

# Melanotrichoblastoma: A Rare Pigmented Variant of Trichoblastoma



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## PRACTICE POINTS

- Pigmented trichoblastoma is a histologic variant of trichoblastoma characterized by the presence of melanin pigment.
- At least some pigmented trichoblastomas contain melanocytes and have been referred to as melanotrichoblastomas.
- The presence of melanocytes within pigmented trichoblastomas should not be confused as representing an example of colonization or a collision tumor.

Trichoblastomas are rare cutaneous tumors arising from the hair bulb and mesenchyme. Although they are benign, they can pose a diagnostic dilemma for the clinician and pathologist because they clinically and histologically mimic more common lesions such as basal cell carcinomas (BCCs) and trichoepitheliomas. It is important for the clinician and pathologist to be aware of such tumors and their variants. We present a case of a melanotrichoblastoma, an exceedingly rare variant of trichoblastoma, as well as review the current literature on the clinical presentation and histologic differentiation of these unique tumors with their more commonly seen mimics.

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Trichoblastomas are rare cutaneous tumors that recapitulate the germinative hair bulb and the surrounding mesenchyme. Although benign, they can present diagnostic difficulties for both the clinician and pathologist because of their rarity and overlap both clinically and microscopically with other follicular neoplasms as well as basal cell carcinoma (BCC). Several

classification schemes for hair follicle neoplasms have been established based on the relative proportions of epithelial and mesenchymal components as well as stromal inductive change, but nomenclature continues to be problematic, as individual neoplasms show varying degrees of differentiation that do not always uniformly fit within these categories.<sup>1,2</sup> One of these established categories is a pigmented trichoblastoma.<sup>3</sup> An exceedingly rare variant of a pigmented trichoblastoma referred to as melanotrichoblastoma was first described in 2002<sup>4</sup> and has only been documented in 3 cases, according to a PubMed search of articles indexed for MEDLINE using the term *melanotrichoblastoma*.<sup>4-6</sup> We report another case of this rare tumor and review the literature on this unique group of tumors.

## Case Report

A 25-year-old white woman with a medical history of chronic migraines, myofascial syndrome, and Arnold-Chiari malformation type I presented to dermatology with a 1.5-cm, pedunculated, well-circumscribed tumor on the left side of the scalp (Figure 1). The tumor was grossly flesh colored with heterogeneous areas of dark pigmentation. Microscopic examination demonstrated that within the superficial and deep dermis were variable-sized nests of basaloid cells. Some of the nests had large central cystic spaces with brown pigment

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**FIGURE 1.** Pedunculated lesion on the left side of the scalp with areas of pigmentation.

within some of these spaces and focal pigmentation of the basaloid cells (Figure 2A). Focal areas of keratinization were present. Mitotic figures were easily identified; however, no atypical mitotic figures were present. Areas of peripheral palisading were present but there was no retraction artifact. Connection to the overlying epidermis was not identified. Surrounding the basaloid nodules was a mildly cellular proliferation of cytologically bland spindle cells. Occasional pigment-laden macrophages were present in the dermis. Focal areas suggestive of papillary mesenchymal body formation were present (Figure 2B). Immunohistochemical staining for Melan-A was performed and demonstrated the presence of a prominent number of melanocytes in some of the nests (Figure 3) and minimal to no melanocytes in other nests. There was no evidence of a melanocytic lesion involving the overlying epidermis. Features of nevus sebaceus were not present. Immunohistochemical staining for cytokeratin (CK) 20

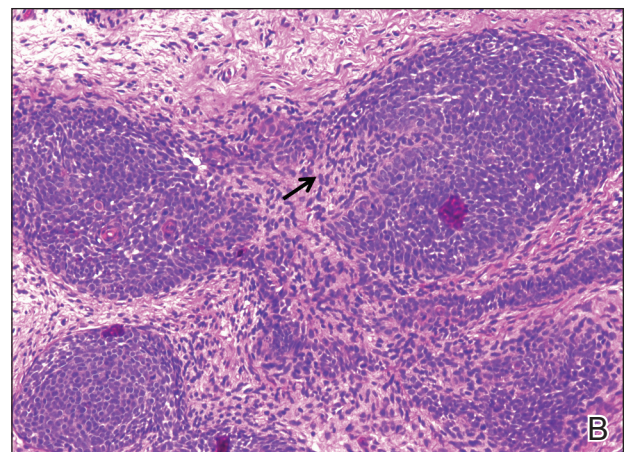
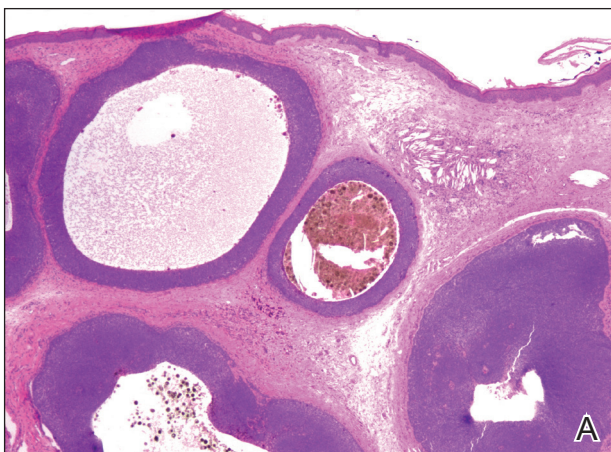
was performed and demonstrated no notable number of Merkel cells within the lesion.

### Comment

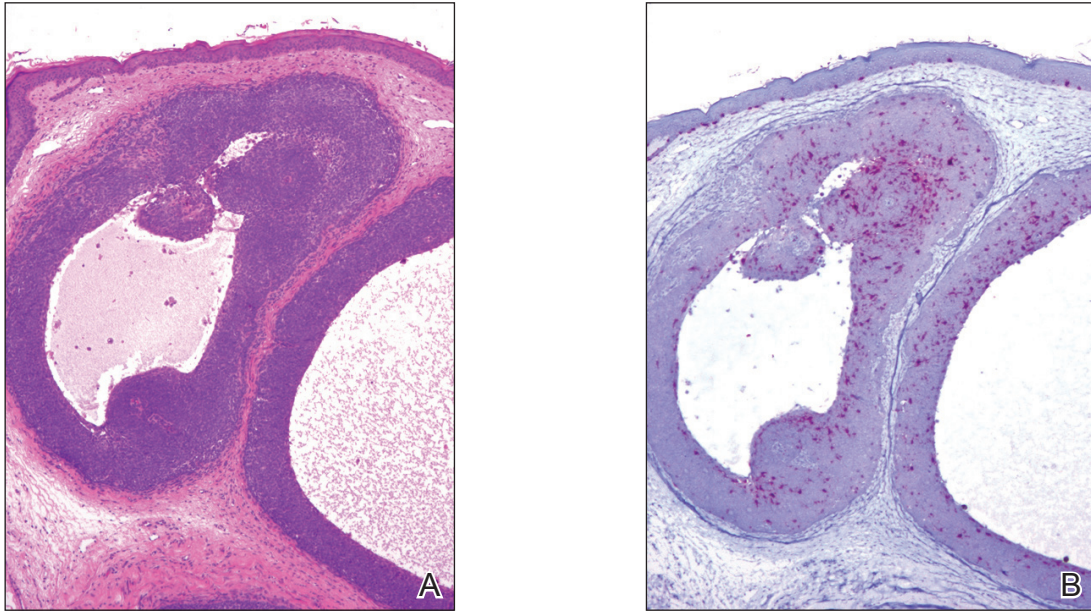
**Overview of Trichoblastomas**—Trichoblastomas most often present as solitary, flesh-colored, well-circumscribed, slow-growing tumors that usually progress in size over months to years. Although they may be present at any age, they most commonly occur in adults in the fifth to seventh decades of life and are equally distributed between males and females.<sup>7,8</sup> 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.<sup>9</sup>

**Histopathology**—Histologically, these tumors are well circumscribed but unencapsulated and usually located within the deep dermis, often with extension into the subcutaneous tissue. An epidermal connection is not identified. The tumor typically is composed of variable-sized nests of basaloid cells surrounded by a variable cellular stromal component. Although peripheral palisading is present in the basaloid component, retraction artifact is not present. Several histologic variants of trichoblastomas have been reported including cribriform, racemiform, retiform, pigmented, giant, subcutaneous, rippled pattern, and clear cell.<sup>5</sup> Pigmented trichoblastomas are histologically similar to typical trichoblastomas, except for the presence of large amounts of melanin deposited within and around the tumor nests.<sup>6</sup> A melanotrichoblastoma is a rare variant of a pigmented trichoblastoma; pigment is present in the lesion and melanocytes are identified within the basaloid nests.

The stromal component of trichoblastomas may show areas of condensation associated with some of the basaloid cells, resembling an attempt at hair bulb formation.



**FIGURE 2.** Melanotrichoblastoma histopathology with variable-sized nests of basaloid cells with central cystic spaces. Pigmentation is associated with some of the nests (A)(H&E, original magnification ×20). A nest of basaloid cells on the right (black arrow) demonstrated an area of indentation in which there was peripheral palisading and condensation of stromal cells consistent with papillary mesenchymal body formation (B)(H&E, original magnification ×100).



**FIGURE 3.** Melanotrichoblastoma nests of basaloid cells (A)(H&E, original magnification  $\times 40$ ) with corresponding immunoperoxidase stain for Melan-A demonstrated the presence of the melanocytes (B)(original magnification  $\times 40$ ).

Staining for CD10 will be positive in these areas of papillary mesenchymal bodies.<sup>10</sup>

In an immunohistochemical study of 13 cases of trichoblastomas, there was diffuse positive staining for CK14 and CK17 in all cases (similar to BCC) and positive staining for CK19 in 70% (9/13) of cases compared to 21% (4/19) of BCC cases. Staining for CK8 and CK20 demonstrated the presence of numerous Merkel cells in all trichoblastomas but in none of the 19 cases of BCC tested.<sup>11</sup> However, other studies have reported the presence of Merkel cells in only 42% to 70% of trichoblastomas.<sup>12,13</sup> Despite the lack of Merkel cells in our case, the lesion was interpreted as a melanotrichoblastoma based on the histologic features in conjunction with the presence of the melanocytes.

**Differential Diagnosis**—The clinical and histologic differential diagnosis of trichoblastomas includes both trichoepithelioma and BCC. Clinically, all 3 lesions often are slow growing, dome shaped, and small in size (several millimeters), and are observed in the same anatomic distribution of the head and neck region. Furthermore, they often affect middle-aged to older individuals and those of Caucasian descent, though other ethnicities can be affected. Histologic evaluation often is necessary to differentiate between these 3 entities.

Histologically, trichoepitheliomas are composed of nodules of basaloid cells encircled by stromal spindle cells. Although there can be histologic overlap between trichoepitheliomas and trichoblastoma, trichoepitheliomas typically will display obvious features of hair follicle differentiation with the presence of small keratinous cysts

and hair bulb structures, while trichoblastomas tend to display minimal changes suggestive of its hair follicle origin. Similar to trichoblastomas, BCC is composed of nests of basaloid cells; however, BCCs often demonstrate retraction artifact and connection to the overlying epidermis. In addition, BCCs typically demonstrate a fibromucinous stromal component that is distinct from the cellular stroma of trichoblastic tumors. Immunoperoxidase staining for androgen receptors has been reported to be positive in 78% (25/32) of BCCs and negative in trichoblastic tumors.<sup>14</sup>

**Melanotrichoblastoma Differentiating Characteristics**—An exceedingly rare variant of pigmented trichoblastoma is the melanotrichoblastoma. There are clinical and histologic similarities and differences between the reported cases. The first case, described by Kanitakis et al,<sup>4</sup> reported a 32-year-old black woman with a 2-cm scalp mass that slowly enlarged over the course of 2 years. The second case, presented by Kim et al,<sup>5</sup> described a 51-year-old Korean man with a subcutaneous 6-cm mass on the back that had been present and slowly enlarging over the course of 5 years. The third case, reported by Hung et al,<sup>6</sup> described a 34-year-old Taiwanese man with a 1-cm, left-sided, temporal scalp mass present for 3 years, arising from a nevus sebaceous. Comparing these clinical findings with our case of a 25-year-old white woman with a 1.5-cm mass on the left side of the scalp, melanotrichoblastomas demonstrate a relatively similar age of onset in the early to middle-aged adult years. All 4 tumors were slow growing. Additionally, 3 of 4 cases demonstrated a predilection for the head, particularly the scalp, and grossly showed well-circumscribed lesions with notable pigmentation.

Although age, size, location, and gross appearance were similar, a comparable ethnic and gender demographic was not identified.

Microscopic similarities between the 4 cases were present. Each case was characterized by a large, well-circumscribed, unencapsulated, basaloid tumor present in the lower dermis, with only 1 case having tumor cells occasionally reaching the undersurface of the epidermis. The tumor cells were monomorphic round-ovoid in appearance with scant cytoplasm. There was melanin pigment in the basaloid nests. The basaloid nests were surrounded by a proliferation of stromal cells. The mitotic rate was sparse in 2 cases, brisk in 1 case, and not discussed in 1 case. Melanocytes were identified in the basaloid nests in all 4 cases; however, in the current case, the melanocytes were seen in only some of the nests. None of the cases exhibited an overlying junctional melanocytic lesion, which would argue against a possible collision tumor or colonization of an epithelial lesion by a melanocytic lesion.

Although the histologic features of our cases are consistent with prior reports of melanotrichoblastoma, there is some question as to whether it represents a true variant of a pigmented trichoblastoma. There are relatively few articles in the literature that describe pigmented trichoblastomas, and of those, immunohistochemistry staining for melanocytes is uncommon. In one of the earliest descriptions of a pigmented trichoblastoma, dendritic melanocytes were present within the tumor lobules; however, the lesion was reported as a pigmented trichoblastoma and not a melanotrichoblastoma.<sup>3</sup> It is possible that all pigmented trichoblastomas may contain some number of dendritic melanocytes, thus negating the existence of a melanotrichoblastoma as a true subtype of pigmented trichoblastomas. Additional study looking at multiple examples of pigmented trichoblastomas would be required to more definitively classify melanotrichoblastomas. It is important to appreciate that at least some cases of pigmented trichoblastomas may contain melanocytes and not to confuse the lesion as representing an example of colonization or collision tumor. A rare case of melanoma possibly arising from these dendritic melanocytes has been reported.<sup>15</sup>

## Conclusion

Trichoblastomas are uncommon tumors of germinative hair bulb origin that can have several histologic variants. A well-documented subtype of trichoblastoma characterized by melanin deposits within and around tumor

nests has been identified and classified as a pigmented trichoblastoma. Four cases of melanotrichoblastoma have been reported and represent a variant of a pigmented trichoblastoma characterized by the presence of melanocytes within the lesion. Whether they represent a true variant is of some debate and additional study is required. Although these tumors are exceedingly rare, it is important for the clinician and pathologist to be aware of this entity to prevent confusion with other similarly appearing follicular lesions, most notably BCCs, because of the difference in treatment and follow-up.

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