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Syringomatous adenoma of the nipple: Case report

Meme başının siringomatöz adenomu: Olgu sunumu

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Abstract

Syringomatous adenoma is a rare disease of the nipple that was mostly easily confused nipple eczema or erosive nipple adenoma. Although, mammographic findings indicate carcinoma, this condition has benign, locally infiltrative characteristics. Herein, a patient with complaints of pruritus and minimal enlargement of right nipple was firstly evaluated as nipple eczema or erosive adenoma of nipple, but no response was detected with topical treatments. Patient was diagnosed with syringomatous adenoma of nipple with histopathologic examination findings that were double layered epithelial ductus proliferations which some of them had tear-drop, comma-like appearance and nipple area was excised. This rare condition is discussed with literature.

Keywords: Breast, Nipple, Syringomatous adenoma

Öz

Siringomatöz adenom, çoğunlukla meme başı ekzeması ya da meme başının eroziv adenomu ile kolayca karışabilen, meme başının nadir görülen bir hastalığıdır. Mammografik bulgular karsinomu düşündürmesine rağmen, bu tablo lokal infiltratif özellik taşımaktadır. Burada 1 yıldır sağ meme başında kaşıntı ve hafif büyüme şikayeti olan hasta öncelikle "nipple ekzema" ya da "meme başının eroziv adenomu" olarak değerlendirilmiş, ancak topikal tedavilere yanıt alınamamıştır. Hastaya, histopatolojik incelemede dermiste iki sıra epitel ile döşeli, bazıları "gözyaşı damlası - tear-drop" ve "virgül şeklinde - comma-like" görünümde duktus proliferasyonu ile meme başının siringomatöz adenomu tanısı konuldu ve meme başı eksize edildi. Bu nadir olgu literatürler eşliğinde tartışıldı.

Anahtar kelimeler: Meme, Meme başı, Siringomatöz adenom

Introduction

Syringomatous adenoma of the nipple (SAN) is a very rare benign tumor that was first reported by Rosen in 1983 [1]. SAN is originated from eccrine sweat glands and usually located over the areola area or more specifically nipple. It is histologically similar to a syringoma, a benign tumour originating in the ducts of the dermal sweat glands, and importantly needs to be distinguished from a tubular carcinoma [2]. Tumoral lesions of the nipple area accompanying bleeding, discharge or ulceration are common in female patients. Unilaterally located SAN usually shows locally infiltrating growth but does not metastasize. Nonetheless, in some cases, this tumor may be misdiagnosed as a malignancy owing to its locally aggressive infiltrating growth pattern. Best treatment option is local excision of the tumor [2.3].

Case presentation

A 38-year-old woman presented with a 1-year history of mild itching sensation on her right nipple. She has been previously given various treatments; however her symptoms did not improve. Both her past personal and family medical history was unremarkable, excluding any history of breast cancer.

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On physical examination, there was an induration over the right nipple area, together with 4 or 5 millia like white papules and desquamation. No discharge or nipple retraction was detected. She was diagnosed as having nipple eczema and started on mid-potent topical corticosteroid ointments and moisturizers. In two weeks follow-up, symptoms improved markedly. Three months later, the signs and symptoms recurred and a biopsy was planned, however the patient did not consent the procedure. Thus physical and mammography exams were performed and fibrocystic changes were detected. Seven months later, she was in 28 weeks of her pregnancy and was admitted with similar complaints together with enlargement of the lesion on her right nipple (Figure 1).

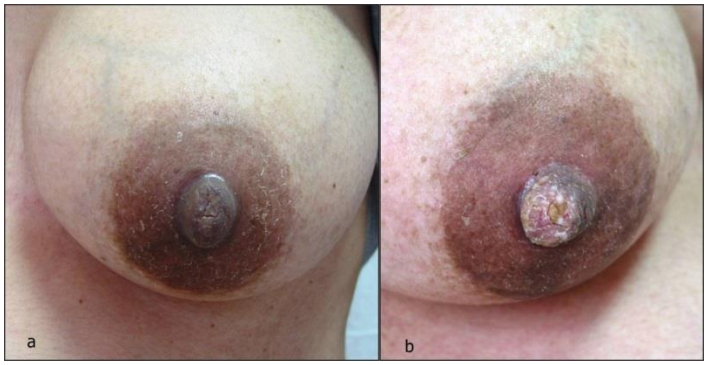

Figure 1: a: Left nipple areolar complex was nonpathological appearance, b: There was an induration over the right nipple area, together with 4 or 5 millia like white papules and desquamation.

A biopsy with a suspicion of nipple eczema, Paget disease, Bowen disease and erosive adenoma of the breast was performed. Histopathological examination showed tear-drop or comma-like ductal proliferation in dermis, lined by double-layered epithelia, consistent with SAN (Figure 2). There were no atypical mitosis or necrosis and estrogen receptors were negative. At the end of the pregnancy period, local excision of the lesion was performed. The written consent was received from the patient who was presented in this study.

Discussion

SAN is a rare, locally aggressive benign tumor of the nipple and subareolar region. These tumors commonly manifest as solitary firm masses in the subareolar or nipple region of the unilateral breast, and may also occur within the breast parenchyma [2-6].

Lesions are generally 1-3 mm in size and they may be clinically asymptomatic, tender and painful on palpation, and/or present with itching and ulceration. Nipple inversion or discharge is noted on occasion [2].

Upon mammographic imaging, SAN generally appears as a high-density mass in the subareolar region with an irregular outline, spicule formation, and microcalcification foci, while ultrasonography shows an ill-defined mass with heterogeneous internal echoes [2,7]. Imaging findings of SANs resemble those of malignant tumors, and, therefore, they may be indistinguishable from carcinoma with mammography, ultrasonography, and MRI. Fine needle biopsy examination of the lesions may not be adequate to fully understand the locally aggressive growth pattern of the lesion and thus may lead to unnecessary mastectomies or axillary dissections [2,5,6]. Incisional or excisional biopsies have been helpful for the diagnosis of 31 patients, with ages ranging between 11 and 76 [2].

In most of the cases, making the diagnosis of syringomatous adenoma clinically is difficult and pathological examination is essential. Histologically, the lesion is composed of tubules, ductules, and strands of small, uniform, generally basophilic cells infiltrating the dermis of the surrounding skin and the stroma of the nipple. Proliferating ducts, which are lined by a single or multiple layers of metaplastic squamous cells, may be present. These double-layered cell nests have a specific teardrop, comma-shaped or tad-pole appearance, and the tumor cells can infiltrate the stroma between the smooth muscle bundles, and even in the perineural region [2,3,8]. Hyperplastic changes can be detected in the breast tissue surrounding the tumoral lesion. While making the histopathological diagnosis of syringomatous adenoma; 1- tumoral invasion of the dermis and subcutaneous tissue around the nipple or areola region 2- Comma-shaped tubular proliferation infiltrating smooth muscle or neural bundles 3- Presence of myoepithelial cells around the tubules 4- Stratified squamous epithelia lining cysts, filled with keratinous material 5- Absence of mitotic activity or necrosis should be considered [2]. In our case, the dermis of the tumor was typically lined with double layered epithelia, together with tear-drop and comma-shaped ductal proliferation. Absence of mitosis and necrosis also supported the diagnosis.

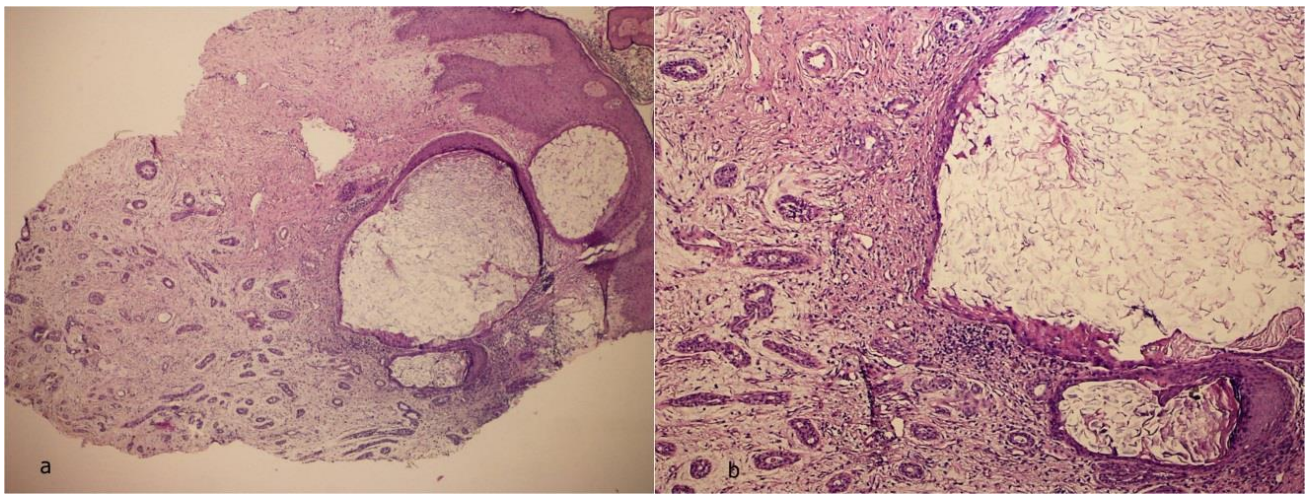

Figure 2: Histopathological examination showed tear-drop or comma-like ductal proliferation in dermis, lined by double-layered epithelia, consistent with syringomatous adenoma of the nipple (H&E x40, x100)

Nipple duct adenoma and well-differentiated tubular carcinoma or low-grade adenosquamous carcinoma are often confused with SAN. These entities, however, exhibit some distinctive characteristics. Nipple duct adenoma often ulcerates the skin, is usually better circumscribed, and does not invade the perineural region or smooth muscle bundles. It consists of epithelial hyperplasia of the lactiferous ducts and does not show stromal invasion, unlike syringomatous adenoma.

Tubular carcinoma tends to occur deeper in the breast with positive estrogen receptors and is commonly located in the upper outer quadrant or away from the nipple. On the other hand, low-grade adenosquamous carcinoma tends to metastasize with both glandular and squamous differentiation. Syringoma, a benign tumor of the eccrine glands, also exhibits clinical features that distinguish it from SAN, including presentation as a solitary lesion in the nipple. Itching and ulceration of the nipple could also be observed in case of Paget's disease. However, histopathological examination of these lesions reveals large and clear cells, also known as Paget cells, with wide, oval shaped nucleus and edematous cytoplasm. Histopathological examination is the gold standard method for the differentiation of all these entities [2-6,8,9].

SAN has a tendency to show local recurrence when resected incompletely. Thus optimal initial management demands complete resection with histologically negative margins [2,3]. There was no recurrence of the lesions with histologically negative margins, for 1-6 years of follow-up periods [2]. There have been no reports of SAN with distant metastasis, so mastectomy or axillary dissection is not routinely performed. For better cosmetic outcomes, tumor resection with nipple preservation should be considered [2,3,10].

In the literature, there have been 3 cases of SAN during pregnancy and lactation, one case during lactation period and 4 cases as soon as the lactation period finishes [8,10-12]. These findings may suggest the possible relation of the tumor and maternal hormonal effects [10]. Increase in the size of the tumor during pregnancy in our case can also support this hormonal influence. In routine practice, local excision of the tumor is the treatment of choice. However, there is not a consensus in the literature about the approach during pregnancy, thus local nipple excision after pregnancy period was preferred in this case.

In conclusion, every chronic lesion of the nipple or subareolar region deserves to be examined histopathologically. Based on the features of this case and a review of the literature, infiltrating SA of the nipple is a rare benign but locally invasive tumor which importantly needs to be distinguished from malignant tumors. Appropriate management involves a consideration of the tendency to recur locally when incompletely excised. Instead of radical approaches, optimal surgical management involves complete excision of the lesion and confirmation of histologically negative margins.

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