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Original Article

Beyond the Usual: Review of Rare Cases of Primary Breast Mesenchymal Sarcoma from a Tertiary Institution

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Abstract

Introduction: Primary breast sarcomas (PBSs) account for <1% of all breast cancer cases, typically affecting patients aged 55–59 years. They are difficult to differentiate from other types of breast cancer based on the clinical and imaging results and are associated with a poor prognosis. It is important to recognize them owing to differences in treatment options. We here present four cases of rare PBSs diagnosed at our center.

Aims and Objectives: The purpose of presenting this case series is to familiarize the pathologist with these rare entities as they clinically mimic epithelial malignancy of the breast but are heterogeneous and totally follow a different clinical course and treatment protocol.

Materials and Methods: A collection of four rare breast mesenchymal tumors is presented. Clinical details of patients, including follow–up, were obtained from the electronic medical record of hospital.

Results/Cases: A total of four cases were identified. All were female with age group of presentation being 14–71 years with mean age being 44 years. The tumor size ranged from 1.5 cm to 20 cm. Microscopically, a diagnosis of Rhabdomyosarcoma was rendered in one case, one was diagnosed as malignant peripheral nerve sheath tumors, one as osteosarcoma, and the last as atypical cartilaginous tumor. A varied panel of immunohistochemistry (IHC) markers was used to reach the final diagnosis, including SMA, Desmin, CD34, S-100, H3K27Me3, Myogenin, MyoD1, and SATB2 among others. Most common differential in all these cases was a malignant phyllodes tumor.

Conclusion: This case series describes unique cases of PBSs. Histopathological analysis has great importance in the diagnosis of these tumors, along with an IHC panel that is valuable in excluding phyllodes tumors and differentiating among the various sarcomas. In our setting, IHC was a major input in diagnosis. This study supports and builds on global data on rare breast neoplasms, while offering valuable regional insights.

Key words: Primary breast chondrosarcoma, Primary breast malignant peripheral nerve sheath tumor, Primary breast osteosarcoma, Primary breast rhabdomyosarcoma, Primary breast sarcoma

INTRODUCTION

Breast sarcoma is an exceptionally rare and varied group of cancers, accounting for under 1% of all breast cancers and fewer than 5% of all soft-tissue sarcomas.[1] Breast sarcoma can be classified into two types: *De novo* (primary) and post-therapy (secondary). The distribution of histologic subtypes varies across studies, but angiosarcoma is the most common. Other subtypes, such as liposarcoma, fibrosarcoma, and undifferentiated pleomorphic sarcoma, have also been reported.[2] While the clinical features of primary breast sarcoma (PBS) resemble those of mammary adenocarcinomas to some degree, it is associated with a higher risk of recurrence and a notably worse prognosis.[3] PBS is usually diagnosed during the fifth or sixth decade of life.[4] When encountering a spindle cell tumor in a glandular organ like breast, it is crucial to exercise utmost caution and

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astute pathologic acumen before rendering a diagnosis of sarcoma. In such cases, meticulous histomorphological and immunohistochemistry (IHC) appraisal with appropriate antibodies is vital in deducing the accurate diagnosis. Herein, the authors highlight the institutional experience on four unusual cases of PBSs, deliberating on the diagnostic challenges faced.

MATERIALS AND METHODS

This retrospective study was conducted in the department of oncopathology from January 2019 to December 2024. A total of 50 breast sarcomas were identified through an initial electronic search and departmental records. All cases were evaluated according to the categories outlined in the World Health Organization Blue Book, with those cases selected as rare PBS not mentioned in the Blue Book under mesenchymal tumors of the breast. The clinical and radiological data were collected, and histological slides were reviewed by two independent pathologists. Since this was a retrospective study, no additional interventions or invasive procedures were performed on any patient, ensuring no ethical violations occurred.

RESULTS

A total of 4 rare PBS were identified. These included one case each of rhabdomyosarcoma (RMS), malignant peripheral nerve sheath tumor (MPNST), extraskeletal osteosarcoma, and chondrosarcoma. The age of the patients ranged from 14 to 72 years, with a median age of 55 years. A summary of these rare histological subtypes, including their clinical presentation, histopathological and IHC findings, treatment, and outcomes, is provided in Table 1.

Case 1

A 14-year-old girl presented to the pediatric oncology department with a left breast mass and palpable left axillary lymph nodes. Positron emission tomography scan revealed a Fludeoxyglucose (FDG)-18 avid, heterogeneously enhancing soft-tissue mass involving nearly the entire left breast, with a maximum standardized uptake value (SUV_{max}) of 11.67. The mass was abutting the overlying skin anteriorly and the chest wall posteriorly, with an unclear fat plane. Low-grade FDG uptake was observed in the left axillary lymph nodes, with the largest measuring 0.9 cm and an SUV_{max} of 1.75. Cytomorphology from the breast conducted at a peripheral center suggested a lymphoproliferative disorder. However, the biopsy performed at our institute indicated a grade 2 spindle cell neoplasm, with a potential diagnosis of phyllodes tumor, to be confirmed on resection. A left modified radical mastectomy (MRM) with axillary clearance was performed, and sample was received for histopathological examination. A gray-white, fleshy tumor was received in the pathology department, measuring 14 × 12 × 7 cm and occupying all the quadrants of the breast. Microscopic examination revealed a malignant mesenchymal tumor consisting of monomorphic oval cells arranged in sheets and nests, separated by fibrovascular septa of varying thickness. The individual cells displayed oval to irregular nuclei, coarse chromatin, indistinct nucleoli, and scant cytoplasm. Cells with clear cytoplasm and occasional large cells were also

Table 1: Clinicopathological parameters of 4 cases of primary breast sarcoma.

S. No	Age	Size (cm)	Imaging	Biopsy	Procedure	Resection	Positive IHC	Negative IHC	Follow-up
1	14/F	14	Heterogeneously enhancing soft-tissue mass involving almost entire left breast.	Borderline phyllodes	Modified Radical Mastectomy	Rhabdomyosarcoma	Desmin, MyodD-1 and Myogenin	AE1/AE3, SOX10, NKX2.2, p63	Palliative treatment Lost to follow-up.
2	65/F	10	Large irregular heterogeneously enhancing ulcerative lesion with lobulated margins-involving the entire right breast.	1. Spindle cell Sarcoma, High grade 2. Malignant Phyllodes	Simple Mastectomy with axillary clearance	Malignant Peripheral Nerve Sheath Tumor	S-100 and SOX-10 (focal)	SMA, Desmin, HMB45	Adjuvant CT Currently doing well.
3	72/F	7.5	Ill-defined, minimally heterogeneously enhancing mass-upper inner quadrant of left breast	1. Primary chondroid neoplasm 2. Heterologous component of Phyllodes	Radical Mastectomy	Extraskeletal Chondrosarcoma	ND		Adjuvant RT Currently doing well
4	45/F	3	Architectural distortion and skin thickening	Malignant phyllodes with osteoclastic giant cells	Radical Mastectomy	Extraskeletal Osteosarcoma	SATB2	H3G34W	Adjuvant CT-Ifosfamide and Adriamycin Currently doing well.

IHC: Immunohistochemistry, CT: Computed tomography

observed, along with myxoid stroma in certain areas. Although the tumor displayed brisk mitosis (12–15 per high-power field [HPF]), necrosis was absent. There were no signs of proliferating epithelial components in the entire specimen, thus precluding the primary concern of malignant phyllodes tumor (MPT). IHC analysis evinced the indubitable reactivity of the tumor cells for desmin, myoD-1, and myogenin, thus confirming a diagnosis of RMS [Figure 1]. Nodal metastasis was also discovered. The tumor was classified as Stage III according to the modified site, size, metastasis staging system for RMS due to its location in an unfavorable site (breast), size >5 cm, and presence of regional lymph node involvement. The patient was lost to follow-up after surgery, so no adjuvant treatment could be administered.

Case 2

A 65-year-old female visited the surgical oncology department with a lump in her right breast, which she had noticed 2 years ago. On examination, a 10 × 10 cm hard, irregular mass was found, extending from the 1 o'clock to 6 o'clock position in the upper-inner and lower-inner quadrants of the right breast, approximately 4 cm from the areola margin. Imaging revealed a large, irregular, heterogeneously enhancing ulcerative lesion with lobulated margins, involving the entire right breast, measuring 10 × 6.2 cm, extending to the overlying skin and nipple-areola complex, not involving the pectoralis muscle, along with indeterminate right axillary lymph nodes. The biopsy suggested a primary diagnosis of spindle cell neoplasm, with the possibility of phyllodes tumor. The patient subsequently underwent a right radical mastectomy, and the specimen was sent for histopathological examination. A gray-white, solid, firm growth measuring 10 × 8 × 5.5 cm was observed, occupying the upper outer and lower quadrants, histology of which demonstrated an infiltrative, cellular tumor composed of cells arranged in sheets and short, intersecting fascicles. The cells were spindle-shaped, with highly pleomorphic nuclei, coarse chromatin, prominent nucleoli, and scant to moderate cytoplasm. Dense lymphoplasmacytic infiltrates were present, along with tumor giant cells. Mitosis ~24/10HPF, along with focal areas of necrosis, was noted. Epithelial elements were not visualized. S100 and SOX10 played up the tumor cells on IHC, while they were negative for SMA, desmin, and HMB45 [Figure 2]. These features were compatible with a diagnosis of MPNST, high grade, despite retention of H3k27me3 in the tumor cells. Chemotherapy with gemcitabine and docetaxel was administered subsequently. At 6-month follow-up, the patient was noted to have stable disease.

Case 3

A 45-year-old female presented with a chief complaint of a left breast lump. She had previously undergone lumpectomy at an outside facility and was referred to our tertiary care center for further evaluation. On local examination, a scar from the previous surgery was observed, but no abnormalities were detected. Computed tomography (CT) scan revealed postoperative changes in the left breast, including architectural distortion and skin thickening, with no obvious suspicious residual lesions or left axillary lymph node involvement. The outside histopathology report from the lumpectomy suggested a diagnosis of MPT, with clear margins. However, the patient defaulted thereafter without undergoing any adjuvant treatment, nor slide block review of the outside histopathology could be done. Nine months later, the patient returned with local recurrence, and review of the outside histology slides suggested a diagnosis of high-grade sarcoma with osteoclastic multinucleated giant cells; no epithelial elements were discerned. MRM with axillary clearance was performed. Microscopic examination of the surgical specimen revealed a mesenchymal neoplasm composed of malignant spindle to epithelioid cells, exhibiting pleomorphic hyperchromatic nuclei, coarse chromatin, and moderate eosinophilic cytoplasm. Numerous osteoclastic giant cells were observed, along with malignant osteoid formation and focal areas of necrosis. No epithelial component was identified, and extensive hemorrhagic areas were noted. To rule out other potential breast mesenchymal tumors, IHC analysis was conducted. The tumor cells were positive for SATB2 and negative for AE1/AE3 and H3.3G34W [Figure 3]. Based on these findings, a final diagnosis of extraskeletal osteosarcoma was proffered. The patient is currently doing well and has completed three cycles of chemotherapy with ifosfamide and adriamycin.

Case 4

A 72-year-old female presented with a unilateral left breast lump, present for 1 year, with noticeable rapid growth in the last month. She had no prior medical history and no family history of breast cancer. Physical examination revealed a non-tender, firm mass measuring 9 × 8 cm in the central quadrant of the left breast, fixed to the chest wall, with no evidence of axillary lymphadenopathy. CT scan revealed an ill-defined, minimally heterogeneous mass of approximately 8.3 × 5.7 cm, extending to the underlying chest wall muscles but without rib involvement, along with scattered areas of suspicious microcalcifications. Subcentimeter-sized reactive nodes were observed in the left axillary region. Radiological impression suggested a phyllodes tumor. Biopsy of the breast mass revealed a chondroid lesion with lobules of atypical chondrocytes in a chondromyxoid background, with mildly increased cellularity. No mitoses or necrosis were observed, and no breast parenchyma was found. The histopathological report indicated either a primary chondroid tumor or a heterologous component of a phyllodes tumor. A 99 m

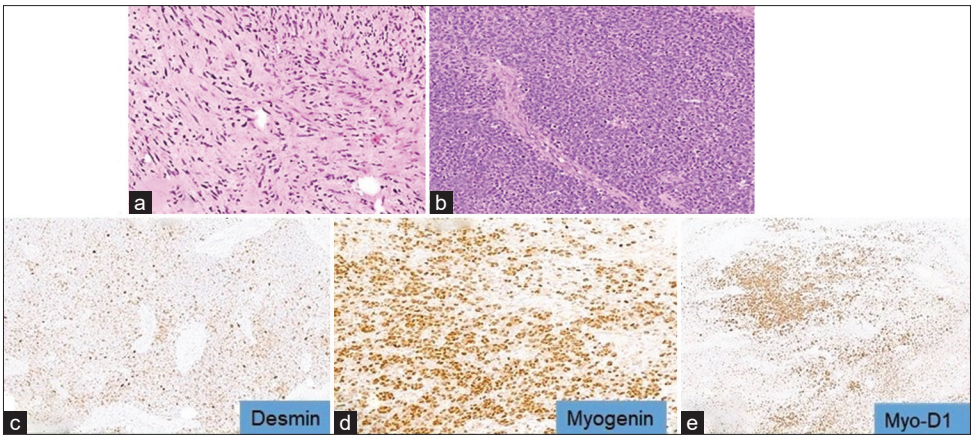

Figure 1: Rhabdomyosarcoma. (a) The section shows a spindle cell tumor arranged in loose fascicles and singly scattered (×10, Hematoxylin and Eosin [H&E]). (b) Section shows neoplasm comprising monomorphic oval cells arranged in sheets and nests separated by variably thick fibrovascular septae. Individual cells exhibit moderate anisocytosis, comprising oval to irregular nuclei, coarse chromatin, indistinct nucleoli, and scant cytoplasm (×40, H&E). (c-e) Immunohistochemistry (IHC) for Desmin, Myogenin, and Myo-D1 shows positive staining (×10, IHC)

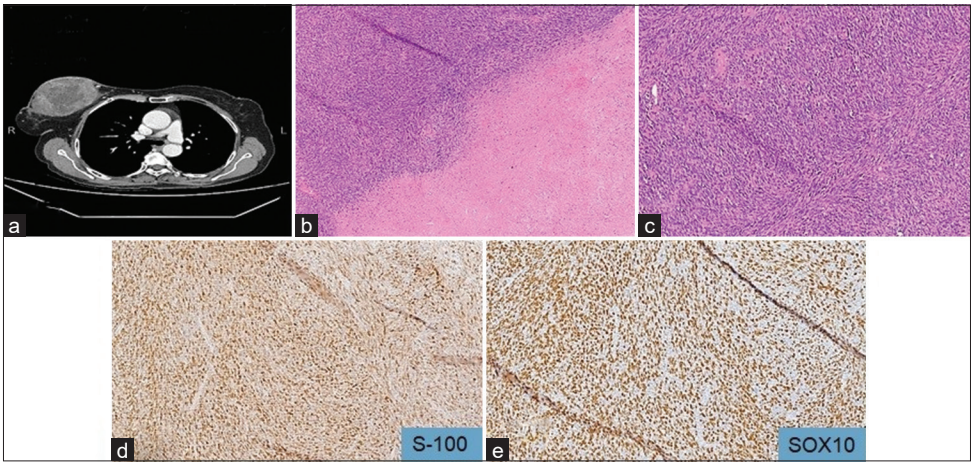

Figure 2: Malignant peripheral nerve sheath tumor. (a) Heterogeneously enhancing mass seen in the left breast. (b and c) Section shows an infiltrative cellular tumor composed of cells arranged in sheets and short intersecting fascicles. The cells are spindle-shaped with markedly pleomorphic nuclei, coarse chromatin, conspicuous nucleoli, and scant to moderate cytoplasm (×10, Hematoxylin and Eosin [H&E]; ×40 H&E). (d and e) Immunohistochemistry (IHC) shows nuclear positivity for S-100 and SOX-10 (×40, IHC)

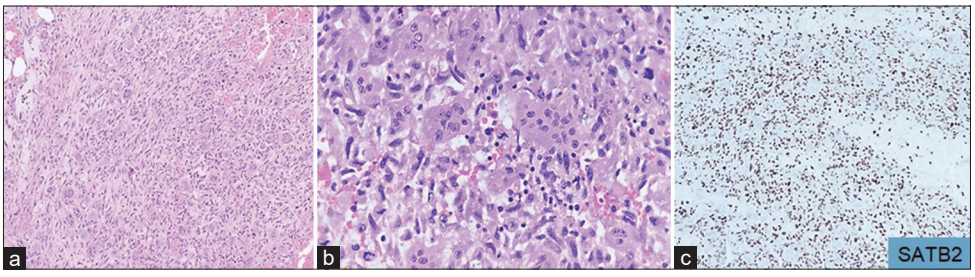

Figure 3: Extraskeletal osteosarcoma. (a and b) Section shows a mesenchymal neoplasm composed of intersecting long fascicles of spindle-shaped malignant cells. These cells display pleomorphic hyperchromatic nuclei with coarse chromatin and moderate eosinophilic cytoplasm. Numerous osteoclastic giant cells are identified (×10, Hematoxylin and Eosin [H&E]; ×40 H&E). (c) Positive nuclear staining for SATB2 (×40, immunohistochemistry)

Technetium-Methylene Diphosphonate (MDP) (99 m Tc-MDP) bone scan unveiled no signs of skeletal involvement or metastases. The patient underwent a left MRM with nodal axillary clearance of levels I and II. On gross examination, the cut section revealed a solid, lobulated, firm, pearly white tumor measuring 7.5 × 7.5 × 5.5 cm, with areas of degeneration and hemorrhage. An infiltrating tumor composed of lobules of hyaline cartilage with

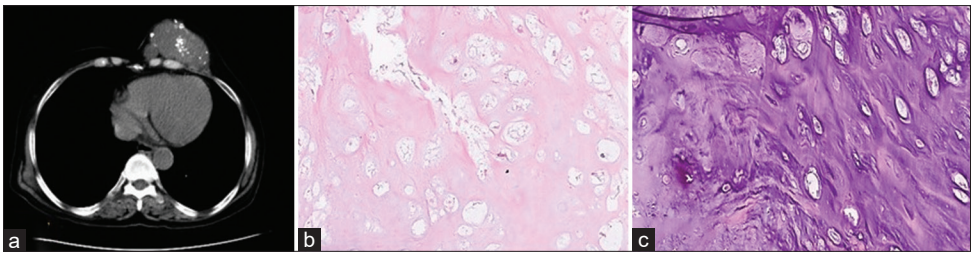

Figure 4: Atypical cartilaginous tumor/chondrosarcoma grade 1. (a) Computed tomography thorax shows an ill-defined, minimally heterogeneously enhancing mass involving the upper inner quadrant of the left breast, measuring approximately 8.3 × 5.7 cm. It involves the underlying chest wall muscles. However, the underlying rib is unremarkable. (b and c) High-powered (Hematoxylin and Eosin, ×20) view demonstrating a cartilaginous tumor with interspersed nodular and lobulated islands of well-differentiated cartilage. Pleomorphic chondrocytes occupy lacunar spaces with hyperchromatic nuclei, some showing binucleation

atypical pleomorphic chondrocytes, accompanied by focal hemorrhage and necrosis, was divulged on histopathology. Mitosis was sparse. There was no evidence of stromal overgrowth, epithelial hyperplasia, ductal carcinoma in situ, malignancy, or any other mesenchymal elements. The adjacent breast tissue exhibited a few compressed terminal duct lobular units. The entire specimen was extensively sampled multiple times, primarily in the search for proliferative epithelial components, to exclude phyllodes tumor with heterologous component or metaplastic breast carcinoma (MBC). IHC testing disclosed S100 positivity in the atypical chondrocytes [Figure 4]. The final diagnosis conferred was atypical cartilaginous tumor/chondrosarcoma Grade 1. The patient was discharged in a stable condition after surgery and was scheduled for adjuvant radiotherapy, but was lost to follow-up.

DISCUSSION

Breast cancer is the most prevalent form of cancer among women globally and now represents 1 in 8 cancer diagnoses, with a total of 2.3 million new cases diagnosed in both men and women each year.[5] In 2020, it is estimated that 685,000 women succumbed to breast cancer, accounting for 16% or one in every six cancer-related fatalities among women.[6] The situation in developing countries is more severe due to limited awareness of screening programs and practices. The scenario is further compounded when rare and diagnostically difficult entities are encountered, which tend to be underdiagnosed and/or misdiagnosed. Cancers of mesenchymal origin in the breast, which are quite uncommon and more specifically PBSs, stand out as a noteworthy paradigm in this regard. Moreso, data on such rare neoplasms is limited in Indian context. Through this study, the authors endeavor to shed light on such unusually encountered diseases, explore the nuances of histopathological and IHC evaluation, and offer a valuable diagnostic framework tailored to the regional population. Soft-tissue sarcomas are not so frequent diseases, and those incriminating the breast are even rarer. Although angiosarcoma is the most frequently reported type of breast sarcoma, almost any form of sarcoma can develop in the breast.[7] Due to their scarcity, large-scale analyses have been limited, and PBS is not known to have a definite etiology. A brief review of the four uncommon PBS is presented henceforth.

RMS is among the most frequently occurring sarcomas in children and young adults aged 15–35 years, with a slightly higher incidence in young women, showing a female-to-male ratio of approximately 3:2 in this age group.[8] However, primary RMS of the breast is extremely rare, although it primarily affects children and young adults. A study conducted by the Intergroup RMS Study Group found primary breast RMS in just 6 (0.17%) of 3,500 patients examined between 1972 and 1992.[9] At present, approximately 30 cases of primary breast RMS have been reported in the literature. Breast RMS is an aggressive tumor, with a long-term survival rate of approximately 50% for primary cases and around 20% for those with metastatic disease.[10] Metastases commonly occur in the lungs, bones, and brain, with reports also describing spread to the vagina, orbits, and colon.[8] Lesions that mimic RMS encompass malignant small round cell tumors, which are defined by small, dark, and round undifferentiated cells. These characteristics closely resemble those found in Ewing sarcoma, synovial sarcoma, hepatoblastoma, granulocytic sarcoma, desmoplastic small round cell tumors (especially in the abdomen), small-cell osteosarcoma, small-cell carcinoma, primitive neuroectodermal tumors, and non-Hodgkin lymphoma.[9] Differential diagnosis for RMS varies depending on the subtype, but distinguishing it from MPT with sarcomatous overgrowth is always crucial, regardless of the RMS subtype.[11] The crux of this differentiation is extensive tissue sampling on gross pathologic examination, as the epithelial component of MPT may be very small and harder to identify.[7] Age is another useful factor in differentiating these malignancies, as MPTs typically occur in the fifth or sixth decade of life, while primary breast RMS predominantly affects younger patients. This is of particular concern, since young

women with a breast lump are commonly thought to be harboring fibroadenoma, unless proved otherwise. While mammography and ultrasound are handy for locating breast lesions, they cannot differentiate RMS from a fibroadenoma. In this context, the recommendation of Hays *et al.* that teenage girls with breast tumors larger than 2 cm ought to undergo fine-needle aspiration and cytology analysis for a precise diagnosis cannot be overemphasized.

Due to its rarity, a definitive treatment protocol for breast RMS has not been established. In general, axillary dissection is not recommended for PBS, as sarcomas typically do not spread to lymph nodes, except in the instance of palpable lymphadenopathy. Nevertheless, given that previous studies have documented metastasis to the axillary lymph nodes, lymph node dissection is recommended in cases of RMS.[12] In our case too, nodal metastasis was ascertained.

MPNSTs arise from either a major or minor peripheral nerve branch or its surrounding sheath. Common sites of origin include the extremities and trunk, particularly from the sciatic nerve, brachial plexus, and sacral plexus. While these tumors can occur spontaneously, 5–42% of cases are associated with neurofibromatosis type 1.[13] They have replaced older terms such as malignant schwannoma, malignant neurilemmoma, and neurofibrosarcoma, and account for about 10% of all soft-tissue sarcomas.[14] MPNST of the breast is extremely rare, with only 16 cases reported in the literature. MPNST must be differentiated from MPT, fibrosarcoma, and leiomyosarcoma. A diagnosis of MPT is given credence, which provided that proliferating ductal epithelial component is manifest, even if scarce or compressed epithelial-lined leaf-like spaces typically seen in low-grade phyllodes is apparent, which were absent in this case despite thorough sampling. Morphologically, MPNST resembles fibrosarcoma, but it is characterized by spindle cells organized in dense cellular regions interspersed with hypocellular myxoid areas. These spindle cells have wavy nuclear contours, which help distinguish it from fibrosarcoma. In contrast, the spindle cells of leiomyosarcoma have a more distinct eosinophilic cytoplasm and blunted nuclei, not conspicuous in this case. Differentiating MPNST from a benign nerve sheath tumor can also be challenging, as some neurofibromas may appear quite cellular and contain occasional pleomorphic cells.[14] Immunohistochemically, Schwann cells in MPNSTs typically show positive staining for S-100, SOX-10, CD-57, laminin, and calretinin, while being negative for EMA, actin, and various cytokeratins. A key point to note is that MPNSTs exhibit focal, rather than diffuse, expression of S-100 protein. If a strong and diffuse pattern of S-100 expression is observed, the diagnosis would likely lean toward schwannoma or metastatic melanoma rather than MPNST.[15] The loss of trimethylation at lysine 27 of histone-H3 (H3K27me3), as detected by IHC, has recently been reported in MPNSTs. While this loss is highly sensitive for diagnosing high-grade MPNST, its specificity remains a subject of debate.[16] In addition, the presence of retained H3K27me3 does not exclude the possibility of MPNST. Partial loss of H3K27me3 is frequently observed in other spindle cell neoplasms that are considered in the differential diagnosis of MPNST, which limits its diagnostic value for MPNST.[17] MPNST generally has a poor 5-year survival rate in most cases. Due to its rarity, there are no established guidelines for managing MPNST of the breast. Successful treatment relies on complete surgical resection. Adjuvant therapy is recommended after wide excision unless the tumor is small, superficial, and low-grade. If histologically positive margins or incomplete excision are present, further surgery or adjuvant therapy (if surgery is not possible) should be pursued. Dissection of the axillary tail is not recommended for patients with clinically negative axilla, as, like other sarcomas, the tumor typically spreads hematogenously. While radiotherapy can help with local control and may delay recurrence, it has minimal impact on long-term survival.[18]

Primary osteosarcoma breast (POB), an example of extraskeletal osteosarcoma, is another uncommon, invasive, and diverse tumor. The origin of POB remains unclear. Some researchers propose that primary breast rhabdomyosarcoma (POB) originates from totipotent mesenchymal cells of the mammary gland or mesodermal remnants. However, Omranipour *et al.* demonstrated that nearly all cases of POB are derived from epithelial cells that undergo metaplastic transformation.[19] The average age of patients with breast osteosarcoma is 64.5 years, whereas skeletal osteosarcoma typically occurs in younger individuals.[20] The exact cause of POB remains unclear. It has been proposed that factors such as previous breast radiation therapy, trauma, exposure to vinyl chloride in the workplace, and the presence of artificial implants may contribute to its development.[21] However, the current case did not present any of these known risk factors. POB usually manifests as a rapidly growing mass, with only a small proportion (18%) of cases being linked to pain.[22]

The primary histological differentiation of POB involves distinguishing it from MBC and MPT with osteoid or bone components. MBC typically shows immunoreactivity to epithelial markers, whereas POB lacks markers for epithelial differentiation. In this case, the epithelial marker, such as AE1/AE3, was negative, confirming the tumor's non-epithelial nature. Unlike POB, MPT involves hyperplasia of both the breast stroma and epithelium. SATB2, a sensitive and specific marker for osteoblast differentiation and mainly expressed in benign and malignant bone tumors with osteoblastic differentiation, as well as soft-tissue

tumors with heterologous bone differentiation, was positive in our case on IHC. Given the rarity of POB, the optimal treatment approach may differ. However, wide excision with negative margins is mandatory. Axillary lymph node dissection is not recommended, as POB is locally aggressive and primarily metastasizes through the bloodstream rather than the lymphatic route. In our case, however, an axillary dissection was performed because radiological findings indicated suspicious nodes, and histological examination revealed metastatic deposits in several lymph nodes.

A primary breast chondrosarcoma (PBC) includes chondrosarcomatoid areas that develop from mammary tissue. Chondrosarcomatoid elements can present in three distinct forms: as a standalone tumor (pure chondrosarcoma), as part of the stromal component in a MPT, or as chondrosarcomatous differentiation within MBC. The case we encountered was a histologically pure PBC. These tumors are typically large and occur in women over the age of 40. They generally do not invade the overlying skin. Axillary adenopathy is observed in 14–29% of cases, with most instances evincing reactive hyperplasia. Our case supports the clinical findings of previously reported cases.[23] Differentiating PBC from MPT with prominent chondrosarcomatoid features can be quite difficult. MPTs are generally characterized by a leaf-like architecture, stromal overgrowth, and a biphasic composition of both epithelial and stromal elements. The stromal component frequently displays atypia, such as elevated cellularity and the presence of mitotic figures, and may contain regions that mimic chondrosarcoma. MBC with heterogenous mesenchymal differentiation is another condition that poses a diagnostic challenge, for which extensive grossing as outlined earlier is essential. In MBC, the epithelial component must be both proliferative and malignant, though a direct transition between the carcinomatous and sarcomatous areas may not always be apparent. Even when the epithelial component is not clearly identifiable under the microscope, MBC can still be a valid diagnosis if IHC testing demonstrates epithelial differentiation.[24] Lastly before conferring the diagnosis of a PBC, it is also paramount to rule out an adjacent non-mammary site of origin, particularly the rib, both clinically and radiologically. Rib chondrosarcoma commonly appears as a mass arising from the rib and extending into adjacent breast tissue, with imaging modalities such as CT or MRI providing clearer visualization. It often shows evidence of bone involvement, including rib remodeling or destruction.[25,26] The primary treatment for PBC is complete surgical excision with negative margins, often accompanied by reconstructive procedures. Although multimodal therapy has shown benefits in reducing local and systemic recurrence in somatic soft-tissue sarcomas, its role in breast sarcomas is still unclear. The application of chemotherapy and radiotherapy is limited and typically considered on an individual basis. Continuous follow-up and a multidisciplinary team approach are crucial for the successful management of these rare tumors.

CONCLUSION

This study is unique in systematically analyzing rare PBSs in an Indian cohort and bringing forth the histopathological and IHC nuances. Distinguishing them from other breast tumors, particularly phyllodes tumors, can be challenging, yet is critical in view of the prognostic implications. The incredible utility of meticulous histopathological examination and judicious IHC appraisal in the veracious recognition of such rare entities cannot be understated in this regard. Increased awareness among clinicians about these tumors is also the need of the hour for determining the best treatment approach and achieving better outcomes. Exposition of PBS s through a study such as ours can be a harbinger to foster valuable genomic research in this genre in the future.

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AUTHORS' CONTRIBUTIONS

All authors contributed equally to the conception, design, data acquisition, and manuscript preparation.

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