Case Report

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Granular Cell Tumor of the Axillary Accessory Breast: A Case Report

액와부 부유방에 발생한 과립 세포 종양: 증례 보고

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Granular cell tumors (GCTs) are rare benign soft tissue tumors that can occur throughout the body, particularly the head and neck; only 5%–8% of GCTs occur in the breast. We report a case of a GCT of the axillary accessory breast, which is a rare location of this tumor. A 50-year-old woman had a 2-month history of a palpable mass in the left axilla. Physical examination, as well as mammographic and ultrasonographic findings suggested a breast malignancy. Histopathological examination showed a benign GCT, and wide local excision was performed. The patient has remained disease-free over 2 years post-operatively. Although most GCTs are benign, wide complete resection of the tumor and follow-up are required considering the possibility of recurrence. The radiologist should know the characteristics of GCTs as a differential diagnosis of breast and axillary lesions to prevent unnecessary treatment.

Index terms Granular Cell Tumor; Accessory Breast Tissue; Axilla; Mammography; Ultrasound

INTRODUCTION

A granular cell tumor (GCT) is a soft tissue neoplasm derived from the Schwann cells of the nerve sheath (1). GCTs may occur throughout the body, especially in the head and neck, and 5%–8% of GCTs occur in the breast (2). It is necessary to distinguish this tumor from breast cancer because they show similar clinical and radiological features. However, diagnostic imaging presentation of GCT of the breast is variable (2). In order to delineate the diagnostic challenges and the therapeutic options of GCT, we report a case of a patient who developed a GCT in the axillary accessory breast, which is a rare location.

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CASE REPORT

A 50-year-old woman came to our hospital with a palpable mass in the left axillary area with no skin change in the overlying skin for 2 months. Physical examination showed that the lump was approximately 1 cm in diameter, had poor mobility, a smooth surface, and no tenderness. A provisional diagnosis of left axillary lymph node enlargement was made and additional examinations were performed.

A circumscribed, round, and isodense mass that had not been observed 2 years earlier on mammography was discovered in the left axillary region on a recent mammogram (Fig. 1A). US examination of the axilla revealed a 1.1 cm, non-parallel, round, hypoechoic mass with a circumscribed margin and an echogenic halo (Fig. 1B). The mass was located within a small mammary gland-like structure in the axillary region. A color doppler image additionally revealed increased blood flow within the mass. The mass was adjacent to the skin layer on mammography and US. It was impossible to rule out breast carcinoma developing in the accessory breast tissue of the axilla. An US-guided core-needle biopsy of the mass was subsequently performed. Histologic examination of the core biopsy showed a benign GCT. Surgical excision was recommended, and wide local excision was performed. Intraoperatively, an axillary specimen of 3.2 cm × 1.5 cm × 1.5 cm was resected. The size of the attached skin was 2.6 cm × 1.0 cm × 0.1 cm. The cross-section showed a well-defined mass itself with a clear resection margin, measuring 1.3 cm × 1.2 cm × 1.1 cm in size. The mass was less than 0.1 cm apart from the skin margin. Histopathologic examination confirmed the diagnosis of a GCT of the axillary accessory breast. A surgical specimen showed tumor cells with a distinctive granular eosinophilic cytoplasm associated with typical nuclei, without an increase in nuclear division or another sign of malignancy (Fig. 1C). A diagnosis of GCT was established based on the immunohistochemistry (IHC) results. IHC analysis of the tumor showed strong S-100 positivity, CD68 positivity, and pancytokeratin negativity (Fig. 1D). The patient remains disease-free 2 years after surgery.

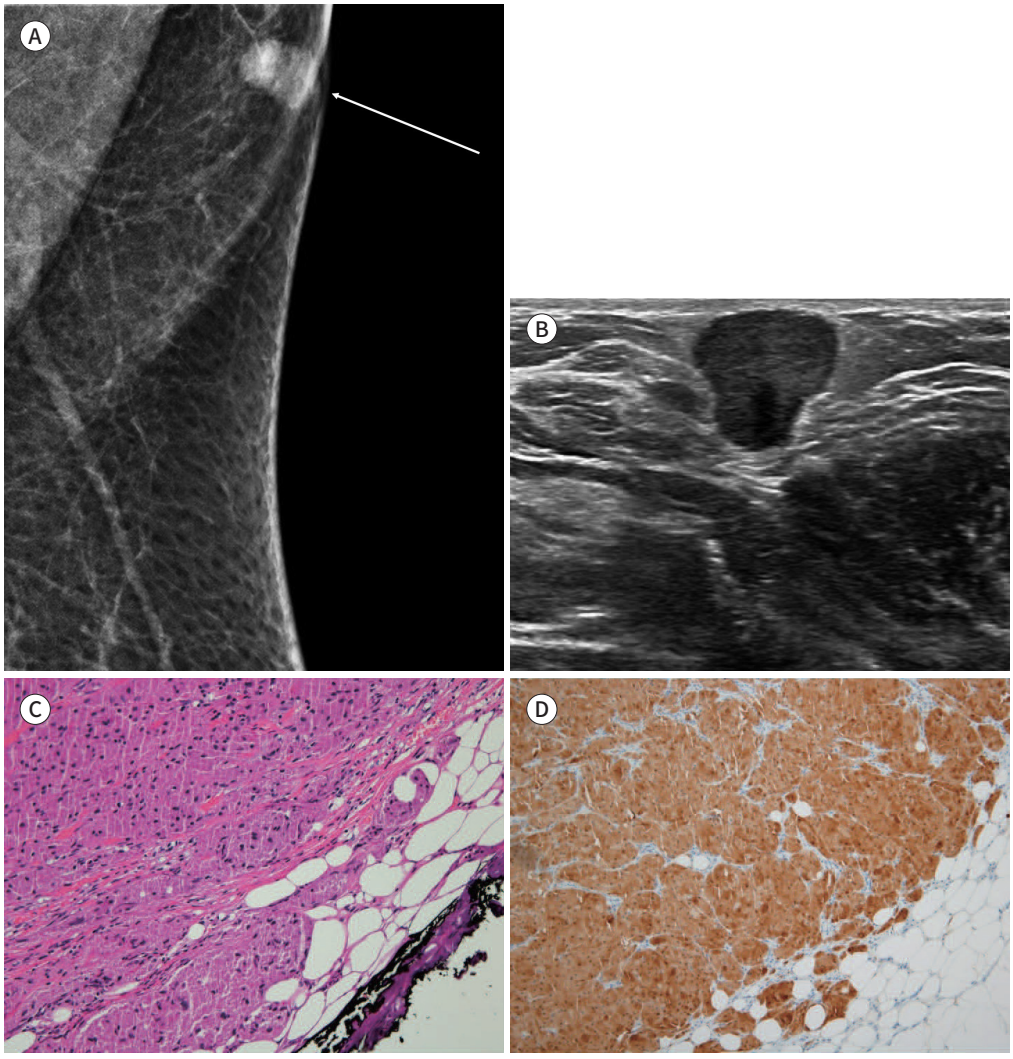
This study was approved by the Institutional Review Board of our institution, and the requirement for informed consent was waived (IRB No. 05-2022-122).

DISCUSSION

GCTs are rare benign soft tissue tumors that can occur anywhere in the body, especially in the head and neck. GCT was considered a myogenic tumor, however, IHC confirmed that GCTs were soft tissue tumors derived from nerve tissue. Currently, GCTs are accepted to be derived from the Schwann cells of the nerve sheath.

GCTs of the breast account for 5%–8% of all cases (3). In contrast to other breast tumors that frequently occur in the upper outer quadrant, GCTs of the breast most commonly occur in the upper inner quadrant, consistent with the location of cutaneous sensory territory of the supraclavicular nerve (4). There have also been reports of GCTs in other parts of the breast in the literature. However, it is known that GCTs in the accessory breast tissue are extremely rare. A literature search found only two cases of accessory breast tissue GCT. Liu et al. (5) described GCT of the accessory breast, which showed a hypoechoic lesion with a

Fig. 1. A granular cell tumor involving the left axillary accessory breast in a 50-year-old woman.
A. Mammography shows an isodense mass with a well-circumscribed margin adjacent to the skin layer in the left axillary region (arrow).
B. Ultrasonography shows a round hypoechoic mass with a well-circumscribed margin and non-parallel orientation and echogenic halo.
C. Photomicrograph of a histopathological specimen shows nests of large cells with granular eosinophilic cytoplasm containing typical small nuclei (hematoxylin & eosin stain, × 200).
D. Immunohistochemical analysis of the resected specimen shows cells with immunopositivity for the S-100 protein (× 100).



blurred margin and posterior shadowing on US and increased blood flow inside the mass on a color doppler image. Olivier et al. (6) described another patient with a GCT in the accessory axillary breast tissue that was also observed as an ill-defined, hypoechoic mass on US. The US findings in our case, in contrast to the two previously reported cases, revealed a round mass with a circumscribed margin and an echogenic halo.

GCTs can mimic breast cancer clinically and radiologically (3, 7). Clinically, they present as painless round nodules that are often palpable (1). Mammography has shown various findings, including round and circumscribed masses to indistinct or spiculated lesions (3). Mi-

crocalcifications have not been reported in GCT. On US, GCTs can be seen as spiculated or indistinct marginated masses with posterior shadowing or seemingly benign well-circumscribed masses (3). Because GCTs are often located subcutaneously, finding the epicenter of the lesion may provide a diagnostic clue, although they can be found at several depths (8). In our case, the mass was located in the subcutaneous fat layer, adjacent to the skin. MRI findings are not specific enough to differentiate the GCT from breast cancer (7, 8). Therefore, GCTs cannot be diagnosed based on radiological findings alone. The histological assessment of a GCT is essential, and core biopsy specimens are sufficiently representative to provide the pathological diagnosis (8).

GCTs of the breast are macroscopically solid, firm, homogeneous masses with a white to tan color without necrotic areas (7). Their histogenesis remains unclear. However, GCTs stain positive for S-100, and this protein is also found in Schwann cells, neural cells, and melanocytes. Although GCTs of the breast are sensitive to S-100, 10% of breast malignancies also stain positive for S-100. Therefore, other markers such as CD68 and neurospecific enolase are required to diagnose GCTs. The CD68 protein demonstrates the presence of lysosomal activity and stains positively for perineural Schwann cells and 90% of GCTs (9). Neurospecific enolase is found in the cytoplasm of neurons and neuroendocrine cells (10). The hypothesis that GCTs originate from the Schwann cells of distal nerves ending in the breast tissue is supported by these findings.

The treatment of benign GCTs in the breast is a wide local excision with free margins. Sentinel lymph node biopsies, lymph node dissections, and mastectomy are not required due to the benignity of the majority of GCTs. If the surgical margin is negative, the recurrence rate is 2%–8%; it is more than 20% when the surgical margin is positive. Therefore, it is necessary to follow up annually with clinical examination for about 10 years (9).

In conclusion, we report a rare case of an axillary accessory breast GCT with suspicious features of breast cancer, observed clinically and radiologically. The final diagnosis is guided by pathological assessment. Radiologists and clinicians should be aware of these findings of GCT in the differential diagnosis of breast and axillary lesions to avoid unnecessary overtreatment.

Author Contributions

Conceptualization, N.K.J.; data curation, L.K.; funding acquisition, N.K.J.; supervision, C.K.S.; writing—original draft, J.Y.J.; and writing—review & editing, N.K.J.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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액와부 부유방에 발생한 과립 세포 종양: 증례 보고

정윤주[1] · 남경진[2*] · 추기석[2] · 이계영[2]

과립 세포 종양은 신체의 모든 부위에서 발생할 수 있으나, 특히 두경부에서 주로 발생하는 드문 양성 연부 종양이며, 이 중 5%–8%가 유방에서 발생한다. 저자들은 드문 부위인 액와부 부유방에 발생한 과립 세포 종양 1예를 보고하고자 한다. 50세 여자에게 2개월 전부터 좌측 액와부에 만져지는 종괴가 있었다. 이학적 검사, 유방 촬영술 및 초음파 소견에서 유방의 악성 종양을 먼저 생각하였다. 조직 검사에서 양성 과립 세포 종양으로 진단되었으며 이후 광범위 국소 절제술을 시행하였다. 환자는 수술 후 2년의 추적 관찰 기간 동안 재발하지 않았다. 대부분의 과립 세포 종양은 양성이지만, 재발의 가능성 때문에 종양의 광범위한 완전 절제와 추적 관찰이 필요하다. 영상의학과 의사는 불필요한 치료를 막기 위해 유방 및 액와부 병변의 감별 진단으로 과립 세포 종양의 특징에 대해 알고 있어야 한다.

양산부산대학교병원 [1]외과, [2]영상의학과