Glycogen Rich Clear Cell Carcinoma of The Breast: A Rare Case Report

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Abstract: Glycogen rich Clear cell carcinoma of the breast is a rare subtype of breast carcinoma occurring in 1.4 to 2.7 % of breast carcinoma patients. The significance of understanding this carcinoma lies in the fact that they usually have an aggressive course and an adverse prognosis. Here we present a case of 50 yearsold female with a left sided breast lump for 2 yearswhich histologically showed the features of clear cell carcinoma breast and was confirmed by PAS staining. This tumor was also positive for pancytokeratin and Her2immunohistochemically.

Keywords: Clear cell carcinoma Breast, Histopathology, Immunohistochemistry

I. Introduction

Worldwide, breast cancer is the most common invasive cancer in women. It affects about 12% of women worldwide¹. There are many histological subtypes of breast carcinoma and a knowledge of each subtype aids in the prognosis of patients. Glycogen rich clear cell carcinoma of breast is a very rare subtype and is associated with variable hormone receptor positivity and worse prognosis. Clear cell carcinoma can also occur in other organs most notably Kidney(most common form of renal cell carcinoma), Ovary, Cervix, Lung, Endometrium and Salivary glands².

II. Case report

A 50 year old female came to our Institution with a complaint of left sided breast lump from 2 years which was gradually increasing in size without any associated pain. On examination a well circumscribed breast lump was palpated in the left upper quadrant which was firm to hard measuring $3\times3\times2.5$ cm³, immobile, fixed to underlying structures but not to skin. Axillary lymph nodes were not present on examination. FNAC of the lump was reported as ductal carcinoma breast and MRM of the breast was done. Specimen was sent for histopathology.

On gross examination the tumor measured $3.5 \times 3.5 \times 2.5$ cm³, yellowish to brown in color, firm in consistency with occasional hemorrhagic areas.

Microscopically,the tumor showed predominantly ductal pattern of growth characterized by averagesized polygonal cells, with well-defined borders. Foci of linear and trabeculargrowth patterns were visible. The cytoplasm was clear and hyperchromatic nuclei with a low mitotic count (2 mitoses per 10HPF) were detected. There was absence of necrosis and no lymphovascular invasion was noticed(fig.1).The tumour also showed PAS positivity(fig.2).

Immunohistochemically,this case was positive for pancytokeratin and Her2/neu, and negative for ER,PR and S100.

III. Discussion

Glycogen-rich clear cell carcinoma of the breast is a rare tumor occurring in 1.4 to 2.7 % of breast carcinoma patients^{3,4}. The first known report of clear cell carcinoma is by Hull et al in 1981³. Fisher et al gave the criteria of at least 50% of the tumor having clear cell morphology should be classified as clear cell carcinoma of the breast⁵. Removal of the water-soluble glycogen during histologic processing causes the cytoplasm to become completely clear in conventional hematoxylin and eosin (H&E)-stained sections, and this phenomenon has led pathologists to designate these tumors as clear cell carcinomas⁶. Age of presentation ranges from 32 to 81 years, usually with a mass accompanied by skin dimpling, nipple retraction, or pain in some cases⁶. Histologically, Glycogen-rich clear cell carcinoma. Our case presented here shows the feature of ductal pattern of growth. These tumors are PAS positive but diastase sensitive.PAS positivity was present in this case. Immunohistologically, clear cell carcinomas have a variable positivity for hormone receptors ER, PR, Her2/neu^{7,8,9,10,11,12}. The frequency of ER positivity varies from 35% to 62%⁷. This case was ER negative .PR is also observed to be positive in upto 43% of cases⁷. We found PR was negative in this case. Her2/neu has

been observed to be positive in 20% to 43% of cases in different researches^{7,11}.Her2/neu was positive in this case.S100 was found to be negative in this case which ruled out the possibility of myoepithelial carcinoma. Pancytokeratin positivity proved the epithelial or ductal origin of this case.

The prognosis of glycogen-rich clear cell carcinoma of the breast is reported to be not particularly favorable and may be similar to or worse than that of ordinary invasive ductal carcinoma, when compared on a stage-matched basis¹³.But our patient had a history of 2 years, and post surgery and chemotherapy, she is having a disease free survival for 6 months.



Fig.1-Tumor shows predominantly ductal pattern of growth with clear cell morphology-H&E(x20)



Fig.2-Tumour showing PAS positivity confirming the case to be glycogen rich clear cell carcinoma(PASx20)

V. Conclusion

Glycogen rich Clear cell carcinoma of breast is a very rare subtype of breast carcinoma with an aggressive clinical course. This case report emphasizes that this carcinoma may have an indolent course. Furthermore, immunohistochemically,this case was ER and PR negative but Her2 positive.

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