Case Report

Recurrent Syringomatous Adenoma of the Nipple Following a Misdiagnosis: A Case Report

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Abstract

Infiltrating syringomatous adenoma of the nipple (SAN) is a rare benign neoplasm of the breast that is often misdiagnosed. SAN may present with a subareolar lesion and clinical, mammographic, and ultrasonographic findings associated with malignancy. We present the case of a 60-year-old woman with a painful, firm, solid tumor in her left breast and deformation of the left nipple-areolar complex (NAC). Histopathological test results were conflicting. The tumor, including the NAC were locally excised. Postsurgical immuno-histochemical tests revealed squamous histology, whereas myoepithelial cells were present in the resected specimen, a feature consistent with SAN. The pathologist noted microscopically positive surgical margins. Three months after surgery, tumor recurrence occurred. The patient underwent revision surgery with wide excision of the skin and gland around the lesion, followed by immediate breast reconstruction, using a pedicled myocutaneous latissimus dorsi (pLD) flap. Extreme care should be taken when diagnosing SAN to ensure proper treatment and prevent recurrence.

Keywords: *Adenoma, breast, latissimus dorsi muscle flap, syringomatous*

Introduction

Syringomatous adenoma of the nipple (SAN) is an extremely rare benign tumor of sweat duct origin that was first described by Rosen *et al*. in 1983.[1] SAN shows locally infiltrative expansile proliferation but no metastasis.[2] As its infiltrative appearance is comparable to that of invasive carcinoma, SAN might be misdiagnosed and over treated.[1] Previously documented surgical treatment includes local excision and mastectomy with axillary lymphadenectomy.[3]

We present a case of SAN recurrence following local resection due to incomplete tumor excision. Breast reconstruction with a pedicled myocutaneous latissimus dorsi (pLD) flap, the first documented method of treatment for this disease was performed.

Case Report

A 60-year-old woman presented with a painful, firm, and solid tumor in her left breast (5 × 5 × 4 cm) and deformation of the left nipple-areolar complex (NAC) [Figure 1]. She had first noticed the nodule more than 20 years prior. At her first visit in 1997, no abnormality in her right breast or regional lymphadenopathy was detected. Physical examination and bilateral breast ultrasonography revealed no parenchymal abnormality and confirmed that the tumor was benign. Mammograms obtained 3 years later were unremarkable. Seven years later, the patient consulted a breast surgeon following an increase in nodule size. Fine-needle aspiration cytology revealed fibroadenoma and glandular tissue hyperplasia with uncertain malignant potential. The patient refused the proposed surgery and adjuvant therapy and sought a second opinion, which involved needle core biopsy. At that time, histological analysis revealed hamartoma. The patient declined surgical treatment. Nine years later, she was referred to our clinic because of an increase in tumor size, and progressive breast deformation. Incisional breast

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biopsy revealed a tumor of unknown origin; however, its features suggested hamartoma with fibromatosis. Because of the possibility of malignancy, we locally excised the tumor, including the NAC. Postsurgical examination revealed p63 [Figure 2] and cytokeratin 5/6 (CK5/6) positivity [Figure 3], suggesting a squamous histology. In addition, myoepithelial cells were present in the resected specimen. On the basis of these findings, the tumor was diagnosed as SAN. The pathologist noted microscopically positive surgical margins associated with a very high recurrence risk. Three months after the surgery, the patient presented with tumor recurrence. On physical examination, a firm, 2-cm tumor was found in the middle portion of the postoperative scar, alongside a deformation of the inferior pole of the breast [Figure 4]. We performed revision surgery with wide excision of the skin and gland around the lesion and immediate breast reconstruction using a pLD flap. The postoperative

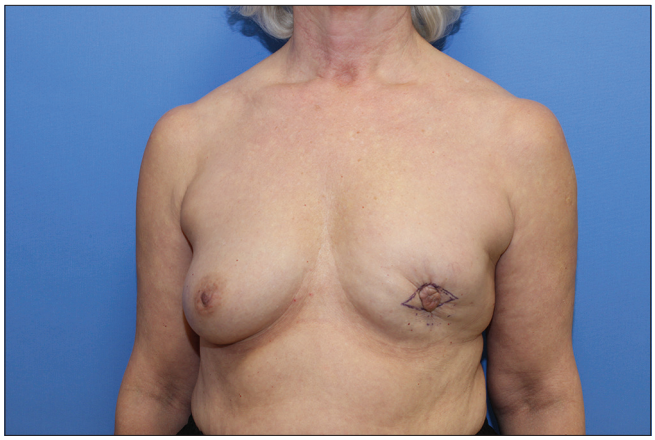

Figure 1: Preoperative view showing the markings for the nipple-areolar complex excision

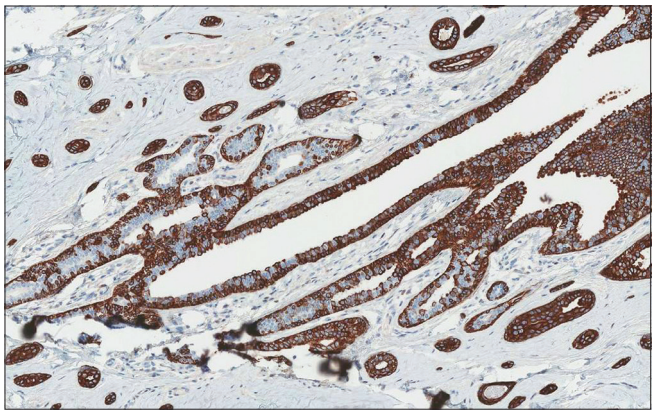

Figure 3: Immunohistochemical analysis of the resected specimen (cytokeratin 5 staining) (x 100 magnification)

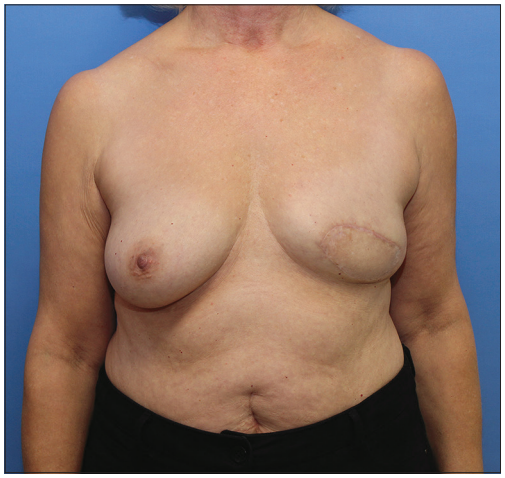

Figure 5: Results at 1-year follow-up

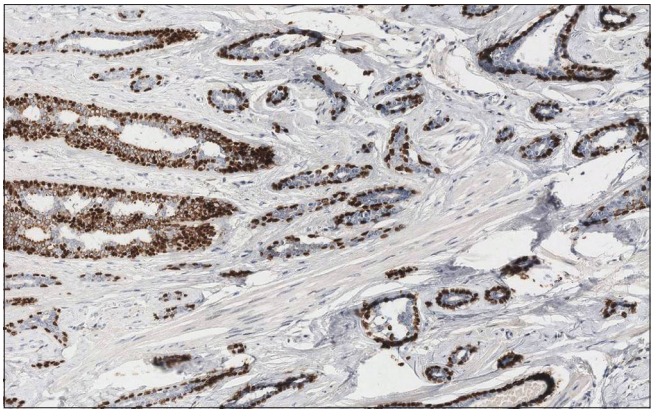

Figure 2: Immunohistochemical analysis of the resected specimen (p63 staining) (x 100 magnification)

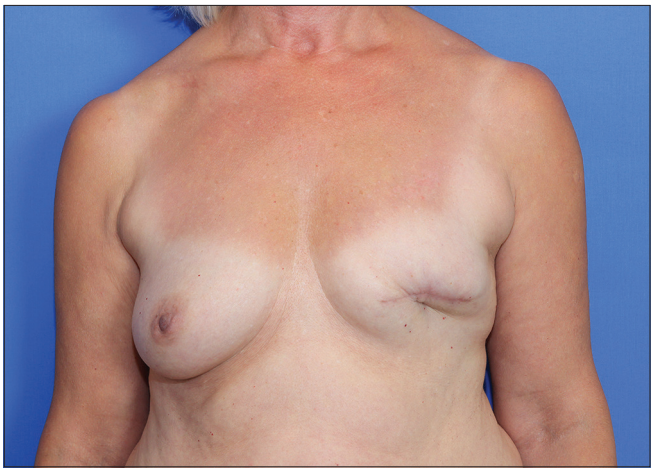

Figure 4: Recurrence of syringomatous adenoma of the nipple

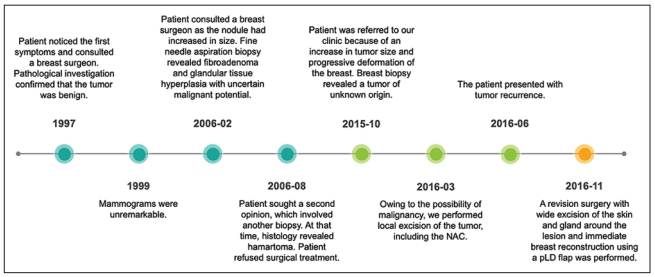

Figure 6: Timeline of case development

period was uneventful. Histopathological examination of the excised mass revealed margins free from neoplastic involvement. One year post surgery, the patient showed no recurrence [Figure 5]. Figure 6 shows the case report timeline.

Discussion

SAN is defined as a locally recurrent and invasive tumor of the nipple and areolar region of the breast that originates from the sweat gland ducts.[4,5] To our knowledge, less than 45 cases have been reported to date, including 2 in men.[2,6] The average age at presentation is 40 years (range, 11–87 years).[2] Only 2 of these previously reported cases were bilateral, and none were associated with regional lymphadenopathy.[7] The tumor size in previous case reports varied from 0.5 to 4.5 cm.[7] Therefore, our case appears to be the largest tumor reported in the literature to date.

SAN typically presents as a solitary, firm mass in the subareolar or nipple region of the unilateral breast.[8] Pain, tenderness, itching, crusting, nipple inversion, or discharge may occur.[8] Ulceration and erosion have been reported in a previous case report.[9] Our patient presented with a painful, firm mass in her breast.

Because of its rarity and similar presentation to that of other pathological processes, SAN can easily be misdiagnosed.[5] Histopathologically, differential diagnoses of SAN include florid papillomatosis, adenosquamous carcinoma, adenoid cystic carcinoma, tubular carcinoma, and sclerosing syringomatous carcinoma. Moreover, it has no typical radiologic or ultrasonographic features.[7] Its classical histological characteristics include two cell layers often resembling a teardrop or comma.[7] It may present as a disorganized assortment of curved, angulated, and compressed cordlike tubes and ducts with a clear layer of myoepithelial cells between the epithelial cells and fibrous stroma.[3]

In our case, SAN was initially misdiagnosed as fibroadenoma and later as hamartoma with fibromatosis. In one of the previously reported cases, an early biopsy also revealed fibroadenoma.[5]

Immunohistochemical analysis is a valuable tool in the diagnosis of SAN.[7] In our case, many cells were positive for two main markers, p63 and CK5/6, indicative of squamous histology, and there were myoepithelial cells in the resected specimen, a feature consistent with SAN.[7]

Reported treatment options include various surgical techniques.[9] To our knowledge, there are no previous reports of wide excision with immediate breast reconstruction using a pLD flap.

The aim of surgical management is complete excision of the lesion and the confirmation of histologically negative margins. This frequently requires excision of the NAC. If the tumor is not excised completely, SAN is likely to recur. A study of 11 cases revealed a 45% recurrence rate after local excision, occurring 1.5 months to 4 years after the initial surgery.[10] To date, there have been 6 reported cases of SAN recurrence associated with incomplete excision.[5]

Here, we report a case of a 60-year-old woman with recurrent SAN. Multiple biopsies over the course of tumor progression yielded different results, which complicated the diagnostic process and led to treatment with an inappropriate surgical method, i.e. local excision, including the NAC. The correct diagnosis, by immunohistochemical analysis, was made only after postoperative biopsy of the excised tumor. Three months after surgery, the patient experienced tumor recurrence. Revision surgery involved wide excision of the skin and gland around the lesion, with immediate breast reconstruction using the pLD flap. Although SAN is a benign tumor that does not metastasize, it is crucial to perform thorough diagnostic testing, including immunohistochemical analysis, to correctly diagnose SAN and avoid tumor recurrence. Wide excision with negative surgical margins appears to be the gold standard treatment for SAN. Reported immediate breast reconstruction using the pLD flap appears to be well suited for recurrent tumors.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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