Melanotrichoblastoma of Nose: A Case Report

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Abstract

Melanotrichoblastoma represents a rare variant of trichoblastoma in which neoplastic epithelium is colonized by dendritic melanocytic cells. We report in this paper a rare case of melanotrichoblastoma located in the nose, diagnosed in a 67-year-old female.

Keywords: Epithelial cells; Melanocytes; Melanotrichoblastoma; Trichoblastoma

Introduction

Trichoblastoma are benign skin adnexal tumors, under the category of trichogenic tumors. It shows proliferation of epithelial and mesenchymal cells which recapitulate the hair follicle development [1]. Melanotrichoblastoma is a rare variant of pigmented trichoblastoma. Only few cases have been reported in literature [2]. We report a rare case of melanotrichoblastoma of nose and we specify similarities with other neoplastic lesions of skin that can lead to diagnostic dilemma.

Case Report

A 67-year-old female patient, with a history of diabetes and high blood pressure, presented with a gradually enlarging mass over the wing of the nose for last 7 years. The patient reported a trauma one year ago, with the increase of the size of the lesion since. Clinical examination revealed an ulcerated, well-circumscribed and pigmented nodule measuring, approximately 15 mm in diameter. It was firm in consistency. The rest of the somatic examination was without abnormalities (Figure 1). The dermoscopy revealed the presence of an arborizing vessel, chrysalis, blue-gray ovoid nests, and ulceration (Figure 2).

The diagnosis of a pigmented Basal Cell Carcinoma (BCC) was initially evoked and the patient was referred for surgical resection. The patient was operated under general anesthesia. She had an excision of the tumor with a sliding flap. The deep cuts were without abnormalities on the extemporaneous examination.

The postoperative course was simple. There was no recurrence after a follow up of one year.

On microscopic examination, the nodule described macroscopically corresponds to a tumoral proliferation of the dermis arranged in masses in places connected to the epidermis. The tumor cells have a basaloid appearance and reduced cytoplasms sometimes loaded with melanin pigments and are provided with nuclei monomorphs taking a palleriadic disposition at the periphery. The figures of mitoses were present. The stroma was reduced and fibrous. The dendritic melanocytes were also positive for HMB45, and for Melan A (Figure 3). Based on the overall histopathology features, a diagnosis of melanotrichoblastoma was made.
Discussion

Trichoblastoma is a rare benign tumor showing differentiation toward the primitive hair follicle [3]. The typical clinical appearance is a solitary, flesh-colored, well-circumscribed, slow-growing tumor that usually progress in size over months to years [4,5]. It can grow up to 3 cm. Some cases can reach up to 8-10 cm in size [3]. Although they may be present at any age, they most commonly occur in adults in the fifth to seventh decades of life and are equally distributed between males and females. They most often occur on the head and neck with a predilection for the scalp. Although they behave in a benign fashion, cases of malignant trichoblastomas have been reported [5].

Melanotrichoblastoma, also called as pigmented trichoblastoma represents a very rare variant of trichoblastoma in which neoplastic epithelium is colonized by dendritic melanocytic cells [6]. Although melanotrichoblastomas seem to be the most common tumors developing on the nevus sebaceous of Jadassohn, they are usually solitary and may develop in association with other adnexal neoplasms [1]. Due to its relative rarity, its molecular landscape has not been completely understood [6].

According to our knowledge, our case corresponds to the third case of nasal melanotrichoblastoma reported in literature [2,7]. The first case, described by Kamat et al., [2] was reported in a 50-year-old man with a 4.5 cm mass over the tip of the nose that slowly enlarged over the course of 6 years. The second case, presented by Sathyaki et al., [7] was described in a 65-year-old Korean woman with a 1 cm mass on the right side of the nose that had been slowly enlarging over the course of 1 year. Comparing these clinical findings with our case demonstrate a relatively similar age of onset in the early to middle-aged adult years. All 3 tumors were slow growing. Additionally, the 3 cases showed well-circumscribed lesions with notable pigmentation.

The differential diagnosis of the pigmented variant includes several epithelial neoplasms with pilar differentiation such as basal cell carcinoma, pigmented pilomatrixoma and melanocytic matricoma [6].

Dermoscopic aspect of trichoblastoma described in the literature is based on small series. We don’t know a specific description for melanotrichoblastoma, given probably the scarcity of this tumor. The presence of ovoid nests, blue-gray cells, and ulceration appears more common in the case of BCC. In our patient, dermoscopy does not differentiate between a melanotrichoblastoma and a BCC [8].

Histologically, pigmented trichoblastomas have distinct melanin pigment at the center of lobules, and dispersed melanocytes between the tumoral cells. These melanocytes show positivity for S100 protein, HMB45, Melan A, and tyrosinase [3]. Epidermal connection, atypia or mitosis is absent. Myxoid stroma and stromal retraction or clefting around basaloid islands, characteristic of basal cell carcinoma, were also absent.

Pigmented trichoblastoma can be a part of a rare syndrome called phacomatosis pigmentokeratotica which is due to mutation of HRAS G123R gene. Treatment consists of surgical resection. No cases of recurrence were noted among the cases reported in the literature [9].

Conclusion

This case of melanotrichoblastoma, is being presented because of its rarity and increased chances of clinical misdiagnosis as melanotic neoplasm, nevus and melanoma.

Conflict of Interest

The authors declare no conflicts of interest.

References

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