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An Extremely Rare Malignant Glomus Tumor of the Breast: A Case Report and Review of the Literature

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1. Abstract

1.1 Introduction: Glomus tumor is a rare neoplasm that originates from the glomus body in the dermis or subcutaneous of the extremities. In rare instances, glomus tumors have been observed in visceral organs. Here, we report an extremely rare case of primary malignant breast glomus tumor.

1.2. Presentation of the Case: A 75-year-old female presented to the hospital with a complaint of a painful 4.7cm diameter lump on the upper-outer quadrant of the right breast. Diagnostic image revealed circumscribed tumorous lesion. The tumor was recommended for removal based on presentation of monotonous cell proliferation, clots, and hemosiderin deposits by core needle biopsy. Pathologic analysis of the resected tumor with hematoxylin and eosin stain, and immunohistochemical studies corresponded with malignant glomus tumor of the breast.

1.3. Conclusion: Glomus tumor is primarily found in extremities but this case reports on an extremely rare, malignant glomus tumor arising from the breast.

2. Introduction

A glomus tumor is a rare neoplasm with an estimated 1–2% among soft tissue tumors. Most glomus tumors originate from glomus body in the dermis or subcutaneous of the extremities in areas rich in glomus bodies. Although the major affected sites of glomus tumor are fingertips and the pulp, glomus tumor can in rare instances occur in various organs, including gastrointestinal tract, lung, mediastinum, liver, ovary, bone, and deep soft tissue[1]. There have been few reported cases of glomus tumor of the breast since the first case reported in 2009[2]. Here, we report an extremely rare case of malignant glomus tumor of the breast and successful treatment with no recurrence after 1 year.

3. Case report

A 75-year-old woman with no history of cancer presented to the hospital with a tender mass on the upper outer quadrant of her right

breast that was first noticed by her 6 months prior to the present visit. She appeared otherwise healthy, and she did not have a family history of cancer.

Mammography showed a large hyperdense mass with round circumscribed partially obscured margins and no microcalcification in the upper area of the right breast (Figure 1).

Breast Sonogram of both transverse and sagittal scans revealed smooth and well-circumscribed round tumor, heterogenous internal echo partly showing hypoechoic regions. These findings presumed intra-cystic tumor or papillary lesions. Sonographic maximum diameter was 47,8mm in the transverse plane (Figure 2).

Breast magnetic resonance imaging, contrast-enhanced T1-weighted image showed ball-like tumor, rim enhancement and segmental internal enhancement on the late phase image. The time intensity curb following administration of gadolinium showed a rapid-plateau pattern and initially suggested a benign tumor (Figure 3).

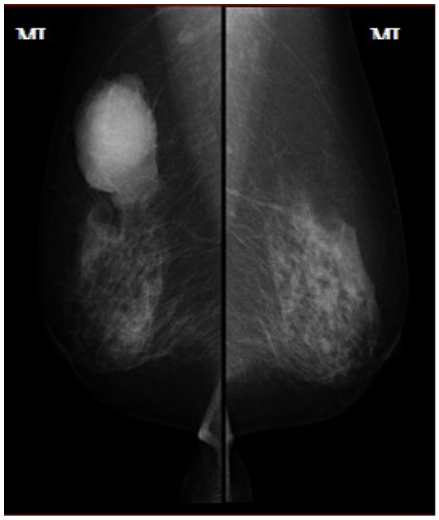

Figure 1: Mediolateral oblique (MLO) mammogram showed ball-like, round hyperdense mass in the upper area of the right breast with partially obscured margins and no microcalcification.

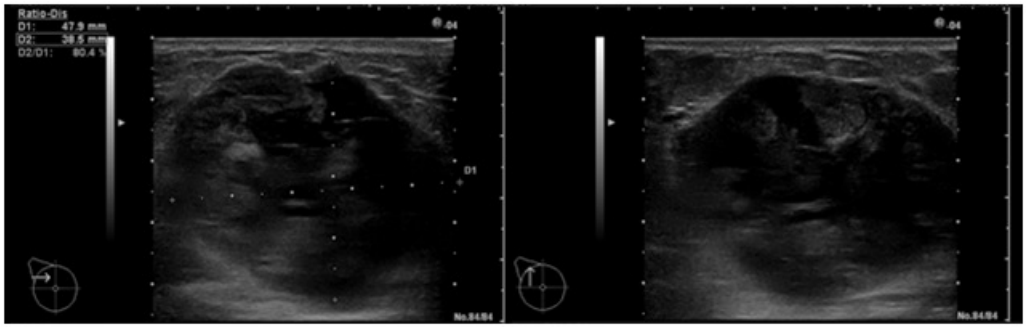

Figure 2: Sonography of both transverse (left) and sagittal (right) scan revealed smooth and well circumscribed round tumor, heterogenous internal echo partly showing hypoechoic regions with an echogenic rim. Maximum diameter being in the transverse plane 47,8mm. Intracystic tumor was speculated.

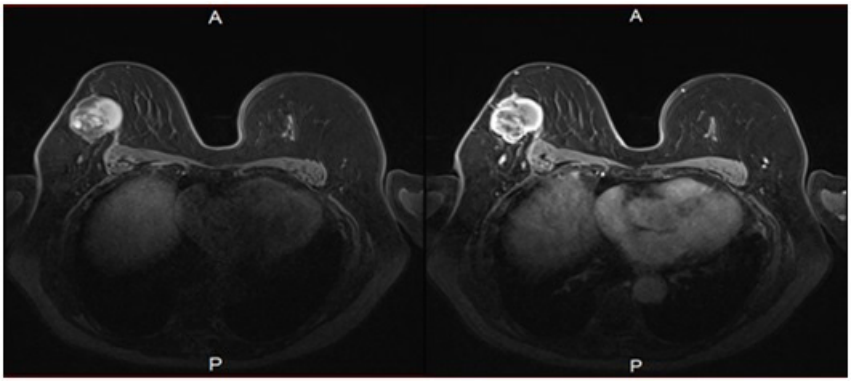

Figure 3: Breast magnetic resonance imaging (MRI), contrast-enhanced T1-weighted image showed ball-like tumor, rim enhancement, and segmental internal enhancement. The time intensity curve following administration of gadolinium showed rapid-plateau manifestation appearing like a benign tumor.

The core needle biopsy (CNB) of the tumor showed proliferation of many monotonous mild atypical round and spindle shaped cells. The diagnosis of the CNB specimen was suspicious of low-grade malignancy (Figure 4). The pathologists required extirpation of the tumor to define pathological diagnosis.

With patient's consent, lumpectomy and sentinel lymph node biopsy was done to elucidate tumor malignancy.

The gross pathology and the hematoxylin and eosin stained (HE) histological appearance showing encapsulated tumor consists of clotted blood in the central area and proliferation cells in the capsulated tissue (Figure 5). The surgical margin was free from the disease.

Low-power view of HE shows well circumscribed tumor as well as blood vessels of various-sizes (Figure 6A). Middle-power view of HE indicates infiltration of the tumor cells to the periphery of the capsulated tissue (Figure 6B). Tumor cells display monotonous round to ovoidal morphology with relatively clear cell borders, implying myoid features. Mitosis counting by high-power HE shows numerous mitotic figures, up to sixteen per 50 high power fields (HPF) (Figure 6C and D). Silver staining shows fine argyrophilic fibers between individual tumor cells with strongly positive immunohistochemistry findings of vimentin and smooth muscle actin (α-SMA) and weak or focal immunohistochemistry findings of synaptophysin and desmin (Figure 7). The Ki-67 was highly expressed around 70% of tumor cells, and the EMA, ER, S-100 protein, Factor VIII, and CD34, were negative (Table 1).

This tumor was characterized with the following: large (over 4cm in diameter), tender, highly angiogenetic, both α-SMA and vimentin immunohistochemistry positive, and highly proliferative (i.e., infiltrating glomus tumor cells, high mitotic index, and around 70% of Ki-67 labeling index). According to these results encompassing numerous vascular networks, myoid cell features, and the expression of α-SMA, the tumor was diagnosed as a malignant glomus tumor of the right breast. One year after the surgery, we found no sign of recurrence.

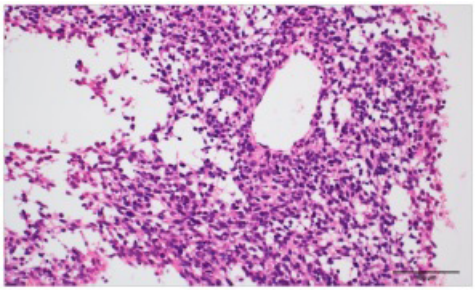

Figure 4: The breast core needle biopsy specimen showed proliferation of many monotonous mild-atypical round and spindle shaped cells. Low grade malignant tumor was suspected. (Hematoxylin and eosin staining. HE staining, scale bar 100μm)

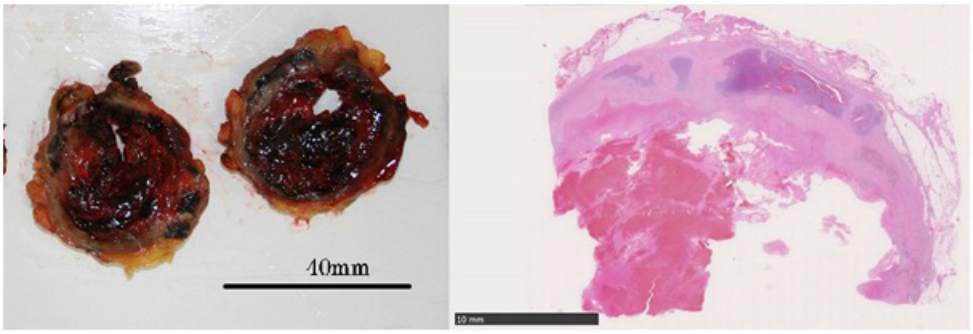

Figure 5A: Gross appearance of transverse section of the tumor showing encapsulated tumor zonal solid area enclose surrounding clots. 5B: Histologic appearance of the HE staining specimen showing central bleeding area and peripheral cell proliferation in the capsulated tissue (scale bar 10mm).

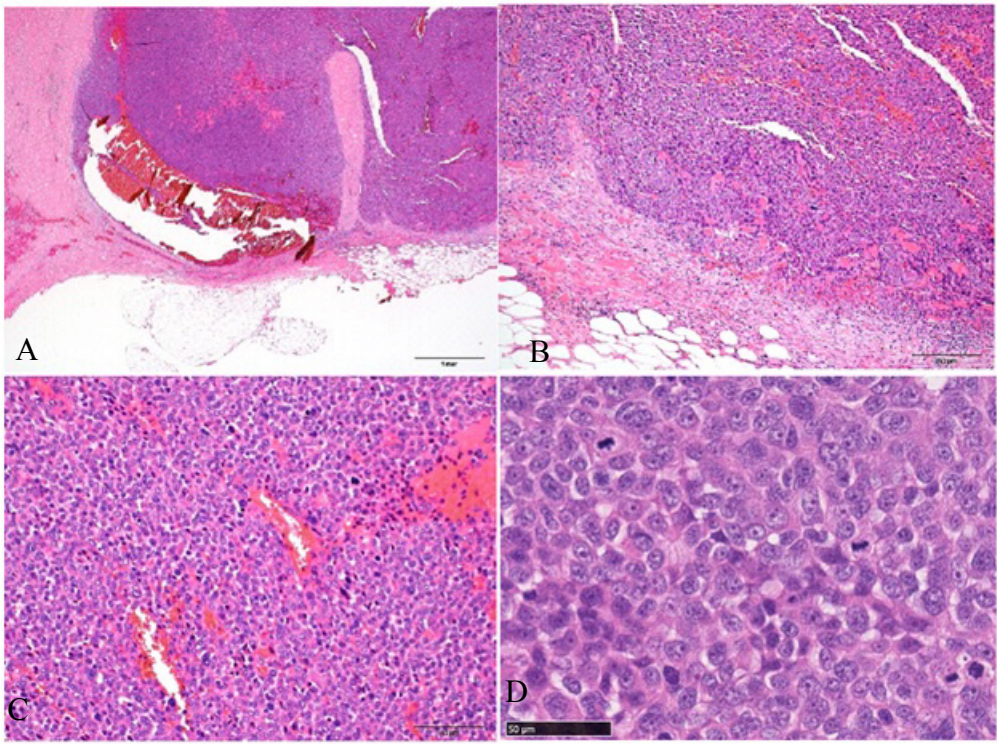

Figure 6: A. field lens x2, B. x10, C. x20, D. x40

A: Low-power HE staining shows well circumscribed tumor and many macro and micro blood vessels (scale bar 1mm). B: Middle-power HE staining indicates locally infiltrative tumor cells to the encapsulated tissue (scale bar 200μm). C and D: High-power HE staining shows high cellularity, marked nuclear pleomorphism and many mitotic figures, sixteen per 50 HPF (high power fields, scale bars 100μm and 50μm, respectively).

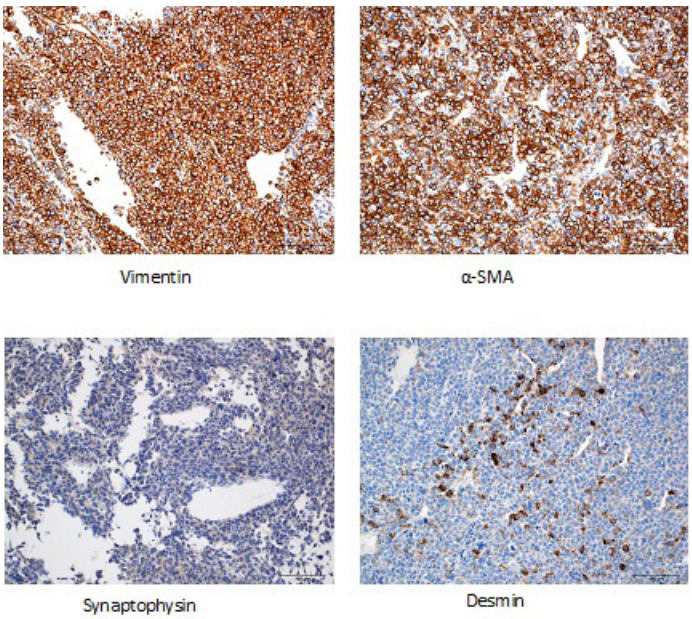

Figure 7: Microphotographs of strong positive vimentin, strong positive α-smooth muscle actin (α-SMA) immunohistochemistry, weak positive synaptophysin, and focal positive desmin.

Table 1: Immunohistochemical characteristics of the tumor cells.
List of immunohistochemical findings.

Antibody to	Tumor cells from the breast
CK AE1/3	Negative
CK CAM5.2	Negative
EMA	Negative
Vimentin	Positive
Chromogranin A	Negative
Synaptophysin	Weakly positive
CD56	Negative
ER	Negative
CD20	Negative
CD79a	Negative
CD3	Negative
CD10	Negative
S-100 protein	Negative
Ki-67 L.I.	About 70%
Desmin	Focally positive
α-SMA	Positive
H-Caldesmon	Negative
D2-40	Negative
Factor VIII	Negative
CD31	Negative
CD34	Negative
ERG	Negative
Collagen type IV	Difficult to evaluate
BRAF V600E	Negative

Abbreviations: CK AE1/3, Cytokeratin AE1/AE3. CK CAM 5.2, Cytokeratin CAM 5.2. EMA; Epithelial membrane antigen. ER; Estrogen Receptor.

Ki-67 LI; Ki-67 labeling index, α-SMA; α-Smooth muscle actin. ERG; Ets related gene.

4. Discussion

Glomus tumor commonly arises from the altered smooth muscle cells surrounding the thermo-regulating arteriovenous anastomosis in the glomus bodies which primarily exists within the subungual region of the fingertips. Glomus bodies regulate body temperature and control of blood pressure. Rarely, extracutaneous glomus tumors were reported in visceral organs including gastrointestinal tracts, liver, lung, mediastinum, ovary etc. in which the glomus body is scarce or absent [3]. Based on a PubMed, Google Scholar, and Crossref database search from March 2009 to April 2021 using keywords “glomus tumor” and “breast.” Only a few cases reports have been reported after the first case report by Yalcin et al. in 2009 [2,4,5,6]. Vasilevska-Nikodinovska et al. (2019) reported, a 38-year-old woman with uncertain malignant glomus tumors in multiple lesions in the muscle compartment, kidney, heart, and breast. The authors suggested an Ewing sarcoma of the left kidney and reported multiple lesions formed as metastases including the breast [7]. The other cases reports were a rare benign glomus tumor in the breast in male patients [8, 9]. Folpe (2001) defined the criteria for classifying malignant glomus tumors as a deep localization with size greater than 2cm, an atypical mitotic index, or a high mitotic activity >5 mitosis /50 fields/50HPF [10]. In our case, the patients represented right breast tumor with tenderness, over 4cm in size. Moreover, HE staining showed high cellularity, marked nuclear pleomorphism, and elevated mitotic index of sixteen per 50 HPF (high power fields). The high mitotic index matches thehigh rate of the Ki-67 labeling index (70% of tumor cells).

Immunohistochemically, malignant glomus tumors are similar to benign glomus tumors. Like our case, they express the vimentin and smooth muscle actin (α-SMA). They may lack type IV collagen and caldesmon expression yet weakly demonstrate positive synaptophysin and focal positive desmin. Other markers including cytokerarin, EMA, CD34, S-100 etc. are negative as usual (Table 1). After careful analysis of the clinical and pathological examination, we concluded this tumor is an extremely rare case of malignant glomus tumor arising from the breast.

Treatment in this case is surgical with complete excision. Insufficient resection can lead to recurrence in 1–2% of cases [11]. The literature mentions a successful outcome by multidisciplinary approach with radiotherapy and chemotherapy to a malignant glomus tumor in the cervical region after surgery [12]. Surgical removal has been the only proven therapy for malignant glomus tumor up to now and in our case the patient has not seen a recurrence one-year post-surgery.

5. Conclusion

This report presented an extremely rare case of malignant glomus tumor arising from the breast.

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