

# History Questions

An 18-month-old boy presented with a slow growing left anterior orbital lump medially.

Examination revealed a palpable mass beneath the skin at the inferonasal aspect without any visual impairment. A CT scan (Figure 1) followed by an excisional biopsy was performed.

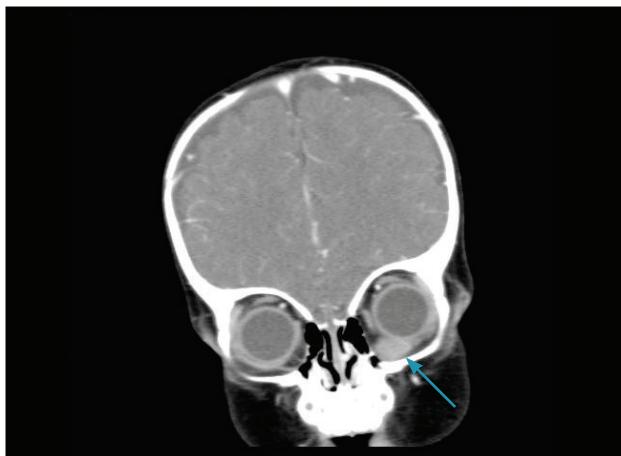


Figure 1

1. What does the arrow show (Figure 1)?
2. What does Figure 2 (H&E) show and what are the arrows pointing to on Figure 3 (PAS)?
3. Figure 4 shows immunohistochemistry for S100 and GFAP in which the brown staining represents positive expression by the epithelial cells. Which other markers may also be positive in this condition?
4. Based on clinico-pathological features what is the diagnosis?
5. How is this lesion formed?

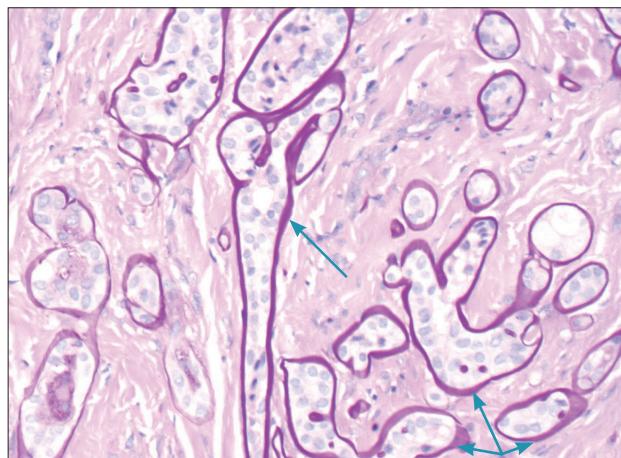


Figure 3

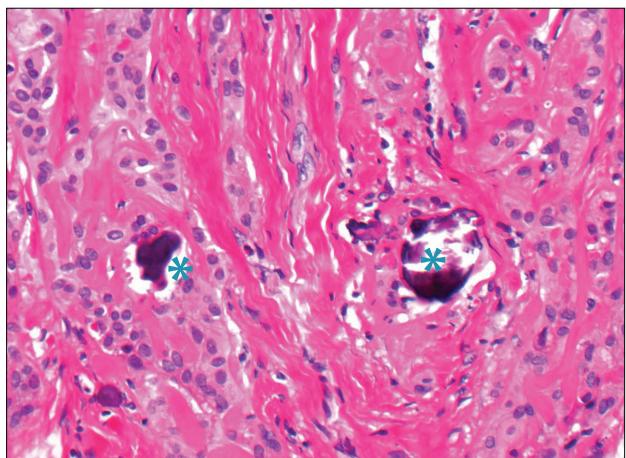


Figure 2

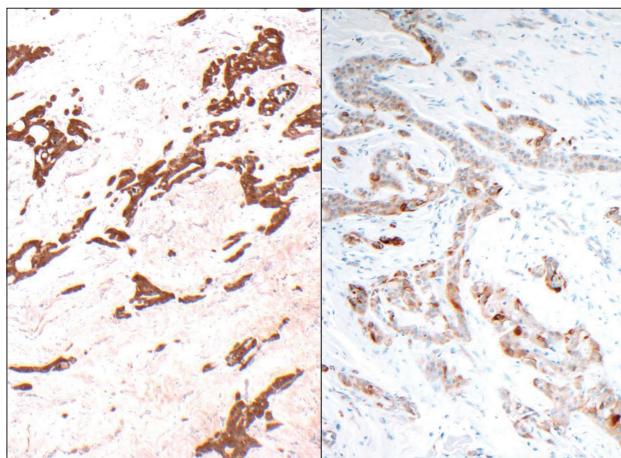


Figure 4

## SECTION EDITOR



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1. Coronal CT scan showing homogeneous mass in left inferonasal anterior orbital quadrant.

2. Figure 2 shows cords of epithelial cells in a dense collagenous matrix. Focal dystrophic calcification is indicated by the blue marks. Figure 3 demonstrates thick basement membrane.

3. Epithelial cells of lenticular arrangement express both S100 and GFAP but are also positive for membranous material produced by and enclosing the epithelial cells.

4. Phakomatous choristoma (also known as Zimmerman's tumour). Coverally, lenticular epithelial cells do not express usual epithelial markers. Vimentin and other antibodies against lens specific proteins (e.g. -lens protein, -crystallin).

5. Phakomatous choristoma is a congenital condition typically noticed in the first few months of life beneath the lower eyelid inferonasally. It consists of nests of lens epithelial-like cells present in an abnormal location (choristoma). The PAS-positive basement membrane of the lens placode into the deeper mesodermal tissue of the lower eyelid. The usual clinical presentation is a dermoid cyst.

## ANSWERS