Glial choristoma: an unusual case of a congenital tongue lesion in a neonate

Background

Glial choristoma is an uncommon entity that usually presents in the head and neck region, with nasal presentations being the most common. Occurrences in the tongue is rare and can be associated with airway and feeding complications. We describe an unusual case of glial choristoma in a neonate who presented with a large congenital lateral tongue lesion with feeding difficulties.

Case Study

A female neonate was brought in by her parents with a large right lateral tongue lesion at 3 weeks of life. The mass was incidentally discovered by her parents, who noticed difficulty in the baby's feeding due to poor latch and suck. The baby was born at term via uncomplicated vaginal delivery and had no abnormalities identified on baby check.

On examination, a large 1.5cm mass was noted arising from right lateral border of the tongue, sparing tongue base and floor of mouth. The mass was firm, immobile and non-pulsatile.

The neonate was admitted for overnight oximetry, which found no desaturations. There was no acute airway compromise, however light snoring was noted during admission.

Patient underwent a non-contrast MRI head, which found a 12mm ovoid, intermediate signal lesion at the right lateral tongue. There was no large arterial pedicle evident or suggestion for intrinsic hypervascularity. Differentials based on the MRI findings included vascular malformation or rhabdomyosarcoma.

Due to the potential having a malignant soft tissue tumour, the patient underwent an incisional biopsy under general anaesthesia. Intraoperatively, a fleshy, poorly vascular lesion was noted. An incisional biopsy of the lesion was performed, where bulk of the lesion was removed and sent for histopathology. There was minimal bleeding encountered during the operation.

Histopathology examination found the mass to be a glial choristoma. The lesion was poorly defined and lined by squamous non-keratinising epithelium, with mild chronic inflammation. It consisted of fibroconnective tissue with a vascular component, small collections of vacuolated cells adipocytes, irregular, scattered bundles of skeletal muscle with scattered small collections of cells with elongated nuclei and pale eosinophilic cytoplasm arranged in strands and lining cracks or clefts separating the fibroconnective tissue. There was no lobular arrangement of vessels.

These cells stained with S-100. Immunohistochemistry for Glut1 did not stain the endothelium, although there was a small component of lymphatics staining with the 240. CD34 stained the endothelial cells as well as fibroelastic component, but not the interspersed eosinophilic cells. This component did not stain with NSE, desmin, Factor XIIIa, B-catenin, CD68, smooth muscle actin or myogenin.

Discussion

• Glial choristoma is an unusual benign developmental lesion thought to be caused by displaced neuroectoderm in the embryonic period.
• They are found in the nose mostly, but rarely have been reported in the tongue.
• Currently only a few case reports have described this congenital malformation on the tongue.
• The clinical behavior of oral glial choristoma varies depending on the age at onset as well as the location and size of the mass.

Conclusion

Glial choristoma is a rare and interesting diagnosis for congenital tongue lesions. It is benign in nature but there is a risk of airway compromise and obstructive sleep apnoea in neonates and infants. Physicians should be aware of it as a differential in congenital tongue lesions and its management options.