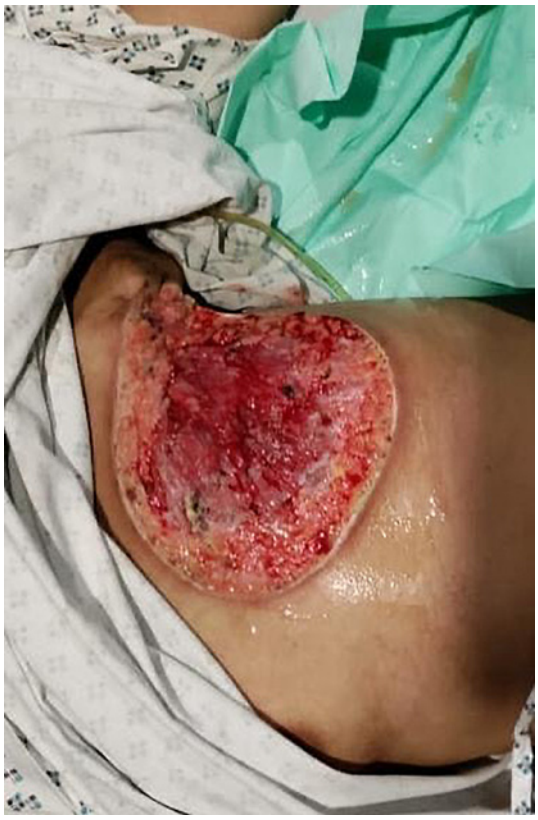

Fig. 6. Postoperative image.

Table 1. Immunohistochemical marker positivity in the mass

Marker	Presence
CD-99	Focal and weakly +
WT-1	–
Myogenin	–
Desmin	–
p63	Focal and weakly +
Cytokeratin	–
Synaptophysin	–
CD-34	–

with other breast lesions and mixed histological patterns [1, 2]. We present a case of a 15-year-old female with a high-grade spindle cell sarcoma differentiated as PT of the breast upon detailed histological examination, focusing on the difficulties associated with diagnosing and treating these rare complex lesions.

The case we present here is distinctive from other cases of breast cancer in that these usually present in older women; however, in our case it is an adolescent who presented with such a case [5]. This makes it difficult to plan the diagnosis and management considering that adolescent and pediatric group require to be managed with greater care as compared to adults. Moreover, the breast lesion in our patient progressed rapidly and was complicated by purulent discharge and bleeding which indicated the high-grade and complexity of spindle

cell sarcoma (involvement of regional lymph nodes also highlighted the aggressive course of the lesion and differentiated our case from other cases of spindle cell sarcomas which did not report spread to lymph nodes) [4, 7, 8].

Histopathology and immunohistochemical staining are essential to differentiate spindle cell lesions from other similar lesions which are included in the differential diagnoses such as myofibroblastoma, fibromatosis, pseudoangiomatous stromal hyperplasia, nodular fasciitis, inflammatory myofibroblastic tumor, PT, several primary sarcomas, and metastases [4, 9]. Histological examination of sections from the cores revealed a malignant neoplasm arranged in fascicles with intervening epithelial elements. Few areas showed stromal overgrowth with increased cellularity. The individual neoplastic cells in these areas showed an epithelioid to spindled morphology moderate to severe nuclear pleomorphism and hyperchromasia with scant cytoplasm and irregular nuclear membranes – high-grade malignant spindle cell neoplasm favoring malignant PT. These findings are comparable to other reports on spindle cell lesions. Moreover, immunohistochemical staining revealed weakly positive focal CD-99 and p63 along with negative WT-1, Myogenin, Desmin, Cytokeratin, Synaptophysin, EMA, S-100, CD-34, and CD-31. These findings aligned with the normal immunoprofile of spindle cell sarcomas.

Treatment of PT involved surgical removal of the affected breast via MRM. This procedure involves the removal of skin, breast tissue, areola, nipple, lymph nodes involved, and lining of the underlying chest muscles [10]. As compared to other procedures, MRM provides an option of reconstruction surgery after mastectomy [10]. To manage the case discussed in this article, we also opted for MRM with reconstruction plan after assessing patient for recurrence via regular and timely follow-up. This case report highlights that spindle cell sarcomas of breast, such as PT as in this case, although very rare, can occur even in adolescents and considering the challenges associated with diagnosis of these lesions it is imperative to investigate such rare lesions in greater detail to enhance the diagnostic modalities to establish early detection and develop an efficient management plan.

Conclusion

This case report contributes to the scare literature available on spindle cell sarcomas of the breast. The case presented highlighting a 15-year-old female with a high-grade spindle cell sarcoma histologically differentiated as PT of the breast shows that these rare lesions can occur in younger age group as well. Rapid progression of the breast lesion is shown by increase in the size of the mass and onset of a purulent discharge and bleeding. These manifestations warrant early detection of this lesion to provide timely management, early recovery, and fewer complications. Histopathological investigations along with immunohistochemistry provide a firm diagnosis by excluding other differentials which include metaplastic carcinoma, and primary breast sarcoma. MRM remains the standard management strategy for these lesions which also provide cosmetic benefits through reconstruction surgery. Further research on this topic is imperative to enhance diagnosis, early detection, and treatment approaches for spindle cell sarcomas of the breast.

Acknowledgments

Open access funding is provided by the Qatar National Library.

Statement of Ethics

Written informed consent was obtained from the parent of the patient for publication of the details of their medical case and any accompanying images. Ethical approval is not required for this study in accordance with local or national guidelines. Ethical approval is not required for this study in accordance with local or national guidelines.

Conflict of Interest Statement

None declared.

Funding Sources

This study was not funded.

Author Contributions

Z.H.T. and Z.H. took lead in writing the manuscript. Z.H., Y.A., and S.I. collected data from the patient. Y.A., Z.H.T., and S.I. edited and designed the final report. A.J.N. gave critical comments and reviewed the final manuscript. All authors provided critical feedback and helped shape the final report.

Data Availability Statement

All data generated or analyzed during this study are included in this article and its online supplementary material files. Further inquiries can be directed to the corresponding author.

References

1 Ayyaz F, Ayyaz F. Spindle cell carcinoma in the breast: a case report. Cureus. 2022;14(6):e25702.
2 Hou Y, Li Z. Spindle cell lesions of the breast. Hum Pathology Reports. 2021;26:300565.
3 Alexander JS, Hashmi H, Reynolds SB. Debilitating metastatic spindle cell carcinoma of the breast. Cureus. 2019;11(1):e3864.
4 Arora S, Rana D, Pujani M, Chauhan V. Malignant spindle cell tumor breast—a diagnostic dilemma. Indian J Surg Oncol. 2018;9(3):387–90.
5 Łukasiewicz S, Czeczelewski M, Forma A, Baj J, Sitarz R, Stanisławek A. Breast cancer-epidemiology, risk factors, classification, prognostic markers, and current treatment strategies-an updated review. Cancers. 2021;13(17):4287.
6 Cancer Today n.d. https://gco.iarc.fr/today/fact-sheets-cancers (accessed October 8, 2023).
7 Cil T, Altintas A, Pasa S, Buyukbayram H, Isikdogan A. Primary spindle cell sarcoma of the breast. Breast Care. 2008;3:197–9.
8 Choi A, Carpenter PM, Chopra S, Lara KM, Tseng WW, Patel DB, et al. Spindle cell carcinoma of the breast managed with neoadjuvant AIM: a case report. Rare Tumors. 2020;12:2036361320977021.
9 Carter MR, Hornick JL, Lester S, Fletcher CDM. Spindle cell (sarcomatoid) carcinoma of the breast: a clinicopathologic and immunohistochemical analysis of 29 cases. Am J Surg Pathol. 2006;30(3):300–9.
10 Mastectomy (breast removal surgery): types, what to expect, recovery n.d. https://www.webmd.com/breast-cancer/mastectomy (accessed October 8, 2023).