Secondary Plaque Over Primary Plaque on Scalp

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A 34-year-old lady presented with a solitary, slightly elevated, asymptomatic yellowish plaque on the vertex of the scalp since birth. There was no history of any overt growth or verrucous surface changes during puberty or thereafter. However, for the past six months, she reported the development of a small non-painful, yellowish swelling over the plaque. On examination, there was involvement of the vertex in the form of an irregular shaped, yellowish greasy plaque of size 3 × 3 cm^2. Hair was characteristically absent from the plaque, with the alopecic patch extending beyond into the adjacent scalp. Overlying the plaque, there was a soft to firm, cystic, non-tender, light yellowish translucent swelling of size 1.5 × 1.5 cm^2 [Figure 1]. Dermoscopy of the cystic lesion showed a translucent homogenous whitish area with a pinkish hue and an arborizing vascular pattern [Figure 2]. An excisional biopsy was done from the plaque and cystic lesion. Histopathological examination showed irregular acanthosis with prominent and crowded sebaceous lobules high up in the dermis. Apocrine glands and abortive hair follicles were also noted. The centre of the section revealed a large unilocular cyst, with apocrine glands abutting it at one edge. The cyst was itself lined by columnar and myoepithelial cells with "decapitation secretion" noted in the lumen [Figure 3a and b].

Question

What's the diagnosis?

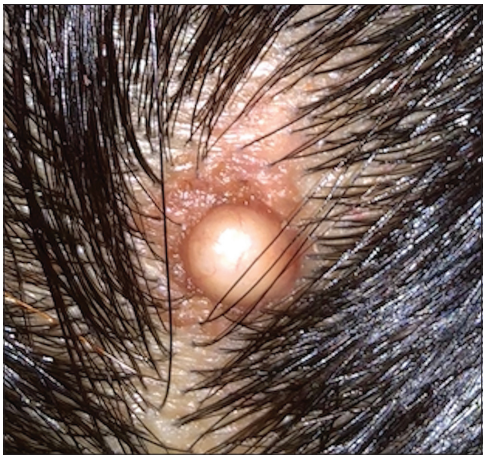

Figure 1: An irregular shaped, yellowish greasy plaque of size 3 × 3 cm^2 with absence of hair over vertex of the scalp with overlying soft to firm, cystic, light yellowish translucent swelling of size 1.5 × 1.5 cm^2

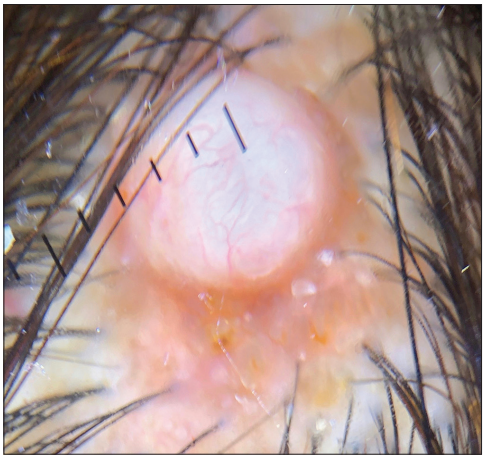

Figure 2: On dermoscopy of the cystic lesion (Dermlite DL4™, polarising, ×10X), a translucent homogenous whitish area with a pinkish hue with an arborizing vascular pattern seen

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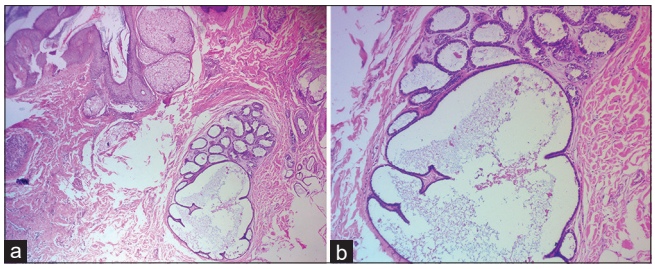

Figure 3: (a) On histopathological examination, prominent and crowded sebaceous lobules high up in the dermis. A large unilocular cyst with apocrine glands abutting it at one edge. (H&E, ×40) (b) Higher magnification showing the large unilocular cyst with apocrine glands. The cyst is lined by columnar and myoepithelial cells with "decapitation secretion" noted in the lumen. (H&E, ×100)

Answer

Nevus sebaceous with apocrine hidrocystoma.

Discussion

Nevus sebaceous of Jadassohn is an organoid hamartoma, originating from the pilosebaceous follicular unit.[1] The common embryological origin of the pilosebaceous-apocrine unit supports the divergent histological findings of follicular, apocrine, and sebaceous units along with epidermal hyperplasia in nevus sebaceous. It commonly presents on the scalp as greasy yellowish warty hairless plaque, with increased growth around puberty secondary to hormonal influences.[2] There can be secondary neoplasms associated with nevus sebaceous in around one-fourth of the cases.[3]

Most of the reported secondary neoplasms are benign. In a study of 707 cases of nevus sebaceous, secondary benign neoplasm was found to be in 18.9% cases. However, 2.5% of cases had a malignant neoplasm. Trichoblastoma followed by syringocystadenoma papilliferum were the common secondary tumors encountered in nevus sebaceous. The malignant neoplasms included basal cell carcinoma, which is the most common, followed by squamous cell carcinoma and sebaceous carcinoma. Other tumors found were apocrine/eccrine adenoma, trichilemmoma, desmoplastic trichilemmoma, sebaceoma, adenomyoepithelioma, apocrine carcinoma, microcystic adnexal carcinoma.[3] Apart from tumors, other conditions like verruca, melanocytic nevi, infundibular cyst, actinic keratosis, keratoacanthoma were also encountered.

Reports in the literature regarding apocrine hidrocystoma over the scalp are limited and to the best of our knowledge, only one case of apocrine hidrocystoma arising over nevus sebaceous has been reported before.[4]

Apocrine hidrocystomas or cystadenomas are benign cystic tumors that arise from the secretory portion of apocrine sweat glands. They present as a dome-shaped, translucent cystic swelling usually with a bluish hue. They are mostly solitary and have a predilection for the head and neck area.[4] Dermoscopy of an apocrine hidrocystoma shows a translucent to opaque, skin-colored to yellowish homogenous area. Two distinct vascular patterns are described, the arborizing vessels and the linear irregular vessels.[5] On histopathology of apocrine hidrocystoma, unilocular or multilocular dermal cysts are noted. These are lined by an outer layer of myoepithelial cells (small cuboidal) and an inner layer of tall columnar secretory cells with a notable presence of 'decapitation secretions.'[6]

Nevus sebaceous is a hamartoma involving epidermal and dermal components, making it a fertile ground for various neoplasms and proliferations to arise.[3] Ours is the second case of apocrine hidrocystoma arising over nevus sebaceous to be reported. Thus, adds another benign neoplasm to the list of conditions arising from nevus sebaceous.

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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