

Pigmented Cystic Masses on the Scalp

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A 67-year-old man presented to the dermatology clinic with 3 asymptomatic pigmented papules on the scalp. The patient reported that he was unaware of the lesions until they were pointed out weeks earlier by his primary care physician during a routine visit. He then was referred to dermatology for follow-up. Physical examination at the current presentation revealed clustered firm, smooth, well-circumscribed, pigmented papules on the scalp measuring 5 to 8 mm. The patient reported no personal or family history of skin cancer but stated that he spent a lot of time outdoors and had a history of 6 blistering sunburns in his life. A punch biopsy of each lesion was performed.

WHAT'S YOUR DIAGNOSIS?

- a. apocrine hidrocystoma
- b. blue nevus
- c. melanoma
- d. pigmented basal cell carcinoma
- e. seborrheic keratosis

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THE DIAGNOSIS: Apocrine Hidrocystoma

Histology for all 3 lesions demonstrated similar cystic structures lined by a dual layer of epithelial cells, with the outermost layer composed of flattened myoepithelial cells and the inner layer composed of cells with apocrine features (Figure 1). Based on these findings, a diagnosis of apocrine hidrocystoma was made. The patient underwent successful surgical excision shortly thereafter without recurrence at follow-up 1 year later.

Apocrine hidrocystomas are rare benign cystic lesions that are considered to be adenomatous proliferations of apocrine glands. They typically manifest as solitary asymptomatic lesions measuring 3 to 15 mm.¹ They tend to appear on the face, usually in the periorbital region, but also have been described on the neck, scalp, trunk, arms, and legs.²⁻⁴ Multiple apocrine hidrocystomas can be a marker of 2 rare inherited disorders: Gorlin-Goltz syndrome and Schopf-Schulz-Passarge syndrome.⁵ Apocrine hidrocystomas may be flesh colored or may have a blue, black, or brown appearance due to the Tyndall effect, in which light with shorter wavelengths is scattered by the contents of the lesions.² Histologically, apocrine hidrocystomas are cysts lined by a dual layer of epithelial cells. The inner layer is composed of cells with apocrine features, and the outer layer is composed of flattened myoepithelial cells. Due to their range of colors and predilection for sun-exposed surfaces, apocrine hidrocystomas may be mistaken for various malignant neoplasms, including melanoma.^{6,7}

The differential diagnosis for our patient included agminated blue nevi, melanoma, pigmented basal cell carcinoma (BCC), and seborrheic keratosis. A blue nevus is a dermal melanocytic lesion that manifests as a well-demarcated, blue to blue-black papule that typically appears on the face, scalp, arms, legs, lower back, and buttocks. Although there are several histologic subtypes, the common blue nevus usually manifests as a solitary lesion measuring less than 1 cm, often developing during childhood to young adulthood.⁸ Histologically, common blue nevi are characterized by a dermal proliferation of deeply pigmented bipolar spindled melanocytes embedded in thickened collagen bundles, often with scattered epithelioid melanophages, and no conspicuous mitotic activity (Figure 2).⁹ There are other types of blue nevi, including cellular blue nevi, which tend to be larger and manifest commonly on the buttocks and sacrococcygeal region in early adulthood.⁹ Histologically, cellular blue nevi contain oval to spindled melanocytes with scattered melanophages forming a well-demarcated nodule typically in the reticular dermis. There may be bulbous extension into the subcutaneous adipose tissue. Occasional mitoses may be seen.^{9,10} Melanoma can arise from common or cellular blue nevi, though it

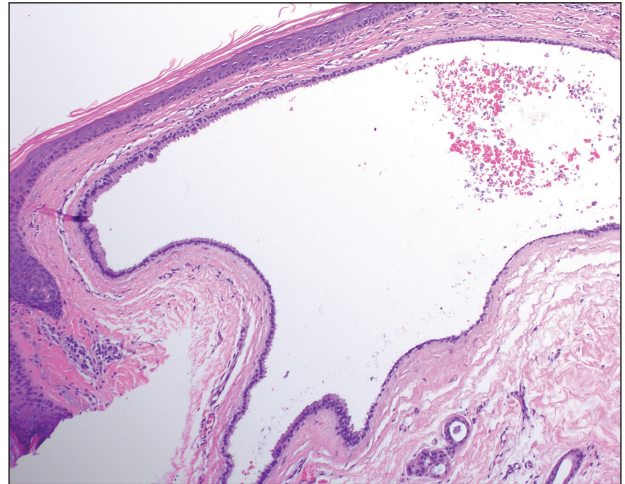


FIGURE 1. Apocrine hidrocystoma. Cystic space lined by a dual layer of epithelial cells with the outermost layer composed of flattened myoepithelial cells and the inner layer of cells with apocrine features (H&E, original magnification $\times 10$).

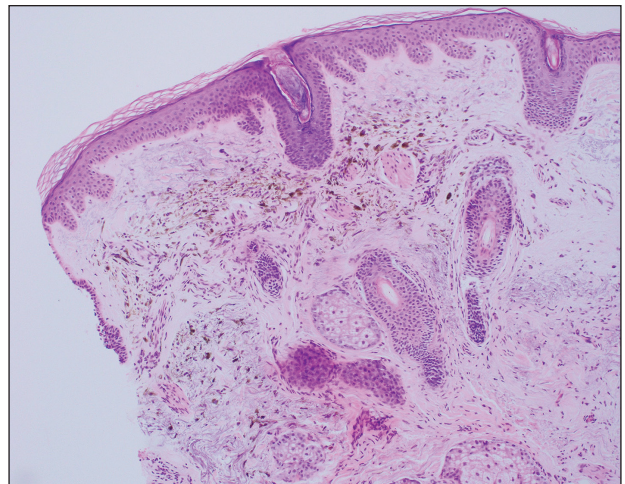


FIGURE 2. Common blue nevus. Deeply pigmented dermal spindle cell proliferation separated from the overlying epidermis by a Grenz zone (H&E, original magnification $\times 10$).

more frequently occurs with cellular blue nevi. Other subtypes of blue nevi have been described, including the sclerosing, plaque-type, combined, hypomelanotic/amelanotic, and pigmented epithelioid melanocytoma.¹¹ However, they typically have features of the common blue nevus or cellular blue nevus, such as oval/spindle cell morphology, some degree of melanin, and biphasic architecture, but are classified according to their dominant histologic characteristics.

Given the location of our patient's lesions on the scalp and his extensive history of sun exposure, malignancy was high in the differential. Multiple synchronous primary melanomas including nodular melanoma, blue nevus-like metastatic melanoma, and metastatic melanoma were considered. The leg and the scalp have the highest reported incidence of cutaneous metastases of melanoma, with many cases presenting as dermal or subcutaneous nodules and eruptive blue nevus-like papules, similar to our patient's clinical presentation.^{12,13} Nodular melanoma (NM) is one of 4 major types of melanoma, accounting for approximately 15% to 30% of cases in the United States.¹⁴ Nodular melanoma typically manifests as a smooth, raised, symmetric, well-circumscribed lesion with variable pigmentation, from very dark to amelanotic. Histologically, NM is defined as a dermal mass, either in isolation or with an epidermal component, not to exceed 3 rete ridges beyond the dermal component.¹⁵ Tumor cells have a high cell density with pleomorphism, usually with atypical epithelioid cells with vesicular nuclei and irregular cytoplasm, and occasionally spindle cells (Figure 2).¹⁶ Mitoses and necrosis are frequent. Scalp location independently is responsible for worse survival, both overall and melanoma specific.¹⁷ Nodular melanoma tends to have greater Breslow thickness at diagnosis than other melanoma subtypes and often carries a worse prognosis.

Malignant melanomas that develop from or in conjunction with or bear histologic resemblance to blue nevi are termed *blue nevus-like melanoma* or *blue nevus-associated melanoma*. These malignancies are exceedingly rare, accounting for only 0.3% of melanomas in one Turkey-based multicenter study.¹⁸ The histologic criteria for diagnosing blue nevus-like melanoma are poorly defined, and terminology of these lesions has led to some debate in naming conventions.¹⁹ Nevertheless, unlike blue nevus, blue nevus-like melanoma demonstrates histologic features of malignancy, including pleomorphism, prominent nucleoli, mitotic activity, vascular invasion, and potential necrosis.¹⁰ The lack of an inflammatory infiltrate, surrounding fibrosis, junctional activity, and pre-existing nevus can help distinguish cutaneous melanoma metastases from primary nodular melanoma. Immunohistochemical stains such as S100, Melan-A/MART1, or SOX-10 can help confirm melanocytic lineage.¹²

Pigmented BCC is a clinical and histologic variant of BCC characterized by increased melanin pigmentation due to melanocytes admixed with tumor cells. Dermoscopically, the pigment can have a maple leaf-like appearance with spoke-wheel areas, in-focus dots, and concentric structures at the dermoepidermal junction, which is more characteristic of superficial and infiltrating BCC.²⁰ In nodular BCC, the pigment occurs as blue-gray ovoid nests and globules in deeper layers of the dermis.²⁰

Seborrheic keratoses (SKs) can vary widely in clinical appearance, with pigmentation ranging from flesh

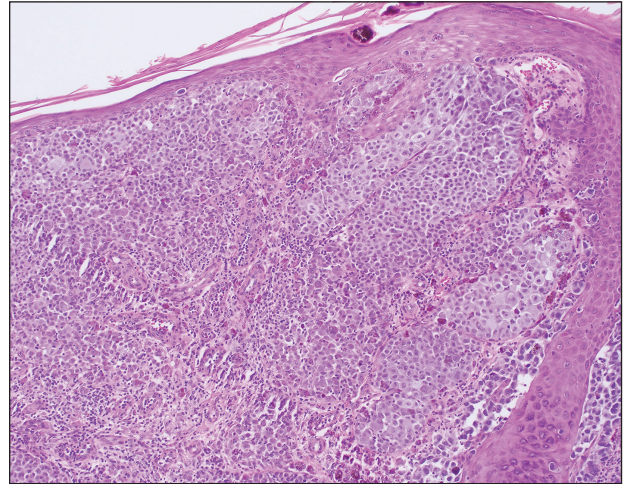


FIGURE 3. Nodular melanoma. Prominent vertical growth into the dermis with cytoplasmic melanin present (H&E, original magnification $\times 10$).

colored to yellow to brown to black. Melanoacanthomas are acanthotic SKs that are highly pigmented due to intermixed epidermal melanocytes and subepidermal melanophages.²¹ Dermoscopy can help distinguish cutaneous malignancies from SKs, which often demonstrate fissures and ridges, comedolike openings, and miliumlike cysts. Biopsy sometimes is required to assess for malignancy, as was the case in our patient. The classic histologic features of SKs include acanthosis, papillomatosis, and hyperkeratosis.²²

This case highlights the need to consider apocrine hidrocystoma, along with malignancy, in the differential diagnosis of pigmented cystic masses of the face and scalp. Because apocrine hidrocystomas are benign, they do not need to be treated but often are surgically excised for cosmesis or complete histopathologic examination. Destruction via electrodesiccation, carbon dioxide ablation, trichloroacetic acid chemical ablation, botulinum toxin injection, and anticholinergic creams sometimes is used, especially for cosmetic treatment of multiple small lesions.⁵ Our patient was treated with surgical excision with no evidence of recurrence on follow-up 1 year later.

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