CASE REPORT
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Syringocystadenoma papilliferum located at the nipple: Description of an extremely rare case with review of the literature

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ABSTRACT

Syringocystadenoma papilliferum (SCAP) is a rare, benign tumor of the apocrine sweat glands, and only two nipple-located SCAP cases have been reported. Very few cases of malignant transformation and metastasis have been reported. We share our experience with SCAP located at the nipple that recurred with intraductal papilloma (IP). A female patient aged 26 years presented to our clinic with a mass at the posterior of the left nipple. The mass was excised, and the pathology report revealed SCAP. The patient had no recurrence for 1 year, but the mass recurred later in the same location. Re-excision was planned and conducted. Diagnosis of the second excised mass according to the pathology report was florid-type ductal epithelial hyperplasia and IP. SCAP may be located in female genitals, extremities, and trunk but these are rarer than in the head and neck. This is the third case reporting SCAP at the nipple. SCAP may be related to nevus sebaceous, resulting in basal cell carcinoma or syrigocystadenocarcinoma papilliferum; however, no data have been reported about the relation of SCAP with IP. The relation may be due to microscopic characteristics of SCAP, including the presence of papillary processes between two epithelial alignments. As a conclusion of this case presentation, SCAP of the nipple must be followed up for IP transformation or recurrence. Further evaluation may be needed for this dark side of the rare and little-known pathological entity; however, because of its rareness, it seems troublesome to diagnose.

Keywords: Intraductal papilloma, syrigocystadenocarcinoma papilliferum, syringocystadenoma papilliferum

INTRODUCTION

Syringocystadenoma papilliferum (SCAP) is a rare benign tumor of the apocrine sweat glands and is usually located in the head and neck region (75% of the cases) which commonly arises at the second decade of life. More frequently, SCAP is a congenital lesion. It was first described by J. H. Stokes in 1917 (1). Breast-localized (especially nipple-localized) SCAP is extremely rare. SCAP may be classified at three forms including plaque, nodular or linear; however, there is no consensus about the classification because of the rarity of the cases (2). SCAP is characteristically described macroscopically as erythematous symmetrical lesions. It may be misdiagnosed with many lesions but more frequent with basal cell carcinoma macroscopically and intraductal papilloma (IP) microscopically. Treatment of both lesions is excision, and excisional biopsy is the best technique for diagnosing the lesion as either SCAP or IP.

This study aimed to report the clinical presentation of a female patient with SCAP of the nipple recurring with intraductal papilloma (IP), whose microscopic features are similar and create a dilemma for the pathologists in accordance with the literature.

CASE REPORT

A white female patient aged 26 years presented to our clinic with an exophytic growing mass for 3 months, located just to the right of her left nipple. The mass measured approximately 0.5 cm in diameter with palpation; it was mobile, and there was no ulceration on the lesion. Patient's laboratory tests were totally in normal range. The patient underwent excisional biopsy under local anesthesia. Final pathology of the specimen revealed SCAP with benign papillary formations.

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One year after excision, the patient presented to our clinic with recurrence of the mass in the same location, this time more deeply located and less exophytic. Physical examination showed a mobile subcutaneous mass 0.9 cm in width, without any cutaneous alternations. Ultrasound revealed a hypoechoic mass measuring 0.9 x 0.8 cm with increased vascularity. Local excision of the mass was planned and conducted. Final pathology of the second excisional biopsy revealed IP, florid hyperplasia, and fibrocystic alternations (fibrosis, periductal inflammation, apocrine metaplasia, macro-microcysts) (Figures 1-3). The patient was discharged and has been followed up for approximately a year without recurrence.

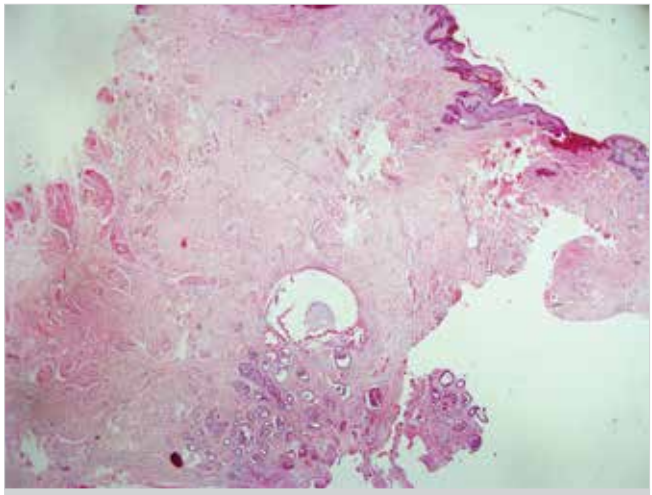

Figure 1. There are cystically dilated lactiferous ducts showing florid type intraductal proliferation. There is also another dilated duct with cystic apocrine metaplasia adjacent to the ducts with usual duct hyperplasia. (H&Ex20).

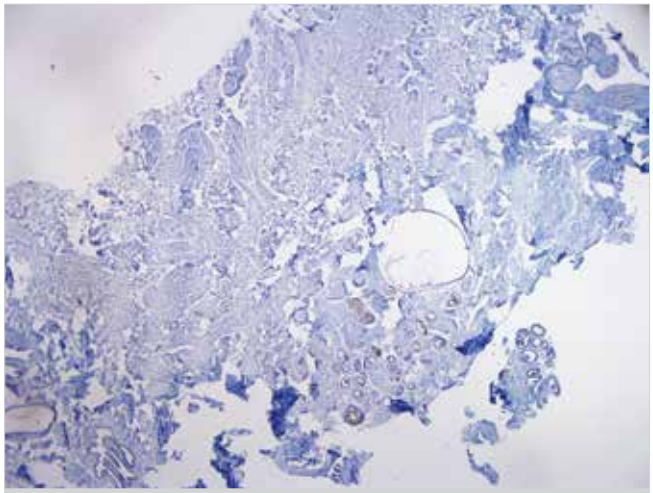

Figure 2. Focal weak ER positivity within the intraductal proliferation (ERx20).

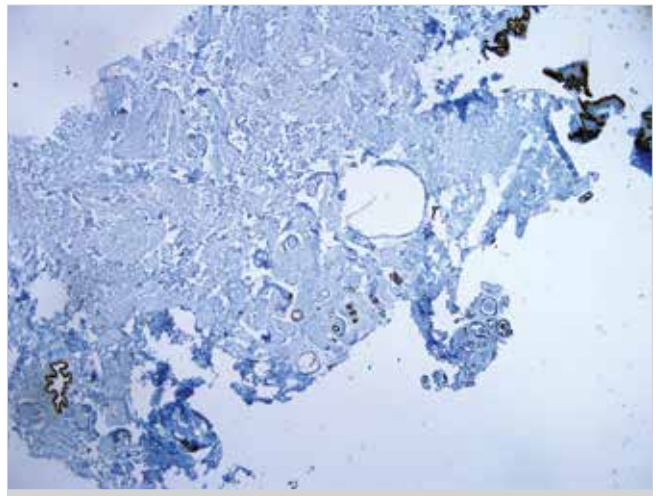

Figure 3. Patchy and focal staining with CK5/6. (CK5/6x20).

DISCUSSION

SCAP is described as a rare dermatological benign lesion. The most common localization for SCAP is the head and neck. Other locations are rare and only one case of SCAP localized at the external auditory canal has been reported (3). Only 2 other nipple-located SCAP cases have been reported, to the best of our knowledge (4,5). Although SCAP is a benign lesion, malignant metastatic lesions, known as syringocystadenocarcinoma papilliferum (SCACP), have also been described (3,6). SCACP are mild malignant tumors; only one case has been described for lymphovascular invasion, and very few cases for metastasis (6). Predisposing factors and progression of SCAP and transformation to SCACP are still uncertain. Much work has been done and debates about the malignant transformation of SCAP are ongoing. Parekh et al. have shown that SCACP lesions resulting from SCAP are related to nevus sebaceous of Jadassohn's (NSJ), in agreement with various other studies (6-8). However, SCAP is a rare entity that arises from NSJ. Kamyab-Hesari et al. have reported the rate of SCAP formation after NSJ to be 1.19%, and Hsu et al. have reported it as 2.7% (7,8). Since not all NSJ transforms to SCAP, not all SCAP lesions arise from NSJ, as was the case in our patient. Ayadi et al. have described tubuloapocrine adenoma associated with SCAP, but it is hard to identify which lesion was the precursor of the other or whether they were independent from each other (9). Sporadic SCAP lesions are also described, as in our present case.

Currently, SCAP lesions have no clinical importance except their cosmetic results. However, malignant transformation and malignancy potential for SCACP or basal cell carcinomas are being newly debated, as mentioned above. Shen et al. and Levinsohn et al. have described BRAF and RAS mutations at sporadic SCAP lesions, but none at SCAP lesions that transformed from NSJ. The study has concluded that the Ras-MAPK pathway is active only

for sporadic SCAP lesions (10,11). BRAF mutation is described for many benign and malignant (especially aggressive) human tumors; malignant melanoma is the most frequent malignant tumor having a BRAF mutation and must be emphasized. Unfortunately, because SCACP cases are very rare, the immunohistochemical and mutational properties of these cases are unknown. Thus, debates on the immunohistochemical and mutational properties of SCAP are ongoing (3,10).

The entity of SCAP being rare probably results in the rarity of SCACP. Almost no published reports exist for the management of these cases. Another reason for the rarity of publications on the management of SCAP is that diagnosis of SCAP is only made with a microscope; very few physicians are experienced enough to diagnose it with its characteristic macroscopic features. For the reasons mentioned above, we suggest that SCAP lesions must be completely excised because of the unknown potential for malignant transformation.

Microscopic features of SCAP include glandular proliferation, fibromatous - inflammatory reaction, and papillary formations lined by double-layer epithelium (3,6). Differential diagnosis for SCAP localized at the nipple is reported as IP and nipple adenoma (3). Because of the characteristic microscopic features, SCAP is a diagnostic dilemma for pathologists, as in our case. Our patient had a diagnosis of SCAP after her first excisional biopsy. The recurrent specimen excised from the same location was diagnosed as IP with other benign features because of a lack of papillary formation lined by double-layer epithelium. There are no strict rules for the diagnosis of SCAP, so we decided not to diagnose SCAP without one of its microscopic characteristics, even though other findings refer to SCAP. We think pathologists must not be too eager to diagnose SCAP to chase a rare case and must evaluate cases deeply for the diagnostic dilemma debated above.

The currently presented case may be the third case for SCAP localized at the nipple but the first case for SCAP recurrence with IP. Since there is a close relation between NSJ and SCAP, we want to draw attention to the relation between SCAP and IP because of their similar microscopic features. Unfortunately, cases are too rare for this topic to be debated.

CONCLUSION

In conclusion, our case is the third case report for nipple-originated SCAP but the first case report of SCAP recurring with IP. The etiology of SCAP and malign transformation is debated, and for this reason we suggest local resection for suspicious lesions. Finally, pathologists must keep in mind the entity of SCAP but must not misdiagnose with the differential diagnosis of SCAP.

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OLGU SUNUMU-ÖZET
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Meme başının syringocystadenoma papilliferum lezyonu: Literatür ışığında nadir bir olgunun sunumu

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ÖZET

Syringocystadenoma papilliferum (SCAP), apokrin ter bezlerinin nadir görülen benign bir tümörüdür ve bugüne kadar sadece iki tane meme başı yerleşimli SCAP olgusu bildirilmiştir. Çok az sayıda malign transformasyon ve metastaz bildirilmiştir. Bu olgu sunumunda; intraduktal papilloma (İP) ile tekrarlayan ve meme başı yerleşimli SCAP ile alakalı deneyimlerimizi paylaşmaktayız. Yirmi altı yaşında kadın hasta kliniğimize sol meme başının arkasında kitle ile başvurdu. Kitle eksize edildi ve patoloji raporu SCAP olarak raporlandı. Hastanın bir yıl boyunca nüksü olmadı fakat kitle daha sonrasında aynı lokalizasyonda tekrarladı. Tekrar eksizyon planlandı ve yapıldı. Patoloji raporuna göre ikinci eksize edilen kitlenin tanısı florid tip duktal epitelyal hiperplazi ve İP idi. SCAP, kadınlarda genital bölgede, ekstremitelerde ve gövdede bulunabilir, ancak bunlar baş ve boyundan daha nadirdir. Olgu sunumumuz, meme başında SCAP bildirilen üçüncü olgudur. SCAP nevus sebaseus ile ilişkili olabilir, bazal hücreli karsinom veya srigosistadenokarsinoma papilliferum ile sonuçlanabilir; ancak SCAP'ın İP ile ilişkisi hakkında herhangi bir veri bulunmamaktadır. İki lezyon arasındaki ilişki, iki epitelyal taban arasındaki papiller proçeslerin varlığı da dahil olmak üzere SCAP'ın mikroskopik özelliklerine bağlı olabilir. Bu olgu sunumu sonucunda, meme başının SCAP lezyonu, İP'e dönüşümü veya İP ile tekrarlaması açısından izlenmesini önermekteyiz. Nadir ve az bilinen bu patolojik antitenin karanlık yüzünü aydınlatabilmek için ileri değerlendirmeler gerekebilir; ancak olgu azlığı nedeniyle bunun oldukça zor olduğu değerlendirilmektedir.

Anahtar Kelimeler: İntraduktal papillom, syringokistadenokarsinoma papilliferum, syringokistadenoma papilliferum

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