Case Report

Syringocystadenoma Papilliferum Associated with Naves Sebaceous of Jadassohn and Squamous Cell Carcinoma

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Abstract

Syringocystadenoma papilliferum (SCAP) is a rare skin adnexal neoplasm of apocrine gland. One-third of SCAP cases are associated with nevus of Jadassohn. Its transition to squamous cell carcinoma is a rare entity. A strong clinical acumen, prompt excision, and confirmation by histopathology underline treatment of this cutaneous tumor. We report a case of this rare association in the deltoid region of a 55-year-old female.

Keywords: Nevus sebaceous, squamous cell carcinoma, syringocystadenoma papilliferum

Introduction

Josef Jadassohn in 1895 first described Nevus sebaceus.[1] It is a congenital hamartomatous lesion with an epithelial and adnexal origin present in approximately 0.3% of newborns.[2] It is also known to be associated with syndromes such as linear nevus sebaceous syndrome, or Schimmelpenning syndrome. In subsequent stages of development, several neoplasms and hamartomas may develop secondarily within nevus sebaceus; the most common being the syringocystadenoma papilliferum (SCAP), a sweat gland tumor or hamartoma.[1-3] It may be difficult to distinguish these hamartomas clinically, and a biopsy is usually required for confirmation. It is a rare congenital abnormality that can lead to a malignant transformation in rare circumstances. The cumulative incidence of benign and malignant tumors is 6.1% and 0.5%, most commonly trichoblastoma and basal-cell carcinoma, respectively.[4]

SCAP is an exceedingly rare cutaneous adnexal neoplasm, which is typically located in the head and neck, and perianal area. Very few cases have been reported outside the classical sites of involvement. Only 5% of cases are reported to occur in extremities adding to the rarity of this case.[5] SCAP is known to be associated with nevus of Jadassohn (NSJ) in one-third of cases. Here, we report a case unusual co-existence of syringocystadenomapapilliferum and squamous cell carcinoma (SCC) developing secondarily in a preexisting naevus sebaceus of the left deltoid region, an unusual site for NSJ. However, the sequence of these histopathological entities could not be ascertained definitively due to the absence of any prior medical consultation by the patient. Thus, we report this rare case in which histopathological examination showing SCC component surrounded by areas of NSJ and SCAP and clinical history supports that the malignancy arouse from the transformation of SCAP associated NSJ.

Case Report

A 55-year-female presented to the Department of Surgery with an ulcerative proliferative growth in the left deltoid region. The lesion started as a plaque in childhood, which gradually progressed to become a warty lesion. Since the past 1 year, patient noticed a rapid increase in size with bleeding from the lesion following the trivial insult. Examination revealed a noduloulcerative growth, measuring 6 cm × 3.5 cm × 2.5 cm over the left deltoid region; it bled on manipulation surrounded by small warty lesions and partly excoriated skin due to continued irritation [Figure 1]. There was no regional lymphadenopathy. Rest of the physical examination and radiological evaluation was insignificant. A provisional diagnosis of SCAP associated with NSJ was made. The patient was taken up for surgical excision of the lesion. Histopathological examination revealed SCC component surrounded by areas of NSJ and SCAP.

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diagnosis of SCC was made. Excision biopsy of the lesion was carried out for histopathological evaluation, the specimen was processed, and hematoxylin and eosin stained slides were examined which showed areas of Nevus sebaceous of Jadassohn, SCAP and SCC. Sections showed several primitive hair germ-like proliferations arising from the epidermis with some peripheral palisading in few sebaceous glands consistent with a diagnosis of Nevus sebaceous of Jadassohn [Figure 2].

The epidermis revealed papillomatosis, with mild acanthosis and with cystic invaginations into the dermis, many papillae were lined by two rows of cuboidal to columnar epithelial cells, with oval nuclei and pale eosinophilic cytoplasm. Few cells showed a decapitation secretion. The stroma contained a dense mononuclear cell infiltrate, comprised predominantly of lymphocytes, plasma cells, and some congested blood vessels. The histopathological features were diagnostic of SCAP [Figure 3].

Deeper to areas showing features of NSJ and SCAP, tumor cells were seen. Tumor cells exhibited dyskeratosis and moderate degree of pleomorphism and occasional mitotic activity. The cells were large with abundant eosinophilic cytoplasm, and intercellular bridges and keratin pearls were often readily identified consistent with a diagnosis of SCC [Figure 4]. Thus, a diagnosis of consistent with nevus sebaceous of Jadassohn, SCAP and SCC was made. On follow-up, the patient has since remained asymptomatic with an acceptable cosmetic recovery.

**Discussion**

Nevus sebaceous has the potential to generate different lineages of tumors that are not restricted to the sebaceous differentiation. Nevus sebaceous of Jadassohn is a congenital organoid nevus appearing as a yellowish patch at birth and tend to become raised, papillomatous or verrucous at puberty under the influence of androgens. It evolves over the years going...
through infantile, adolescent and adult stage. Infantile stage presents as a patch of alopecia; during adolescence, under the influence of androgens, the lesion increases in size to become a verrucous hyperkeratotic, hyperpigmented plaque and in the adult stage as large malformed sebaceous glands, ectopic apocrine glands, and prominent epidermal hyperplasia.\(^5\) Malignant change is manifested by a rapid increase in size, the appearance of new lesions, bleeding, and appearance of metastatic lymph nodes.\(^5\)

Kaddu et al.\(^7\) in their study of 316 cases neoplasm arising in Nevus sebaceus of Jadassohn found 7.6% of benign and two cases of malignant cases all occurring in adulthood. Munoz-Perez et al.\(^5\) in their series of 226 cases found 18% of benign tumors in subjects all above the age of 14 years. Zhang et al.\(^8\) and Malhotra et al.\(^9\) reported SCAP with the transition to areas of squamous differentiation.

Both NSJ and SCAP are known to occur predominantly in scalp region followed by face and neck. Trunk and extremities are extremely rare sites with only 5% incidence, and shoulder is one of the rarest reported sites for this lesion.\(^1\) NSJ is known to transform into various benign and malignant neoplasm. Among benign lesions trichoblastoma, SCAP and trichoepithelioma are most common with basal cell and SCC is common malignant lesions.\(^2\) Almost similar transformations are known to occur in SCAP. Malignant transformation of this lesion is possible, usually to basal cell carcinoma in 10% of cases, uncommonly to SCC and rarely to its malignant counterpart: SCAP.\(^2,7,8,12-15\) Clinical diagnosis of SCAP is mostly not feasible due to the various presentations and possible differential diagnosis thus histopathology is the best option. Knowledge of this diagnosis would add a supplementary alternative to differential diagnosis. Treatment for such cases is surgical excision. SCAP should be excised given its future malignant transformation in adults. Thus, excision sufficiently eliminates this risk as well allows cosmetic relief for the patient.\(^7,12\)

The SCC transition from NSJ and SCAP has been mentioned in literature, and the same may be considered in our case. However, definite exclusion of collision tumor is not possible as the patient had no prior medical record for this lesion. To the best of our understanding of patients’ history, NSJ was associated with SCAP which then transformed into malignancy.

### Conclusion

NSJ, a hamartomatous childhood lesion is associated with varied skin lesions most commonly SCAP. Natural history of SCAP is yet to be determined. Thus prevention is not yet an option. Excision of the lesion is the mainstay of treatment. Since most patients describe a long-standing mass with recent change, supporting the idea that SCC arises from malignant transformation of preexisting SCAP, a commonly associated tumor of NSJ. Surgical excision of the lesion is sufficient and systemic therapy is usually not required. Due to the possibility of various presentations of SCAP, histopathological evaluation can prevent misdiagnoses and overtreatment.\(^15\) Hence, once NSJ becomes associated with SCAP, excision should be undertaken considering its pluripotency, appropriate clinical suspicion and histological confirmation of lesion are necessary by treating dermatologist/clinician.

### 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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### Conflicts of interest

There are no conflicts of interest.

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