Newborn with duplication of the pituitary gland (DPG)-plus syndrome and congenital hairy polyp.

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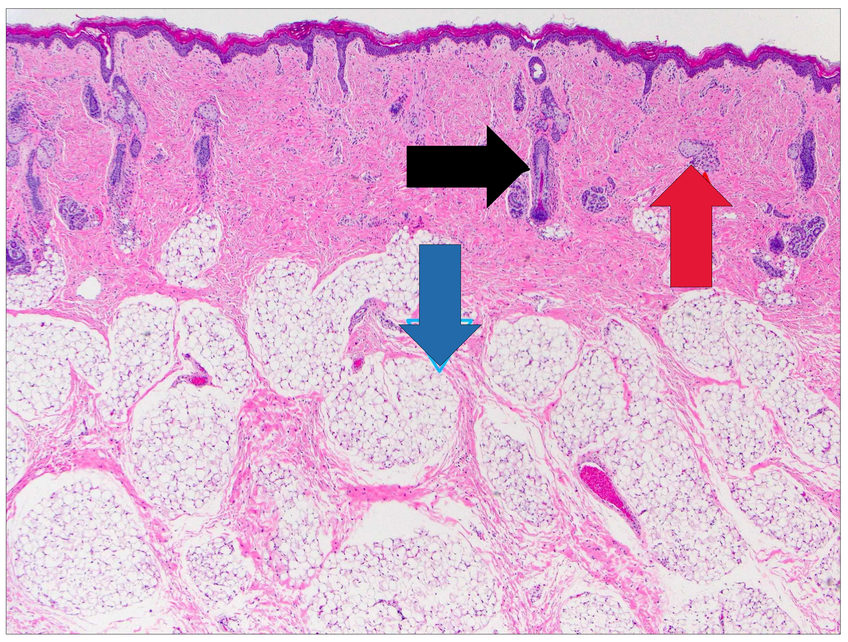
BACKGROUND

Approximately fifty cases of DPG-plus syndrome have been reported worldwide, a small number of which have been associated with nasopharyngeal teratomas.[1,2] A handful of benign hairy polyps have been reported separately, originating within the nasopharynx, esophagus, and eustachian tubes, among other locations.[3] We present what we believe is the first case of DPG-plus syndrome with a congenital hairy polyp.



HISTOLOGIC FEATURES [4]

- Hairy polyps are comprised of tissue derived from ectoderm and mesoderm
- Consist of keratinizing squamous epithelium with underlying adnexa which includes pilosebaceous units
- Mesoderm derivatives include fibroadipose tissue, skeletal muscle, smooth muscle, seromucous glands, and cartilage
- No endodermal elements are present

Key:
Black arrow = hair follicle
Red arrow = sebaceous glands
Blue arrow = fibroadipose tissue

PATIENT PRESENTATION

- Full-term female with prenatally-diagnosed left-sided congenital diaphragmatic hernia with resultant pulmonary hypoplasia (s/p thoracoscopic repair), cleft palate with a nasal/ oral mass, and agenesis of the corpus callosum.
- Subsequent work-up revealed duplication of the pituitary gland (DPG)-plus syndrome.
- The initial plan was to delay resection until the baby was older, however feeding and respiratory support plateaued, with concern that the mass was impacting function.

IMAGING FINDINGS

MRI BRAIN WITHOUT CONTRAST (DOL 6)

- Duplicated pituitary gland.
- Corpus callosal agenesis.
- Cleft palate with incompletely evaluated pedunculated mass arising from the palate/basisphenoid.

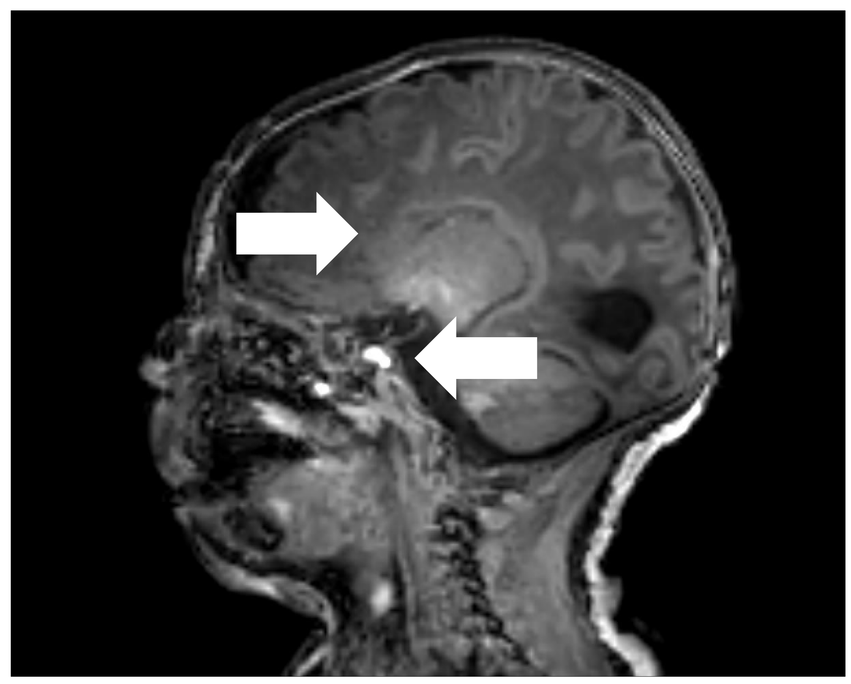
CT SINUS WITHOUT CONTRAST (DOL 7)

- Lobulated mass projecting anteriorly from the basisphenoid through a wide cleft palate into the oral cavity, with features most compatible with congenital nasopharyngeal teratoma.

TREATMENT

- The infant was taken to the OR for a combined trans-oral and trans-nasal endoscopic resection on DOL 28. Intraoperatively, a pedunculated mass arising from the posterior tongue was also resected.
- Feedings did not improve post-operatively, and placement of a gastrostomy tube was ultimately recommended.

Top: Pre-operative image demonstrating a large, smooth soft tissue mass protruding through the patient's cleft palate.
Bottom left: Coronal view of the patient's cleft palate with protruding soft tissue mass (white arrow).
Bottom right: Mid-sagittal image from MRI Brain w/o contrast (DOL 6) demonstrating agenesis of the corpus callosum (top arrow) and duplication of the pituitary gland (bottom arrow).



PATHOLOGY

A. Palate, anterior margin, biopsy:
- Benign fibrovascular tissue with mucinous glands.

B. Soft tissue, nasopharyngeal mass, excision:
- Congenital hair polyp

C. Tongue, midline lesion, excision:
- Lingual hamartoma

Right: Post-operative image (7 weeks s/p endoscopic resection of the patient's hairy cell tumor) showing the patient's cleft palate.

REFERENCES

1) Manjila, S., E. A. Miller, S. Vadera, R. K. Goel, F. R. Khan, C. Crowe and R. T. Geertman (2012). "Duplication of the pituitary gland associated with multiple blastogenesis defects: Duplication of the pituitary gland (DPG)-plus syndrome. Case report and review of literature." Surg Neurol Int 3: 23.
2) Sen, D. and V. Arora (2016). "Duplication of the pituitary gland - plus syndrome." Indian J Radiol Imaging 26(1): 126-130.
3) Tariq, M. U., N. U. Din and M. R. Bashir (2013). "Hairy polyp, a clinicopathologic study of four cases." Head Neck Pathol 7(3): 232-235.
4) Bantumilli S, Maygarden S. Hairy polyp. PathologyOutlines.com website. https://www.pathologyoutlines.com/topic/nasalhairypolyp.html. Accessed September 13th, 2024.

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