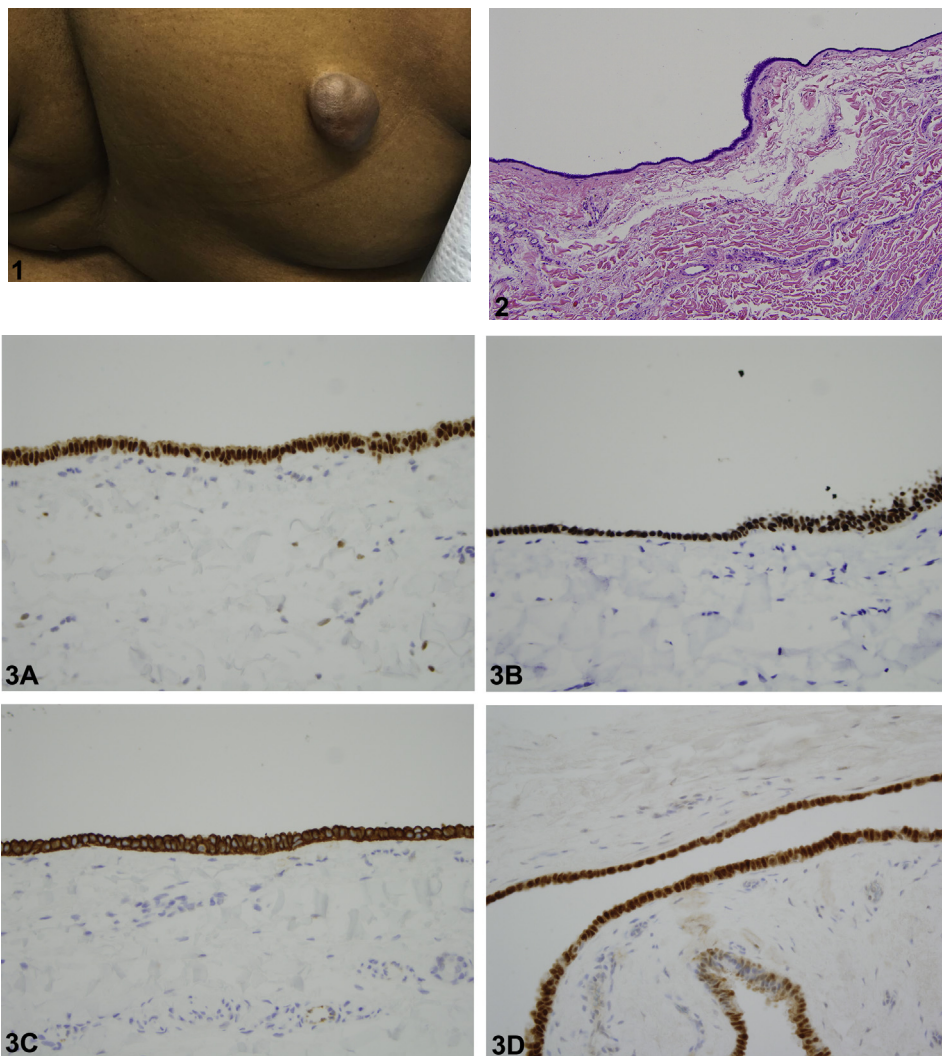


A subcutaneous nodule on the mid back

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A 37-year-old African-American woman presented for evaluation of an enlarging growth on her back that had been present for years but not since childhood. She denied previous trauma to the region. Physical examination found a single 3.5-cm soft but tense painless nodule on the right mid back, just above the bra line (Fig 1). During excisional biopsy, the nodule was punctured and clear fluid was expressed. Histopathologic (Fig 2) and immunohistochemical (Fig 3) staining was performed on the biopsy.

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Question 1: What is the most likely clinical diagnosis?

- A. Epidermal cyst
- B. Cutaneous dermoid cyst
- C. Cutaneous Müllerian cyst
- D. Apocrine hidrocystoma
- E. Hidradenoma papilliferum

Answers:

A. Epidermal cyst—Incorrect. An epidermal cyst is a flesh-colored to yellowish, fluctuant cystic nodule with an overlying punctum representing the follicle from which the cyst develops.¹ They commonly present on the trunk, but these cysts contain keratin debris rather than clear fluid as seen in this patient.¹

B. Cutaneous dermoid cyst—Incorrect. A cutaneous dermoid cyst is a firm, cystic nodule that generally presents in the first 5 years of life and can persist into adulthood. Dermoid cysts are the result of incomplete closure of the embryonic zones.¹ They commonly present on the nasal root or lateral end of the eyebrow.¹ Dermoid cysts can present on the trunk as well. These lesions contain other cutaneous appendages such as hair, sebaceous lobules, and eccrine glands.²

C. Cutaneous Müllerian cyst—Correct. Cutaneous Müllerian cysts are rare, benign, solitary lesions that typically occur on the lower extremities of young women. Rarely, these cysts may present in unusual locations such as the back. They are theorized to develop from heterotopic fimbrial type Müllerian epithelium that form a cyst under the influence of ovarian hormones after puberty.³ Similar cysts occurring in males have been reported, but these are recognized as a distinct subtype of cutaneous ciliated cysts theorized to be of eccrine origin.⁴

D. Apocrine hidrocystoma—Incorrect. Apocrine hidrocystomas are usually solitary lesions with a cystic cavity containing clear fluid and are more common in women. However, most hidrocystomas are smaller, about 1 to 3 mm in size.¹ They generally present around the eyelids but have also been reported on the face and scalp as well.¹

E. Hidradenoma papilliferum—Incorrect. Hidradenoma papilliferum is a benign adenoma arising in the vulvar and perianal areas. Ectopic locations have been reported.² The nodule itself is firm and

not fluid filled despite pathology results showing cyst-like spaces with villi.¹

Question 2: What would you expect to find on histology?

A. A cystic wall lined by pseudostratified, ciliated, columnar epithelium containing goblet cells, seromucinous glands, and underlying fibromuscular connective tissue or cartilage.

B. A cystic wall lined by stratified squamous epithelium with lymphoid follicles and absent smooth muscle.

C. A cystic wall lined by cuboidal, columnar, or stratified squamous epithelium with thyroid follicles.

D. A cystic wall lined by a ciliated cuboidal to columnar epithelium with psuedostratified areas and lacking mucin-secreting cells.

E. A cystic cavity filled with laminated keratin lined by a stratified squamous epithelium that includes a granular layer.

Answers:

A. A cystic wall lined by pseudostratified, ciliated, columnar epithelium containing goblet cells, seromucinous glands, and underlying fibromuscular connective tissue or cartilage—Incorrect. These histologic findings are seen in bronchogenic cysts.^{1,2}

B. A cystic wall lined by stratified squamous epithelium with lymphoid follicles and absent smooth muscle—Incorrect. These histologic findings are seen in branchial cleft cysts.^{1,2}

C. A cystic wall lined by cuboidal, columnar, or stratified squamous epithelium with thyroid follicles—Incorrect. These histologic findings are seen in thyroglossal duct cysts.²

D. A cystic wall lined by a ciliated cuboidal to columnar epithelium with psuedostratified areas and lacking mucin-secreting cells—Correct. These histological findings are seen in cutaneous Müllerian cysts. Histology analysis of our patient's biopsy found an extensive multiloculated cyst lined by benign glandular epithelium. The epithelium was primarily one layer in thickness and ciliated. Histologically, cutaneous Müllerian cysts closely resemble tissue found in the fallopian tube; they are believed to originate from Müllerian duct structures (Fig 2).^{1,4}

E. A cystic cavity filled with laminated keratin lined by a stratified squamous epithelium that

includes a granular layer—Incorrect. These histologic findings are seen in epidermal cysts.²

Question 3: Which of the following immunohistochemistry staining profiles is typically characteristic of this lesion?

- A.** Estrogen⁺/progesterone⁺/cytokeratin 7⁺/PAX-8⁺/CEA[−]/P63[−]
- B.** Estrogen[−]/progesterone[−]/cytokeratin 7⁺/PAX-8⁺/CEA[−]/P63[−]
- C.** Estrogen⁺/progesterone⁺/cytokeratin 7⁺/PAX-8[−]/CEA[−]/P63[−]
- D.** Estrogen⁺/progesterone⁺/cytokeratin 7⁺/PAX-8⁺/CEA⁺/P63[−]
- E.** Estrogen⁺/progesterone⁺/cytokeratin 7⁺/PAX-8⁺/CEA⁺/P63⁺

Answers:

- A.** Estrogen⁺/progesterone⁺/cytokeratin 7⁺/PAX-8⁺/CEA[−]/P63[−] — Correct. Müllerian-derived ciliated cutaneous cysts stain positive for estrogen and progesterone, consistent with fallopian tube epithelium. These cysts also stain strongly for cytokeratin 7 (Fig 3, A-C).^{4,5}
- B.** Estrogen[−]/progesterone[−]/cytokeratin 7⁺/PAX-8⁺/CEA[−]/P63[−] — Incorrect. Negative estrogen and progesterone staining is observed in ciliated cutaneous eccrine cysts, a distinct variant of ciliated cutaneous cysts thought to originate from ciliated metaplasia of eccrine or apocrine glands.⁴

C. Estrogen⁺/progesterone⁺/cytokeratin 7⁺/PAX-8[−]/CEA[−]/P63[−] — Incorrect. Paired-box gene 8 (PAX-8) is a transcription factor involved in the development of Müllerian-derived organs. Cutaneous Müllerian cysts stain positive for PAX-8 (Fig 3, D).⁵

D. Estrogen⁺/progesterone⁺/cytokeratin 7⁺/PAX-8⁺/CEA⁺/P63[−] — Incorrect. Carcinoembryonic antigen is expressed in the cells of eccrine sweat glands. Positive carcinoembryonic antigen staining is typically associated with ciliated cutaneous cysts of eccrine origin.⁴

E. Estrogen⁺/progesterone⁺/cytokeratin 7⁺/PAX-8⁺/CEA⁺/P63⁺ — Incorrect. Positive p63 staining is seen in ciliated cutaneous cysts of eccrine origin while lack of p63 staining is associated with cutaneous Müllerian cysts.⁴

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