CUTANEOUS CYSTS AND RELATED CONDITIONS

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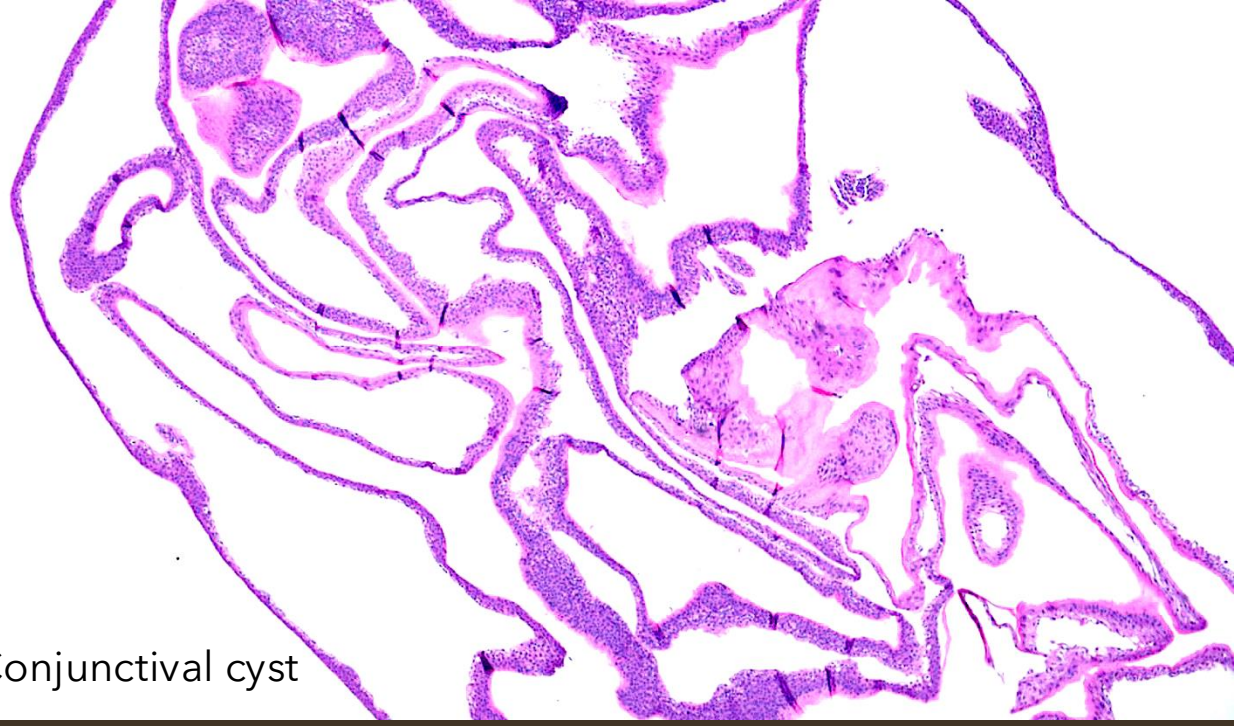


Conjunctival cyst

Introduction: Learning Tips and Pitfalls

- One of the most common specimens

  What is a cyst?

  A cyst: <u>any cavity lined by an epithelium</u> (native to that organ)

- How are cysts classified?

  According to the nature of the epithelial lining

- Recognize the histology of epithelial lining, especially adnexal structures

- Overlapping classification schemes

  Appendageal origin i.e., hair, apocrine, eccrine, or salivary glands

  Derivation i.e., either developmental or non-developmental

- Pitfalls (not true cysts):

  Nodular tumors may deteriorate (cystic degeneration)

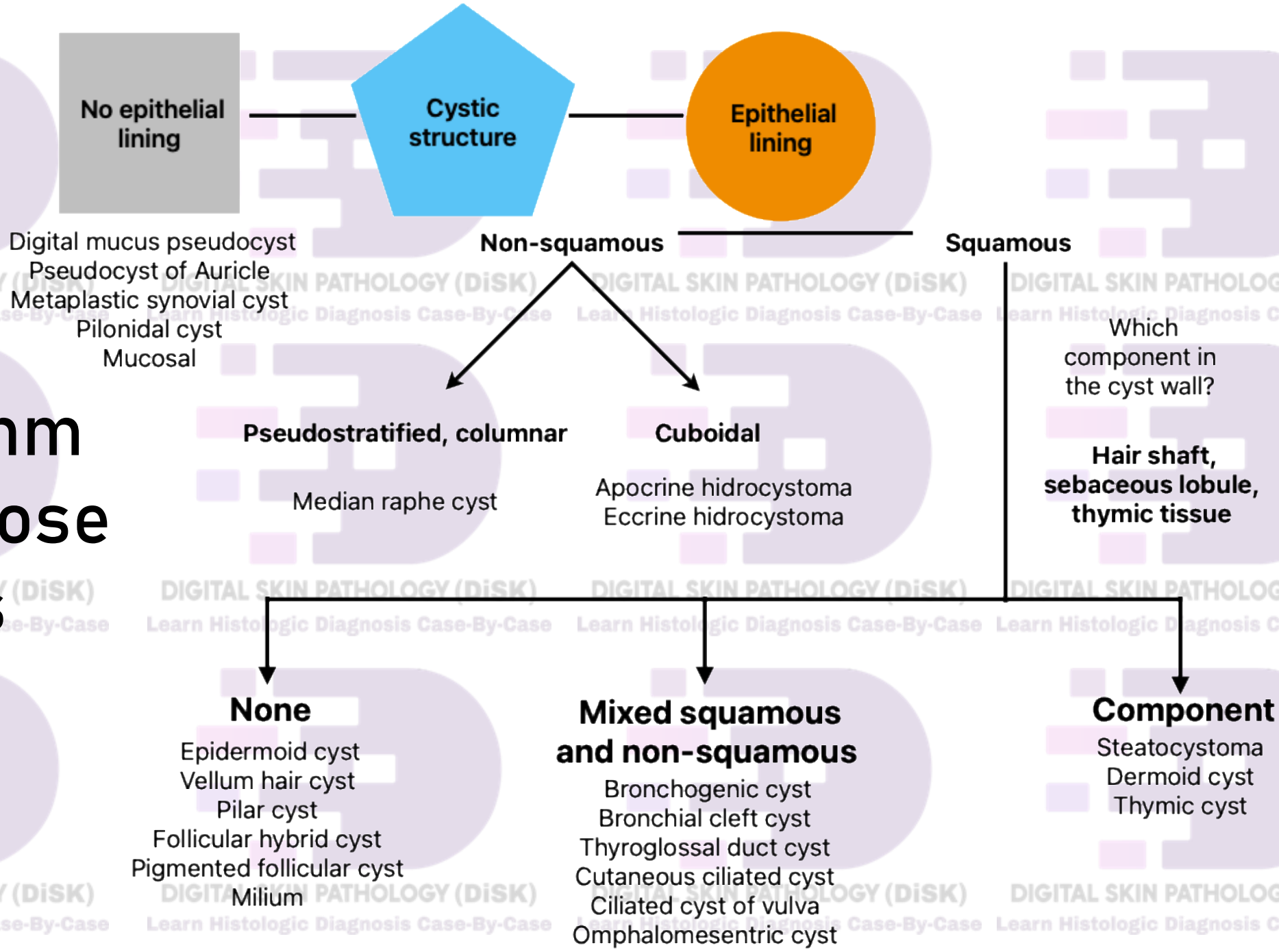
  Empty space (e.g., mucin) or cracking (histologic artifact)

  Scarring and granulomatous reaction (no cyst lining): secondary changes of ruptured cyst

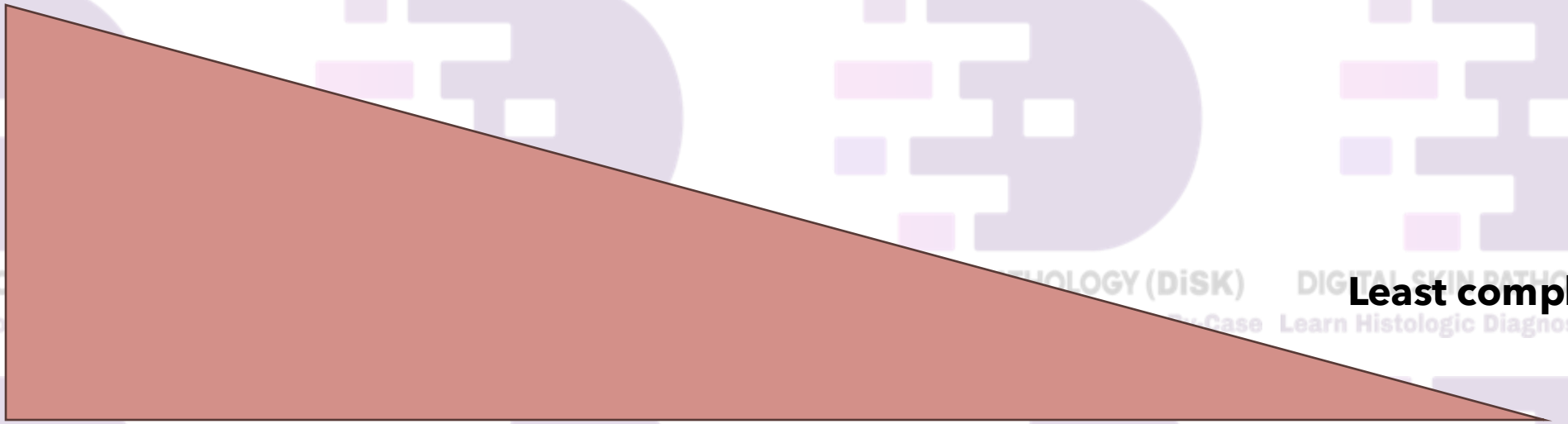
How are cutaneous cysts broadly classified? (types of cyst lining)

- Squamous lining (follicular cysts)
- Glandular lining (glandular cysts)
- Pseudocysts

Algorithm to diagnose cysts



Varying histopathologic complexity in cutaneous cysts



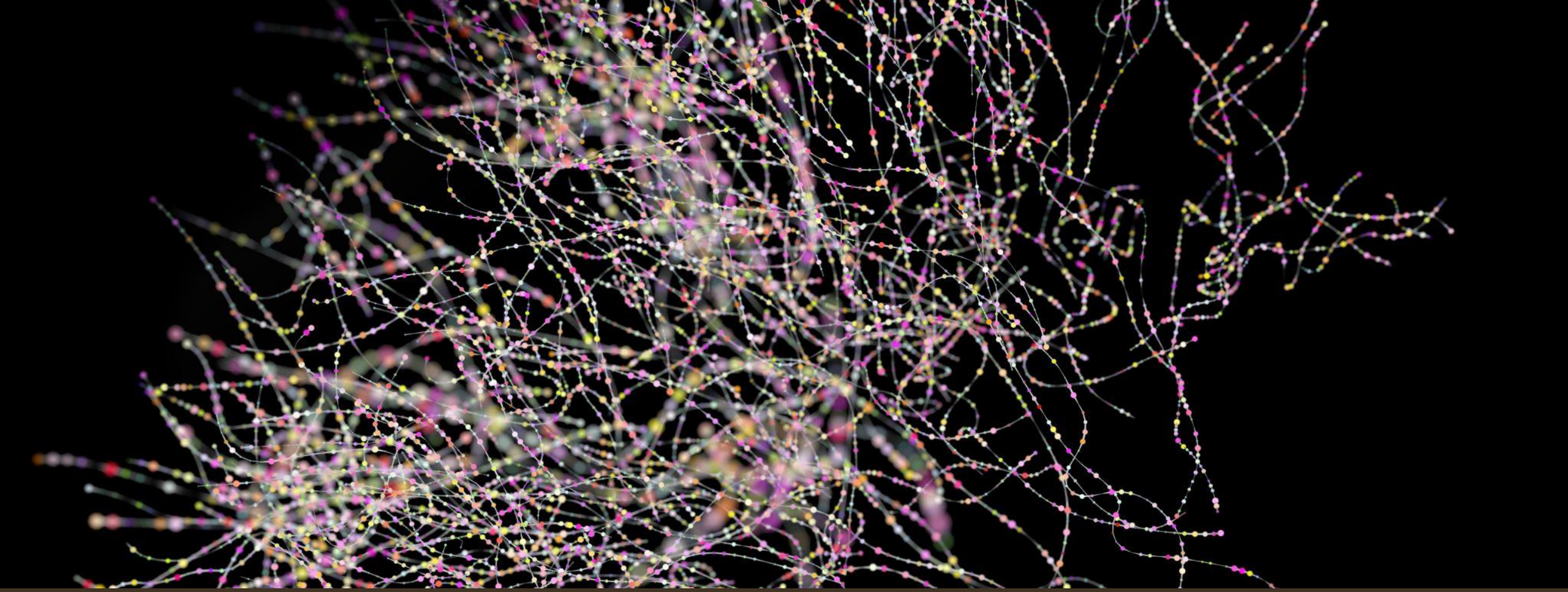
Most complex

Least complex

Proliferating pilar cyst/tumor
Steatocystoma
Dermoid cyst

Mixed squamous
& nonsquamous
ciliated cysts

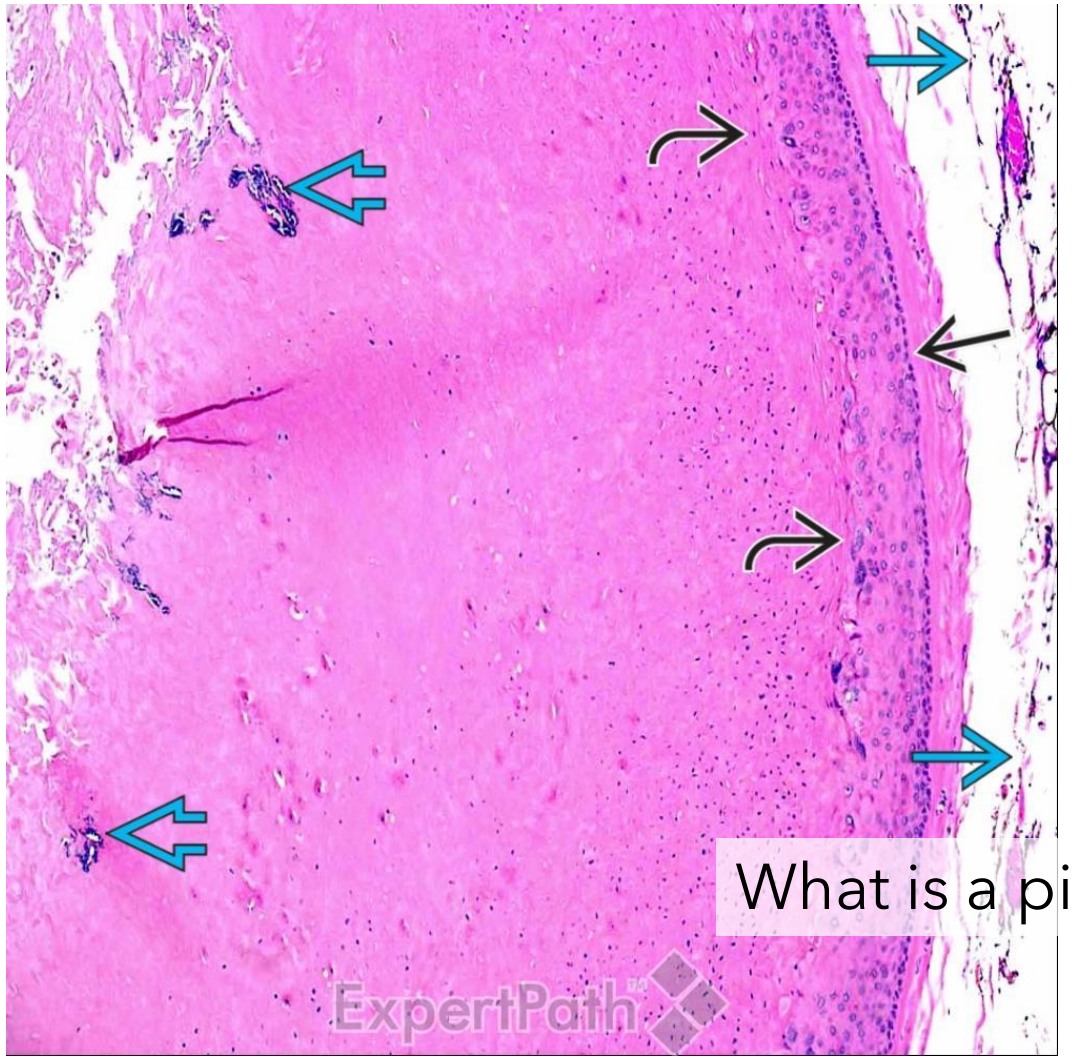
Epidermal cyst
Pilar cyst
Vellus hair cyst



CYSTIC SPACE WITH EPITHELIAL LINING

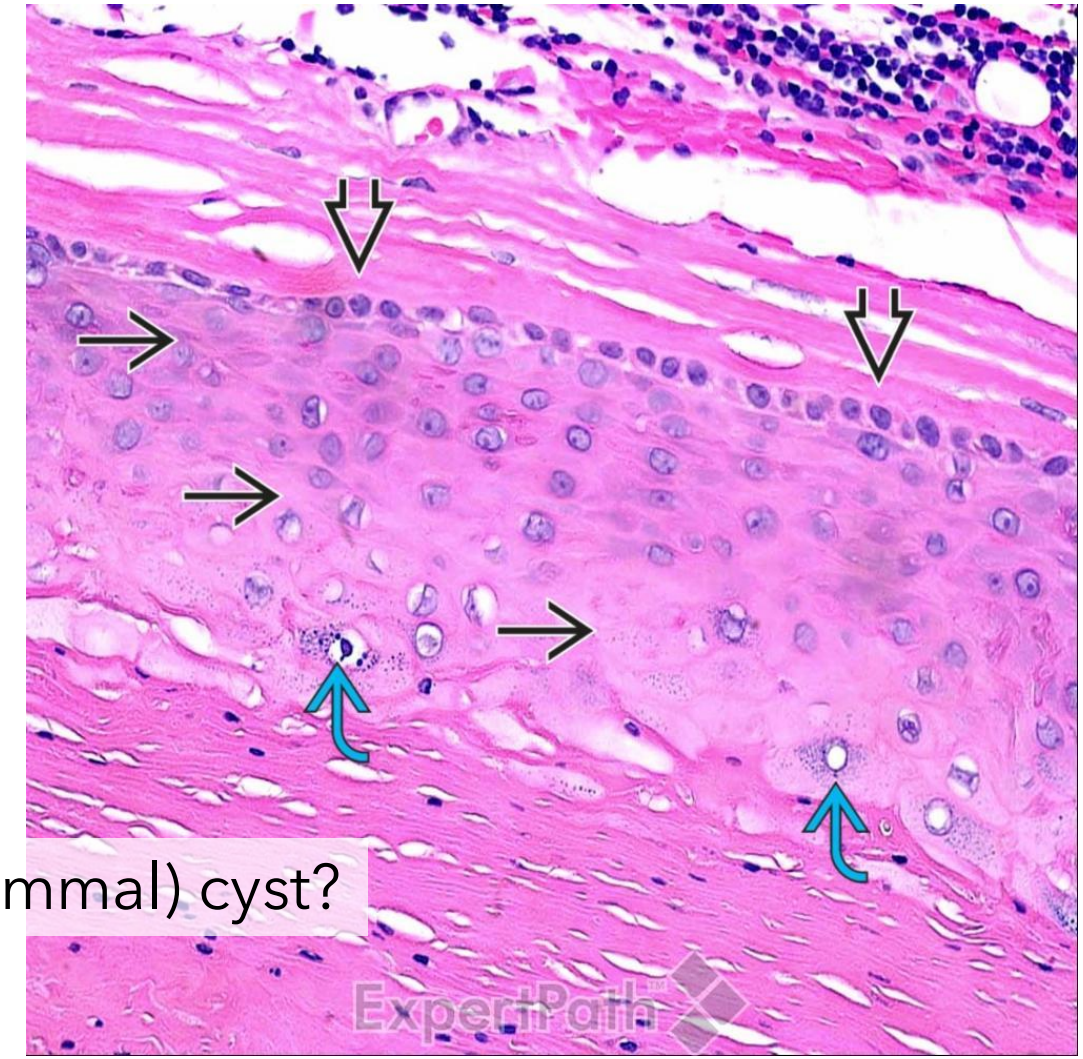
Epidermal inclusion cysts often contain a surface punctum or connection to the surface (black open arrow). The cysts contain laminated keratin or keratin debris.

Epidermoid cyst is lined by keratinized squamous epithelium (black open arrow). The cyst wall is composed of fibrous stroma without adnexal structures. The lumen of the cyst contains keratin debris (black solid arrow). The presence of a granular layer (cyan solid arrow) helps differentiate this cyst from other cystic lesions.



The border of the cyst (black curved arrow) shows a smooth outer cell layer (black solid arrow) at the dermal-subcutaneous junction (cyan solid arrow). Basophilic granular flecks of focal calcification (dark blue) (cyan open arrow) can be seen within the cyst contents.

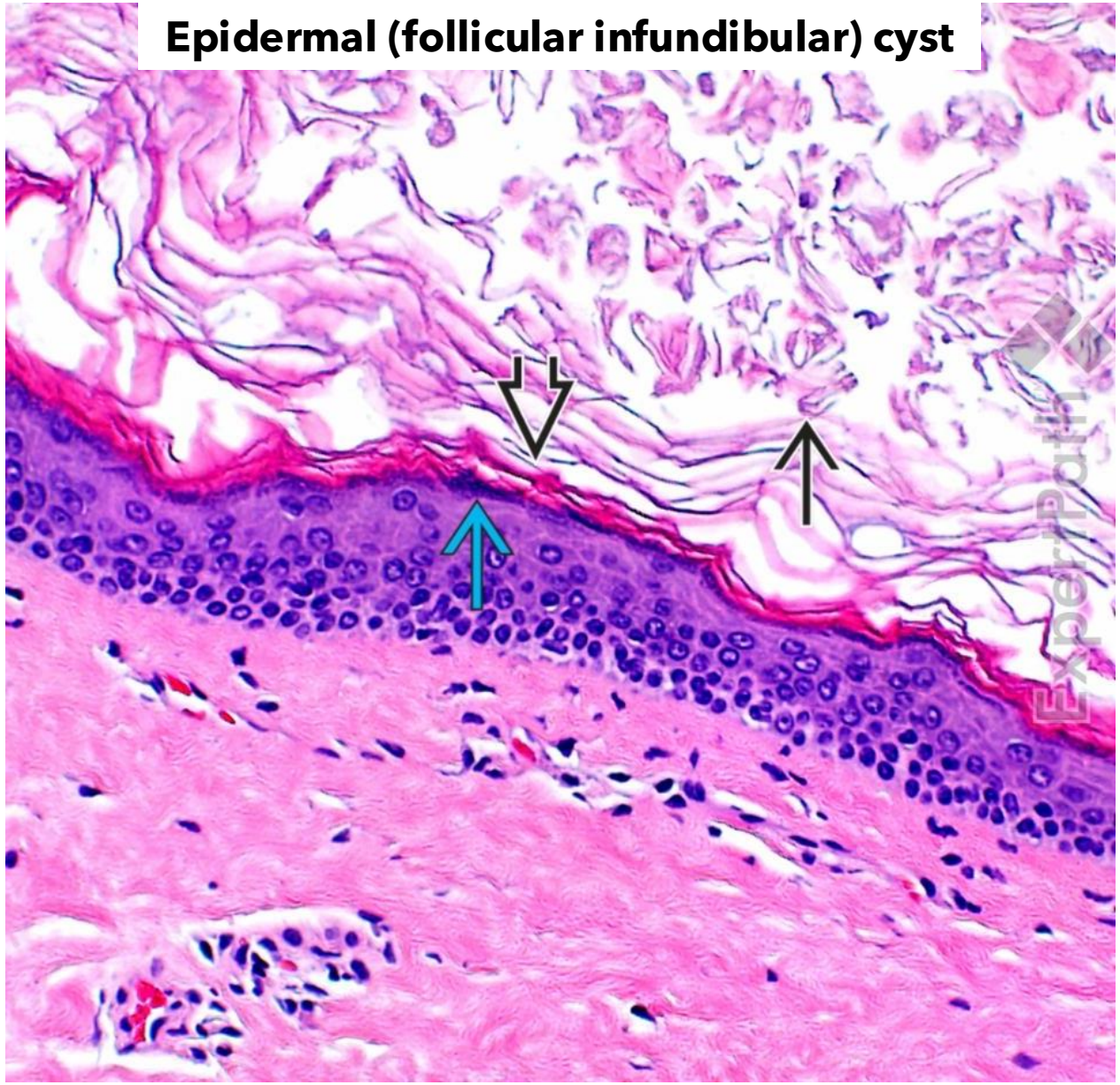
What is a pilar (trichilemmal) cyst?

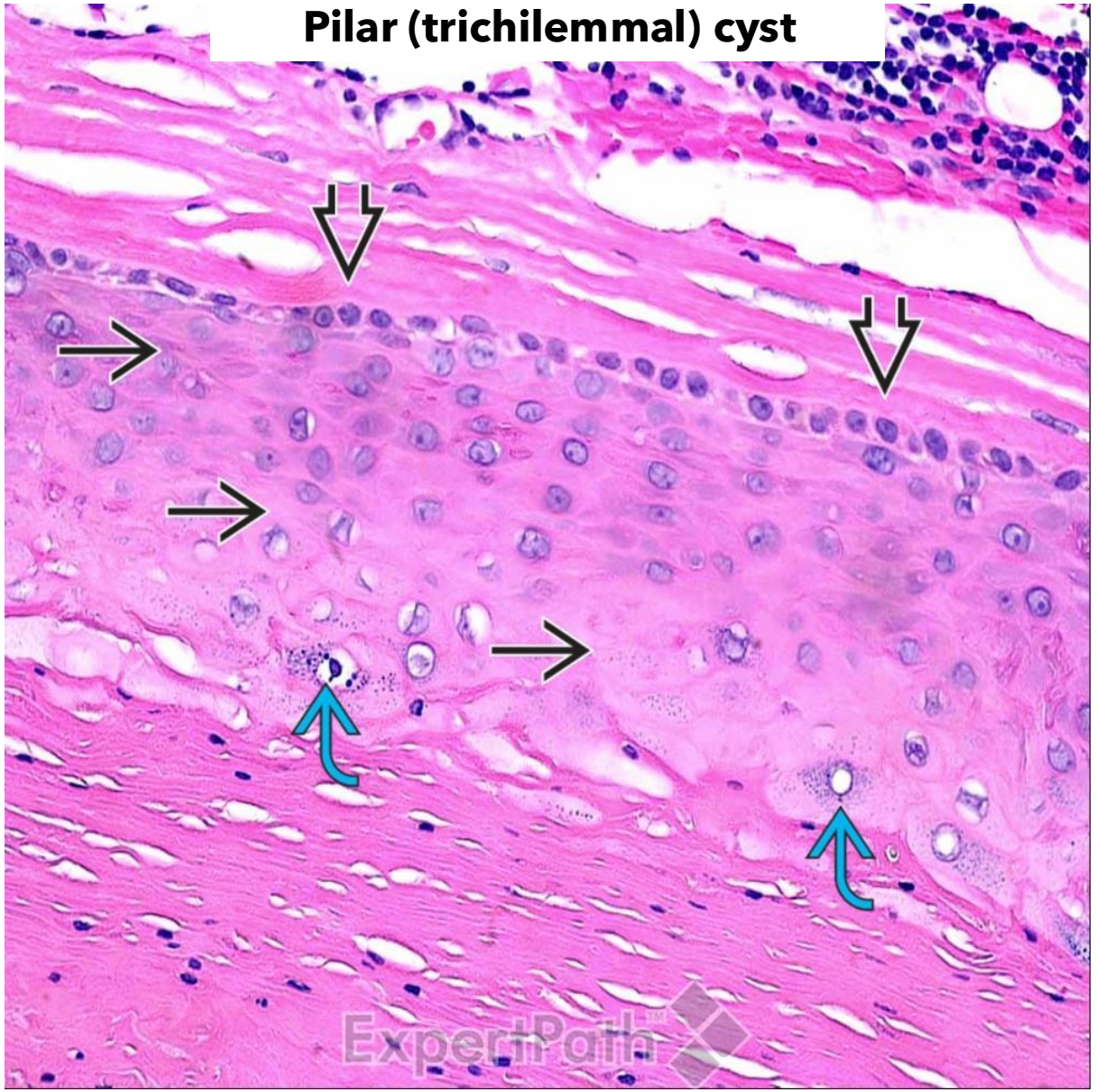


The cyst wall resembles the outer root sheath of the follicle at the level of the isthmus. The wall is composed of several layers of squamous epithelium (black solid arrow) with eosinophilic-staining cytoplasm and a peripheral basal layer (black open arrow). There is no granular layer, and some cells contain keratohyalin granules (cyan curved arrow).

How to differentiate an epidermal cyst from a pilar cyst?

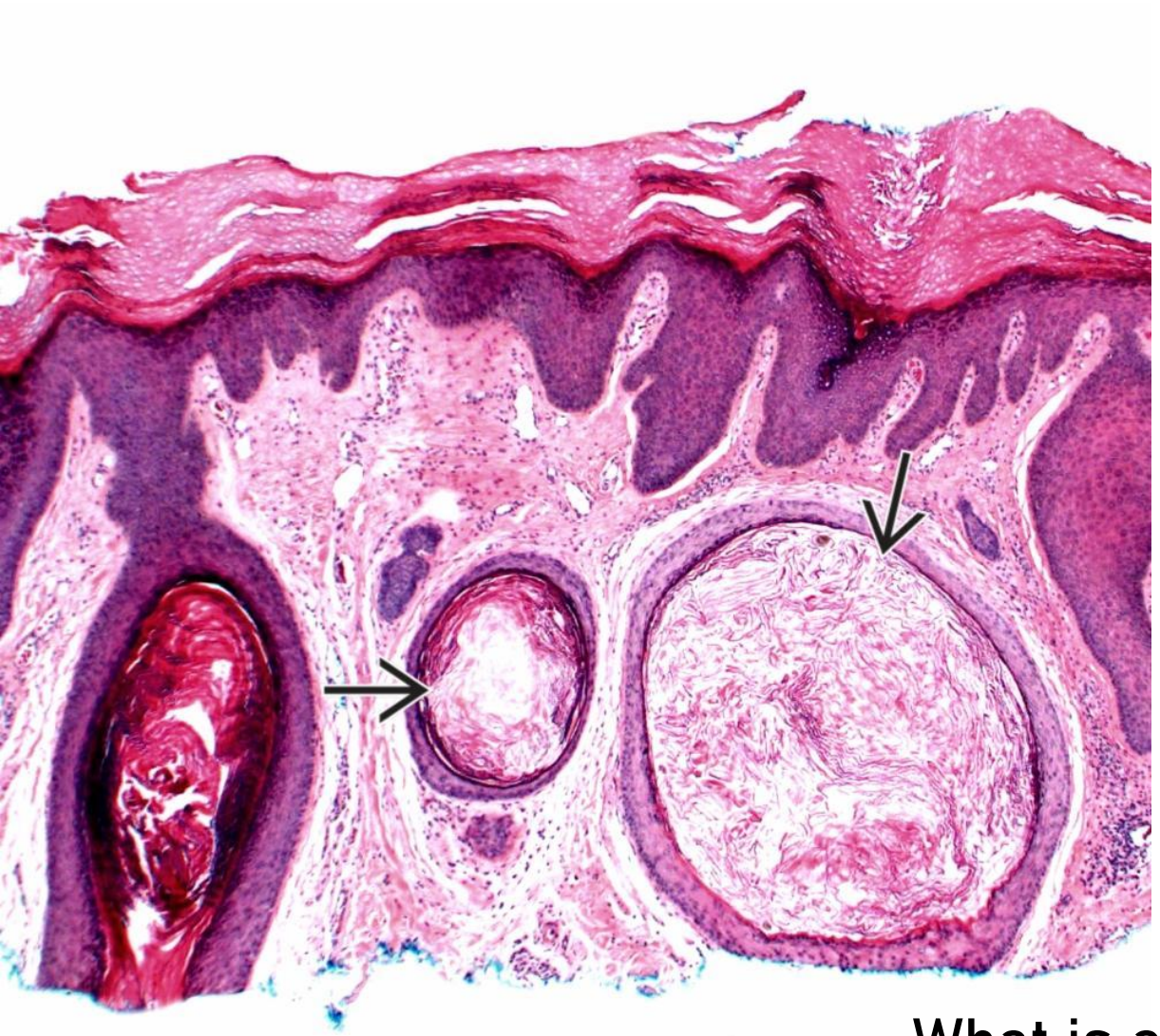
Histopathologic features	Epidermal	Pilar
Granular cell layer	Present	Absent
Keratin quality	Loose, flaky	Compact, homogenous
Palisading of nuclei in basal layer	Absent	Present
Resemblance to which structure	Infundibulum	Isthmus



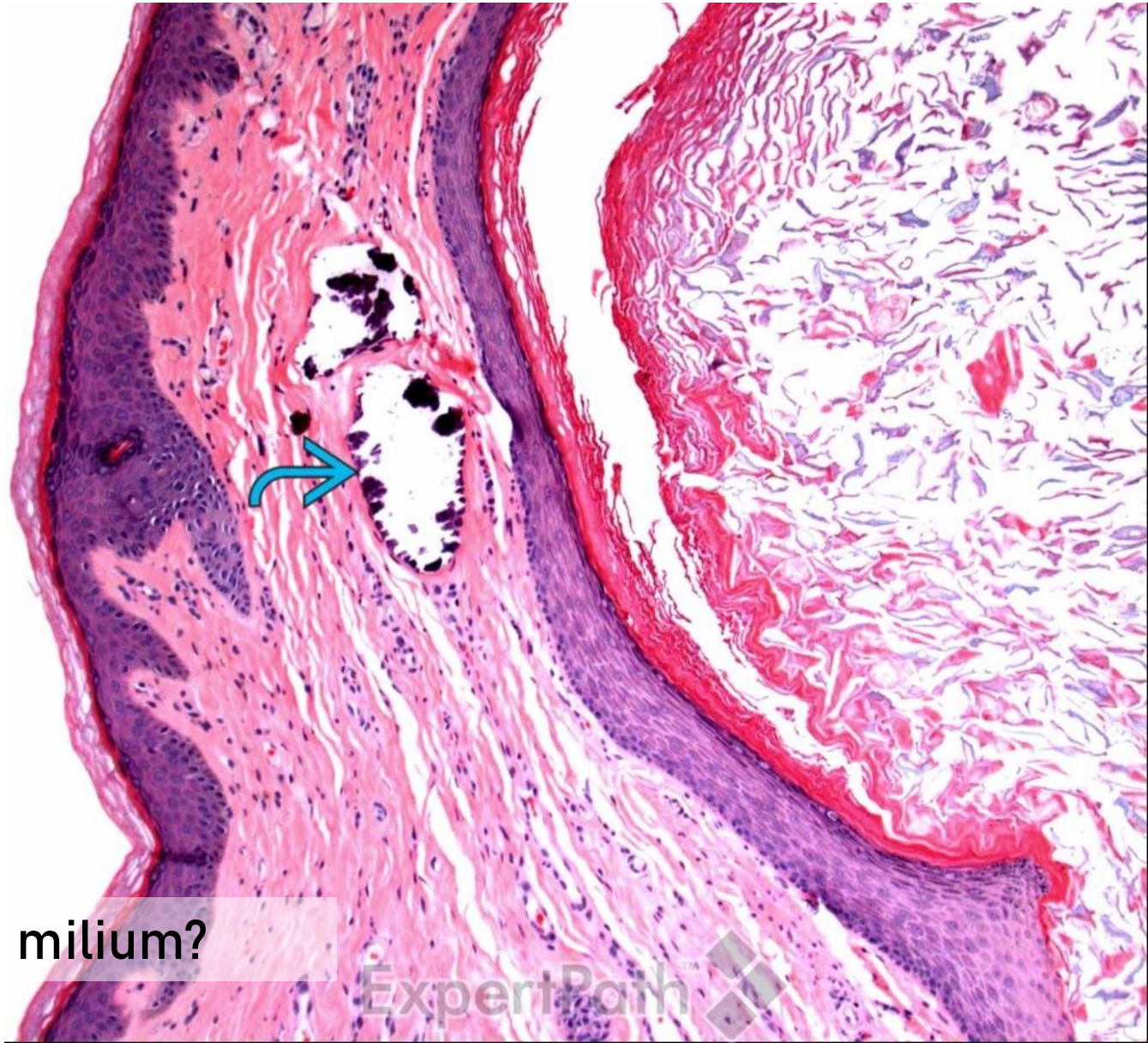


What is the difference between epidermal and pilar cysts?

Low magnification shows multiple small dermal cysts (black solid arrow), which appear essentially identical to miniature epidermoid/epidermal inclusion (follicular, infundibular-type) cysts with loose keratin debris and a preserved granular layer.

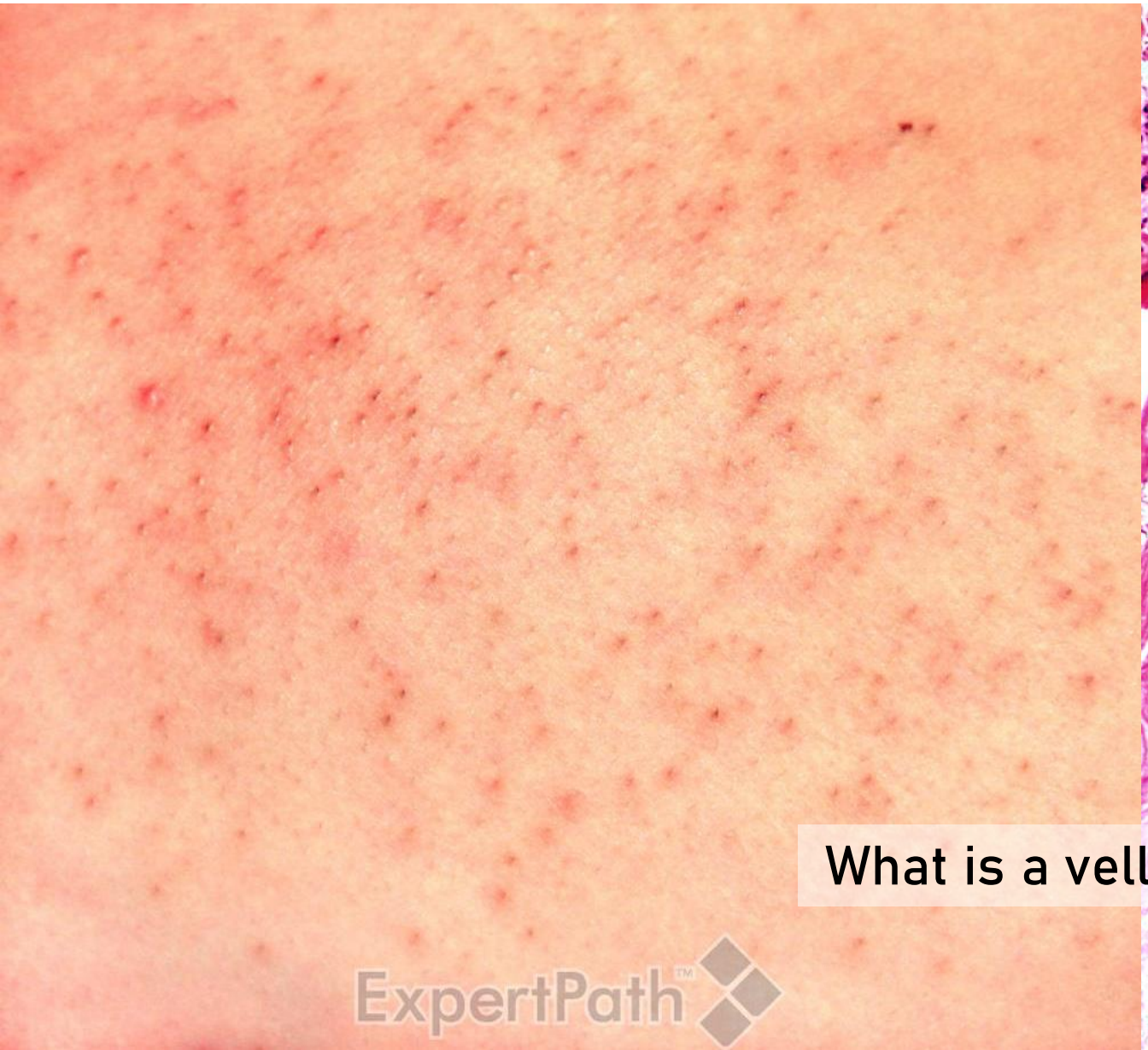


Another example of a milia cyst with adjacent stromal calcifications (cyan curved arrow) and mild fibrosis, changes likely due to previous rupture of the cyst, is shown.

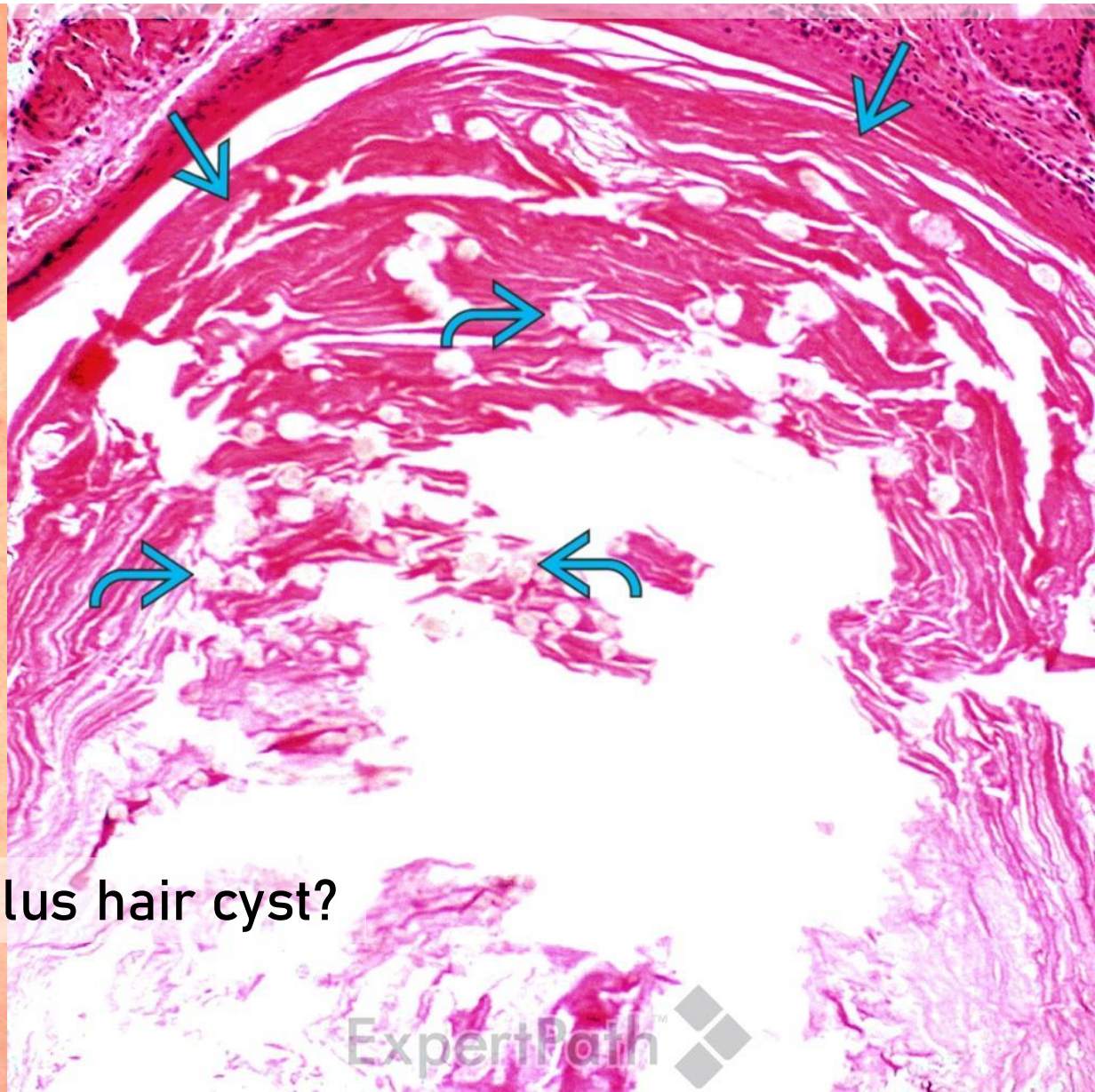


What is a milium?

Multiple slightly brownish, small follicular papules are seen on the chest. This is a classic presentation of **eruptive vellus hair cysts**. (Courtesy J. Wu, MD.)

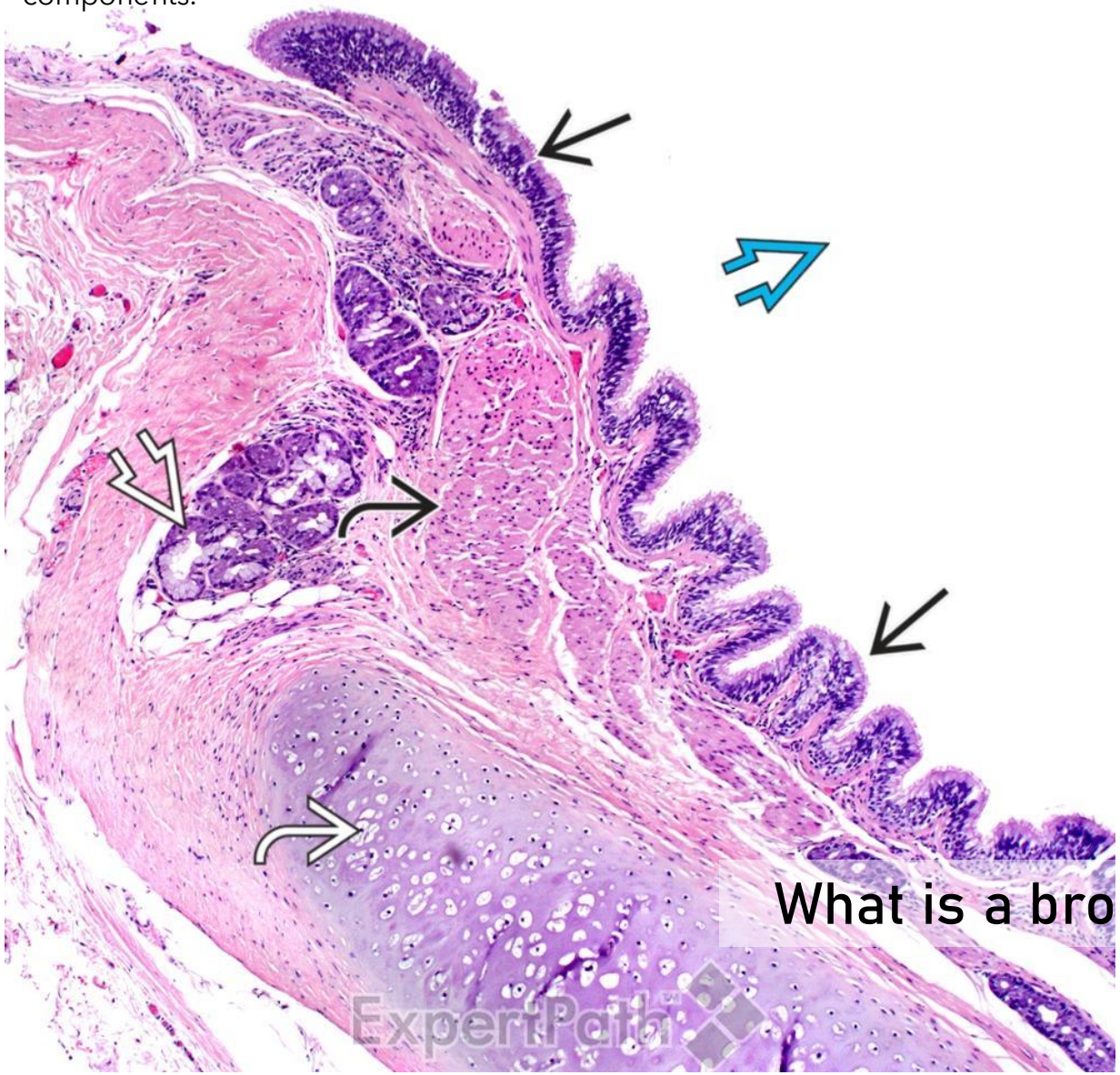


Low magnification shows a cyst lined by a keratinizing squamous epithelium. The cyst contents include laminated keratin (cyan solid arrow) and numerous transversely sectioned hair shafts (cyan curved arrow).

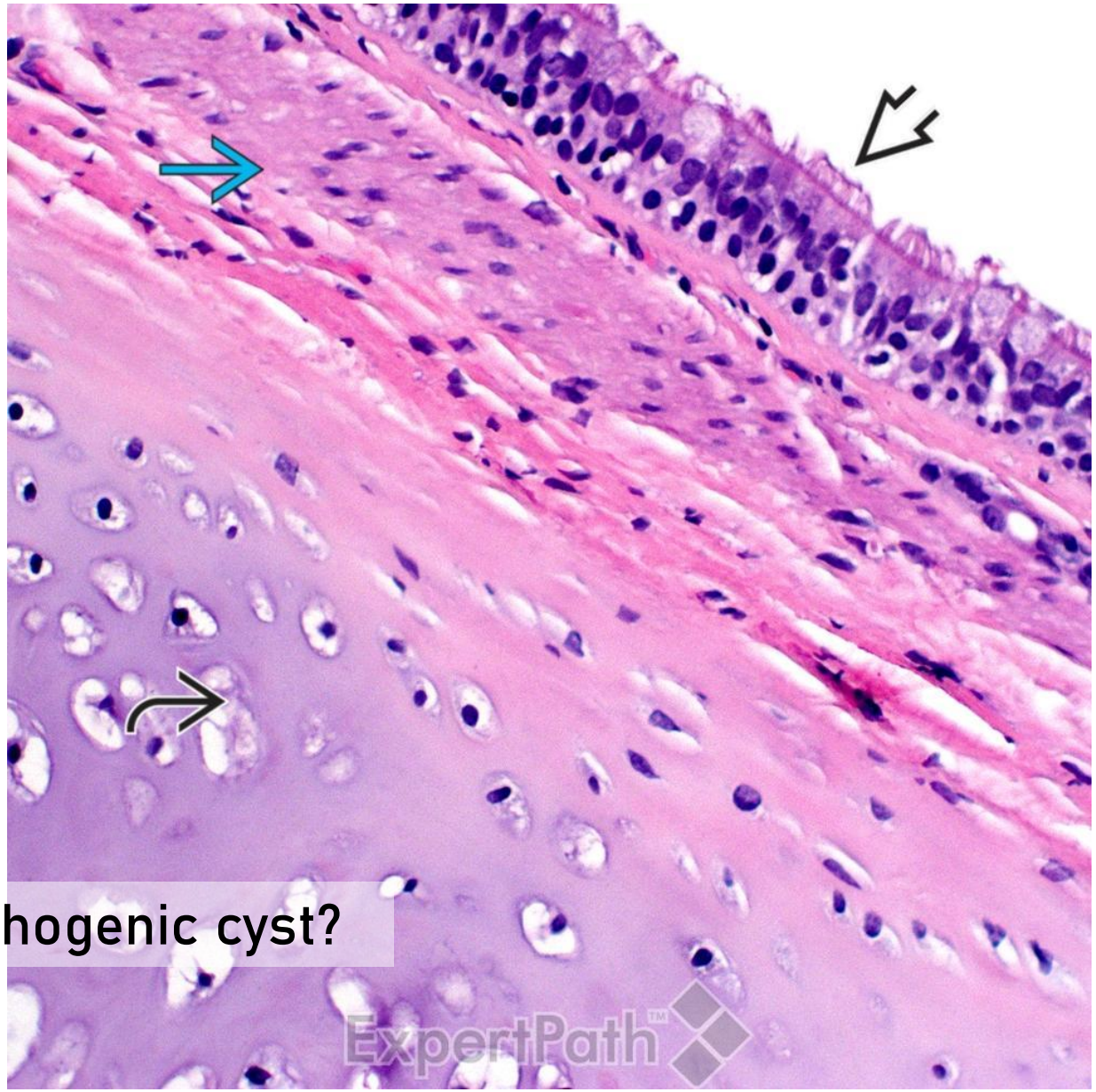


What is a vellus hair cyst?

Under low magnification, all the major features of a **bronchogenic cyst** are evident: Cyst space (cyan open arrow) lined by respiratory-type epithelium (black solid arrow) with a fibrous connective tissue wall that contains mucoserous glands (white open arrow), hyaline cartilage (white curved arrow), and smooth muscle (black curved arrow). Multiple step sections may be needed to identify the different components.

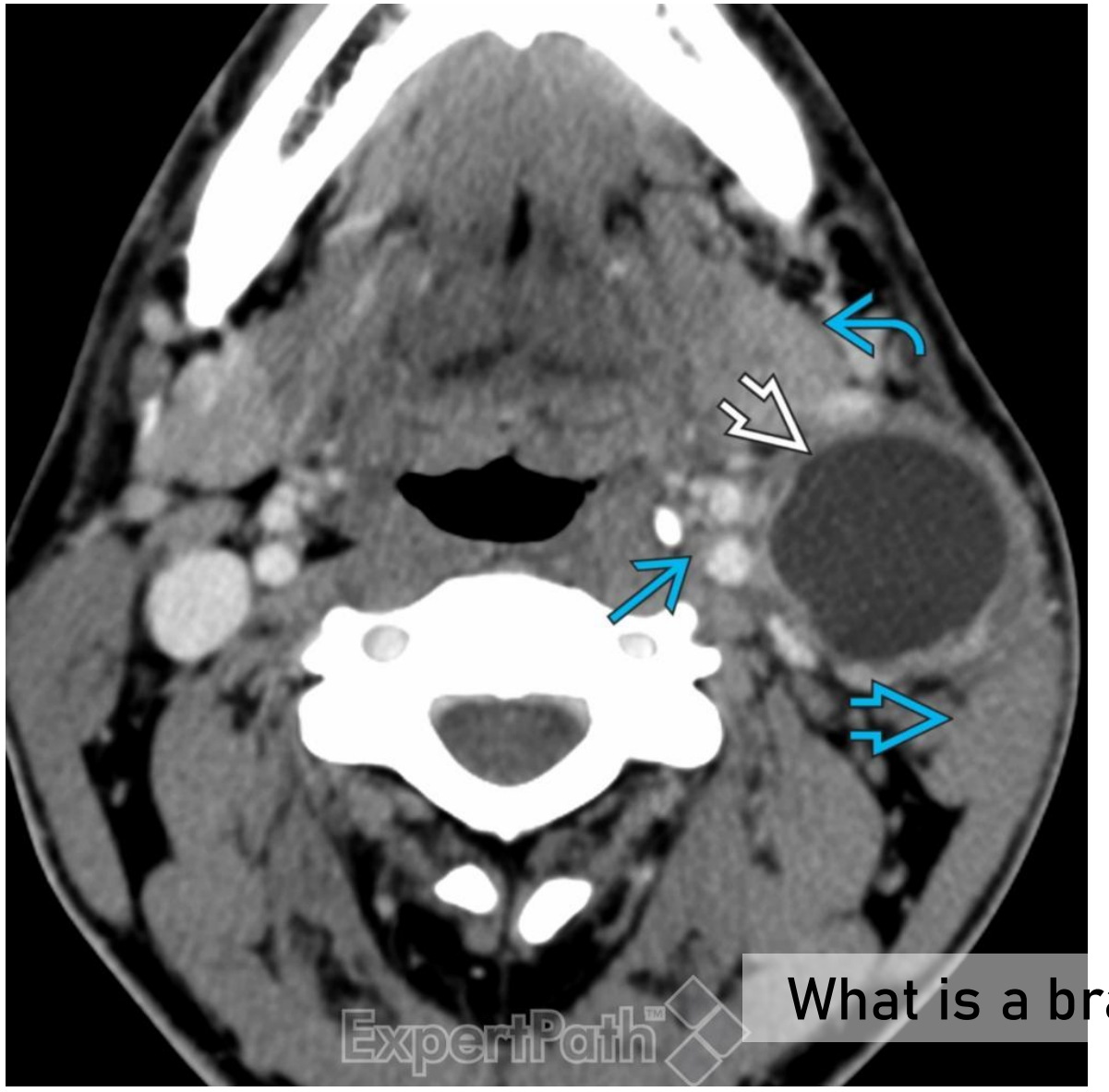


High-power H&E shows ciliated respiratory-type epithelium (black open arrow) overlying a band of smooth muscle (cyan solid arrow) closely associated with cartilage (black curved arrow).

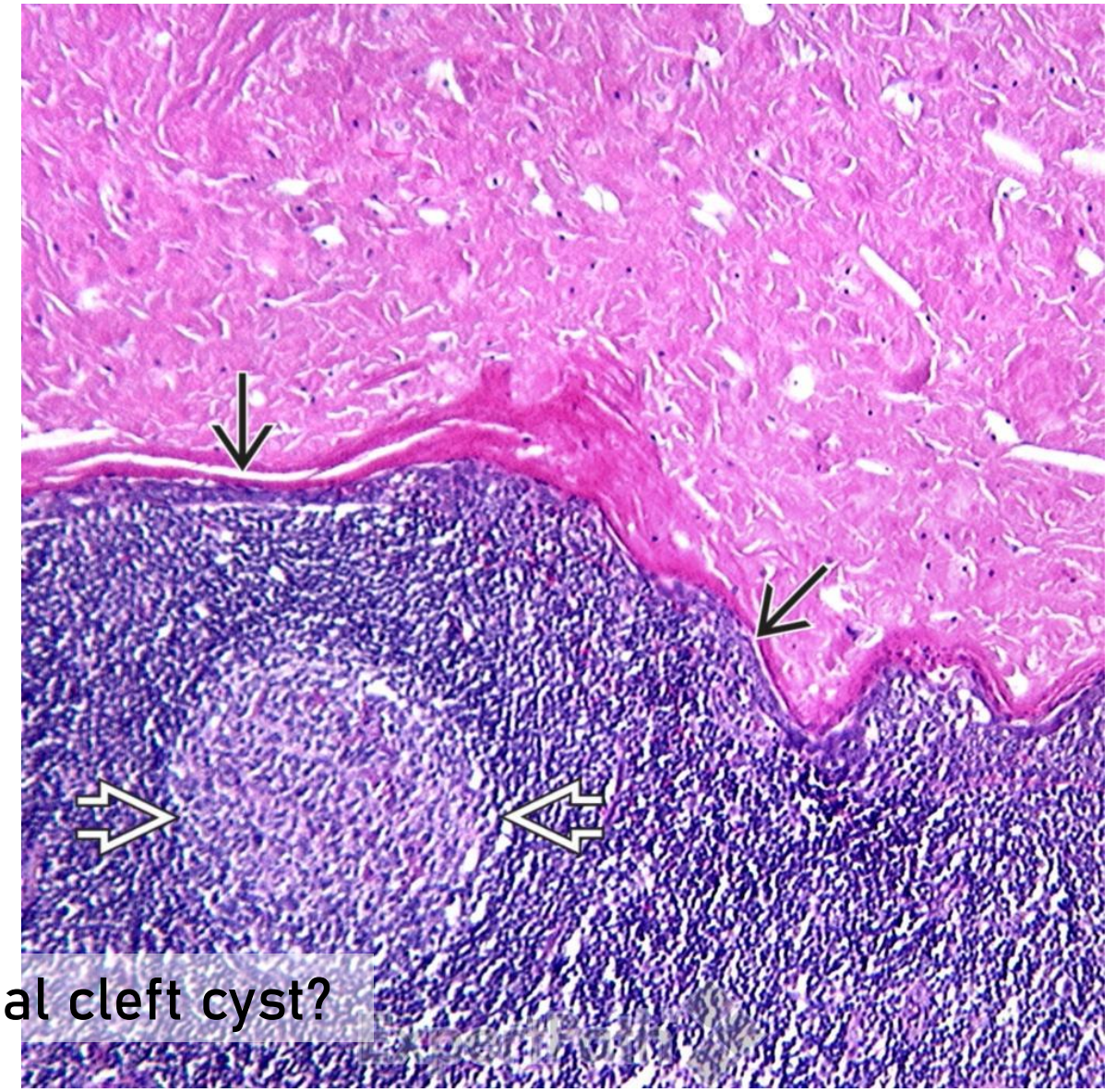


What is a bronchogenic cyst?

Axial CECT reveals a 2nd **branchial cleft cyst** (BCC) (white open arrow) located posterior to the submandibular gland (cyan curved arrow), lateral to the carotid space (cyan solid arrow), and anterior to the sternomastoid muscle (cyan open arrow). Capsule thickening suggests inflammation.

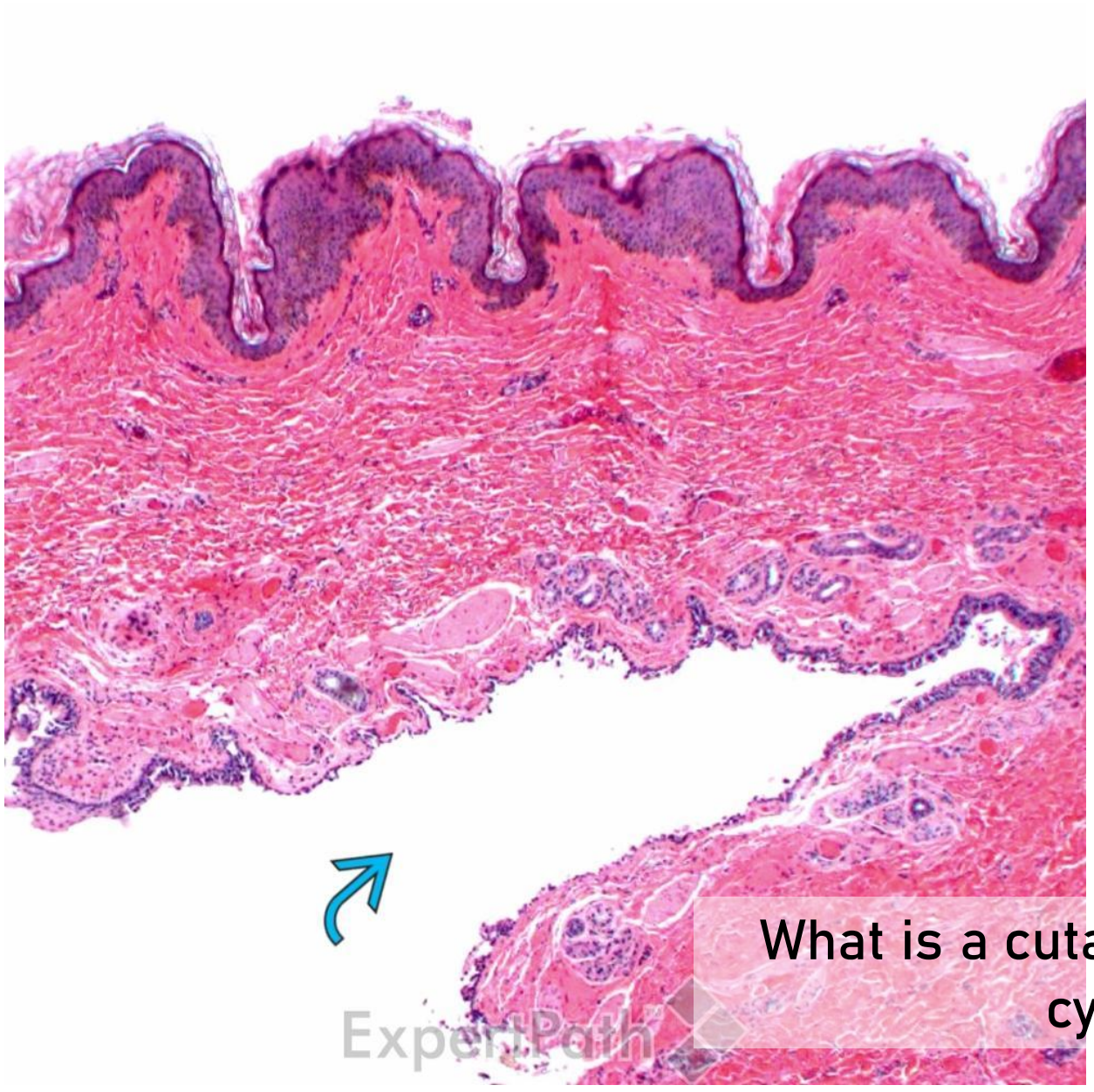


The lumen of this BCC is filled with keratinaceous debris. There is a thin, squamous epithelium (black solid arrow) without any atypia. There is a germinal center (white open arrow) within the associated lymphoid tissue.

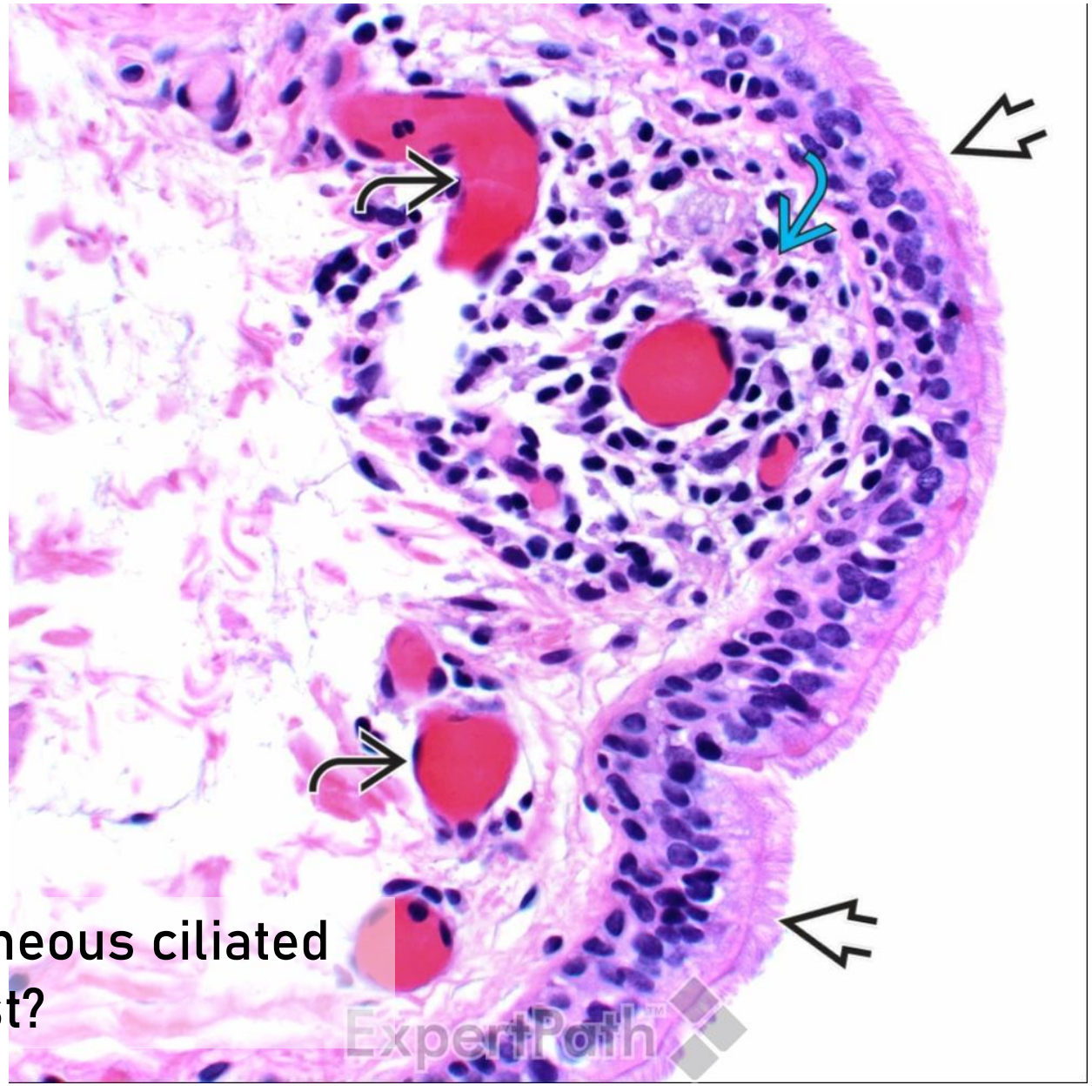


What is a branchial cleft cyst?

Cutaneous ciliated cyst (cyan curved arrow) presents as a unilocular cavity within the dermis. Occasionally, they may be located in the subcutis.

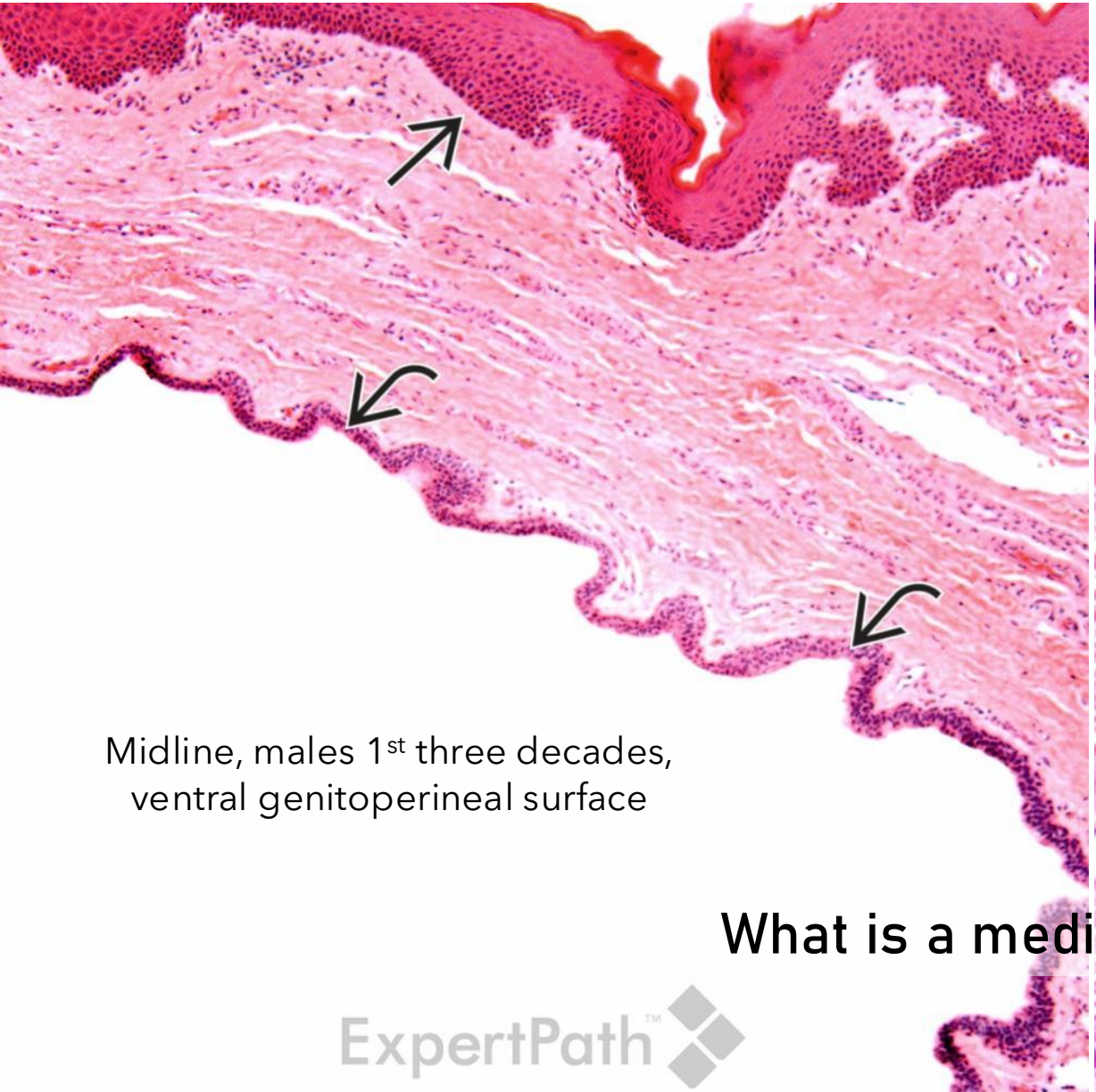


The fibroconnective tissue underlying the cyst wall shows small, dilated blood vessels (black curved arrow) and a surrounding subtle lymphocytic infiltrate (cyan curved arrow). Smooth muscle bundles are usually absent. As expected, a ciliated pseudo-columnar epithelium (black open arrow) lines the surface of the cavity.



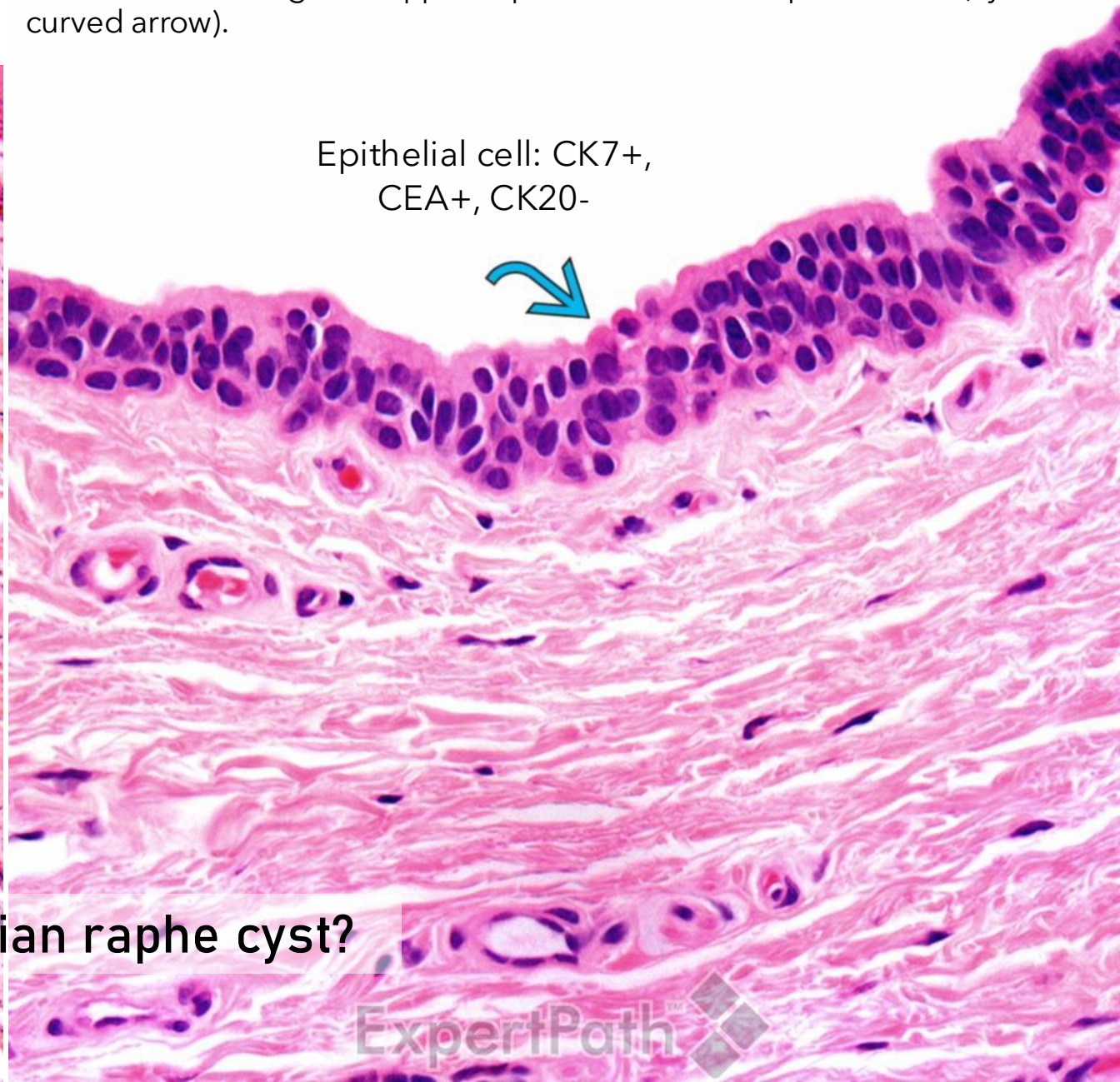
What is a cutaneous ciliated cyst?

This example of a **median raphe cyst**, shown under low magnification, is located in the middermis. There is no connection with the overlying epidermis (black solid arrow). The cyst lining is composed of a several-cell thick layer in the wall (black curved arrow).



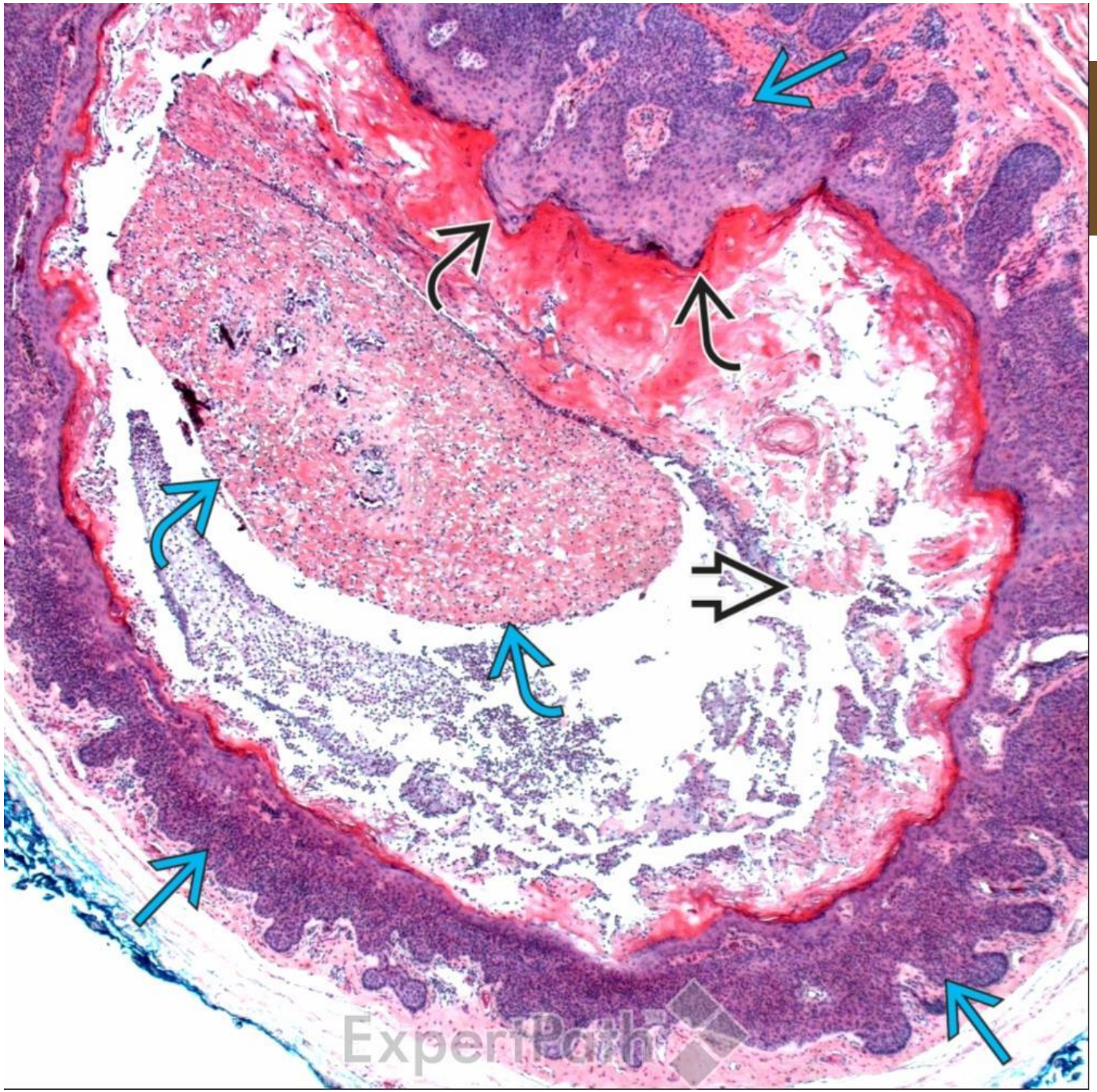
Midline, males 1st three decades, ventral genitoperineal surface

Under higher magnification, the pseudostratified columnar epithelium is shown, which is the characteristic lining of most median raphe cysts. Some of the surface-lining cells appear apocrine with focal apical snouts (cyan curved arrow).



Epithelial cell: CK7+, CEA+, CK20-

What is a median raphe cyst?

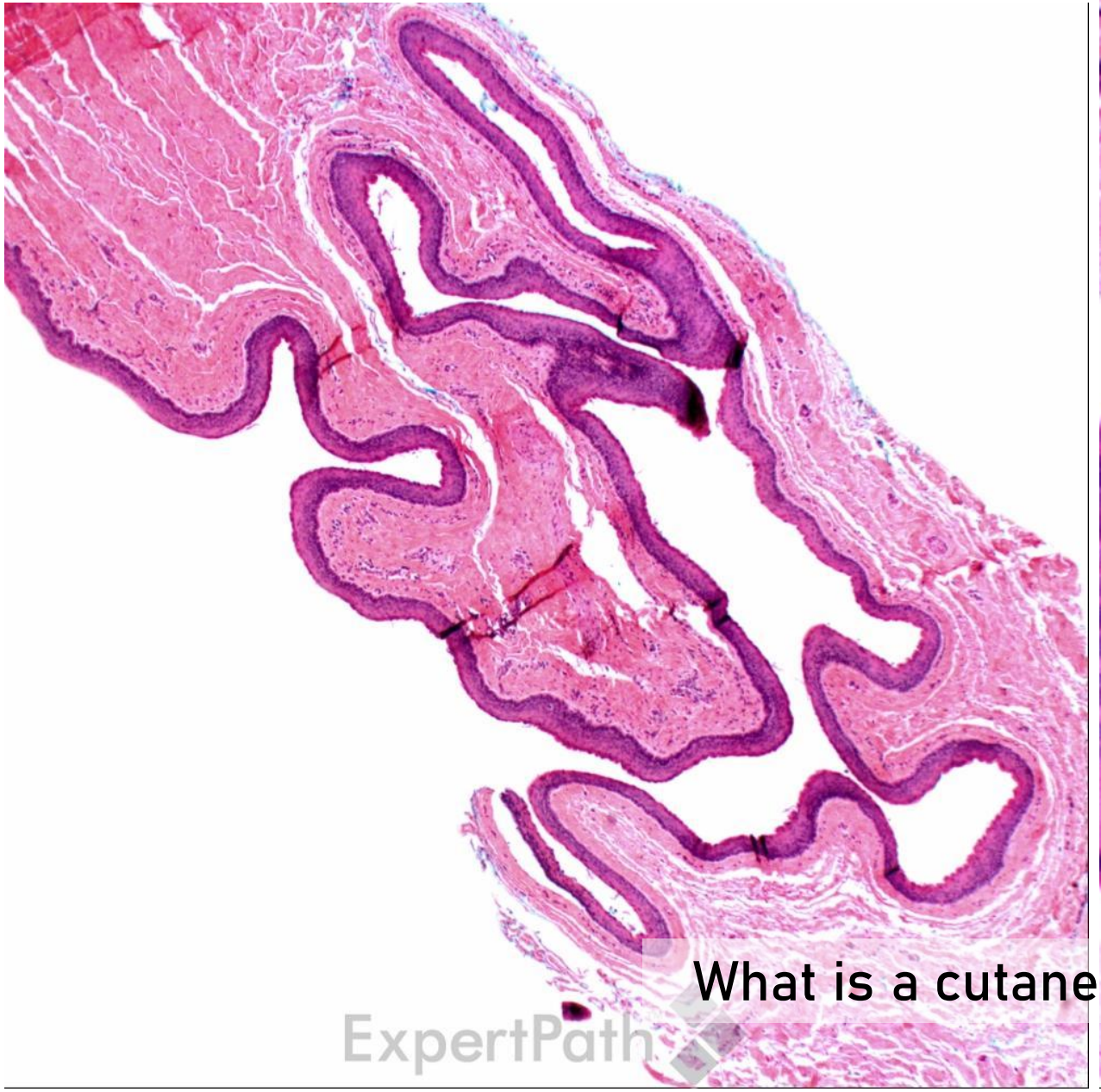


Can a cyst be related to HPV infection?

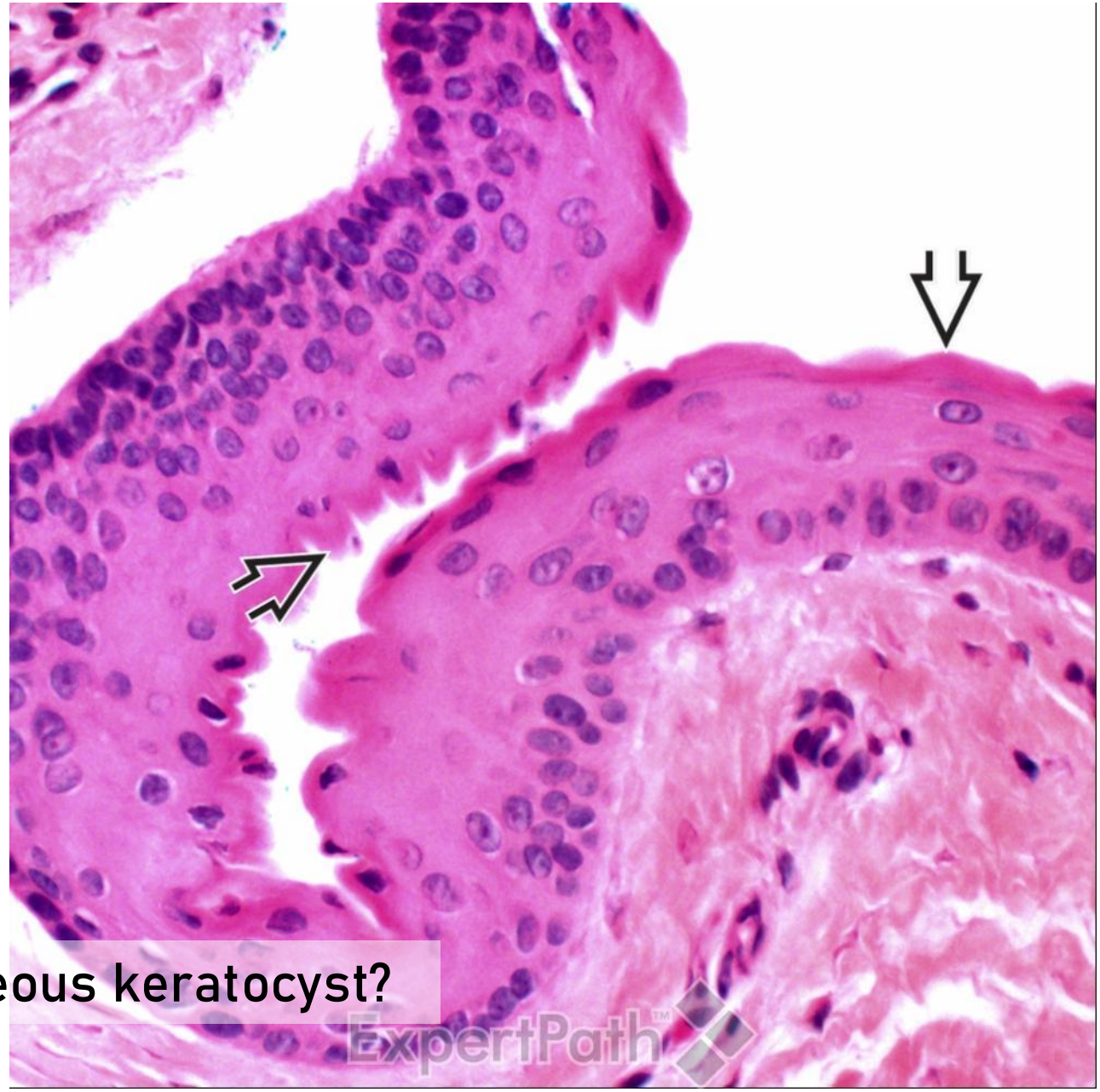
This **verrucous epidermal cyst** shows a thickened cyst wall (cyan solid arrow) lined by benign-appearing squamous epithelium. The surface epithelium displays verrucous hyperplasia with papillomatosis (black curved arrow). The lumen contains keratin (black open arrow) and a degenerating fibrovascular core (cyan curved arrow).

HPV (subtypes 60, 57, 65)

Scanning magnification view of a **cutaneous keratocyst** shows an undulating squamous epithelium lining the irregularly shaped cystic spaces. There are no associated sebaceous glands, unlike in steatocystoma.



High-magnification view of a cutaneous keratocyst shows a bland squamous epithelium with a dense, eosinophilic, jagged-appearing cuticle (black open arrow) lining the cystic space.



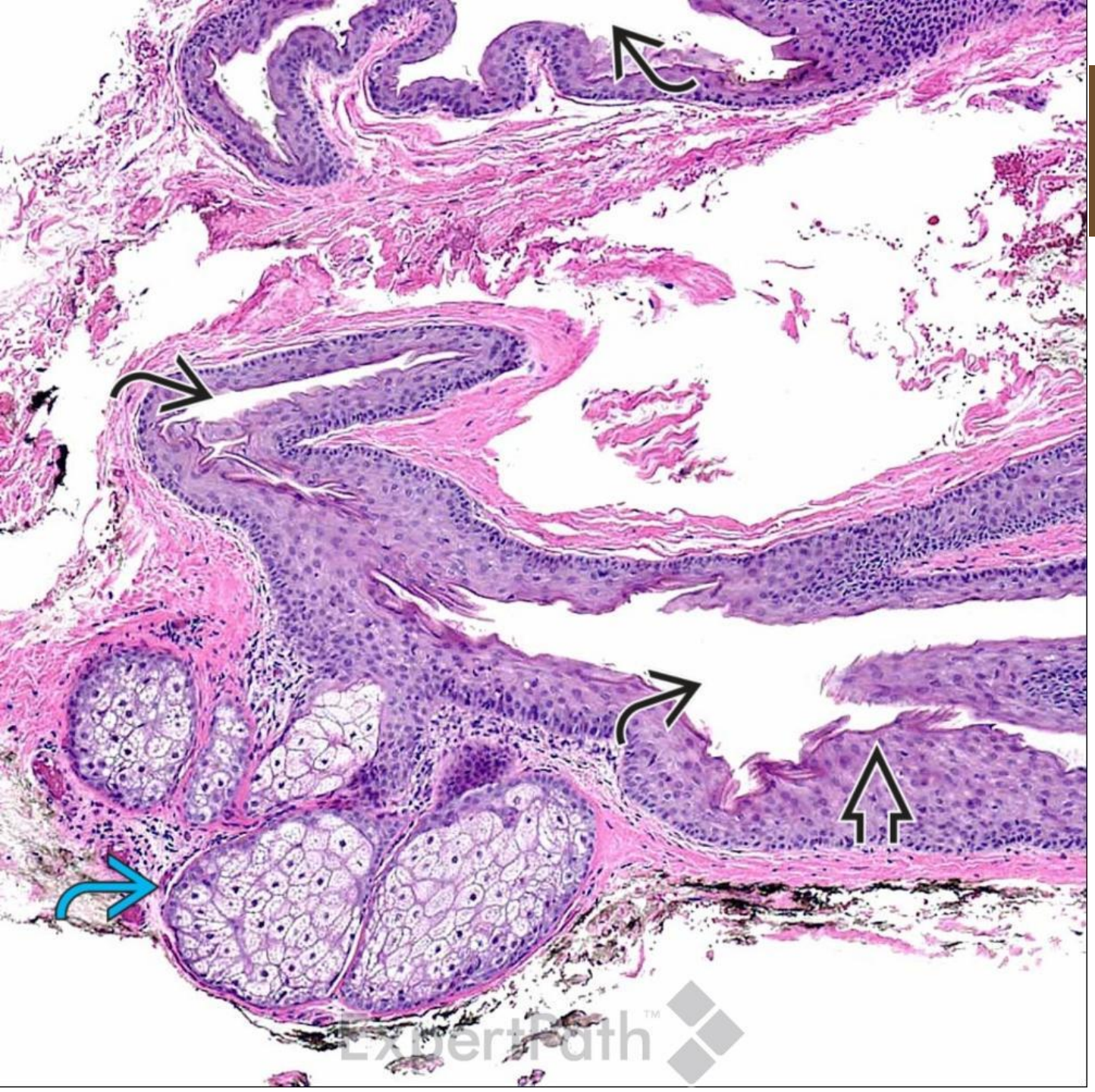
What is a cutaneous keratocyst?

Cysts with Stratified Squamous Linning and Cyst Wall Component

Steatocystoma

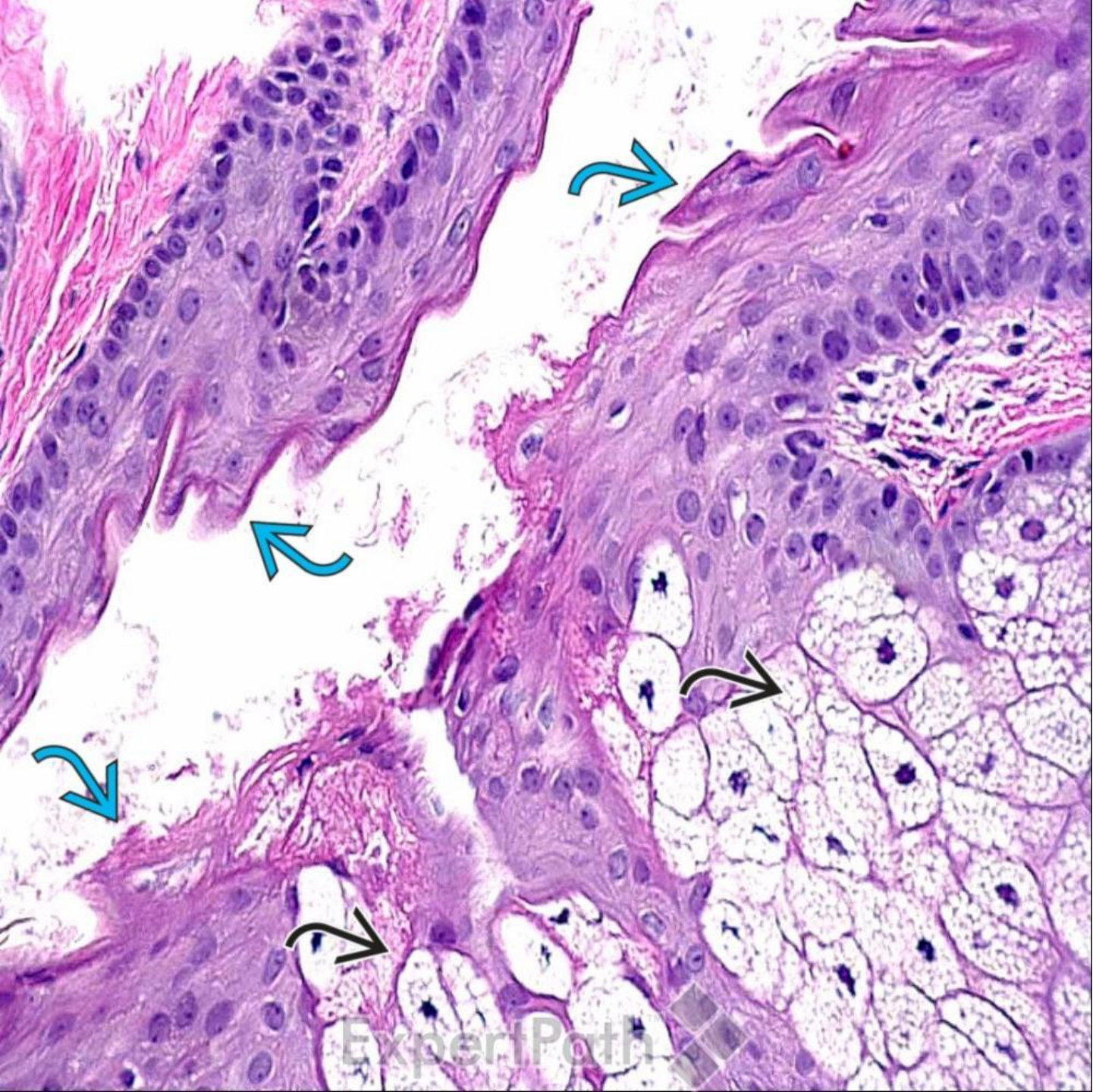
Dermoid Cyst

Thymic Cyst

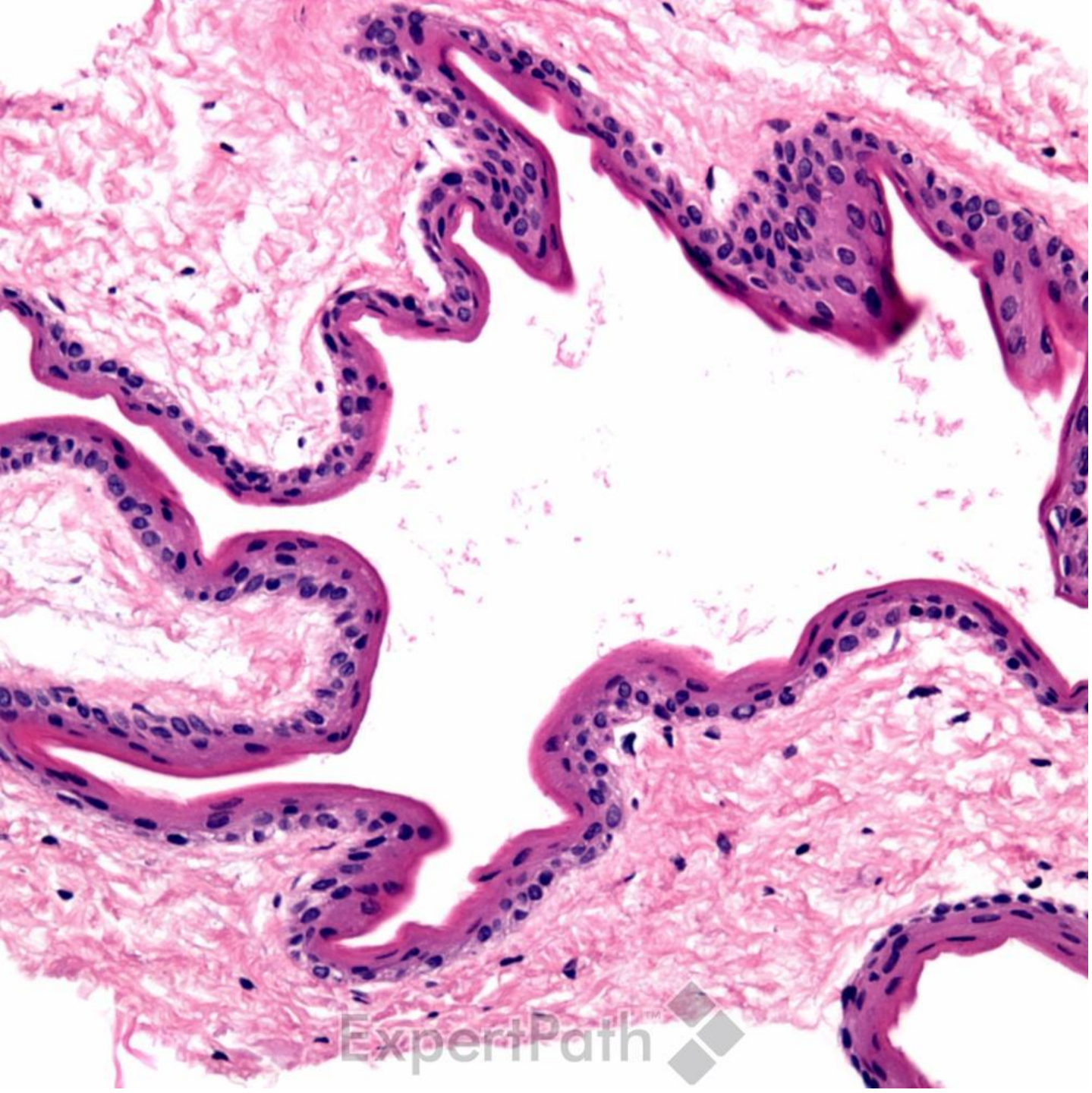


What is steatocystoma multiplex?

A young male patient presented with multiple cystic papules. At first glance, cystic spaces (black curved arrow) are evident. There are sebaceous glands (cyan curved arrow) attached to the cyst. There is no glandular layer in the epidermal cyst lining (black open arrow).

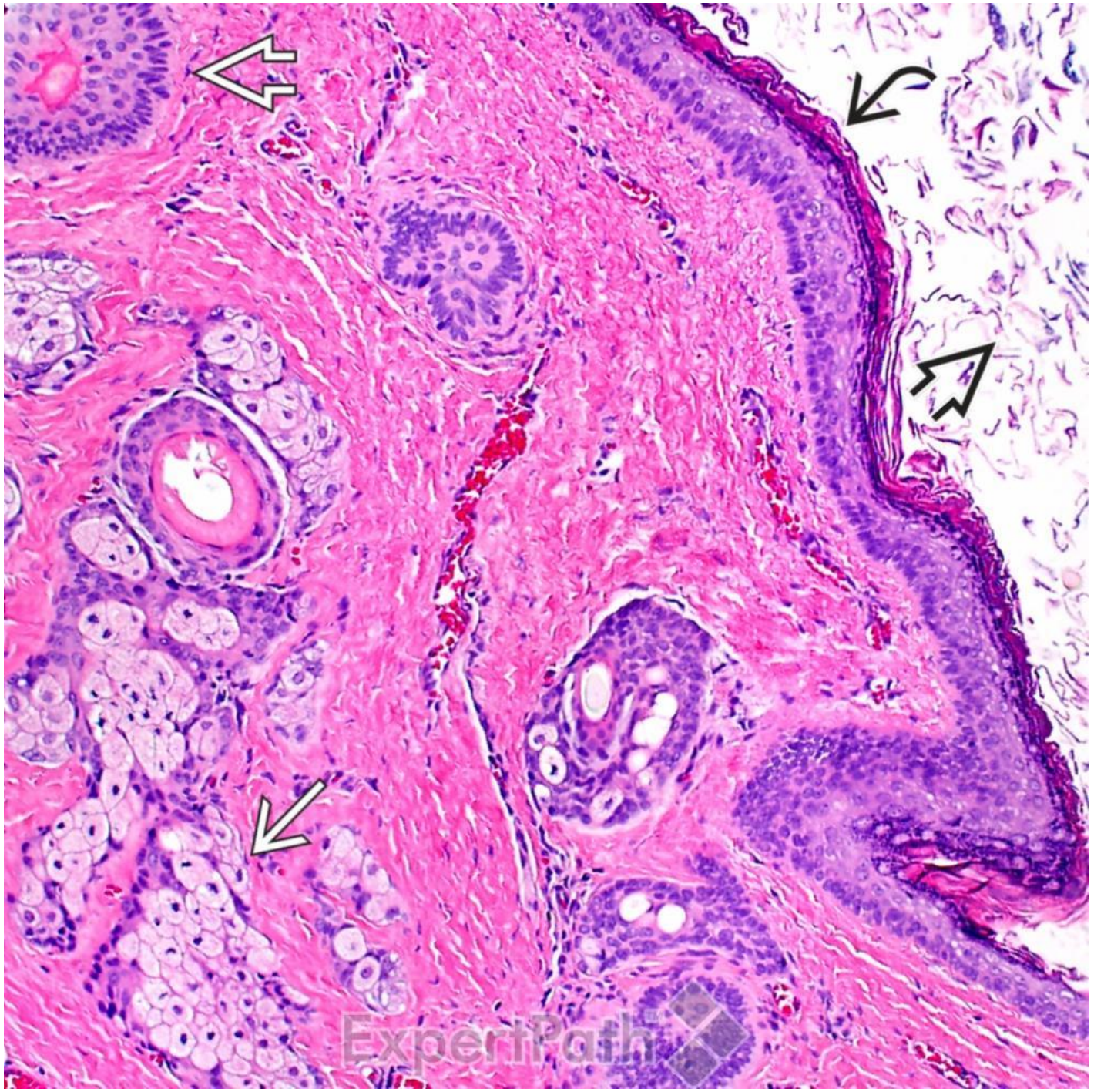


The inner surface of the cyst lining has characteristically eosinophilic jagged or shark tooth appearance (cyan curved arrow). The attached sebaceous glands (black curved arrow) are evident. Fragments of vellus hair were seen in the cystic space (not shown).



In some cases of steatocystoma, sebaceous glands may not be evident in each histologic section. May need deeper sections to demonstrate sebaceous glands.

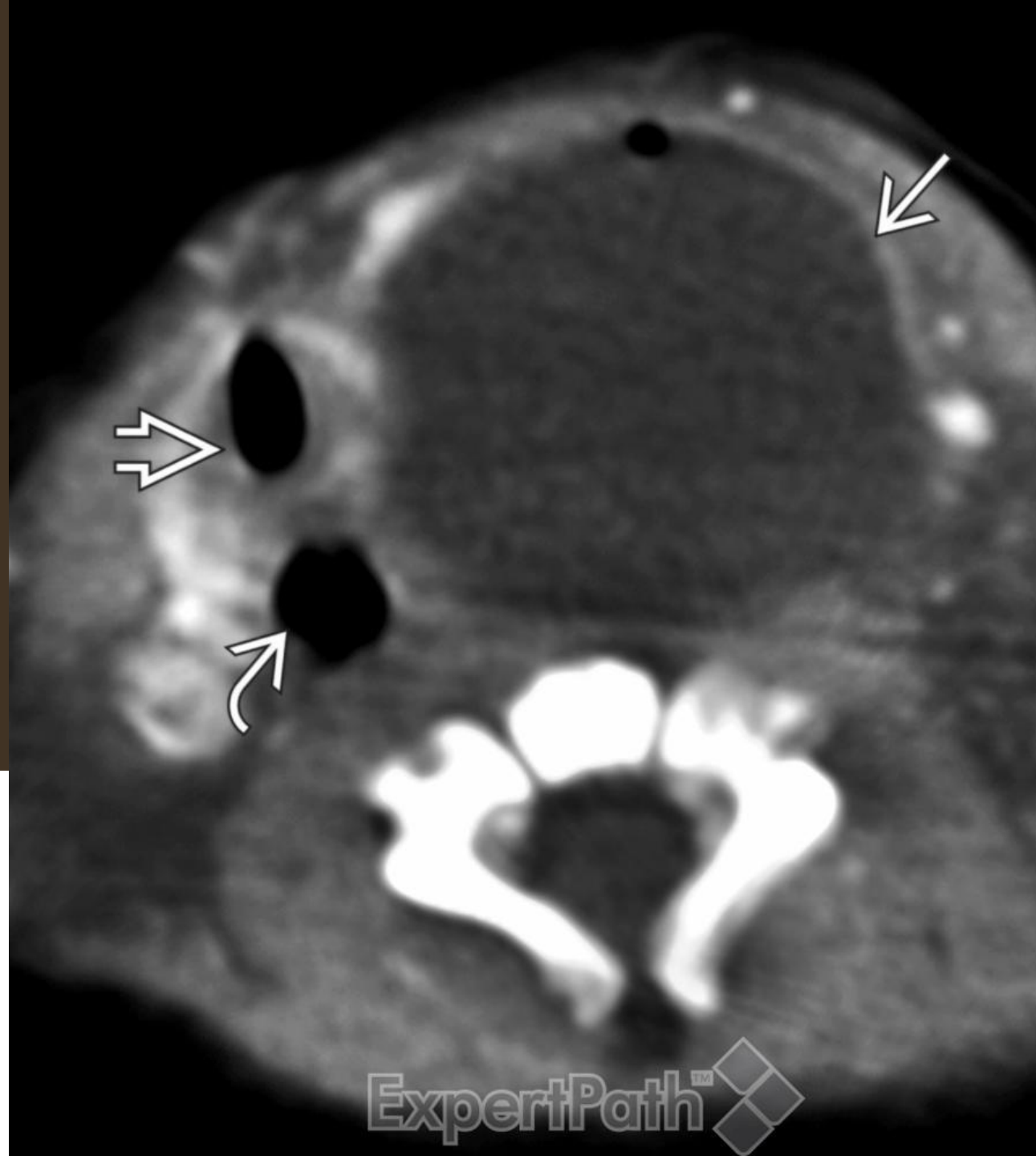
DDX: cutaneous keratocyst



What is a dermoid cyst?

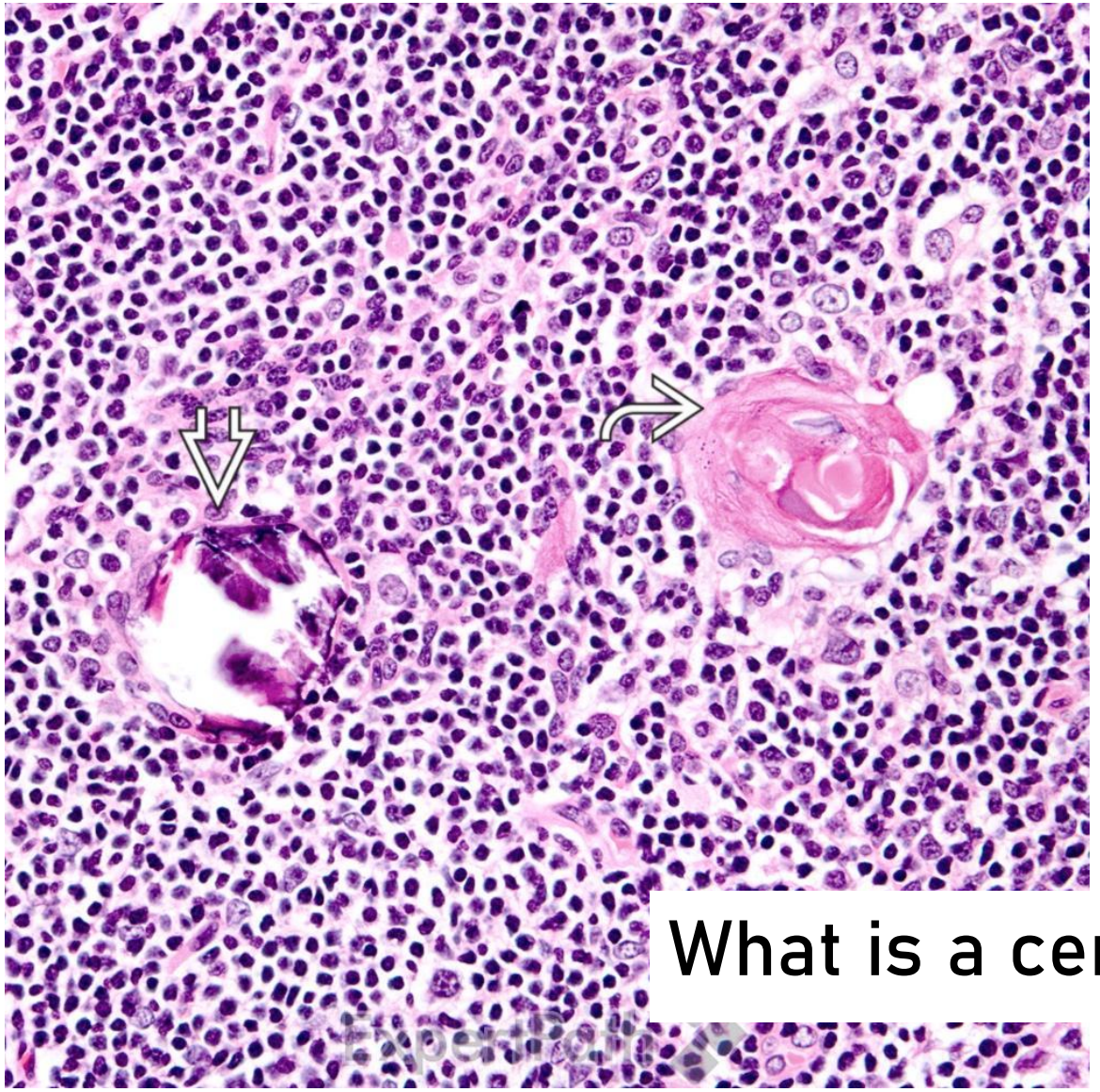
This **dermoid cyst** contains luminal keratin (black open arrow) and is lined by keratinized squamous epithelium (black curved arrow). The cyst wall contains adnexal structures, such as hair follicles (white open arrow) and sebaceous glands (white solid arrow).

Cervical thymic cyst

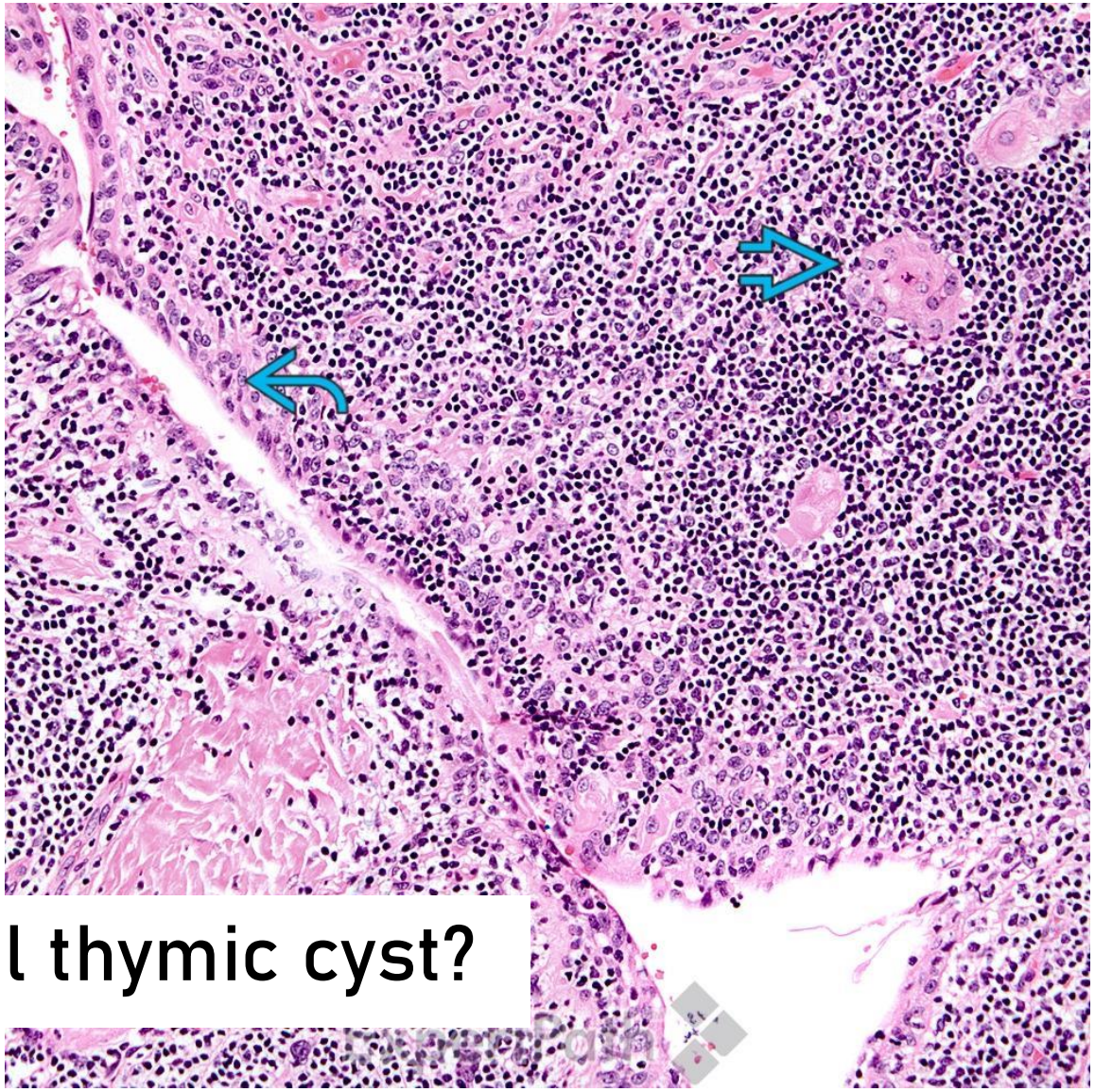


Axial CECT in an infant with an infected thymic cyst demonstrates a small bubble of gas anteriorly within a large, left-sided cystic neck mass (white solid arrow) that causes significant deviation of the airway (white open arrow) and esophagus (white curved arrow) to the right. (From DI: H&N.)

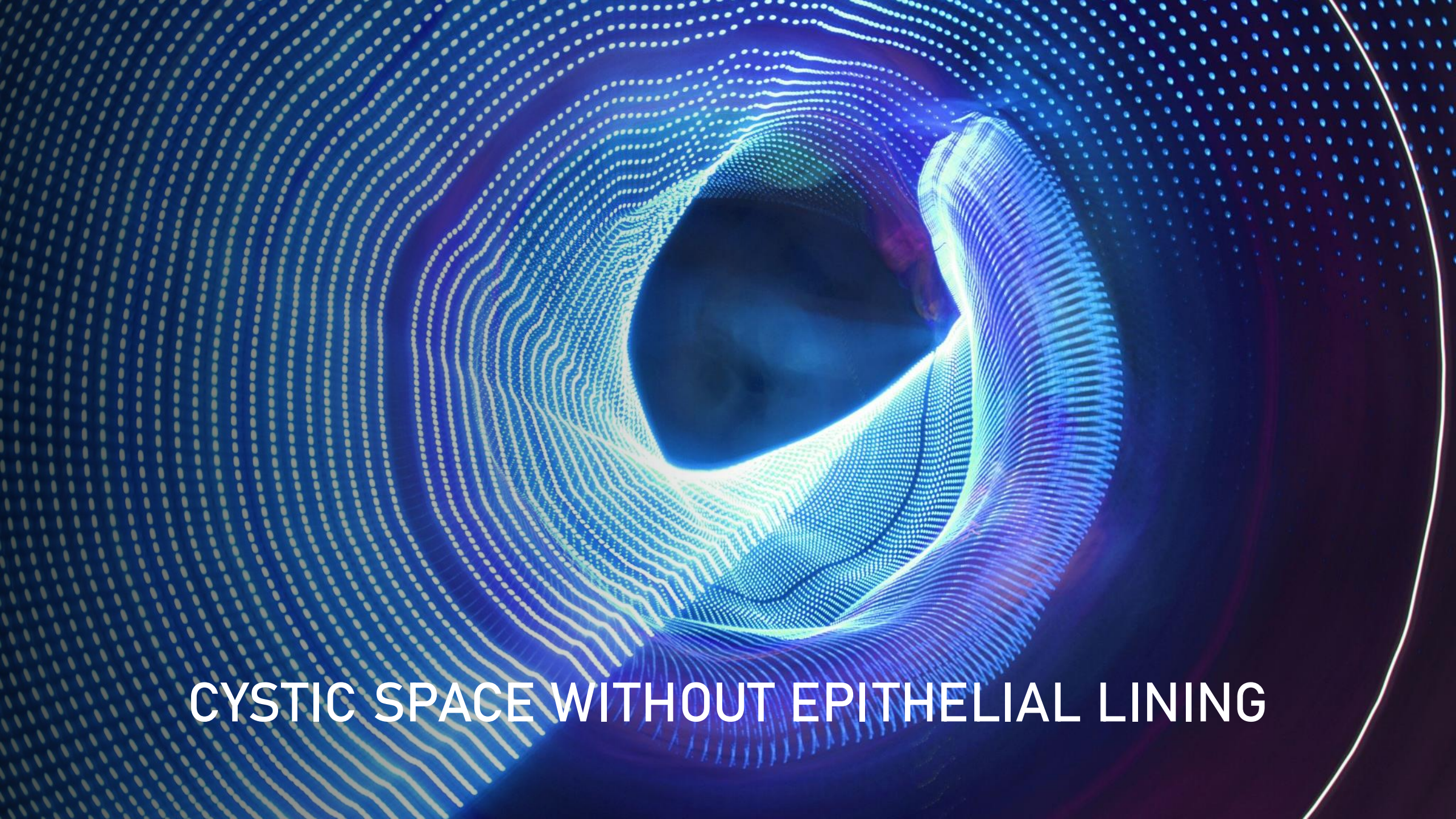
What is a cervical thymic cyst?

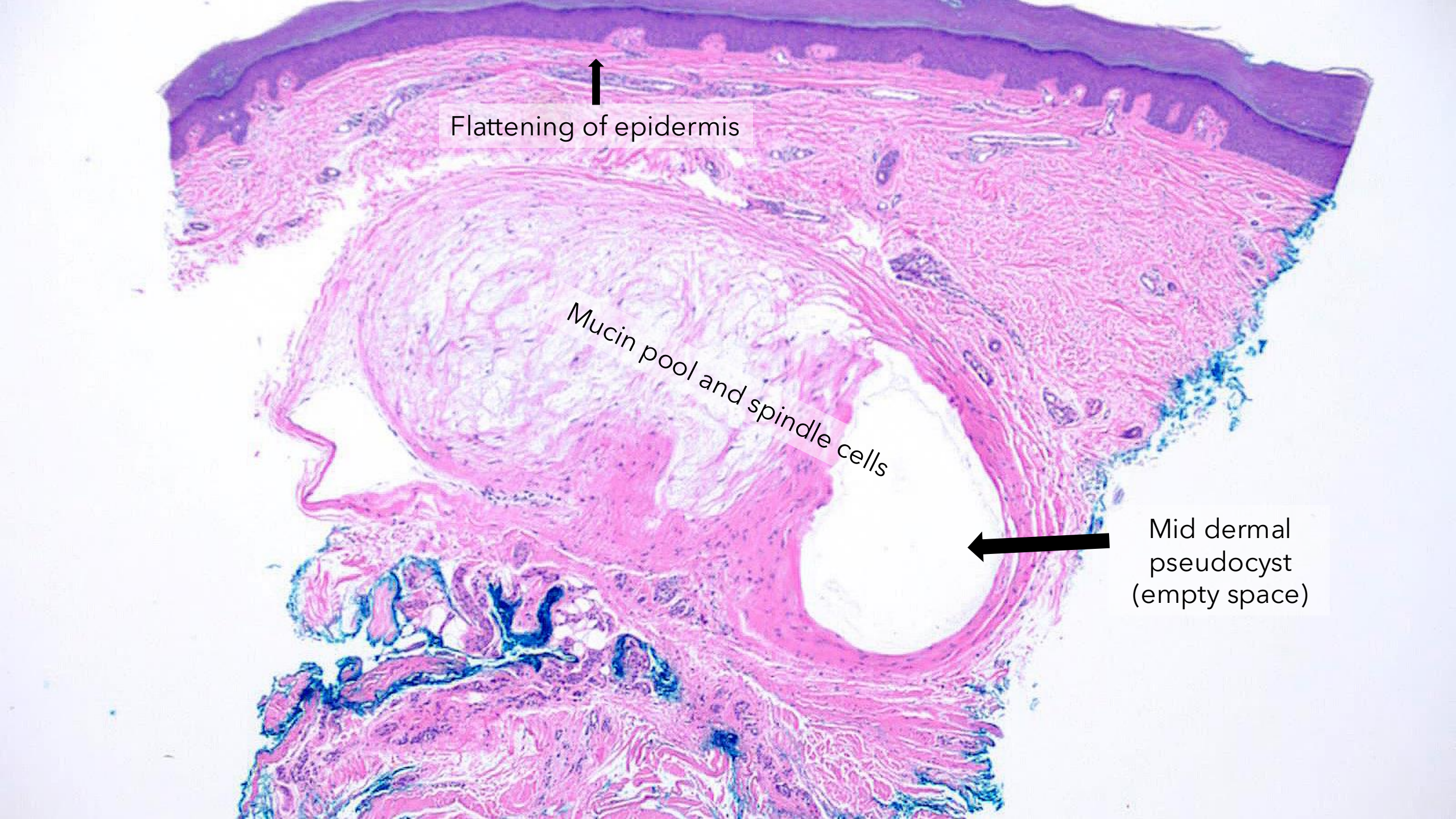


The wall of the cyst includes an identifiable Hassall corpuscle (white curved arrow) characterized by a concentric island of squamous cells with central keratinization; microcalcification (white open arrow) is focally noted. Identification of thymic tissue may require extensive sampling.

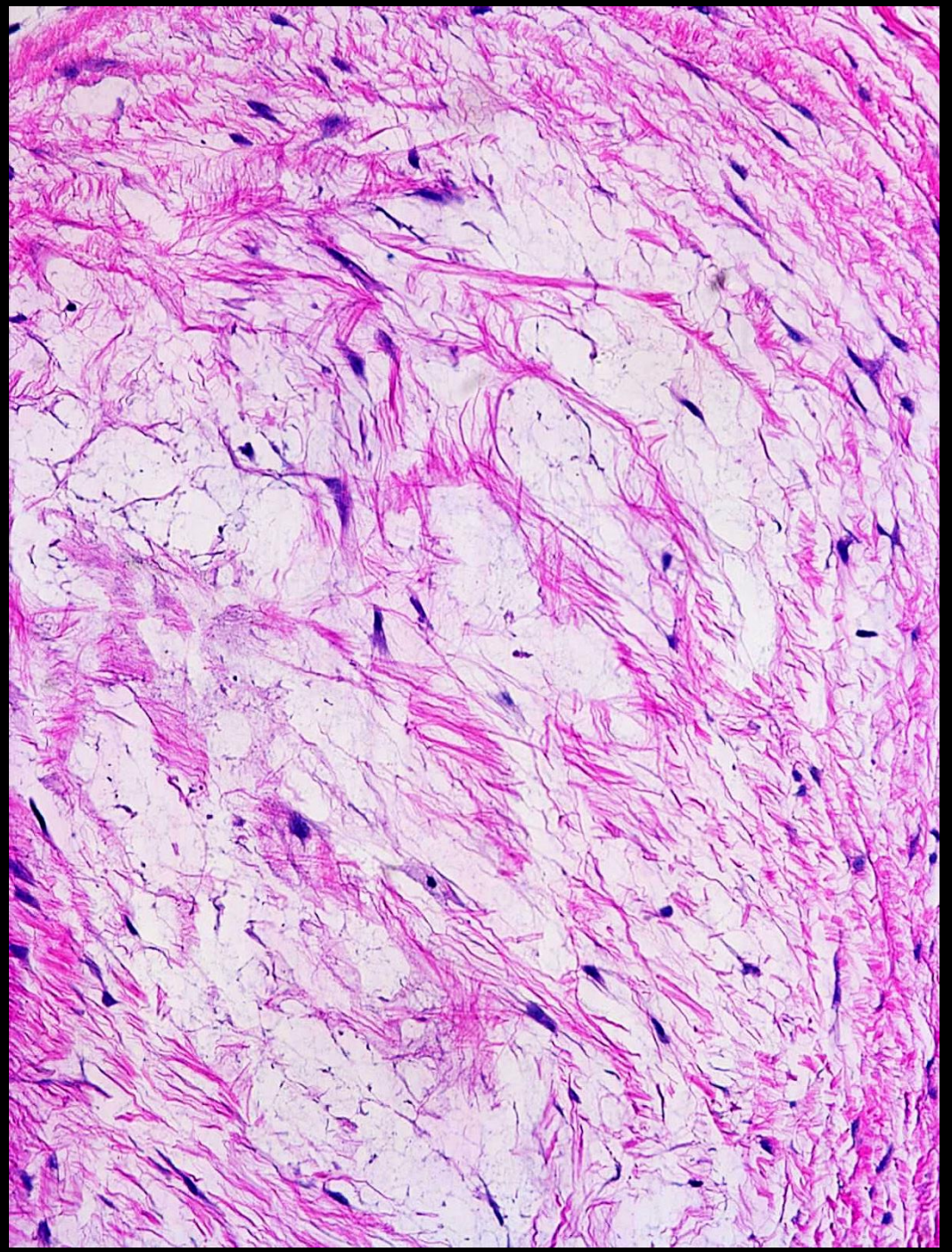


Medium-power hematoxylin and eosin shows an epithelial-lined cyst (cyan curved arrow) with a dense mixed inflammatory cell infiltrate and a Hassall corpuscle (cyan open arrow). These findings in conjunction with a cyst wall are diagnostic of a thymic cyst.

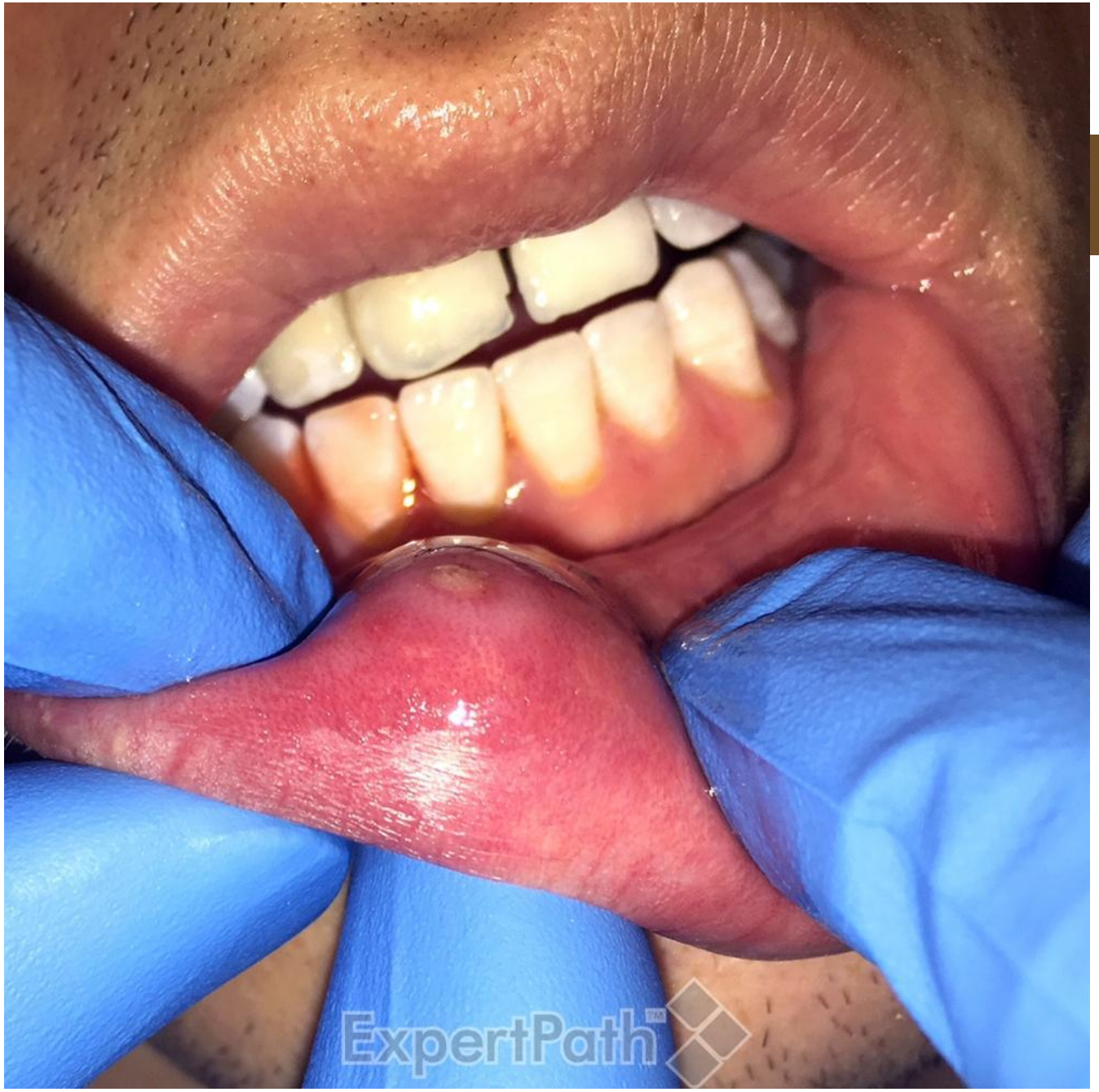

CYSTIC SPACE WITHOUT EPITHELIAL LINING


Flattening of epidermis
Mucin pool and spindle cells
Mid dermal pseudocyst (empty space)

Benign spindled fibroblasts in mucinous background

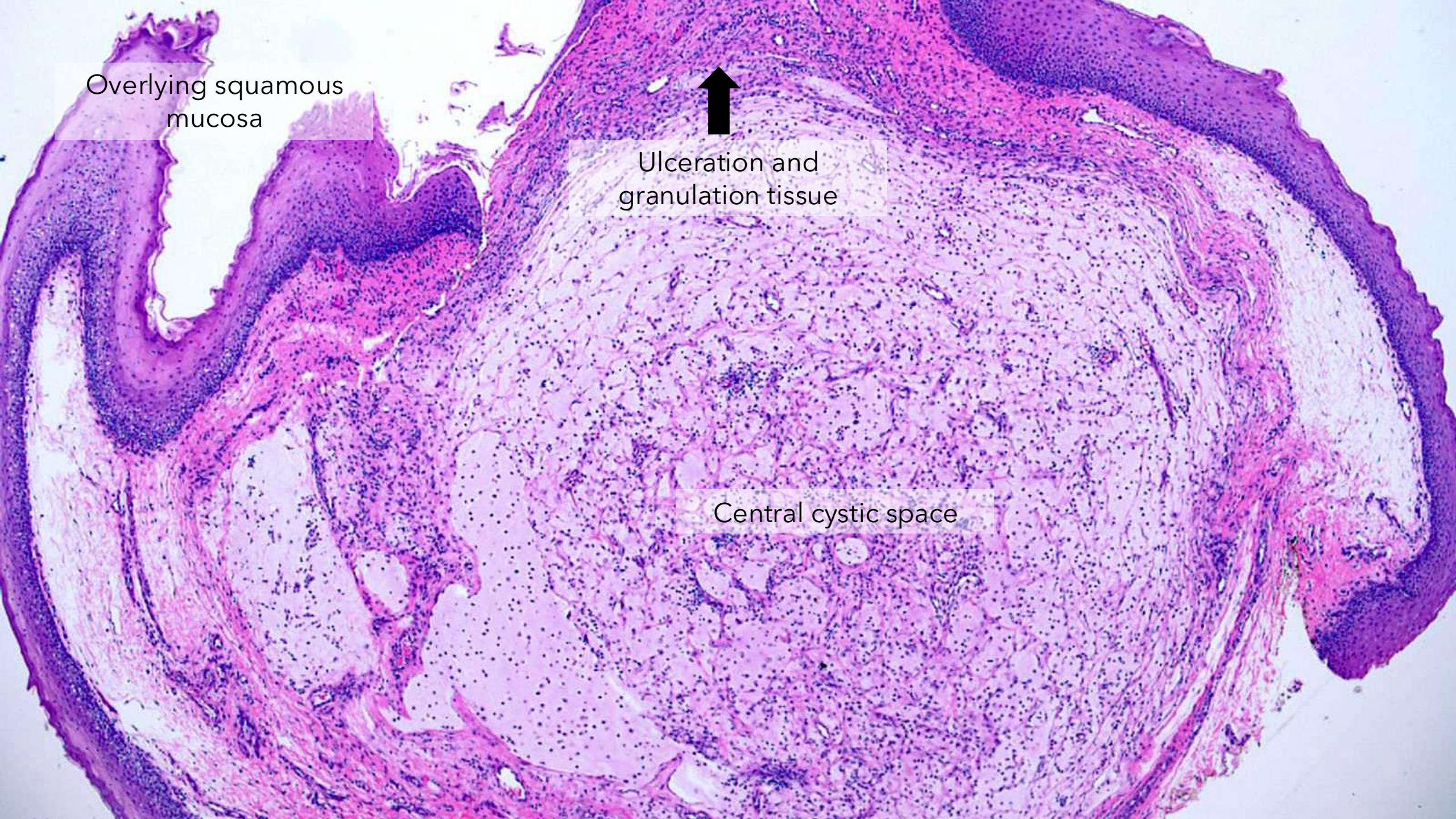


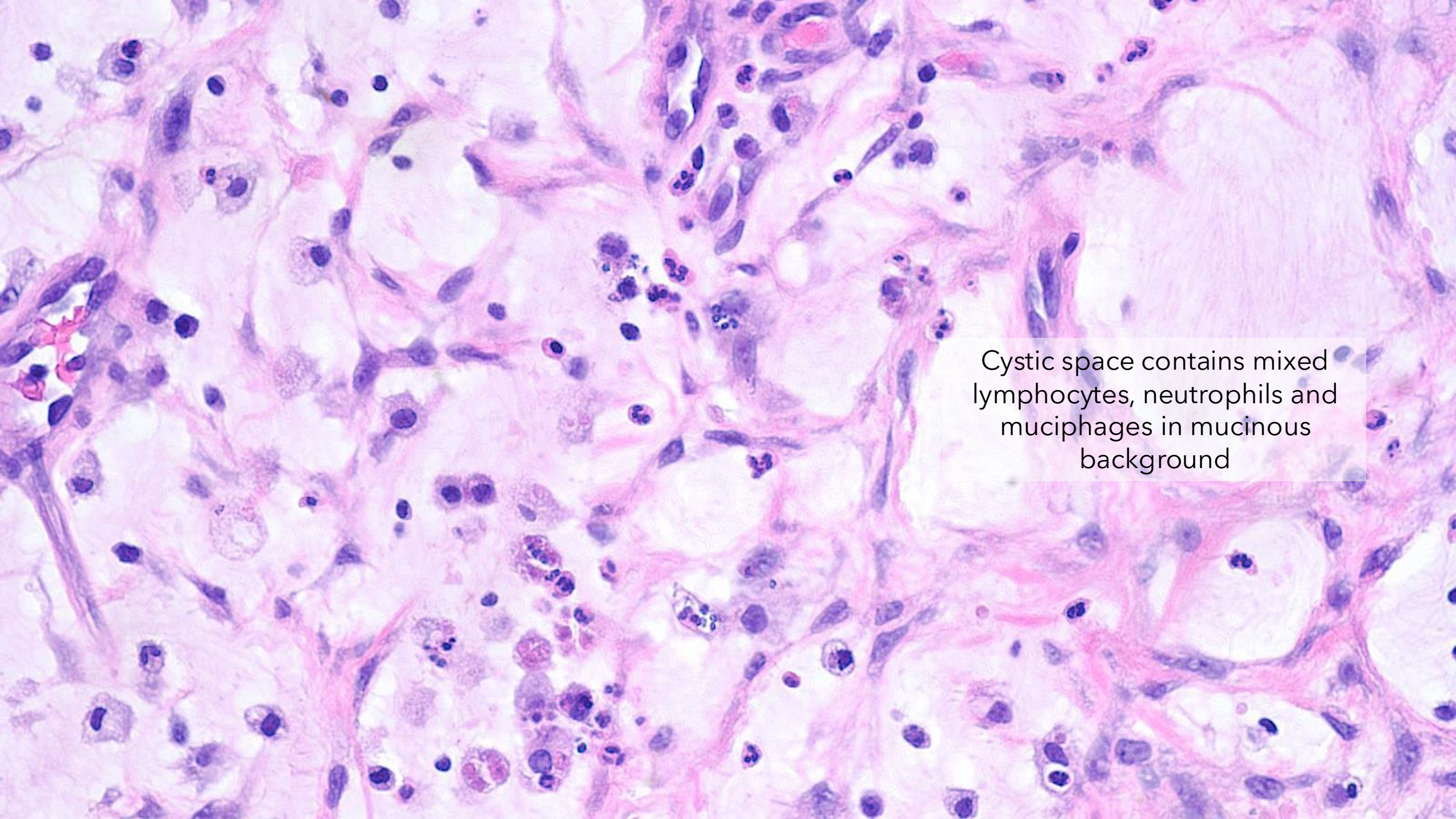
What is a digital mucous pseudocyst (cyst)?



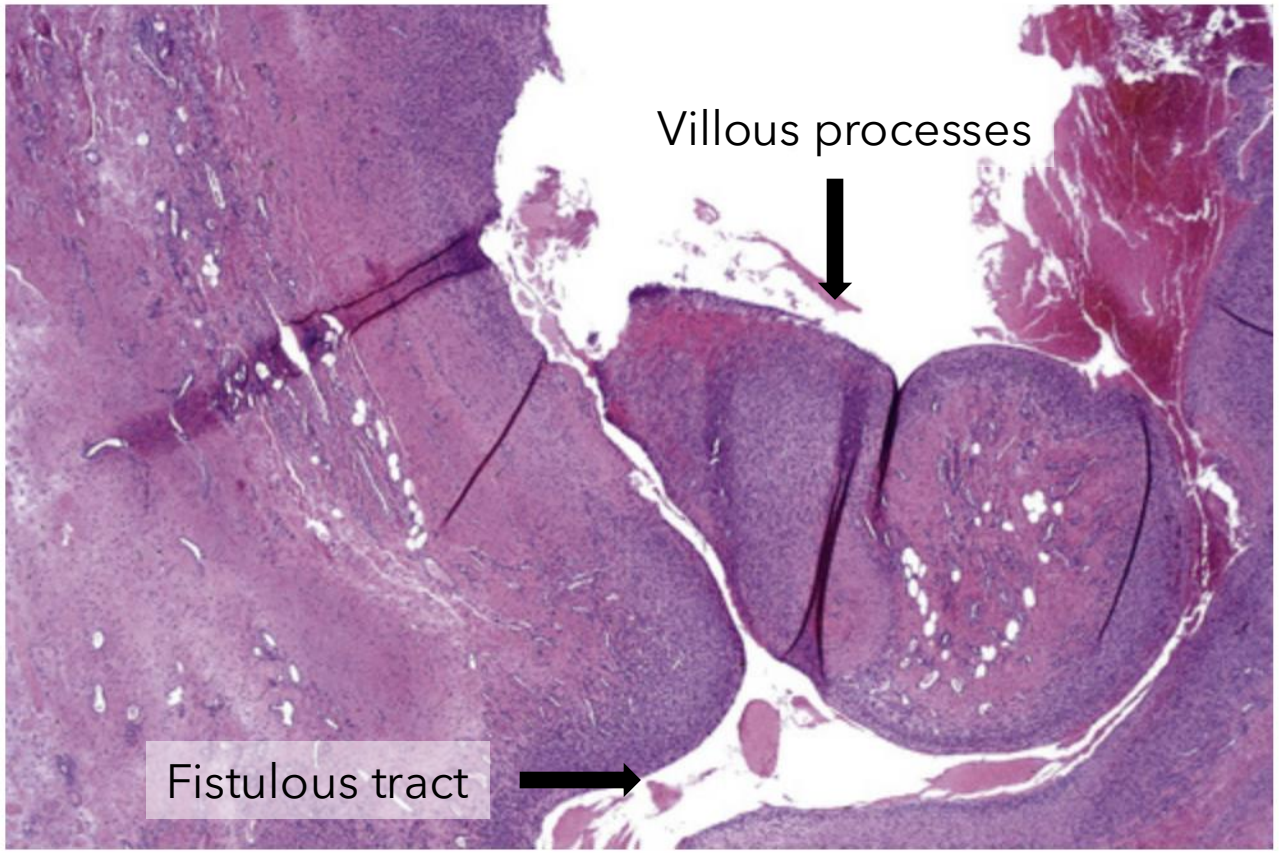
What is a mucocele?

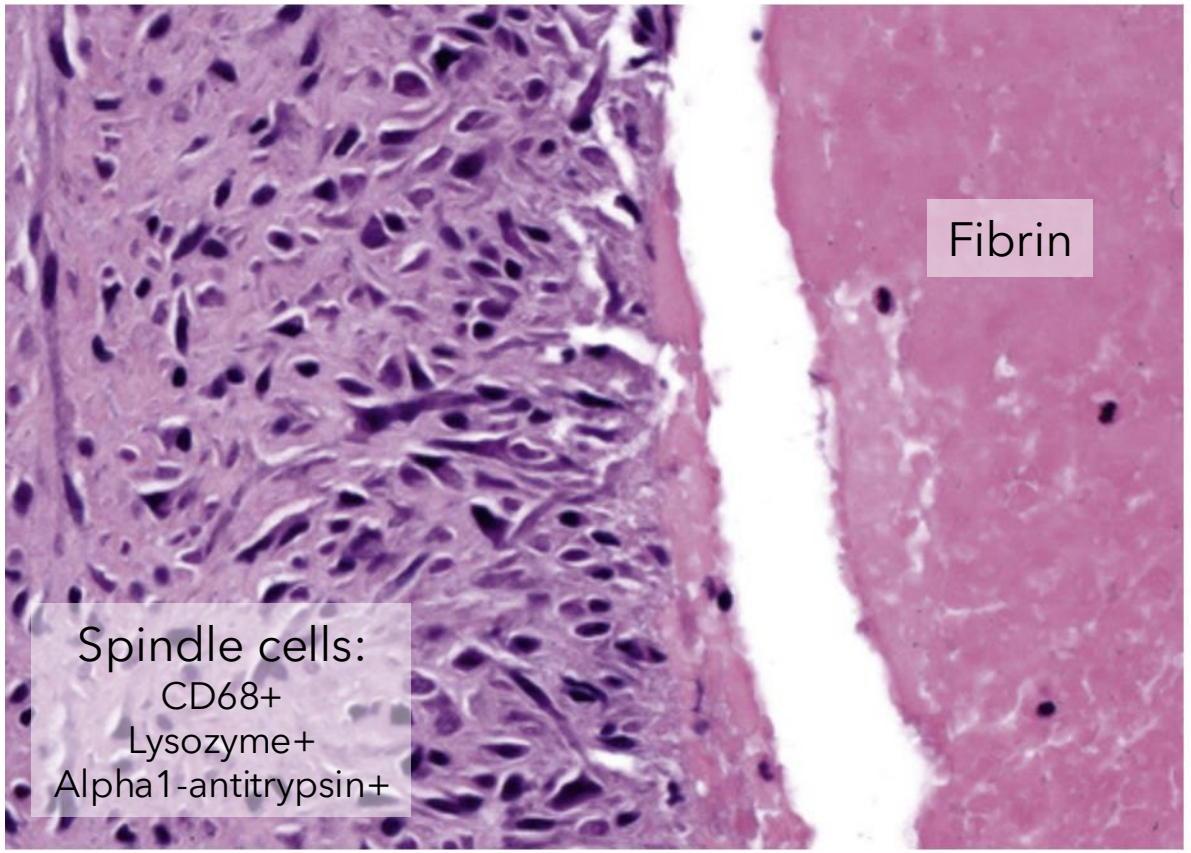
Clinical photo shows a dome-shaped swelling on the lower lip, the most common location for a mucocele. Mucoceles are generally fluctuant, although older lesions may be firm on palpation. Color ranges from mucosal to blue to red and surface ulceration may be present if traumatized.


Overlying squamous mucosa
Ulceration and granulation tissue
Central cystic space

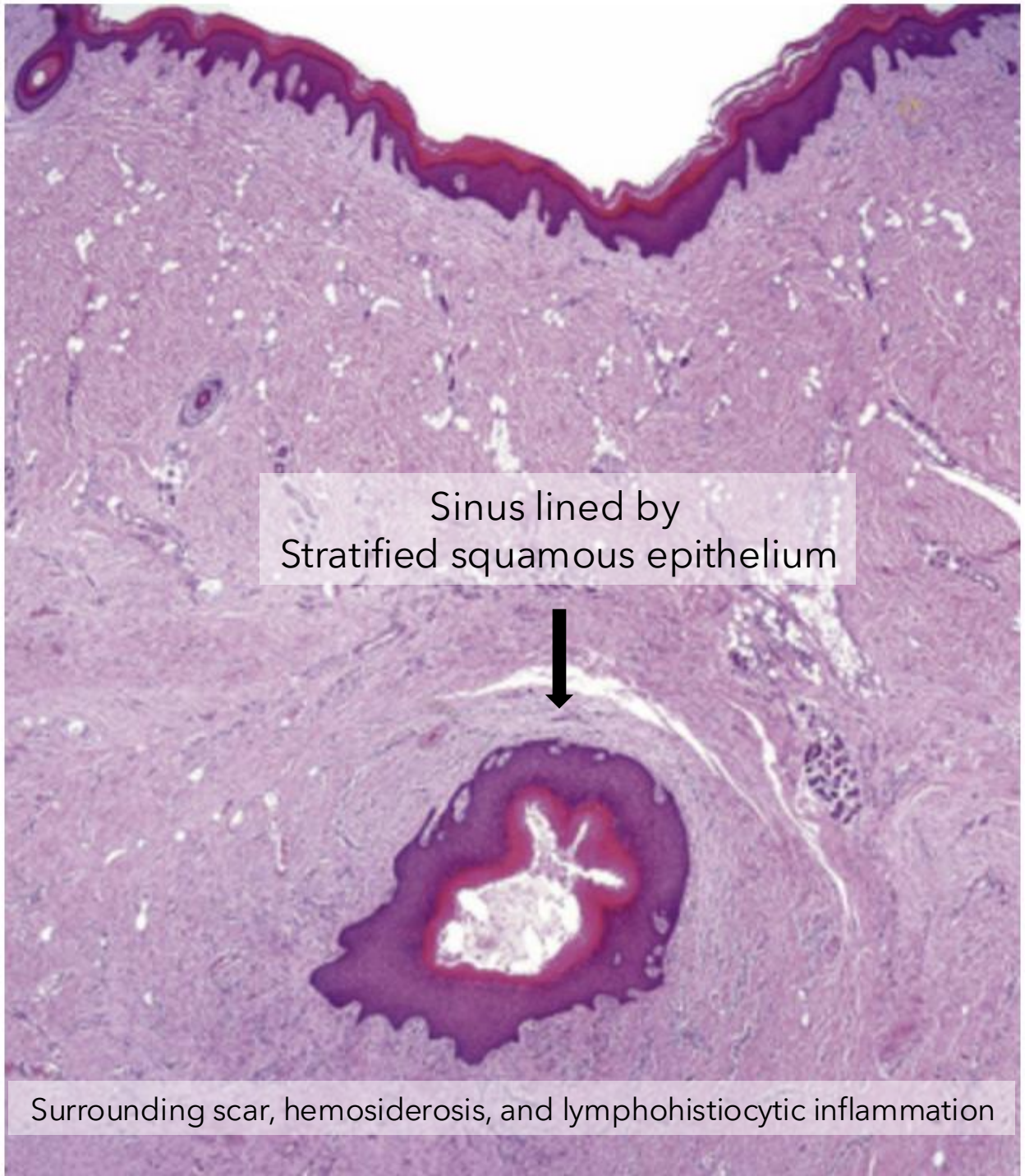

Cystic space contains mixed lymphocytes, neutrophils and muciphages in mucinous background

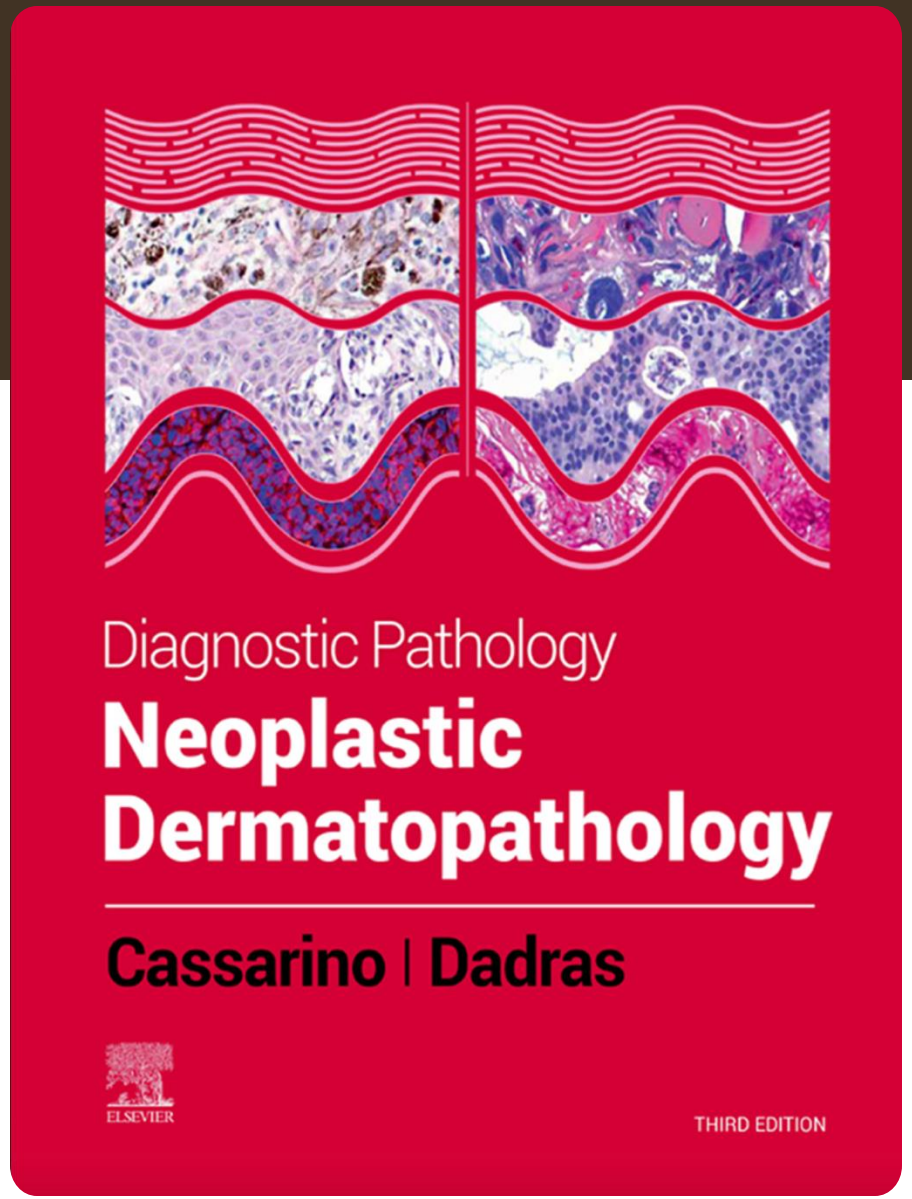
What is a metaplastic synovial cyst?





What is a pilonidal sinus?





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