

Glycogen-Rich Carcinoma of the Breast: A Case Report¹

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Glycogen-rich carcinoma of the breast, defined as one in which more than 90% of neoplastic cells have abundant clear cytoplasm containing glycogen, is very rare. We report a case occurring in a 50-year old woman, and include the mammographic and ultrasonographic findings.

Index words : Breast neoplasms , diagnosis
Breast neoplasms , radiography
Breast neoplasms , US

Carcinomas of the breast that have a clear cell appearance are uncommon neoplasms and may be either primary or metastatic. The majority of primary clear cell carcinomas of the breast are rich in cytoplasmic glycogen. Glycogen-rich clear cell carcinoma of the breast is defined as a carcinoma in which more than 90% of neoplastic cells have abundant, clear, glycogen-containing cytoplasm (1). A rare malignant tumor, it accounts for about 1 - 3% of breast carcinomas (1 - 3), and to our knowledge, only one case report describing its pathologic appearance has appeared in Korea (4). This did not, however, describe the mammographic or ultrasonographic findings. We report a rare case of glycogen-rich carcinoma of the breast, including the mammographic and ultrasonographic findings.

Case Report

A 50-year-old woman presented with a mass in the right breast, which over the preceding six months had

slowly enlarged. Other than her recent use of an estrogen medication for osteoporosis, her past medical history was unremarkable. Palpation indicated that the mass was 5 × 6 cm in size, firm, fixed and tender.

Mammography demonstrated a 3.5 × 4 × 5 cm sized, oval shaped, ill-defined margined, high density mass, with some spiculations, at the upper central portion of the right breast (Fig. 1A). A few central microcalcifications were revealed by spot-magnification mammography (Fig. 1B), and ultrasonography depicted a 2.2 × 2.4 × 4 cm sized, relatively well-defined, multilobulated, oval-shaped, relatively homogeneous hypoechoic mass with central bright echogenicity (Fig. 1C). Posterior enhancement and bilateral edge shadowing were observed. Fine needle aspiration was performed using a 24-gauge needle, resulting in "malignancy". At scintimammography using Tc99m-tetrofosmin (Fig. 1D), considerable focally increased uptake was noted in the right breast.

The patient underwent modified radical mastectomy, and a relatively well-margined, pale tan colored, solid firm mass, 2.9 × 3.3 cm in size, was discovered. Microscopically, it was an invasive carcinoma exhibiting solid growth of tumor cells and central necrosis. The cytoplasm was clear, and small, dark, punctate nuclei were centrally or eccentrically placed (Fig. 2A). Mitotic figures were infrequent. PAS staining revealed abun-

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dant positive granules in the cytoplasm of tumor cells (Fig. 2B); ipsilateral axillary lymph nodes were unaffected by metastasis. The pathologic diagnosis was glycogen-rich carcinoma of the breast.

Discussion

Carcinomas that accumulate abundant cytoplasmic glycogen arise in many organs, including the lungs, salivary glands, vagina, cervix, endometrium and ovary (5, 6). As revealed by routine sections, the extraction of water-soluble glycogen during histological processing gives the cytoplasm an optically clear, vacuolated appearance (6). Glycogen-rich clear cell carcinoma is a rare variant

of breast carcinoma, accounting for 1 - 3% of all breast carcinomas (1 - 3), and since first being reported in 1981 by Hull *et al.*, fewer than 50 cases have been described in the literature (1 - 4, 7).

All patients were women aged 37 to 78 years, and the observed clinical features did not differ from those of the usual invasive or intraductal carcinomas of the breast. Most tumors have measured between 2 and 5 cm; the largest described in a clinical report has a diameter of 10 cm (8). Hormone receptor analysis revealed that approximately 50% of tumors were estrogen receptor-positive, but all lesions studied have been negative for progesterone receptor (7 - 9).

Pathologically, no specific gross features have been

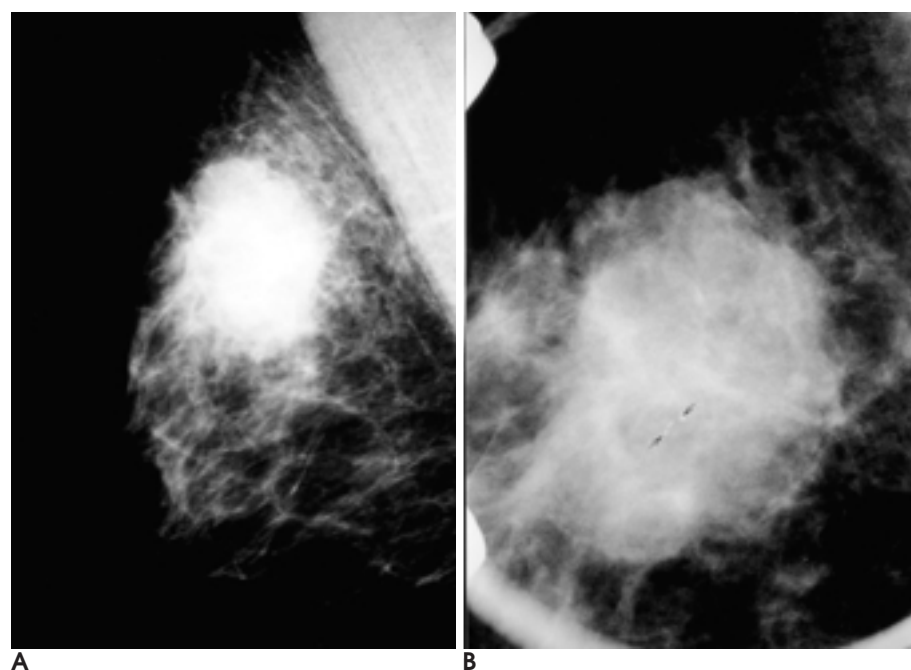
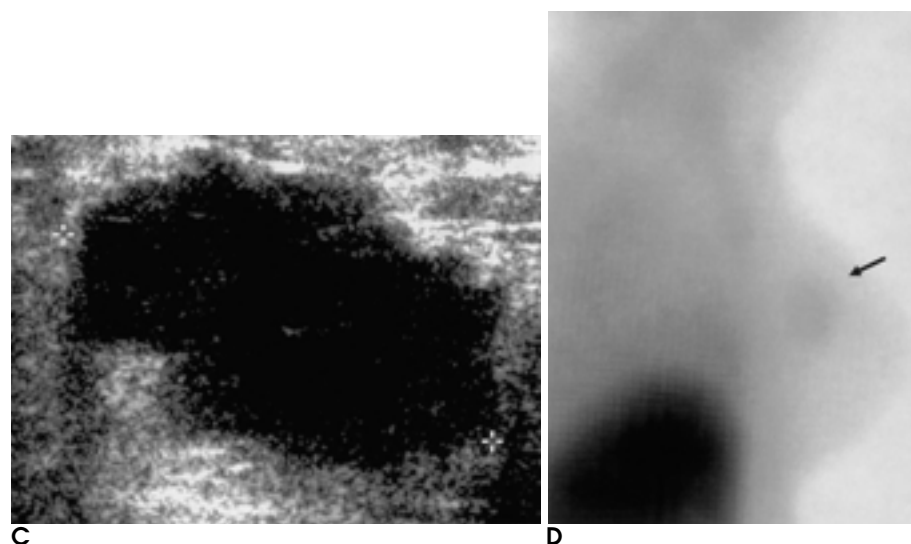


Fig.1. A 50-year-old woman with glycogen-rich carcinoma of the breast. **A, B.** Right mediolateral oblique (**A**) and spot compression magnification (**B**) mammograms show a 3.5 × 4 × 5 cm sized, high-density mass with ill-defined margin, some marginal spiculations and a few tiny central microcalcifications (arrows).

C. Longitudinal ultrasonogram of the right breast shows an 2.2 × 2.4 × 4 cm sized, relatively well-defined, multilobulated, relatively homogeneous hypoechoic mass with central tiny bright echogenicity. Posterior enhancement is noted.

D. Right lateral view of scintimammography shows a focal increased uptake in the right breast (arrow).



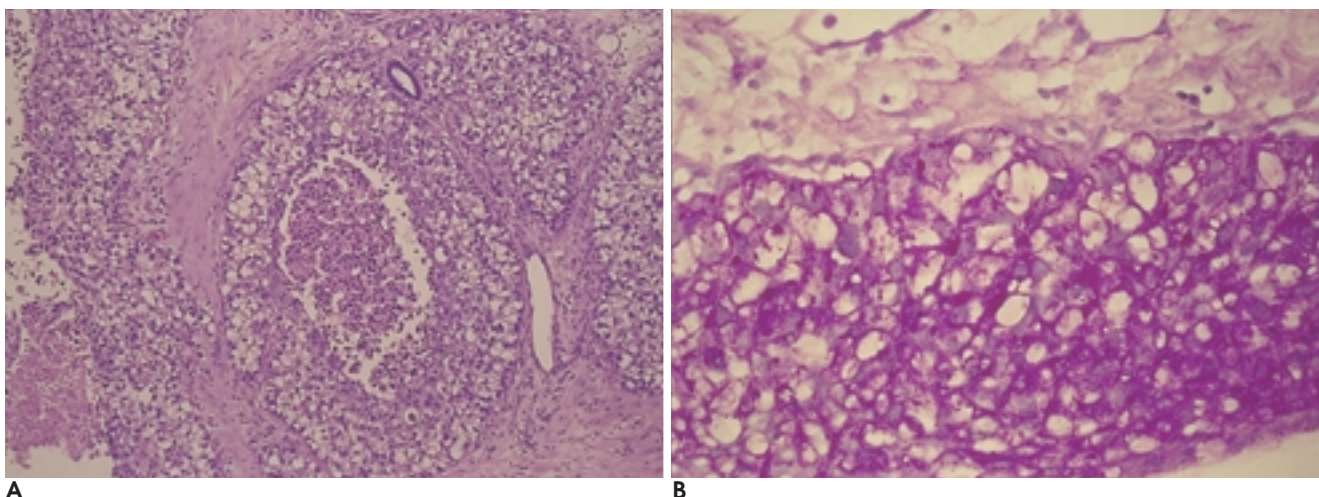


Fig. 2. A. Photomicrograph shows an invasive carcinoma composed of cells with clear cytoplasm and small, dark and punctate nuclei (H & E staining, $\times 40$).
B. Periodic acid Schiff (PAS) staining shows abundant PAS positive granules in the cytoplasm of the tumor cells ($\times 200$).

identified (9). The microscopic features of glycogen-rich carcinoma of the breast resemble the basic structural features of intraductal carcinoma alone or of intraductal and infiltrating duct carcinoma (6). The intraductal component identified in most cases shows a compact solid, comedo, cribriform, or papillary growth pattern (9). In invasive areas, the tumor cells form cords, solid nests, or papillary structures. Cytologically, the cytoplasm is clear or, less often, finely granular or foamy. The central or eccentrically placed nuclei are hyperchromatic, and mitotic figures are infrequent (6). The differential diagnosis includes lipid-rich, secretory, signet-ring and cystic hypersecretory carcinoma of the breast and metastatic clear cell renal carcinoma. The cytoplasm shows a positive diastase-labile reaction at PAS staining. Focally, cells are only Alcian blue- or mucicarmine positive (3, 8), and the oil red O stain for lipid is negative (8).

Although case reports have quite frequently included the pathologic findings (1 - 5, 7 - 9), only two such reports (7, 8) have described those of mammography and/or ultrasonography, using the term "suspicious malignancy" or mentioning the visualization at mammography of "a large, malignant tumour with spicule formation and vague demarcation" and of one suggestive of cystosarcoma at ultrasonography. Our patient presented with a slowly growing palpable mass, and mammography demonstrated a 3.5 \times 4 \times 5 cm sized, high-density mass with an ill-defined margin, and some spiculations, and a few tiny microcalcifications. Ultrasonography depicted a large, relatively well-defined, multilobulated,

relatively homogeneous hypoechoic mass with central, tiny, bright echogenicity. Scintimammography revealed a large focal uptake. We considered that the differential diagnosis should include invasive ductal carcinoma (NOS), medullary carcinoma, mucinous carcinoma, lymphoma, and sarcoma.

In most cases, the appropriate treatment is mastectomy and axillary dissection. Axillary lymph node metastasis is frequent at presentation, occurring in approximately 30% of reported cases. The prognosis of patients with glycogen-rich mammary carcinoma is not particularly favorable and may be similar to or worse than ordinary invasive ductal carcinoma (2, 9).

Although glycogen-rich carcinoma of the breast is an uncommon malignancy, its possibility should be considered during the evaluation of a breast mass.

References

1. Toikkanen S, Joensuu H. Glycogen-rich clear cell carcinoma of the breast: a clinicopathologic and flow cytometric study. *Hum Pathol* 1991;22:81-83
2. Fisher ER, Tavares J, Bulatao IS, Sass R, Fisher B. Glycogen-rich clear cell breast cancer: with comments concerning other clear cell variants. *Hum Pathol* 1985;16:1085-1093
3. Hull MT, Warfel KA. Glycogen-rich clear cell carcinomas of the breast. A clinicopathologic and ultrastructural study. *Am J Surg Pathol* 1986;10:553-559
4. Kim NH, Kim WS, Ko YH, Lee JD. Glycogen-rich clear cell carcinoma of breast : a case report. *Korean J Pathol* 1994;28:316-318
5. Herbst AL, Scully RE. Adenocarcinoma of the vagina in adolescence : a report of 7 cases including 6 clear-cell carcinoma[s-called mesonephromas]. *Cancer* 1970 ; 25 : 745-757

6. Rosen PP. *Rosen's breast pathology*. Philadelphia : Lippincott-Raven, 1997 : 485-487
7. Hull MT, Priest JB, Broadie TA, Ransburg RC, McCarthy LJ. Glycogen-rich clear cell carcinoma of the breast. A light and electron microscopic study. *Cancer* 1981 ; 48 : 2003-2009
8. Soensen FB, Paulsen SM. Glycogen-rich clear cell carcinoma of breast: a solid variant with mucus. A light microscopic, immunohistochemical and ultrastructural study of a case. *Histopathology* 1987 ; 11 : 857-869
9. Hayes MMM, Seidman JD, Ashton MA. Glycogen-rich clear cell carcinoma of the breast: a clinicopathologic study of 21 cases. *Am J Surg Pathol* 1995 ; 19 : 904-911

2002;46:393 - 396

