Carcinosarcoma of the Breast With An Unusual Secretory Carcinoma as the Carcinomatous Component

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Introduction

Carcinosarcoma (CaSa), as defined by the World Health Organization, is a tumour composed of intimately admixed malignant epithelial and stromal components [1]. It is considered a distinct form of metaplastic carcinoma of the breast. Secretory carcinomas of the breast are uncommon tumours, generally encountered in children and are therefore called by some authors as juvenile carcinomas. However, they have also been reported in older patients including post-menopausal women [2]. We report a case of CaSa of the breast with an unusual secretory carcinoma as the malignant epithelial component.

Case Report

A 70 year old post-menopausal lady presented with a painful lump in the left breast of three months duration. The lump was small initially and gradually increased in size. There was no past history of surgery / radiotherapy / chemotherapy. Clinically an irregular, firm and mobile swelling was felt beneath

the nipple and areola in the left breast. The nipple was drawn outwards. The skin was puckered. No peau d'orange was noted. The lump was not fixed to the underlying tissues. Axillary nodes were palpable. The right breast and systemic examination revealed no abnormality. She was diagnosed as carcinoma breast (staged as T3N2M0 - Stage 3B) and underwent a total mastectomy. Gross examination of the specimen showed a 9.0 x 7.0 x 4.0 cm grey-white tumour in the upper outer quadrant impinging onto the nipple and areola. The skin overlying the tumour was ulcerated. Axillary dissection showed a large 3.0 x 3.0 x 2.0 cm metastatic mass alongwith nine other lymph nodes, two of which were grossly metastatic.

Microscopic examination of sections of the breast tumour showed a malignant tumour with carcinoma and sarcoma elements. The carcinomatous component was a secretory type of carcinoma with a tubulo-alveolar pattern, lined by cells with vacuolated cytoplasm and forming lumina filled by eosinophilic PAS + secretions. The carcinomatous component was intimately admixed with a fusiform spindle cell sarcoma at places (Fig 1,2). The sarcomatous areas showed a high

Fig. 1 : Secretory carcinoma intimately admixed with a fusiform spindle cell sarcoma (HE x 100)

Fig. 2 : Secretory carcinoma intimately admixed with a fusiform spindle cell sarcoma (HE x 200)

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Fig. 3 : IHC stain : Secretory carcinoma element showing CK positivity whereas the sarcoma component is CK negative (cytotokeratin stain x 400)

mitotic count of 10/10 HPF. Areas of necrosis were seen. No osseous / cartilagenous matrix / osteoclastic giant cells/ squamous cell carcinoma areas were noted. The nipple and areola were uninvolved. The epidermis of the overlying skin was not involved but the tumour was seen close to the epidermo-dermal junction in the dermis. The metastatic mass and two other lymph nodes in the axilla showed metastasis of an adenocarcinoma with perinodal extension. Immunohistochemical staining of the primary tumour revealed the secretory carcinoma component to be cytotokeratin (CK) positive (Fig 3) and vimentin negative whereas the sarcoma component was vimentin positive (Fig 4) and CK negative.

Discussion

CaSa breast accounts for only 0.1% of all cases [3]. CaSa, as applied to the breast, has been used as a generic term for biphasic neoplasms having both malignant epithelial (carcinoma) and malignant stromal (sarcoma) components [4]. Metaplastic carcinoma is a generic term for tumours of ductal type in which the predominant component has an appearance other than epithelial and glandular and more in keeping with another cell type. The term is too encompassing, imprecise and includes sarcomatoid carcinoma, CaSa, spindle cell carcinoma, carcinoma with osteoclastic giant cells and squamous cell carcinoma [5]. The term CaSa is used where the sarcoma component resembles fibrosarcoma (FS), malignant fibrous histiocytoma (MFH), chondrosarcoma (CS), osteogenic sarcoma (OS), rhabdomyosarcoma (RMS), angiosarcoma (AS) or a combination of these various patterns, and when present with a carcinomatous component. The carcinomatous component usually is the typical ductal type of carcinoma. The term CaSa has not been applied consistently in literature. Some authors use the term CaSa only for those biphasic tumours where at least 50% of the tumour is composed of a cellular malignant appearing spindle cell component with the carcinoma component being contiguous or

Fig. 4 : IHC stain : Sarcoma element showing Vimentin positivity whereas the carcinoma component is Vimentin negative (Vimentin stain x 400)

admixed with the sarcomatous component. The stroma, by definition, had to appear sarcomatous. Other authors maintain that the term CaSa is applicable only to those neoplasms composed of epithelial and mesenchymal cancers without evidence of transition between the two [6]. The presence of a carcinoma not contiguous or admixed with an existing sarcoma is viewed as two separate tumours. Some authors have characterised these biphasic lesions according to the immunohistochemical reactivity of the mesenchymal elements; if positive immunostaining with CK markers is seen they would classify the tumour as metaplastic carcinoma and if none is present, the tumour would be grouped as 'CaSa'.

The main differential diagnosis of CaSa is an infiltrating carcinoma with formation of a pseudo-sarcomatous stroma. Sarcomatoid carcinomas often show separation, albeit a gradual transition between the epithelial and the sarcoma-like component. The sarcoma like element, here, is also CK positive. In our case, the carcinomatous and sarcomatous areas were intimately admixed. The sarcomatous areas showed a high mitotic count and were CK negative and vimentin positive ruling out the possibilities of a pseudosarcomatous stroma and a spindle cell carcinoma. FS, MFH, OS and stromal sarcoma were ruled out because of the presence of an overt malignant glandular component. Cystosarcoma phyllides with malignant transformation of the epithelial component was ruled out because of the absence of curved elongated ducts and stromal leaf-like projections and the presence of an extensive secretory carcinoma component. Matrix producing carcinoma was excluded because of the lack of chondroid and/or osseous stromal matrix. A squamous metaplastic component was not seen.

Secretory carcinoma is rare in adults. Even though

axillary lymph nodal metastasis are seen, systemic metastasis is rare in a pure secretory carcinoma [3]. There is no known hormonal abnormality that would explain the prominent secretory activity in this type of carcinoma.

Metaplastic carcinomas of the breast are a heterogeneous group of neoplasms with CaSa being a distinct form and also the most aggressive form in keeping with its high-grade stroma. These tumours commonly occur in older individuals and display aggressive behaviour. CaSa is reported to have a 5 year survival of 49%. Although controversy still exists regarding their exact histogenesis and their classification remains obscure, pathologists should recognize and diagnose CaSa to assign the correct prognosis and ensure appropriate treatment to the patient.

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