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CASE REPORT

Infiltrating Syringomatous Adenoma of Nipple

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ABSTRACT

Infiltrating syringomatous adenoma of the nipple is a rare, benign lesion with distinct clinical and histological features. Origin from ducts of dermal sweat glands has been postulated. Important differential diagnosis include nipple adenoma, tubular carcinoma and adenosquamous carcinoma. Appropriate local management includes accurate diagnosis and complete excision to avoid local recurrences. This report describes the condition in a 39-year-old parous lady.

Key words: *Infiltrating syringomatous adenoma. Nipple adenoma. Benign nipple neoplasm.*

INTRODUCTION

Syringomatous adenoma of the nipple is a rare, distinct, benign though locally infiltrative clinical entity. Only 31 cases have been reported to-date.[1] Histologically, it is similar to syringoma, a benign neoplasm originating in the ducts of the dermal sweat glands located in the lips, axilla, buttocks or minor salivary glands.[2] Important aspects of management involve its recognition and differentiation from adenosquamous and tubular carcinoma with associated risk of local recurrence when incompletely excised.[3,4] We report the case of infiltrating syringomatous adenoma of the nipple with review of relevant literature.

CASE REPORT

A 39-year-old lady presented with 3 months history of painless lump over left nipple. Although not a regular breast self-examiner, she noticed a recent painless increase in size of left nipple without any associated nipple discharge or ulceration. She did not notice any lumps in either breast. There was no significant history of breast diseases in the past. No risk factors were identified for breast cancer except late age at first delivery.

She initially presented elsewhere where an incisional biopsy of the left nipple was done and the histopathology revealed neoplastic lesion of dermal adnexal origin. On examination, upon refferal to Aga Khan University Hospital, Karachi, the left nipple was enlarged and exuberant, but epithelialized with an indistinct retroareolar palpable mass. Right breast was normal on examination and there was no regional lymphadenopathy. Mammogram did not reveal any parenchymal abnormality and a bilateral breast ultrasound was normal. The histopathology was also reviewed at The Aga Khan University Hospital. It showed ductules, tubules and strands of small uniform cells infiltrating into the dermis. Some ducts showed a characteristic comma-shape. Nuclear pleomorphism, hyperchromasia and lack of mitosis were noted. The findings were consistent with syringomatous adenoma of the nipple.

In addition to routine haematoxylin and eosin stained slides, immunohistochemical markers for demonstration of myoepithelial layer-anti smooth muscle antigen and CD10 were performed. The presence of double layer provided additional evidence in favour of syringomatous adenoma and excluded other differential of tubular carcinoma and low-grade adenosquamous carcinoma.

She underwent wide excision of the nipple areola complex via an elliptical incision. Histopathological analysis revealed widely scattered epithelioid strands in the dermis with eccrine differentiation and papillary hyperplastic changes in the epidermis (Figure 1a, b). The findings were consistent with syringomatous adenoma. The lesion was 0.2 cm from the painted deep margin. She did not develop any recurrence at 6 months follow-up. In view of the unusual nature of this lesion, the patient was advised long-term follow-up.

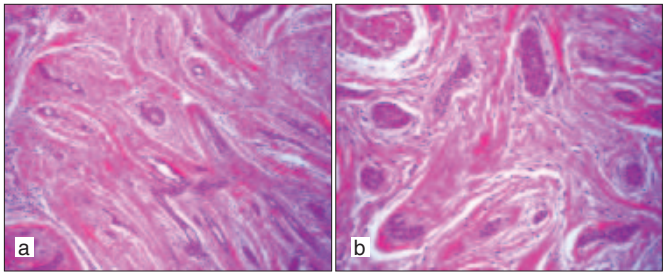

Figure 1 (a and **b):** Histopathology of syringomatous adenoma of the nipple with increasing magnification showing islands of squamous epithelium

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DISCUSSION

Breast neoplasms with squamous differentiation are relatively rare and tumours with specific syringomatous features are sparsely reported in the literature. It was described as a tumour like proliferation of small squamous islands in which no connection could be found between the squamous structures and the surrounding breast parenchyma.[5] The authors interpreted the lesions as a result of squamous metaplasia arising from pre-existing ductal structures. Clinically and pathologically similar lesions to infiltrating syringomatous adenoma but located away from the nipple in the breast parenchyma were also described by Suster *et al.*[6]

Historically termed as "adenoma", the term "syringomatous adenoma of the nipple" was first coined by Rosen in 1983.[7] In a clinical and pathological study of 11 cases in 1989, Jones *et al.* introduced the term "infiltrating" to emphasize the locally infiltrative nature of this benign but peculiar lesion.[4]

Patients with syringomatous adenoma of the nipple have been reported to range in age from 11-76 years with an average age of 40 years.[4,7] Though, there is a female preponderance, it has been reported in men presenting with nipple ulceration.[8]

The clinical presentation of infiltrating syringomatous adenoma of the nipple is variable. The common presentation is that of a solitary, painful nipple mass which may cause nipple inversion, discharge or itching.[4,7,8] Except one, all reported lesions are unilateral and none have been associated with regional lymphadenopathy. There is no specific radiological features characteristic to this unusual condition. Histological and microscopic examination reveals an infiltrative pattern of epithelial islands that are angulated or comma-shaped as well as tubular or solid in configuration. The glandular lumina are small or obliterated. Squamous metaplasia is usually present in variable proportion of epithelial islands, which have an inconspicuous or absent outer myoepithelial layer. The epithelial elements often invade into the smooth muscle of the nipple, mimicking invasive carcinoma.[7,9]

It is important to differentiate syringomatous adenoma from tubular carcinoma and low-grade adenosquamous carcinoma. The glandular structures of tubular carcinoma are mostly angulated with open lumina and are composed of single cell population as opposed to syringomatous adenoma, which has a variable amount of squamous metaplasia and has characteristic "comma" or "tad pole" shapes.[10] Ductal carcinoma *in situ* is often associated with tubular carcinoma. Low-grade adenosquamous carcinoma may be impossible to differentiate from syringomatous adenoma, especially if involving the nipple areola complex.

Surgical management has varied from excisional biopsy to radical mastectomy.[3] Recurrence has been reported from 25-55% in patients treated initially with some form of local excision to subcutaneous mastectomy.[4] Local recurrence has been attributed to be related to incomplete excision. It is evident from the literature that axillary dissection is not indicated as no nodal metastasis has been found in patients who had axillary dissection. Distant metastasis is unknown with syringomatous adenoma on follow-up of as long as 24 years.[4]

Based upon this case report and review of the literature, infiltrating syringomatous adenoma is an unusual, benign though locally infiltrative clinical entity. It should be included in the differential diagnosis of the patients who present with lump in the nipple areola complex. Though non-metastasizing, it may recur locally if incompletely excised. Hence, complete excision with clear margins seems to be an appropriate therapy and extensive surgical procedures should be avoided. Owing to the rarity of this disease, long-term follow-up is warranted.

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