

Malignant Neoplasm Arising in Nevus Sebaceous : A Rarity - two Case Reports

KEYWORDS

Nevus sebaceous, basal cell carcinoma, squamous cell carcinoma

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ABSTRACT Sebaceous nevus is an epidermal appendageal tumour, histologically composed of acanthosis, papillomatosis, and a hamartomatous conglomerate of large sebaceous glands. Defective hair structures consisting of dilated, keratin filled infundibula showing multiple buds of undifferentiated cells are a prominent feature. Sebaceous nevi are known to be associated with secondary benign adnexal tumours such as trichoblastoma, syringocystadenomapapilliferum, and eccrineporoma, but their association with malignant cutaneous neoplasms such as basal cell carcinoma and especially squamous cell carcinoma is rare. Here we report two such cases.

INTRODUCTION:

Fine needle Nevus sebaceous of Jadassohn is a complex hamartoma involving pilosebaceous unit, epidermis and other adnexal structures. It usually involves the scalp or the face. It was in 1895, that Jadassohn first described nevus sebaceous, as a circumscribed hamartomatous lesion predominantly composed of sebaceous glands. However, Pinkus designated this disease organoid nevus, as the changes are confined not only to the sebaceous glands but also involve proliferative changes of hair follicles and sweat glands.²

The morphologic appearances of this lesion vary with the age of the lesion. Hence, there is a significant likelihood of missing the diagnosis if only the typical findings of a well developed nevus sebaceous are looked for.²A good variety of appendageal tumours, may develop within sebaceous nevi. The most commonly reported among these are syringocystadenomapapilliferum and trichoblastoma.³Malignant cutaneous neoplasms are less commonly seen and include basal cell carcinoma and, to a lesser extent, squamous cell carcinoma, trichilemmal carcinoma, sebaceous carcinoma, porocarcinoma, and apocrine carcinoma.²

These two case reports highlight the potential of nevus sebaceous being complicated by malignant tumours.

CASE REPORTS:

We present two cases of malignant neoplasms arising in Nevus Sebaceous :

Case 1:

A 72 year old male presented with ulcerative lesion over the nose for 4 months. Histopathological examination of excised specimen revealed increased number of sebaceous glands, strands and islands of basaloid cells showing peripheral palisading along with cribriform spaces. Tubular and glandular structures were seen formed by the tumour cells. Some of the islands show peripheral palisading with pigmentation. A diagnosis of nevus sebaceous with adenoid basal cell carcinoma was made.

Case 2

A 50 year old male presented with non healing ulcer over left cheek for 8 months. Histopathological examination of excised tissue revealed ulcerated epithelium with nests of atypical squamous cells infiltrating the underlying stroma. Dermis showed increased number of sebaceous glands with immature hair follicles and chronic inflammatory cell infiltrate. A diagnosis of nevus sebaceous with squamous cell carcinoma was made.

DISCUSSION:

Nevus sebaceous presents 0.05–1% of outpatients who consult dermatologists.⁴ Nevus sebaceous occurs with equal frequency in males and females. These lesions are generally sporadic, but in cases of inherited nevus sebaceous, lesions are thought to develop as a result of para dominant inheritance.⁴

Various appendage tumours are known to frequently develop in nevus sebaceous. But these are mostly benign. Less than 1% of these lesions are complicated by malignant tumours according to previous studies. The commonest benign tumours are trichoblastoma and syringocystadenomapapilliferum. Amongst malignant tumours, basal cell carcinoma has an incidence of 10%. Its frequency in reported series varies, primarily due to the differences in interpretation of what really represents basal cell carcinoma. In one study, basal cell carcinomas were found in 0.8% of cases and benign tumours in 13.6% of cases.

Basal cell carcinoma cases, especially the nodular type, should be histopathologically differentiated from nodular hidradenoma, eccrinespiradenoma and trichoepithelioma or trichoblastoma like benign adnexal proliferations. In few cases, trichoblastoma arising in nevus sebaceous can show remarkable histopathologic overlap with BCC or are even found in combination with other neoplasms, such assyringocystadenomapapilliferum, desmoplastictrichilemmoma andsebaceoma. In our case, strands and islands of basaloid cells showing classical peripheral palisading along with cribriform spaces were seen, along with increased number

of sebaceous glands (Fig 1 and 2).

Squamous cell carcinoma developing within nevus sebaceous is extremely rare. Verrucous hyperplasia of the epidermis is well documented in nevus sebaceous. This could potentially mimic a well-differentiated squamous cell carcinoma. In our case, there was seen ulcerated epithelium with nests of atypical squamous cells infiltrating the underlying stroma (Fig 4). Dermis showed increased number of sebaceous glands with immature hair follicles and chronic inflammatory cell infiltrate (Fig 3). Keratin Pearl formation was also seen (Fig 5).

CONCLUSION: These cases have been presented in view of their rarity as well as prognostic implications. Association of nevus sebaceous with basal cell carcinoma is uncommon, and with squamous cell carcinoma, extremely rare. Nonetheless, though malignancy is uncommon, a cautious histologic analysis is mandated in sebaceous nevi as secondary malignancies associated with them affect the prognosis and outcome of disease.

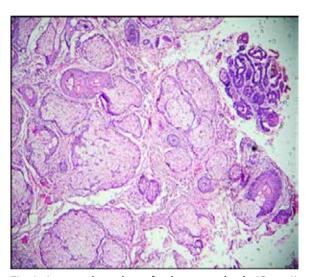


Fig 1: Increased number of sebaceous glands (Case 1) H&E (40x)

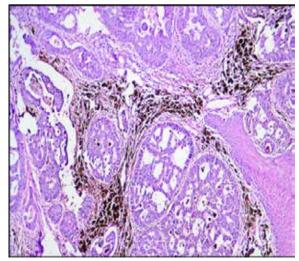


Fig 2: Islands of basaloid cells showing peripheral palisading with cribriform spaces and pigmentation (Case 1) H&E (100x)

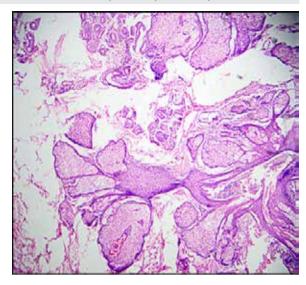


Fig 3: Increased number of sebaceous glands (Case 2) H&E (40x)

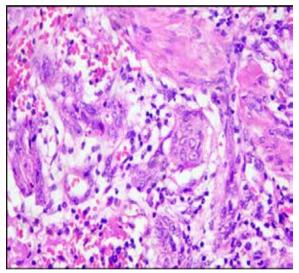


Fig 4: Nests of atypical squamous cells (Case 2) H&E (400x)

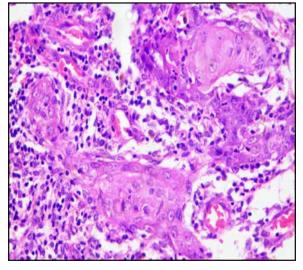


Fig 5: Nests of atypical squamous cells forming keratin pearls (Case 2)H&E (400x)

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