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Recurrent malignant phyllodes tumor of the breast: An extremely rare case of recurrence with only rhabdomyosarcoma components

Jia Han[1,2], Shuice Liu[1,2], Akihoro Shioya[1,2], Motona Kumagai[1,2], Emi Morioka[3], Miki Noguchi[3], Masafumi Inokuchi[3] and Sohsuke Yamada[1,2]

Abstract

Rhabdomyosarcoma is a rare disease that typically occurs in children. Rhabdomyosarcoma seldom occurs in the breast, and its diagnosis and treatment have infrequently been reported. The present case is a rare one of a recurrent malignant phyllodes tumor of the breast with only rhabdomyosarcoma components. A 69-year-old woman received a diagnosis of borderline phyllodes tumor of the left breast and underwent partial mastectomy. During follow-up, a left breast mass was found 1 year and 8 months after the previous surgery. Based on examination findings, it was suspected to be recurrent phyllodes tumor, so total left mastectomy was performed in our hospital. After the surgery, immunostaining failed to determine the epithelial component which may be produced by the proliferative part of stromal cells of previous phyllodes tumors. However, we could not exclude the possibility that this was a new tumor. After comparing samples with specimens from the first operation, it was finally determined to be a malignant phyllodes tumor with a rhabdomyosarcoma component. Therefore, chemotherapy was given, and vincristine, actinomycin D, and cyclophosphamide therapy was introduced. At the same time, radiation therapy was planned. Among phyllodes tumors, cases involving rhabdomyosarcoma components are very rare, especially those where the recurrence morphology only shows the same rhabdomyosarcoma components. This was a rare case with unique characteristics and great reference value.

Keywords

Rhabdomyosarcoma of the breast, breast malignant tumor, phyllodes tumor of the breast, recurrent malignant tumor

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Introduction

Phyllodes tumor of the breast is an uncommon tumor, accounting for no more than 1% of all primary breast tumors.[1] Malignant phyllodes tumor of the breast usually has a variety of biological manifestations and may contain heterologous components, such as rhabdomyosarcoma (RMS).[2] However, among phyllodes tumors, cases involving RMS components are seldom seen.[3]

RMS is an infrequent disease that typically occurs in children.[4] It seldom manifests in the breast, and its diagnosis and treatment have rarely been reported.[5] RMS of the breast, including primary and recurrence, is exceedingly rare, with only about 32 cases reported thus far in the English literature. Over the years, although research concerning malignant tumors has made substantial progress, a few reports have described the diagnosis and treatment of malignant phyllodes tumor of the breast with RMS components.

The presently reported case is an extremely rare one. Indeed, to our knowledge, there have been no other case reports of recurrent malignant phyllodes tumor of the breast with RMS components, especially those involving recurrence with the same RMS components. Therefore, this case with unique characteristics is of great reference value.

[1]Department of Pathology and Laboratory Medicine, Kanazawa Medical University, Kahoku, Japan
[2]Department of Diagnostic Pathology, Kanazawa Medical University Hospital, Kahoku, Japan
[3]Department of Breast and Endocrine Surgery, Kanazawa Medical University Hospital, Kahoku, Japan

Corresponding Author:
Jia Han, Department of Pathology and Laboratory Medicine, Kanazawa Medical University, 1-1 Daigaku, Uchinada, Kahoku 920-0293, Ishikawa, Japan.
Email: hanjia0227@yahoo.co.jp

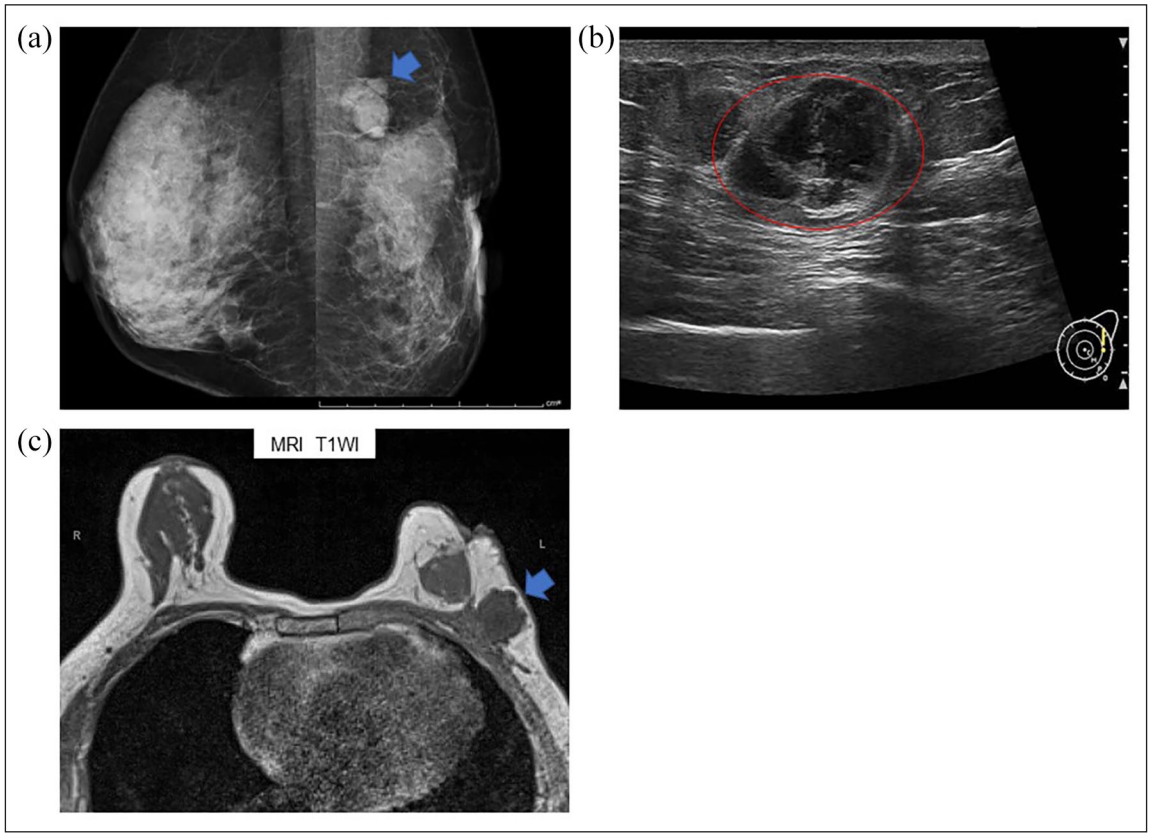

Figure 1. (a) Mammography showed a tumor shadow with a clear boundary in the outer upper quadrant of the left breast. (b) Breast sonography showed a lobulated hypoechoic tumor with a rough boundary. (c) MRI showed a 29-mm neoplastic lesion in the outer upper quadrant of the left breast.

Case presentation

This patient was a 69-year-old female with a history of meningioma (surgically treated, no recurrence), lipid abnormality and arrhythmia, and a family history of gastric cancer (mother) and liver cancer (father) but no history of breast or ovarian cancer. Four years and 6 months ago, she had undergone partial mastectomy with a diagnosis of left breast phyllodes tumor (outer upper quadrant, borderline) at a previous hospital.

Two years and 11 months ago, while still being followed up after surgery, she became aware of a left breast mass and visited our hospital. After a physical examination, a 2 cm × 2 cm elastic, hard mass in the outer upper quadrant of the left breast was found. The mass showed good surface smoothness and mobility. Regarding imaging findings, mammography showed a tumor shadow with a clear boundary in outer upper quadrant of the left breast (Figure 1(a)); breast sonography showed a lobulated hypoechoic tumor with a rough boundary (Figure 1(b)); and magnetic resonance imaging showed a 29-mm neoplastic lesion in the outer upper quadrant of the left breast (Figure 1(c)). Needle biopsy was performed as well. According to the needle biopsy results, the proliferation of atypical cells of the mesenchymal lineage was observed. Since the specimen did not contain a foliate growth pattern with epithelium, recurrence of the phyllodes tumor in the outer upper quadrant was suspected. Therefore, total left mastectomy was performed at our hospital.

The gross specimen could be viewed with the naked eye after the operation, appearing as a 43 mm × 41 mm × 33 mm, well-defined gray–white tumor in the outer upper quadrant. On the cut surface, a multi-nodule-to-lobular structure could be seen (Figure 2). Histologically, it was a tumor with a high cell density, and diffuse proliferation was obvious (Figure 3(a)). It was also conspicuous that the cells were arranged along the fibrotic septum (Figure 3(b)). The tumor cells were mainly spindle-shaped and polygonal with a high N/C ratio and mixed with striated myoblast-like cells with acidophilic cytoplasm (Figure 3(c)). Immunohistochemistry showed that the tumor was negative for estrogen receptor (ER), progesterone receptor (PR) and CD45, and its human epidermal growth factor receptor 2 (HER-2) score was 1+, while its Ki-67 index was 63% (data not shown). The tumor was positive for MyoD1, myogenin, and desmin (Figure 4(a)–(c)) and partly α-smooth muscle actin (α-SMA) positive (Figure 4(d)).

All tumors were sampled, and immunostaining of epithelial markers, for example, cytokeratin (CK) AE1/3 was performed, but no epithelial components were detected (data

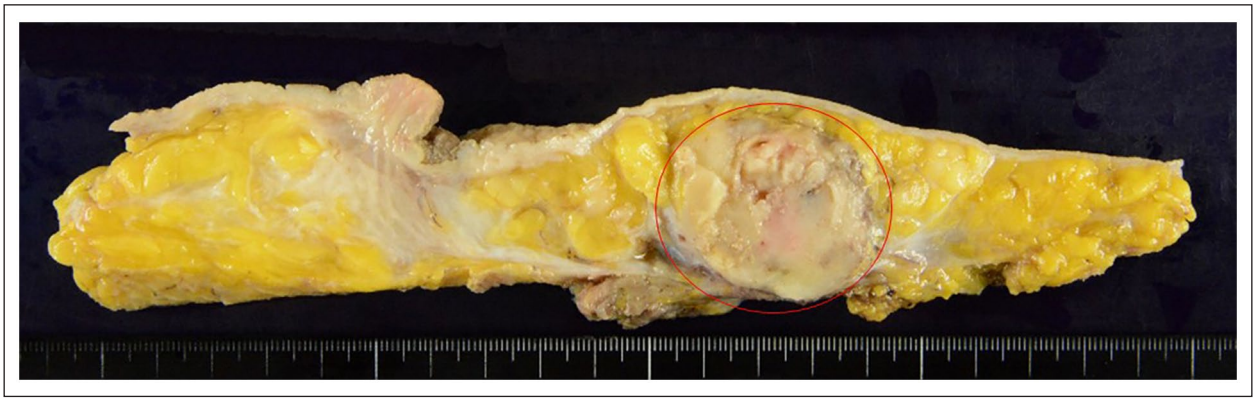

Figure 2. A 43 mm × 41 mm × 33 mm gray tumor with a clear boundary could be seen in the outer upper quadrant of the gross specimen of total left mastectomy. Multi-nodule and lobulated structures could also be seen in this specimen.

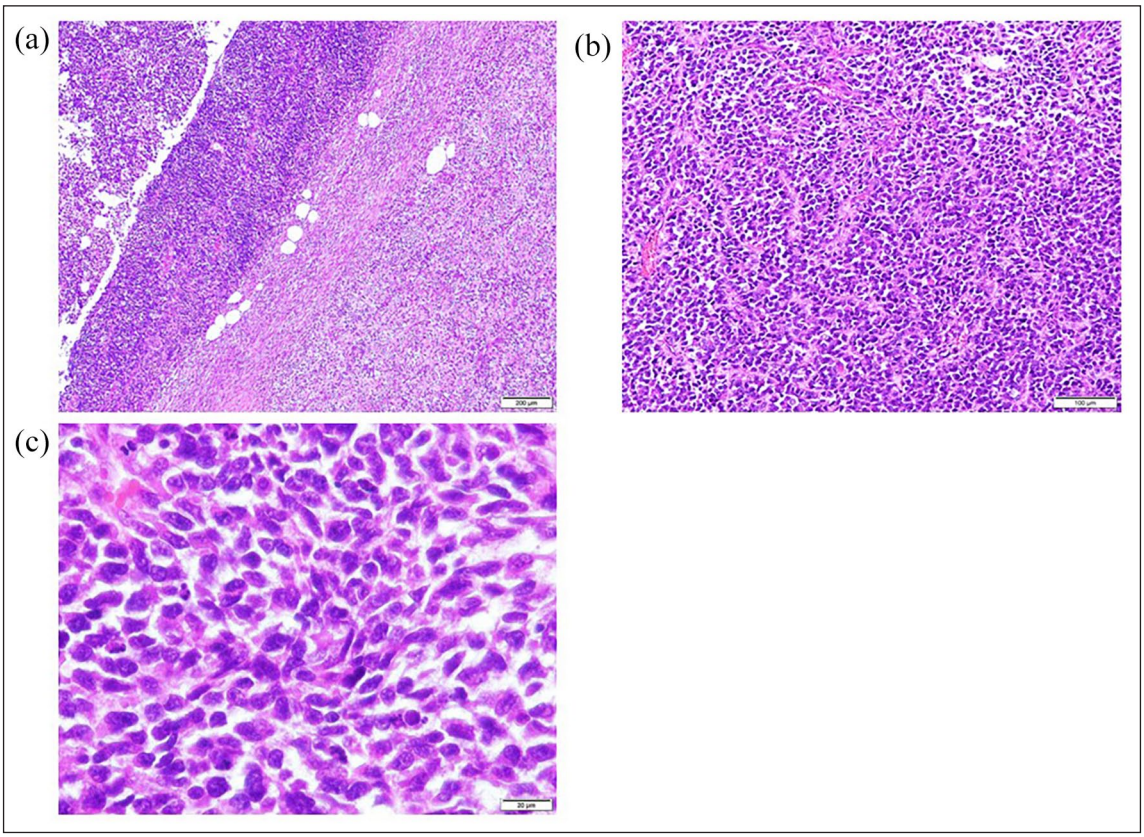

Figure 3. HE-stained images of the surgical specimen. (a) In the low-magnification field of view, a high cell density and diffuse proliferation were seen in the tumor. (b) In the medium-magnification field of view, the arrangement of tumor cells along the fibrous membrane was very obvious. (c) In the high-magnification field of view, the tumor cells were mainly spindle-shaped to polygonal cells with a high N/C ratio, and striated myoblasts were also found.

not shown). We suspected that it might have originated from the proliferative part of the stromal cells of the previous phyllodes tumor, but since new tumor development could not be excluded, a comparison with the specimen from the previous surgery was necessary. The previous surgery samples showed similar histological findings. Interstitial hyperplasia was seen with a leaf-like structure covered by a glandular epithelium with poor atypia and flat epithelial metaplasia epithelium (Figure 5(a) and (b)). In the area with a high cell density, diffuse proliferative images and cells arranged along the fibrotic septum could also be seen (Figure 5(c)). Similarly, the tumor cells were mainly

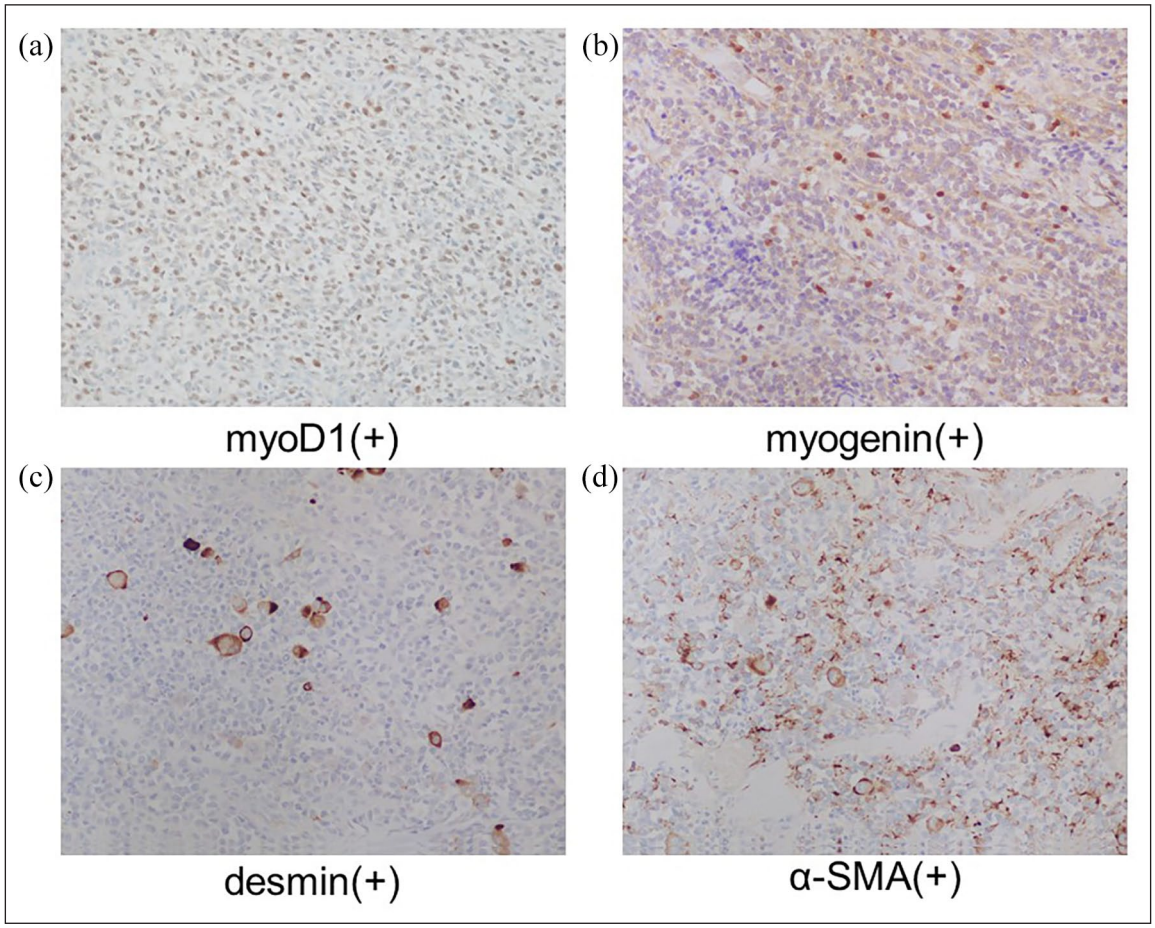

Figure 4. Immunostained images of MyoD1 (a), myogenin (b), desmin (c), and SMA (d).

spindle-shaped to polygonal with a high N/C ratio and mixed with acidic cytoplasm and striated myoblast-like cells (Figure 5(d)).

The microscopic characteristics of tumors meet the diagnostic criteria of malignant phyllodes tumor. There is no epithelial component found in one low power the field of view, nuclear polymorphism, interstitial hyperproliferation, increased mitotic figures, increased cell density, stromal infiltration, and especially, the presence of malignant ectopic components could be seen. Therefore, it was considered to be in the category of malignant phyllodes tumor and was evaluated as a malignant phyllodes tumor with an RMS component. Since the specimen from the previous surgery contained the similar RMS component as well, it was considered that the RMS component had recurred. The postoperative pathological evaluation resulted in a diagnosis of recurrent malignant phyllodes tumor with alveolar RMS.

Postoperatively, it was judged that chemotherapy was indicated, and VAC therapy (vincristine, actinomycin D, cyclophosphamide) was introduced, with subsequent radiation therapy planned. During the 2 years from the operation to the latest follow-up, the patient remains well without recurrence or metastasis.

Discussion

We encountered a case of recurrent malignant phyllodes tumor of the breast with RMS. Reports of phyllodes tumors containing RMS components of the breast are very rare,[6–13] and to our knowledge, reports of recurrence containing only the same RMS component as before are extremely rare.

Actually, pure RMS in the breast is very rare. In the present case, all tumor tissues were sampled, and immunostaining of epithelial markers was also performed, but no obvious epithelial components could be detected in the tumor. Combined with the positive immunohistochemical findings, such as MyoD1, myogenin, and desmin, these characteristics were consistent with alveolar RMS.

Phyllodes tumor needs to differentially diagnose from many tumors, including primary or metastatic sarcoma, desmoid-type fibromatosis, and metaplastic breast cancer.[14–18] To diagnose if it was RMS originating from the breast or not, it is necessary to distinguish it from the recurrence of previous phyllodes tumor. The tumor showed the histological manifestations of RMS as the previous site of the phyllodes tumor in a very short period of time, so that it was considered as a recurrence of the previously diagnosed phyllodes tumor

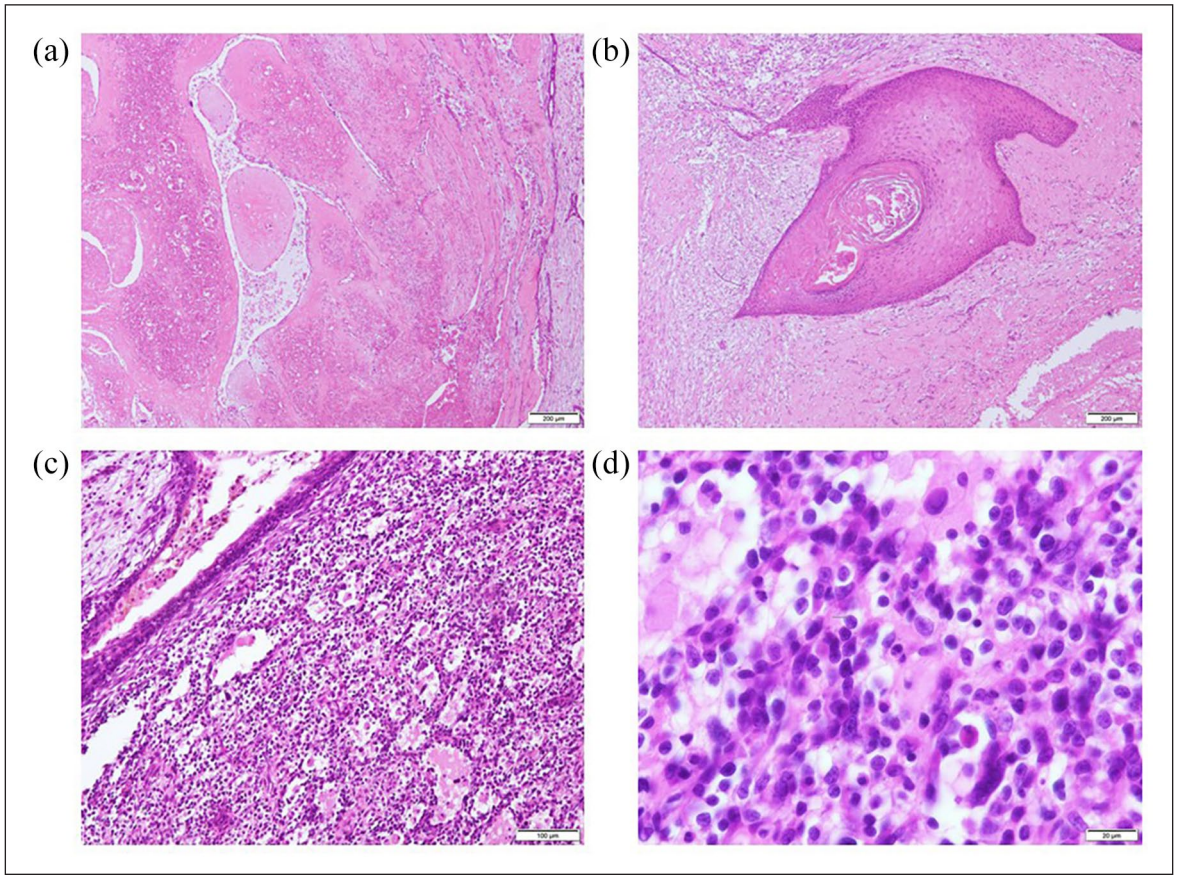

Figure 5. HE-stained images of the first surgical specimen. In the low-magnification field of view, stroma hyperplasia with foliate construction covered with atypical glandular epithelium with infarct (a) and polygonal metaplasia (b) were found. (c) In the medium-magnification field of view, the arrangement of tumor cells along the fibrous membrane was also found. (d) In the high-magnification field of view, the tumor cells were mainly spindle-shaped to polygonal cells with a high N/C ratio, and a mixture of acidophilic cytoplasm and striated myoblast-like cells with striated myoblasts was also found.

with the only RMS component. Although the previous hospital had considered the lesion to be a borderline phyllodes tumor, we considered it to be a malignant phyllodes tumor this time because of the malignant heterogeneous components. The local recurrence rate of phyllodes tumor is about 20% in total, and approximately 23%–30% are malignant.[1,19] Even if a tumor is benign at first, the degree of malignancy may increase with recurrence.[20] Indeed, in the present case, the Ki-67 index was higher at the time of recurrence than it had been as a primary lesion (data not shown). The only treatment of recurrent malignant phyllodes tumor of the breast with RMS at present is basically total mastectomy, as no standard for chemotherapy or radiation therapy has yet been established.[21] In this case, since it was a secondary operation and the local condition was in some sense turned to be uncertain, we decided to carry out VAC chemotherapy [13] and adjuvant radiotherapy. Nevertheless, chemotherapy is controversial because of its low benefit in metastatic and recurrent tumor diseases. While adjuvant radiotherapy appears to be beneficial for malignant tumors which shows a reduction in local recurrence.[22] As the reason that similar cases are very rare, we hope our report can provide corresponding diagnosis, treatment, and prognosis data for malignant phyllodes tumor containing the RMS component of breast. In summary, we encountered an extremely rare case of phyllodes tumor, and the recurrence form also showed unique findings involving the RMS component alone.

Conclusion

In conclusion, we reported a case of recurrent malignant phyllodes tumor containing only RMS component. The disease needs to differentially diagnose from many tumors. Meanwhile, the chemotherapy and radiotherapy, as well as the treatment standard of this disease, still remain to be controversial. The prognosis of each case also varies. In view of our insufficient understanding of recurrent malignant phyllodes tumor containing the RMS component of the breast, we need more data of similar cases and more in-depth scientific research on the disease.

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Availability of data and materials

The data set supporting the findings and conclusions of this case report is included within the article.

Declaration of conflicting interests

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Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

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Informed consent

Written informed consent was obtained from the patient for their anonymized information to be published in this article.

ORCID iDs

Jia Han https://orcid.org/0000-0003-0108-996X
Akihoro Shioya https://orcid.org/0000-0002-1863-5368
Sohsuke Yamada https://orcid.org/0000-0003-2662-0024

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