

Case Report

Desmoplastic trichilemmoma arising in a nevus sebaceous: a case report

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Abstract: Nevus sebaceous is a congenital hamartoma of cutaneous structures, commonly associated with the development of secondary neoplasms. Desmoplastic trichilemmoma is characterized by the typical features of trichilemmoma (proliferation of columnar clear cells surrounded by cells forming a palisade resting on a thickened eosinophilic basement membrane) but contains a fibrous stroma with bands of epithelial cells, generally in the central zone. To the best of our knowledge, only 5 cases of desmoplastic trichilemmoma arising in a nevus sebaceous have been reported previously. Herein we report an unusual case of desmoplastic trichilemmoma arising in a nevus sebaceous and review the literature.

Keywords: Desmoplastic trichilemmoma, nevus sebaceous, skin, neoplasms, fibrous stroma

Introduction

Trichilemmomas are benign adnexal skin tumors related to the outer sheath of pilosebaceous hair follicles, which is a lobular or plate-like growth of palisading clear cells with variable central keratinization. The cells are glycogen-rich by PAS stain. Trichilemmomas were first described by Headington and French in 1962 [1]. Desmoplastic trichilemmoma (DT) is a rare variant of trichilemmoma, first reported by Starink et al. [2]. DT arising in a nevus sebaceous is so rare that only 5 cases have been reported [3-5]. Herein we report such a case and review the related literature.

Case report

A 28-years-old woman presented a plaque with a history of a birthmark above her left ear. The patient noted that the affected area has been increasing in size. The lesion, which appeared nodular and verrucoid (**Figure 1**), was clinically diagnosed as nevus sebaceous and excised.

Pathological findings

On gross examination, the mass was hairless, waxy in appearance, with a smoothly warty surface. Microscopically, the epidermis showed

mild acanthosis and papillomatosis. Follicular dysgenesis and follicular induction were found. Sebaceous glands were aberrantly placed, some of the sebaceous glands were inserting directly onto the epidermis, and also some follicular induction was found. A superficial, well-circumscribed, little solid tumor was also seen, characterized by bands of clear cells connecting to the basal layer of the epidermis (**Figure 2A**). These bands were surrounded by a peripheral palisade of columnar cells resting on a thickened eosinophilic basement membrane (**Figure 2B**). The tumor was composed of cuboidal cells, which had central, round nuclei and clear cytoplasm. The central area of the tumor was cellular, and nets and cords of cells were entrapped in dense collagenous stroma. Centrally, foci of epidermal keratinization and occasionally small keratinous microcysts were found (**Figure 2C**). Beside the solid nodules, follicular induction and micro-epidermoid cysts were found.

Discussion

Nevus sebaceous usually occurs on the head and neck region and is usually clinically apparent at birth. It presents as a well-demarcated skin-colored to yellowish alopecic patch. In a



Figure 1. The lesion appears nodular and verrucoid.

study of nevus sebaceous, a total of 159 secondary proliferations in the 707 specimens were analyzed, and only 3 cases of DT were found [3]. A number of benign and malignant neoplasms might arise in or be associated with nevus sebaceous, such as trichoblastoma, followed by syringocystadenoma, syringoma, spiradenoma, squamous cell carcinoma, keratoacanthoma, and various sebaceous and apocrine, eccrine gland tumors.

DT is a histologic variant that, in addition to classical trichilemmal features, demonstrates thin strands of epithelial cells embedded into a desmoplastic stroma, which are usually found on the face, but other sites might be involved. Occasionally, it arises within a pre-existing nevus sebaceous, and coexistence with basal cell carcinoma has been described [6-9]. Reviewing the literature, 82 cases of DTs have been reported in the English literature. But DTs arising in nevus sebaceous are rare. Superficially, DT shows the changes of a trichilemmoma, but in the center of the lesion are squamoid cells enmeshed in a densely collagenized stroma. Hilliard et al. reported p16 immunostaining was more pronounced in the DTs than in the trichilemmomas [10].

The differential diagnosis at the pathological aspect, according to the basaloid, clear cell and desmoplastic features, has a different differential diagnosis. From the basaloid aspect, DT should differentiate with other basaloid benign or malignant entities, such as basal cell carcinoma, inverted follicular keratosis, trichoepithelioma, tumors of the follicular infundibulum, hydroacanthoma, poroma, and so on. Unlike

basal cell carcinomas, DT has features of glycogen rich basaloid proliferation showing peripheral palisading, lacking the slit-like retraction of a basal cell carcinoma. The absence of both atypia and mitotic activity, and the presence of lobules of clear epithelial cells surrounded by a thickened basement membrane, are the key to a correct diagnosis of DT. A PAS staining with diastase can help to differentiate DT from sebaceous carcinoma by highlighting the glycogen in the clear cells of DT. Some authors suggest that immunochemical staining for antibodies against CD34 may be helpful. The positivity of CD34 can also be used to differentiate it from basal cell carcinoma. Tricholemmomas may share features with inverted follicular keratosis, but the latter lacks hyaline membranes, and clear cell or basaloid features are less common. From the clear cell aspect, DT should differentiate with hydroacanthoma simplex, poroma, hidradenoma, and metastasis renal cell carcinoma. In contrast with a trichilemmoma or DT, hydroacanthoma simplex or poroma shows lumen formation, at least focally. From the desmoplastic aspect, DT should differentiate with morpheiform variant basal cell carcinoma, desmoplastic squamous cell carcinoma, microcystic adnexal carcinoma, and desmoplastic trichoepithelioma. Generally, DT has superficial growth with the features of a typical trichilemmoma, which is composed of columnar cells peripherally palisaded, has a thickened, eosinophilic basement membrane, variable proportions of squamous cells with eosinophilic to clear cytoplasm. Despite its “desmoplastic” aspect, DT has more distinct borders than other desmoplastic adnexal tumors.

DT arising in a nevus sebaceous is very rare. The disease is easily confused with other mimics. The prognosis of DT may be favorable. Therefore, the accurate diagnosis of this type of tumor is important.

Disclosure of conflict of interest

None.

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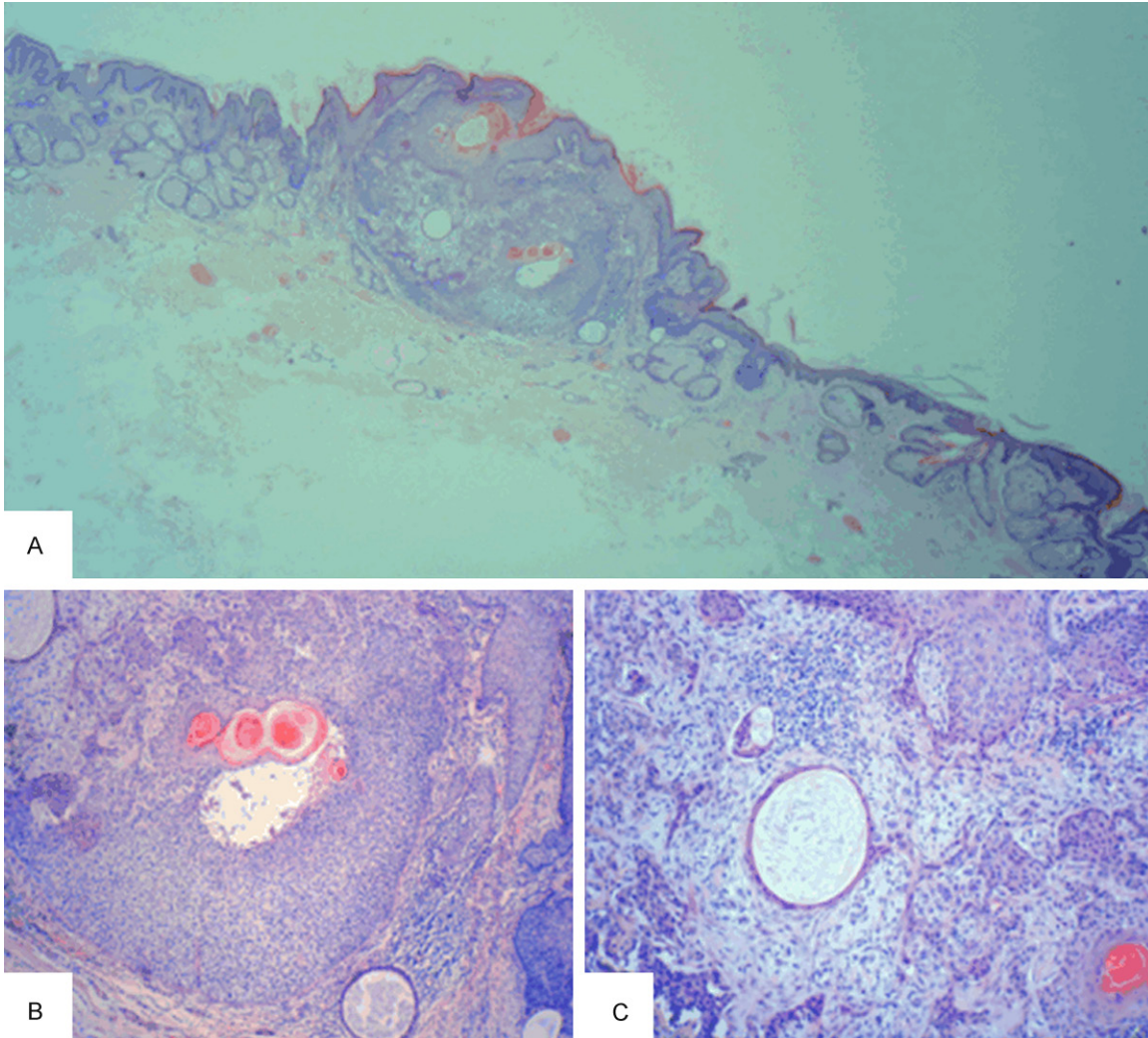


Figure 2. A. A little nodule is apparent, with mild acanthosis and papillomatosis, follicular dysgenesis and follicular induction are also seen. B. The nodule is surrounded by a peripheral palisade of columnar cells, a thickened eosinophilic basement membrane, and micro-epidermoid cysts and follicular induction are also detected. C. At medium power of the central area of the nodule, the tumor contains cellular nets and cords entrapped in a dense collagenous stroma. Centrally, the foci of epidermal keratinization and occasionally small keratinous microcysts are found.

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