

## [ C A S E R E P O R T ]

# Combined Cellular Blue Nevus and Trichoepithelioma

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## ABSTRACT

Biphasic lesions composed of melanocytic and epithelial components are not uncommon. Combined trichoepithelioma and cellular blue naevus is a rare lesion that may mimic melanoma clinically. The authors describe what they believe to be the fourth case of this rare tumor and review the literature. Recent descriptions of malignant basomelanocytic tumors have raised interesting questions surrounding the pathogenesis of lesions composed of these cell lineages.

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The authors describe a combined tumor composed of a cellular blue nevus and a trichoepithelioma. This highly distinctive tumor may not be as uncommon as is reflected in the literature. To the best of the authors' knowledge, there have only been three previous cases reported in the literature (Table 1).<sup>1,2</sup>

## CASE REPORT

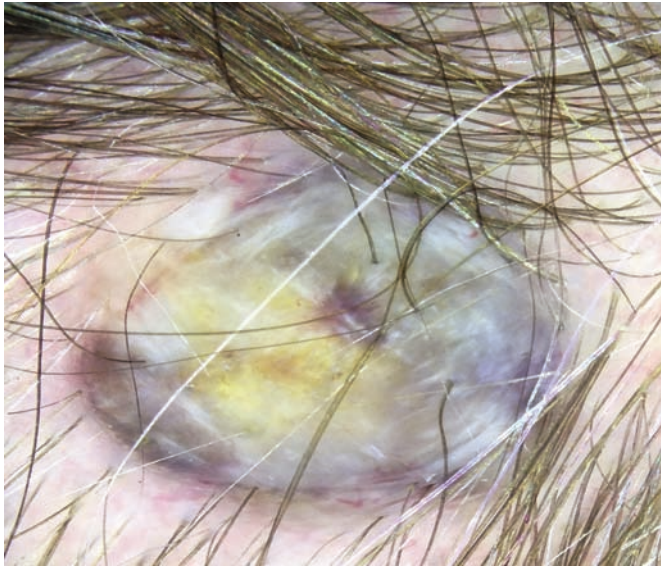
A 56-year-old woman presented with a darkly pigmented lesion on her scalp that her hairdresser had noticed and was concerned it had increased in size. The patient reported the lesion had been present for years. She had no previous skin cancers and there was no family

**TABLE 1. Previously reported cases of combined cellular blue nevus and trichoepithelioma**

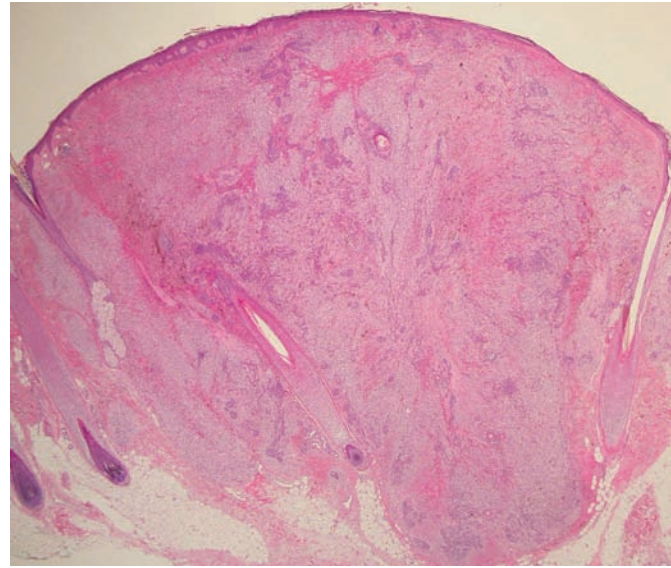
	AGE	SITE	CLINICAL DIAGNOSIS	ADDITIONAL PATHOLOGIC FEATURES
Case 1 (Newton) <sup>1</sup>	83	Scalp	Malignant melanoma	Metaplastic bone
Case 2 (Newton) <sup>1</sup>	39	Temple	Pigmented nevus	Metaplastic bone, "syringomatous areas"
Case 3 (Al-Ghamdi) <sup>2</sup>	61	Scalp	Melanoma	

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**Figure 1.** Dermoscopic image shows a heavily pigmented lesion with yellow areas. This was presented on a dermoscopy blog (International Society of Dermoscopy) and was thought to be either a blue nevus, a melanoma, or a reactive change to a ruptured cyst.



**Figure 2.** A well-circumscribed, biphasic, dermal proliferation involving the full thickness of the dermis and shows tongues of rounded tumor fronts in the deep dermis. The overlying epidermis was uninvolved (H&E stain, 4X)

history of melanoma. Clinical examination revealed Fitzpatrick skin type II with a raised, nodular, variably pigmented lesion on the right posterior scalp measuring approximately 6x5mm. Dermoscopic (DermLite, DL-1) examination with cross polarized light (Figure 1) showed an irregularly colored lesion with focal homogeneous blue black pigmentation and yellow areas. An excision biopsy was performed with 2mm margins as per guidelines.

Histopathological examination revealed a fairly well-circumscribed biphasic dermal proliferation involving the full thickness of the dermis and shows tongues of rounded tumor fronts, which encroached on subcutaneous fat. The overlying epidermis was uninvolved (Figure 2).

The tumor comprised intimately mixed populations of melanocytes and basaloid cells. The melanocytic component consisted of islands of plump spindled cells with an abundant pale cytoplasm and minimal pigment and rarer slender dendritic melanocytes with heavy melanin pigmentation. Regularly intermixed with this, there were nests of basaloid cells anastomosed in a lace-like pattern and forming horn cysts focally (Figure 3). The follicular component was surrounded by a prominent stroma, which condensed around the basaloid islands as hair papilla-like structures (Figure 4).

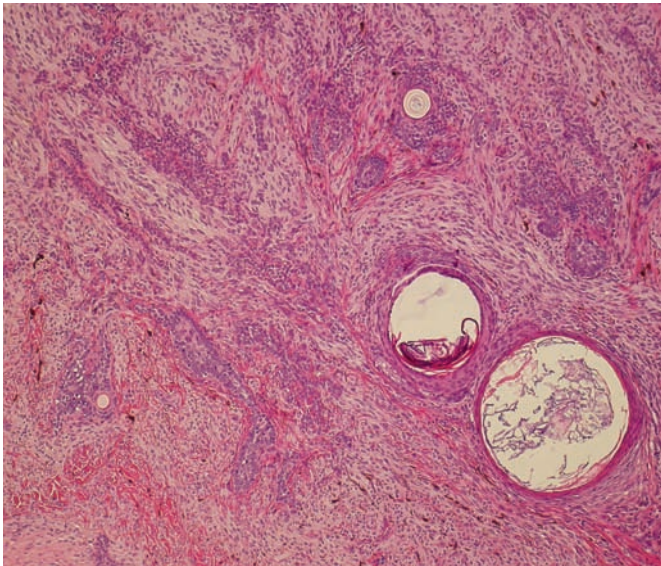
The follicular and melanocytic components stained strongly with p63 and S100, respectively, and there was no cross-over of staining.

The combination of common melanocytic nevi with other tumors of adnexal origin has often been described.<sup>3-5</sup> Melanocytic nevi may be closely associated with eccrine

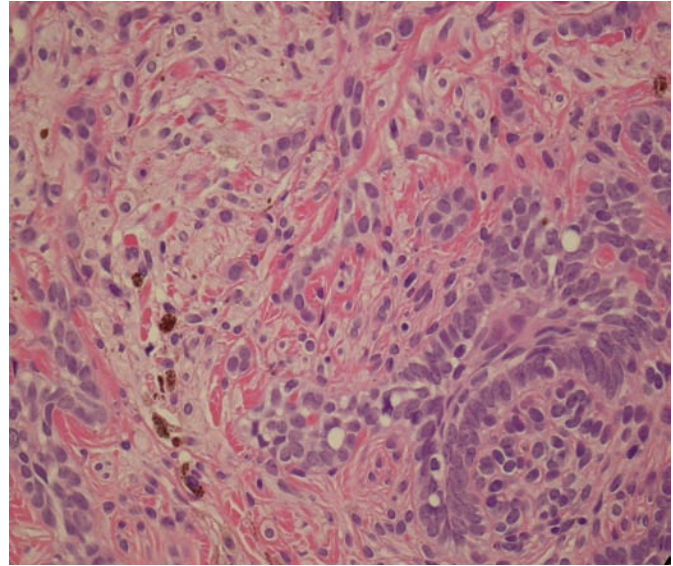
sweat ducts,<sup>6</sup> cystic dilatation of hair follicles, or cysts.<sup>7</sup> The most common association is a dermal nevus with epidermoid cyst. This is so common that in French-speaking countries it has an eponym: Duperrat nevus. Combinations of pigmented nevi and various adnexal proliferations have also been reported.<sup>8</sup> About 10 percent of cases of desmoplastic trichoepitheliomas are associated with a coexisting intradermal melanocytic nevus.<sup>5</sup>

There are several hypotheses to explain these distinct cell types within the same lesion. It has been proposed that pigmented nevi include not only benign neoplasms but also hamartomas in which nevus cells occur together with several different structures of ectodermal origin.<sup>5</sup> Another hypothesis is that the melanocytic lesion elicits an inductive effect on epithelia. Perhaps the most compelling example of this can be seen in an example of a melanocytic nevus stimulating hair follicle formation on the soles.<sup>9</sup> Similarly, epidermal hyperplasia and follicular induction is well known to occur in conjunction with dermal nevi.<sup>10</sup> Keen<sup>11</sup> suggested that the local paracrine effect of cytokines or growth factors secreted by the nevus cells accounts for epithelial proliferation.<sup>11,12</sup> It seems likely that at least some of the benign cystic proliferations associated with nevi may be due to occlusion of follicles rather than some type of chemical paracrine effect.<sup>13</sup>

Malignant tumors with dual follicular and melanocytic differentiation (basomelanocytic tumors) have received some attention in the literature recently. It remains unclear whether these represent tumor collision, or melanocytic colonization, or whether they illustrate a



**Figure 3.** Sheets of plump spindled melanocytes with an abundant pale cytoplasm and rarer collections of dendritic melanocytes with heavy melanin pigmentation are intimately intermixed with nests of basaloid cells anastomosed in a lace-like. Horn cysts are seen focally (H&E stain, 20X).



**Figure 4.** The follicular component is surrounded by the cellular blue nevus and focally the basaloid islands as hair papilla-like structures (H&E stain, 40X)

previously undelineated phenomenon by which pleuropotential stem cell populations can exhibit dual differentiation into epithelial and melanocytic cell lines.<sup>14-16</sup> This later theory is particularly troublesome as the evidence for the neural crest origin of melanocytes is compelling and has been widely accepted.<sup>17</sup> Rodriguez et al<sup>14</sup> advanced an interesting analogy with the neuroendocrine cells of the gastrointestinal tract. Those cells were long believed to be of neural crest derivation, but the occurrence of mixed adenocarcinomas-neuroendocrine carcinomas provided one of the first clues that eventually debunked that theory and led to the currently held belief that those cells are of the same endodermal derivation than the other epithelial cells of the gastrointestinal mucosa.<sup>14</sup>

Though there is no evidence from the current case that there is a single pluripotent cell line, it is interesting to consider that trichoepithelioma is generally regarded as a bona fide tumor rather than a hamartoma or hyperplasia, complete with an oncogene that is convincingly involved in its pathogenesis (CYLD). On the other hand, dendritic melanocytes are thought to be derived from precursor cells which do not complete migration from the neural crest to the epidermis during embryogenesis.<sup>18</sup> How these two populations are interrelated is unclear at this time.

## CONCLUSION

Combined cellular blue nevus and trichoepithelioma is a rarely reported but distinctive biphasic tumor. It can have a striking clinical presentation that mimics melanoma. The pathogenesis of such tumors is unclear.

## ACKNOWLEDGMENT

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