

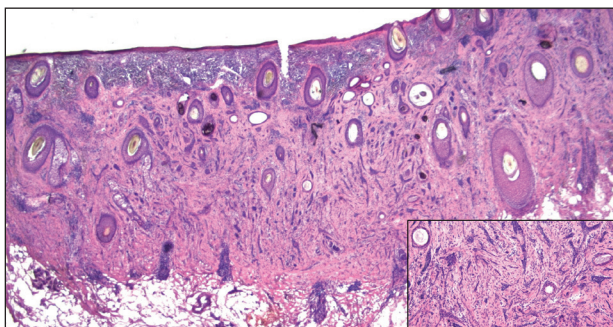
Indurated Plaque on the Eyebrow



Ryan Bogner, MD; Tye Haeberle, MD; Carol Thompson, MD; Timothy Brown, MD

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H&E, original magnification $\times 40$ (inset, original magnification $\times 100$).

A 52-year-old woman presented with an indurated plaque on the right lateral eyebrow that had been slowly enlarging over the last 4 months.

THE BEST DIAGNOSIS IS:

- a. desmoplastic trichoepithelioma
- b. microcystic adnexal carcinoma
- c. morpheaform basal cell carcinoma
- d. syringoma
- e. trichoadenoma

PLEASE TURN TO **PAGE 365** FOR THE DIAGNOSIS

From the Division of Dermatology, University of Louisville, Kentucky.

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Correspondence: Ryan Bogner, MD, 3810 Springhurst Blvd, Louisville, KY 40241 (ryan.bogner05@gmail.com).

THE DIAGNOSIS:

Microcystic Adnexal Carcinoma

Microcystic adnexal carcinoma (MAC) is a rare, low-grade adnexal carcinoma consisting of both ductal and pilar differentiation.¹ It typically presents in young to middle-aged adults as a flesh-colored or yellow indurated plaque on the upper lip, medial cheek, or chin. Histologically, MACs exhibit a biphasic pattern consisting of epithelial islands of cords and lumina creating tadpole-like ducts intermixed with basaloid nests (quiz image). Keratin horn cysts are common superficially. A dense red sclerotic stroma is seen interspersed between the ducts and epithelial islands creating a “paisley tie” appearance. The lesion displays an infiltrative pattern and can be deeply invasive, extending down to the fat and muscle (quiz image, inset). Perineural invasion is common. Atypia, when present, is minimal or mild and mitoses are rare. Although this tumor’s histologic pattern appears aggressive in nature, it lacks immunohistochemical staining such as p53, Ki-67, bcl-2, and c-erbB-2 that correlate with malignant behavior.² A common diagnostic pitfall is examination of a superficial biopsy in which an MAC may be mistakenly identified as another entity.

Syringomas are benign adnexal neoplasms with ductal differentiation.³ They are more common in women, especially those of Asian descent, and in patients with Down syndrome. They typically present as multiple small, firm, flesh-colored papules in the periorbital area or upper trunk. Histologically, syringomas also display comma-shaped tubules and ducts with a tadpolelike appearance and a dense red stroma creating a paisley tie-like pattern.

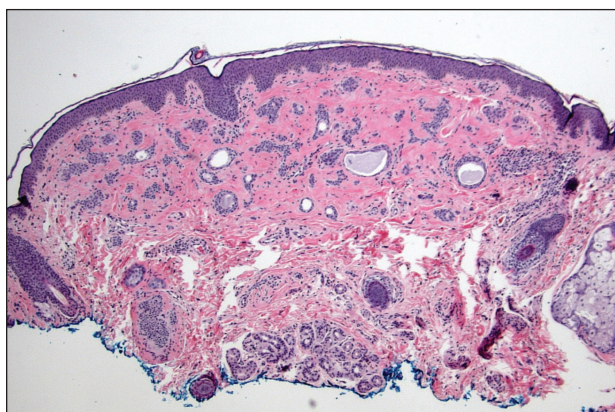


FIGURE 1. Well-circumscribed tumor invading to the depth of the superficial to mid dermis composed of small comma-shaped tubules within a dense sclerotic stroma characteristic of a syringoma. Ductal cells are polygonal or flattened with prominent eosinophilic cytoplasm. Small central lumens are present within some epithelial aggregates. There is no cytologic atypia or mitotic activity (H&E, original magnification $\times 40$).

Ductal cells have an abundant pink cytoplasm. Syringomas are well-circumscribed and more superficial than MACs without an infiltrative pattern. They lack mitotic activity or perineural invasion (Figure 1).

Desmoplastic trichoepithelioma (DTE) is a benign follicular neoplasm.⁴ It presents in adulthood with a female predominance. Clinically, it appears as a solitary flesh-colored to yellow annular plaque with raised borders and a depressed central area, often on the medial cheek. Histologically, DTEs are well-circumscribed with narrow branching cords lined with polygonal cells. A dense red stroma in combination with the epithelioid aggregates also creates the paisley tie-like pattern in this lesion. Retraction between collagen bundles within the stroma can be seen, helping distinguish this lesion from a morpheiform basal cell carcinoma (BCC), which

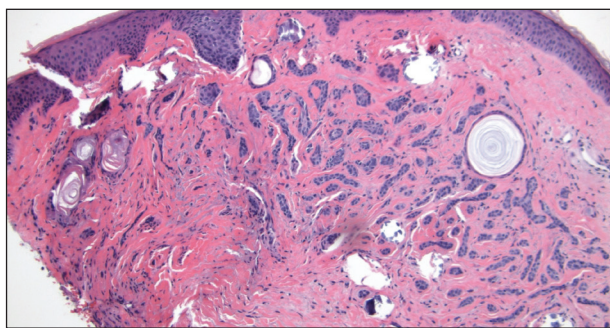


FIGURE 2. Well-circumscribed tumor in the mid dermis with narrow branching cords of compact polygonal cells interspersed within a dense sclerotic stroma characteristic of desmoplastic trichoepithelioma. Numerous keratin horn cysts are present. There is no cytologic atypia or mitotic activity (H&E, original magnification $\times 100$).

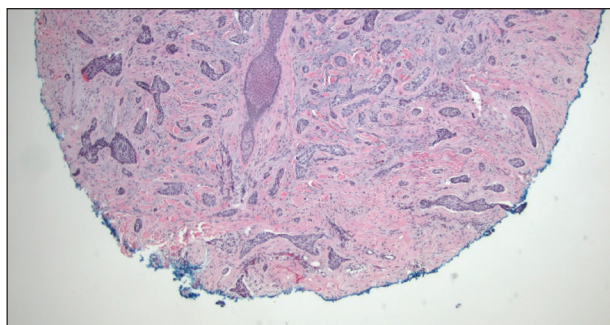


FIGURE 3. Poorly circumscribed, infiltrative tumor with thin elongated strands of basaloid cells within a dense sclerotic stroma characteristic of morpheiform basal cell carcinoma. There is clefting between some epithelial aggregates and adjacent stroma (H&E, original magnification $\times 40$).

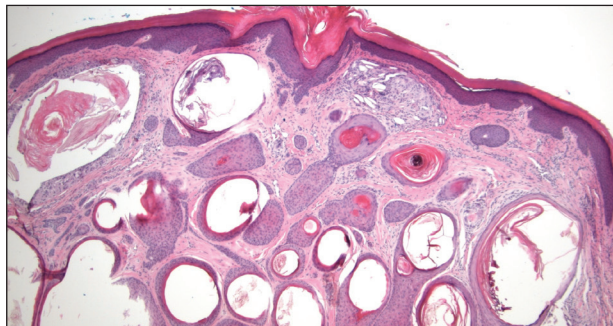


FIGURE 4. Multiple dilated keratin horn cysts lined with cuboidal epithelial cells scattered within a fibroblastic stroma characteristic of trichoadenoma. The epithelial cells contain an eosinophilic or clear cytoplasm without atypia or mitotic activity. There is no attachment to the epidermis (H&E, original magnification $\times 40$).

has retraction between the epithelium and stroma. Immunohistochemistry also can be a useful tool to help differentiate DTEs from morpheaform BCCs in that sparse cytokeratin 20–positive Merkel cells can be seen within the basaloid islands of DTE but not BCC.⁵ Also seen with DTEs are numerous keratin horn cysts that commonly are filled with dystrophic calcifications. Cellular atypia and mitoses are not seen (Figure 2). Compared to MACs, DTEs lack abundant ductal structures and also contain papillary mesenchymal bodies and a more fibroblast-rich stroma.

Morpheaform BCC is an aggressive subtype of BCC. It presents as a scarlike plaque that gradually expands. Thin

infiltrating strands of basaloid cells are seen haphazardly throughout a pink sclerotic stroma. Tadpolelike basaloid islands and rarely horn cysts can be seen scattered superficially, creating the paisley tie–like pattern. This lesion is more infiltrating than a syringoma or a DTE, and perineural invasion is common. Retraction is uncommon, but when present, it is seen between the epithelial cords and adjacent stroma (Figure 3).

Trichoadenoma is another benign neoplasm of follicular differentiation.⁶ It typically presents as a dome-shaped papule or plaque on the head or neck. Histologically it displays numerous dilated cystic spaces that reflect its origin from isthmic and infundibular differentiation. There is no attachment to the overlying epidermis. It can be distinguished from MAC, DTE, and syringoma due to a lack of basaloid aggregates and only a small number of non–cyst-forming epithelial cells (Figure 4).

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