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ADENOID CYSTIC CARCINOMA OF THE BREAST AT ENUGU NIGERIA

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ABSTRACT

Background: (Adenoid cystic carcinoma (ACC) of the breast is a rare type of neoplasm that is histologically indistinguishable from other examples in other sites and generally has a good prognosis).
To characterize the clinical and pathological features of ACC in our environment, as well as the treatment offered to our patients, a review of the clinical records of patients treated at the University of Nigeria Teaching Hospital Enugu was undertaken.

Patients and Methods: Case notes of breast cancer patients stored in the medical records department were retrospectively reviewed with a view to studying those with ACC. Follow up on these patients were documented.

Result: Adenoid cystic carcinoma of the breast was diagnosed in 9 out of 222 patients treated for cancer of the breast, from 1995-2000. Patients aged 34-70years were afflicted with this disease. A lump in the breast led to the initial suspicion of a tumor. Some of them had pain in the breast. Many of the patients came with advanced disease. Surgical treatment ranged from simple mastectomy to modified radical mastectomy with radiotherapy and chemotherapy in some patients.

Conclusion: The disease is rare at Enugu but contrary to findings elsewhere, majority of our patients had advanced disease. Those with early disease appeared to have a good outcome.

Key words: Adenoid cystic carcinoma, Enugu, rare, advanced disease. *(Accepted 19 April 2006)*

INTRODUCTION

It is one of the rare forms of primary breast cancer reported in individual case reports[2, 3] or as a small series of patients, and larger series have tended to originate from major referral centers [4, 5]. Patients with ACC have an excellent prognosis compared to ACC in other locations, mainly in the minor salivary glands [2], with rare instances of local recurrence or distant metastasis[4-6].
Nine cases of adenoid cystic breast cancer seen over a period of 6 years in the University of Nigeria Teaching Hospital Enugu were assembled for study. Since many reports up to now [4,6,-8] describe isolated cases, or small series of patients, one felt that reporting the result of analysis of these 9 cases would be a useful addition to information about this disease. This analysis includes the clinical and pathological features as well as the treatment, to see if the prognosis is as good as described in literature.

Correspondeces: Dr G.E.Njeze

PATIENTS AND METHODS

Records of patients treated for carcinoma of the breast at the University of Nigeria Enugu, over a 6year period (from January 1995-December 2000) were retrospectively reviewed. Those with ACC were sorted out for study. Information collected from the case notes included the age, sex, complaints, clinical findings, treatment offered, and histology reports. Finally the record of follow up visits to the surgical clinics by these patients was documented.

RESULTS

Nine out of 222 (two hundred and twenty two) patients were shown to have ACC of the breast on histological examination. Apart from one patient aged 70years, the rest were below 46 years of age. Four of these patients had other forms of synchronous carcinoma necessitating the diagnosis of multiform carcinoma (Table 1). These included invasive ductal carcinoma, medullary carcinoma, and mucinous carcinoma. Each of these had a breast lump, 2 of them being recurrent lumps, while 5 had pain in the breast. Only 1 patient had a serosanguinous discharge.

Some of these patients waited for over 7 months before presenting to the clinic. Seven patients had palpable axillary lymph nodes and these included all the patients with multiform carcinoma. Four of our patients had distant metastatic disease after mastectomy (Table 1). For surgical treatment, 4 patients were offered modified radical mastectomy, while 5 had simple mastectomy. Two patients had radiotherapy, while 8 had cytotxic therapy (with cyclophosphamide, methotrexate and 5-fluorouracil) in addition to mastectomy (Table 1).

Follow up visits by these patients to the clinic was documented. None of the patients with metastasis shortly after treatment was initiated was seen again after one year in the clinic. One patient whose disease was staged as T_2 N_2 M_0 kept her appointment regularly for 30 months; thereafter she was not seen again. Three patients who had ACC without any other synchronous tumor kept their appointments to the follow up out patient clinic for 5 years. Thereafter they were lost to follow, but when they were last seen, they were in good health.

DISCUSSION

Adenoid cystic carcinoma of the breast is a rare form of primary cancer accounting for less than 1% of tumors of the breast [1,2,7]. In this study, it accounted for 4% of cancers of the breast seen in our hospital. The reason for this higher occurrence is not immediately obvious, although it is known that some patients with breast cancer, in our locality seek treatment from unorthodox healers and churches, rather than attend hospitals. The effect is that some cancer patients living in the geographical area covered by this teaching hospital were not part of this hospital statistics.

ACC most frequently manifests as a tender breast mass, often in the subareolar area, but nipple discharge is an uncommon symptom. One of our patients with mucinous and intraductal carcinoma in addition to ACC, had a serosanguinous nipple discharge. Intraductal carcinoma is often associated with nipple discharge and may have been responsible for the discharge in this patient. ACC of the breast is said to be far more common in women than in men; indeed all our 9 patients were women. Pain is said to be a prominent symptom in these patients just as in those with ACC of the salivary gland. This symptom was recorded in 5 out of our 9 patients. In the salivary gland, ACC is associated with perineural invasion by tumor tissue but that was not seen in our patients, just as in those reported by McClenathan.[4]

Localized pain, which is known to be a prominent symptom in patients with mammary ACC, has been recorded to preceded the appearance of a mass either clinically or radiographically, in some patients.[3,7] The pain actually lasted for up to one year in some of these patients, before the detection of this carcinoma.[4] With this information, it is advisable for the surgeon who sees patients with breast pain to always have the diagnosis of ACC at the back of his mind, while considering other causes of mastalgia, in patients aged 34 years and above. Such patients may have to be followed for up to 1 year, though without frightening them, just to ensure that one does not miss the early diagnosis of ACC. Sheen-Chen et al [7] suggest that the presence of a painful breast lesion without obvious inflammatory evidence while compressed is a meaningful clue, which should lead to the suspicion of ACC.

Lymph node metastasis has been reported to be extremely uncommon in ACC of the breast.[2,8] In this study of 9 patients however, only 2 were free from axillary node metastasis.

These 2 patients had ACC without any other synchronous tumor, and came with early disease (Table 1). The rest either had synchronous tumors or harboured lone ACC for periods of time varying from 7 months to 5 years (Table 1). These 2 factors may have led to the observed high incidence of axillary node metastasis in our patients. Simon et al [1] had shown in a study of 9 cases that some histologic subtypes known as the solid variant of mammary ACC with basaloid features have a greater propensity for axillary lymph node spread, recurrence, and distant metastasis This type of ACC is considered as "high grade" tumor [1] and has been shown to even coexist with small cell carcinoma in the same breast.[9]

Distant metastatic disease has been shown to be uncommon in the conventional mammary ACC, unlike ACC in other locations, mainly in the minor salivary glands.[2]

Leeming et al [5] recorded distant metastasis in only 10 out of 140 patients, while Kleer and Oberman [10] recorded none in 20 patients that they had follow up information on.

In our patients the story was different, as there were metastasis to different sites i.e. (a) liver, lower limb, and sacrum in one patient (b) the other breast, ovary and supraclavicular node in 3 other patients (Table 1). The presence of other cancers occurring synchronously in 4 patients, and the delay until recurrent lumps appeared in 2 patients may have modified ACC in these patients, for a worse outcome.

However, because of the good prognosis noted generally in patients with ACC [2, 3, 6], a lingering and legitimate problem arises as to what treatment is best suited for this disease. It has not been determined. [4, 11] ACC has a relatively well-defined nature, with less surrounding architectural disruption and fibrosis.[7] But local excision alone, based on this nature is followed by unacceptably high rates of recurrence. [1, 4] On the other hand, the reported low incidence of axillary lymph node metastasis has been an argument against lymph node dissection. Radiotherapy has been used rarely to treat it, unlike what obtains in ACC of the head and neck, where radiotherapy is used with success.[13] Recently however, Kasagawa et al. [6] advocate breast conserving surgical procedures with axillary dissection and adjuvant radiotherapy, instead of primary mastectomy or chemotherapy. Simple mastectomy has been widely preferred by many surgeons in first world countries [3, 16] but most of their patients present without axillary nodal disease. Some of these patients depend on advances in imaging techniques like mammography, ultrasonography and MRI for detection of their disease. [4,6,14,-16] These imaging techniques, apart from ultrasonography, are unfortunately not available in most centres in this country where patients with breast cancer are treated.

Our patients, just like the 8 patients studied by Kontos [8] had either modified radical mastectomy, or simple mastectomy with radiotherapy and cytotoxic therapy, depending on the stage of the disease. Since mastectomy is known to be curative for ACC, and neither adjuvant chemotherapy nor hormonal manipulation has been conclusively studied in patients with ACC, [1] modified radical mastectomy may be the advisable option in our setting. This is because many of our cases had nodal disease; besides, some of them even had additional synchronous carcinoma. After all Kasagawa[6] who presently advocates breast conservation surgery advises additional axillary node dissection.

Finally our patients showed a mixed result in favour of poorer prognosis unlike what is described in literature. The presence of lymph nodes, distant metastasis and synchronous tumors worsened their outlook, such that only 3 patients were well 5 years after surgical treatment, when the were last seen.

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