

Solitary Pink Plaque on the Neck

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A 17-year-old adolescent girl presented with a solitary, 8-cm, pink plaque on the anterior aspect of the neck of 5 years' duration. No similar skin findings were present elsewhere on the body. The rash was not painful or pruritic, and she denied prior trauma to the site. The patient previously had tried a salicylic acid bodywash as well as mupirocin cream 2% and mometasone ointment with no improvement. Her medical history was unremarkable, and she had no known allergies. There was no family history of a similar rash. Physical examination revealed no palpable subcutaneous lumps or masses and no lymphadenopathy of the head or neck. An incisional biopsy was performed.

WHAT'S YOUR DIAGNOSIS?

- desmoplastic trichoepithelioma
- inflammatory linear verrucous epidermal nevus
- microcystic adnexal carcinoma
- morpheaform basal cell carcinoma
- plaque-type syringoma

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The authors report no conflict of interest.

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THE DIAGNOSIS: Plaque-type Syringoma

A biopsy demonstrated multiple basaloid islands of tumor cells in the reticular dermis with ductal differentiation, some with a commalike tail. The ducts were lined by 2 to 3 layers of small uniform cuboidal cells without atypia and contained inspissated secretions within the lumina of scattered ducts. There was an associated fibrotic collagenous stroma. There was no evidence of perineural invasion and no deep dermal or subcutaneous extension (Figure 1). Additional cytokeratin immunohistochemical staining highlighted the adnexal proliferation (Figure 2). A diagnosis of plaque-type syringoma (PTS) was made.

Syringomas are benign dermal sweat gland tumors that typically present as flesh-colored papules on the cheeks or periorbital area of young females. Plaque-type tumors as well as papulonodular, eruptive, disseminated, urticaria pigmentosa-like, lichen planus-like, or milium-like syringomas also have been reported. Syringomas may be associated with certain medical conditions such as Down syndrome, Nicolau-Balus syndrome, and both scarring and non-scarring alopecias.¹ The clear cell variant of syringoma often is associated with diabetes mellitus.² Kikuchi et al³ first described PTS in 1979. Plaque-type syringomas rarely are reported in the literature, and sites of involvement include the head and neck region, upper lip, chest, upper extremities, vulva, penis, and scrotum.⁴⁻⁶

Histologically, syringomatous lesions are composed of multiple small ducts lined by 2 to 3 layers of cuboidal epithelium. The ducts may be arranged in nests or strands of basaloid cells surrounded by a dense fibrotic stroma. Occasionally, the ducts will form a comma- or teardrop-shaped tail; however, this also may be observed in desmoplastic trichoepithelioma (DTE).⁷ Perineural invasion is absent in syringomas. Syringomas exhibit a lateral growth pattern that typically is limited to the upper half of the reticular dermis and spares the underlying subcutis, muscle, and bone. The growth pattern may be discontinuous with proliferations juxtaposed by normal-appearing skin.⁸ Syringomas usually express progesterone receptors and are known to proliferate at puberty, suggesting that these neoplasms are under hormonal control.⁹ Although syringomas are benign, various treatment options that may be pursued for cosmetic purposes include radiofrequency, staged excision, laser ablation, and oral isotretinoin.^{8,10} If only a superficial biopsy is obtained, syringomas may display features of other adnexal neoplasms, including microcystic adnexal carcinoma (MAC), DTE, morpheiform basal cell carcinoma (BCC), and inflammatory linear verrucous epidermal nevus (ILVEN).

Microcystic adnexal carcinoma is a locally aggressive neoplasm first described by Goldstein et al¹¹ in 1982

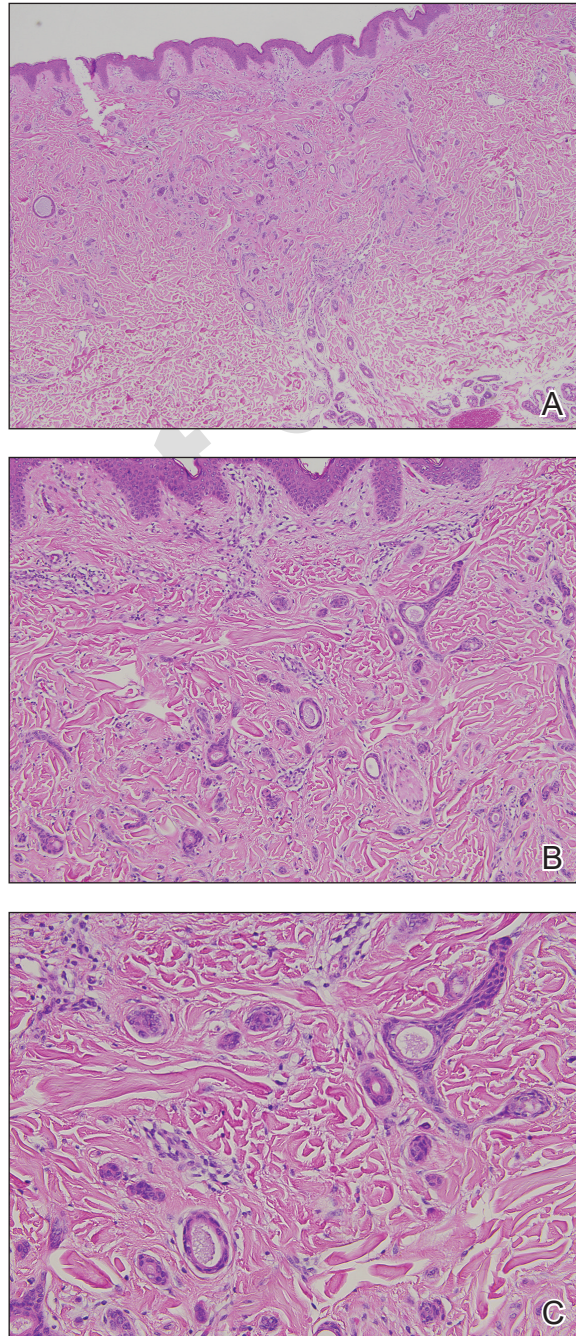


FIGURE 1. A–C, Histopathology demonstrated multiple basaloid islands of tumor cells in the reticular dermis with ductal differentiation, some with a comma-like tail. The ducts were lined by 2 to 3 layers of small uniform cuboidal cells without atypia and contained inspissated secretions within the lumina of scattered ducts with an associated fibrotic collagenous stroma (H&E, original magnifications $\times 40$, $\times 100$, and $\times 200$, respectively).

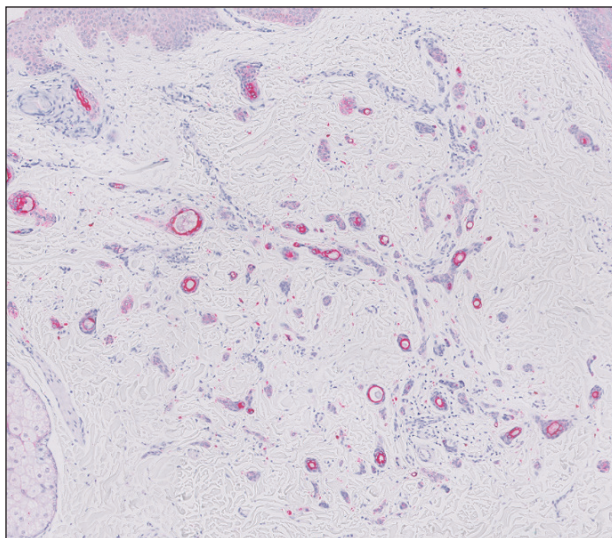


FIGURE 2. AE1/AE3 cyokeratin immunohistochemical staining highlighted the adnexal proliferation (original magnification $\times 50$).

that most commonly presents in middle-aged adults as an indurated, ill-defined plaque or nodule on the face with a predilection for the upper and lower lip. Prior radiation therapy and immunosuppression are risk factors for the development of MAC.¹² Histologically, the superficial portion displays small cornifying cysts interspersed with islands of basaloid cells and may mimic a syringoma. However, the deeper portions demonstrate ducts lined by a single layer of cells with a background of hyalinized and sclerotic stroma. The tumor cells may occupy the deep dermis and underlying subcutis, muscle, or bone and demonstrate an infiltrative growth pattern and perineural invasion. Treatment includes Mohs micrographic surgery.

Desmoplastic trichoepitheliomas most commonly present as solitary white to yellowish annular papules or plaques with a central dell located on sun-exposed areas of the face, cheeks, or chin. This benign neoplasm has a bimodal age distribution, primarily affecting females either in childhood or adulthood.¹³ Histologically, strands and nests of basaloid epithelial cells proliferate in a dense eosinophilic desmoplastic stroma. The basaloid islands are narrow and cordlike with growth parallel to the surface epidermis and do not dive deeply into the deep dermis or subcutis. Ductal differentiation with associated secretions typically is not seen in DTE.¹ Calcifications and foreign body granulomatous infiltrates may be present. Merkel cells also are present in this tumor and may be highlighted by immunohistochemistry with cyokeratin 20.¹⁴ Rarely, desmoplastic trichoepitheliomas may transform into trichoblastic carcinomas. Treatment may consist of surgical excision or Mohs micrographic surgery.

Morpheaform BCC also is included in the clinical and histopathologic differential diagnosis of infiltrative basaloid neoplasms. It is one of the more aggressive

variants of BCC. The use of immunohistochemical staining may aid in differentiating between these sclerosing adnexal neoplasms.¹⁵ For example, pleckstrin homology-like domain family A member 1 (PHLDA1) is a stem cell marker that is heavily expressed in DTE as a specific follicular bulge marker but is not present in a morpheaform BCC. This highlights the follicular nature of DTEs at the molecular level. BerEP4 is a monoclonal antibody that serves as an epithelial marker for 2 glycopolypeptides: 34 and 39 kDa. This antibody may demonstrate positivity in morpheaform BCC but does not stain cells of interest in MAC.

Inflammatory linear verrucous epidermal nevus clinically presents with erythematous and warty papules in a linear distribution following the Blaschko lines. The papules often are reported to be intensely pruritic and usually are localized to one extremity.¹⁶ Although adult-onset forms of ILVEN have been described,¹⁷ it most commonly is diagnosed in young children. Histologically, ILVEN consists of psoriasiform epidermal hyperplasia with alternating areas of parakeratosis and orthokeratosis with underlying agranulosis and hypergranulosis, respectively.¹⁸ The upper dermis contains a perivascular lymphocytic infiltrate. Treatment with laser therapy and surgical excision has led to both symptomatic and clinical improvement of ILVEN.¹⁶

Plaque-type syringomas are a rare variant of syringomas that clinically may mimic other common inflammatory and neoplastic conditions. An adequate biopsy is imperative to differentiate between adnexal neoplasms, as a small superficial biopsy of a syringoma may demonstrate features observed in other malignant or locally aggressive neoplasms. In our patient, the small ducts lined by cuboidal epithelium with no cellular atypia and no deep dermal growth or perineural invasion allowed for the diagnosis of PTS. Therapeutic options were reviewed with our patient, including oral isotretinoin, laser therapy, and staged excision. Ultimately, our patient elected not to pursue treatment, and she is being monitored clinically for any changes in appearance or symptoms.

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