Adenoid Cystic Carcinoma of the Breast A case report and review

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SUMMARY: Adenoid cystic carcinoma of the breast is a malignant tumour of significant incidence with a better prognosis than histologically identical tumours occurring in other sites and as compared to other malignant tumours of the breast. The recognition of the tumour is important especially in the management of the individual patient but also in furthering understanding of growth, spread and biological behaviour of tumours.

Introduction

Adenoid cystic carcinoma of the breast is a rare tumour with an incidence of one to two cases per thousand mammary carcinomas(1-3) and a total of around one hundred and twenty reported in the world literature. It has identical histological features to adenoid cystic carcinoma of the salivary glands, and has to be differentiated pathologically from intra-ductal carcinomas with a cribriform pattern found in the breast, from which it has important distinguishing features; these are summarised in Table I. The widely divergent prognosis of these two mammary neoplasms makes their differentiation particularly important. The favourable prognosis of this tumour in the breast as compared to other sites remains unexplained. The UK cancer registry(4) records over twenty thousand cases of carcinoma of the breast each year, which would indicate an incidence of twenty to forty cases each year of adenoid cystic carcinoma of the breast; it seems likely from the currently reported series that a large proportion of these are not being recognised.

Case Report

A forty three year old white female presented with a five month history of a non-tender lump in the right breast, deep to the areola. There was a three year history of an intermittent bilateral odourless green discharge which varied with her menstrual cycle. It had never contained blood. Two mammograms had been performed in the past, the first nine years previously when the patient had given a history of breast tenderness and the second three years previously as part of an age-related breast screening program. Both were reported as normal.

Having had 3 children, the patient had breast fed for six weeks, four months and six months respectively. Two trials on the oral contraceptive pill were abandoned because of breast tenderness, weight gain and hypertension. There was no past medical history of breast surgery nor a family history of breast disease.

Physical examination showed the right breast to contain an area of nodular breast tissue enclosing, supero-lateral and deep to the areola, a hard mass. The

mobile mass was tender with normal overlying skin and no nipple retraction. There was no axillary or supra-clavicular lymphadenopathy.

At incision biopsy a superficial macroscopically "non-suspicious" cystic lesion was found, deep to which was a hard non-adherent mass, measuring 5cm x 2cm occupying the subareolar area and superolateral quadrant. Histology was reported as adenoid cystic carcinoma of the breast, confirmed following a simple mastectomy with axillary clearance five days later. Figures 1 and 2 demonstrate the histology as found in this patient. There was no evidence of lymph node involvement but apocrine metaplasia and mild benign mammary dysplasia was present. No further treatment was given. The patient remains well and has no clinical recurrence of disease after five years follow-up.

Discussion

Adenoid cystic carcinoma occurs rarely in the breast; it occurs more frequently in the major and minor salivary glands and in mucous and mucoserous glands of other sites, giving rise to tumours in the lacrimal gland, external auditory canal, oesophagus, cervix uteri, prostate, Bartholin's gland and skin(1,2,5,6,7,8).

In extra-mammary sites it has, untreated, a characteristic slowly progressive course, with local recurrence, lymph node involvement and ultimately distant metastasis and death. When occurring in the breast it carries a favourable prognosis; only five cases have been reported causing death(9-12), of these the diagnosis has been questioned in two cases(1,13) and documentation is incomplete in two(9,10). Metastases are uncommon, reported occurring to lymph nodes in two cases(11,14) and more distant haematogenous spread most commonly to lung(15-17), often after a delay of several years. Koller *et al* report a solitary case to the brain after 12 years(18). Ten cases of local recurrence, nine following local excision, are reported(3,5,8,17,19,20,21) accounting for simple mastectomy being the treatment of choice. Deep tumour fixation is rare(19,22) occurring in the presence of large tumour masses. The reported incidence of skin change ranges from none in 21 cases(5) to five of nine

Table 1

Comparison of Pathological Features of Adenoid Cystic Carcinoma and Intraductal Carcinoma

Adenoid Cystic Carcinoma	Cribriform Intraductal Carcinoma
Biphasic Cellular Pattern	
present	— absent
cribriform or reticular	
basophilic cells	
— cuboidal cells	
eosinophilic lining ducts	
Mucin Staining Pattern	
biphasic	
— pseudocysts—Alcian blue +ve	PAS ++
— ducts PAS strong +ve	Alcian blue +ve
Alcian blue weak +ve	
Basophilic Cells Lining Pseudocysts	
— irregular cobblestone appearance	— regular polarised
Cells Comprising Cribriform or Reticular Lobules	
— small, uniform cells with small round or oval nuclei.	large with atypical pleomorphic nuclei
— 1 or 2 small nucleoli	prominent nucleoli
— sparse cytoplasm	abundant finely granular cytoplasm
Mitotic Figures—infrequent	
	— frequent
Necrotic Areas—infrequent	
Histological pattern maintained at periphery on invasion of stroma or fat	— frequent
	Histology develops pattern of infiltrating ductal carcinoma with infiltration of surrounding stroma or fat.

Fig 1. Typical cribriform pattern of tumour islands in connective tissue stroma. Basaloid cells with scanty cytoplasm surround cystic spaces. Haematoxylin and eosin $\times 20$.

Fig 2. High magnification of cribriform pattern of adenoid cystic carcinoma of the breast. Haematoxylin and eosin $\times 400$.

cases(6); fungating tumours have been reported(7,13,17,23). Nipple discharge has been reported only twice before; in each case the diagnosis of adenoid cystic carcinoma has been questioned by Azzopardi(2). In the case currently reported other breast pathology was present, the histology being reported as apocrine metaplasia and benign mammary dysplasia.

Associated tumours are reported in a number of cases; Cavanzo and Taylor(5) state that the prognosis of the patient is that of the other tumour. Associated

carcinoma is reported by a number of authors(1,6,7,24,25,26). Peters and Wolff(16) report a synchronous contra-lateral intra-ductal carcinoma. Also reported are fibroadenoma(6,8,19), sclerosing adenosis(6,27) and cystic spaces lined with metaplastic epithelium(17).

Mammography of cases of adenoid cystic carcinoma of the breast reported by three authors produces no consensus of opinion as to its 'typical' appearance(2,27,28).

Five authors(11,21,24,29,30) report cases in multiple patients.

Conclusion

A further case of this unusual but significant tumour is reported. Adenoid cystic carcinoma of the breast is important by virtue of its behaviour when related to its site and its histology. Adenoid cystic carcinoma of the breast, and of the skin, has a significantly better prognosis than when the histologically identical tumour occurs in other sites. Prioleau et al(7) postulate both anatomical and biochemical explanations for this. As the prognosis is good with only rare cases of distant metastasis(9,11,17,18,19) even following excision, local recurrence or presentation with large locally invasive tumours, it is hard to support an anatomical basis for the differences, especially in view of the apparent frequency of metastasis observed with other breast tumours. The biological and biochemical features of this tumour then gain importance and require further study to evaluate how these are related to relative biological aggressiveness.

Simple mastectomy is advocated as the treatment of choice by most authors(5,7,11,24,27,28,30) to minimise the risk of local recurrence.

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