Glial Heterotopia in ENT—Two Case Reports and Review of Literature

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Abstract The purpose of presenting these case reports is to highlight the occurrence of heterotopic glial tissue of the tongue and nose in children. So far, literature review has revealed few case reports of such lesions in neonates, but our patients presented with this unique lesion at the age of two and a half years and 3 years. This is a rare congenital anomaly in the tongue, which can mimic a lingual thyroid, teratoma, dermoid cyst etc. Surgical excision is mandatory when the lesion causes obstructive symptoms. The authors discuss the problems in diagnosis, pathology and management and review the literature.

Keywords Tongue · Teratoma · Thyroglossal cyst · Lingual thyroid · Glial heterotopia · Nasal glioma · Dysphagia

Introduction

Heterotopic neuroglial tissue represents normal brain tissue in an abnormal location away from the CNS. It is a rare congenital anomaly and the majority of these lesions are diagnosed at birth. Schmidt [1] reported the first extracranial and extraspinal heterotropias of neural tissue. Since

then several reports have appeared in the literature [2–4]. The most common location for heterotopic neuroglial tissue is in the base of nose, where it is traditionally but erroneously termed as Nasal “Glioma”. Other heterotopic sites include skin, palate, orbit, scalp, lung, etc. We encountered two cases, one presented as a globular mass in the posterior third of the tongue in a child of two and a half years, which is a rather infrequent location for this choristomatous lesion and the other child of 3 years presented with a unilateral nasal polyp and watery nasal discharge. Failure to recognize this entity is the principle difficulty in diagnosis and complete surgical excision is curative.

Case Report 1

A two and a half year old male child was referred for management of a swelling in the dorsum of the tongue of one-year duration. About a year earlier, the child’s mother had noticed a swelling on the tongue, which was gradually increasing in size. The child gradually developed dysphagia for solids and coughed sometimes on taking liquids. There was no history of change in voice or stridor. The prenatal history had been unremarkable. As the swelling was gradually increasing in size, the parents consulted an otolaryngologist, who suspected a lingual thyroid. A thyroid scan was done using Technetium 99, which revealed no abnormality. The child was brought to us for further management.

The child was well built with no gross craniofacial or other congenital anomalies. Examination of the oral cavity revealed a smooth submucosal, hemispherical, pale swelling over the dorsum of the tongue in the posterior third, which was firm and non pulsatile. The rest of the head and neck was unremarkable. A lateral view of X-ray of the

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neck showed a smooth hemispherical suprahyoid opacity arising from the base of the tongue. Ultrasound of the neck was not contributory. A CT scan of the neck under sedation was attempted, but the child was restless, hence imaging was not optimal.

The child was taken up for surgery after informed consent. Under general anaesthesia with fiberoptic nasotracheal intubation, the mouth was opened with Doyen's mouth gag. The mass in the dorsum of the posterior third of the tongue was visible, about 2×2 cm in size and firm on palpation (Fig. 1). The mucosa over the swelling was not congested and posteriorly it extended to the vallecula in the midline. The tongue was pulled forward with the help of a stay suture and 2% lignocaine with 1: 200,000 dilution of adrenaline was infiltrated around the mass. With bipolar cautery, a midline incision was made over the swelling and an unencapsulated fleshy mass was encountered, which did not bleed excessively. In order to plan further surgery intraoperatively, an intraoperative pathology consultation was sought. This was reported as a mesenchymal lesion of uncertain nature. Subsequently the rest of the mass was excised and bleeding controlled with bipolar cautery. Nasogastric feeds were started on the first postoperative day; oral feeds on the third day and the child recovered uneventfully.

Histopathological examination showed a lesion in the submucosa in between the underlying muscles and mucous glands, composed of a mass of elongated spindled astrocytic cells with round to oval vesicular pale nuclei, consistent with the appearance of glial tissue (Fig. 2). These cells were S-100 protein (Fig. 3) and glial fibrillary acid protein (GFAP) positive, confirming their glial nature. No neuronal, ependymal or choroidal elements were noted. The final histopathological conclusion was glial heterotopia.

Fig. 1 Peroperative picture showing the swelling in the tongue base

Fig. 2 Mass of glial tissue infiltrating the skeletal muscle (H&E $\times 100$)

Fig. 3 S-100 protein positive astrocytic cells (anti S-100 protein, Immunoperoxidase $\times 400$)

Case Report 2

A three-year-old male child presented to the ENT outpatient department with watery nasal discharge from the left nasal cavity of 3 months duration. There was no history of nasal block, fever or sneezing. On examination, child had rhinitis on left side, but he was not cooperative for a detailed examination. A foreign body in the left nostril was suspected. An endoscopic examination of the nose was done under general anaesthesia. A small polypoidal mass was seen arising from the roof of the nose, above and anterior to the middle turbinate.

An MRI of brain and skull bone done under sedation, showed no evidence of bony defect in the floor of the anterior cranial fossa or the presence of encephalocele, meningocele or CSF leak. A polypoidal lesion was seen in left nasal cavity (Fig. 4). Subsequently the child was taken

Fig. 4 STIR (short time inversion recovery) image-showing polyp in the left nasal cavity (shown by the *pointer*)

up for endoscopic nasal polypectomy, under general anaesthesia. The polyp was found to be arising from left nasal septum close to the roof at the level of the middle meatus; it was excised and sent for histopathological examination. A merocele was kept in the left nasal cavity. HPE showed fragments of polypoid tissue lined focally by respiratory epithelium overlying a loose connective tissue matrix composed of glial tissue mainly and focal sub-epithelial mixed inflammatory infiltrates associated with surface ulceration. The features were consistent with glial heterotopia. Further conformation was done by immunohistochemistry. The report was positive for both S-100 and GFAP.

Discussion

Glial heterotopia of the tongue causing dysphagia in a two and a half year old male child was successfully treated by local excision and another child with a nasal polyp was treated by endoscopic nasal surgery. Histologically both specimens showed pure glial elements and no endodermal or mesodermal components. The lesions therefore were diagnosed as heterotopic glial tissue.

Heterotopias represent isolated rests of nervous tissue without attachment to the nervous system. A thorough review of literature revealed only a few cases of heterotopic glial tissue in the tongue. Ofodille et al. reported the first case of glial choristoma of the tongue in 1982 [5]. Neural heterotopias have been reported in the skin [6], pharynx [7], soft palate [8], paranasal sinuses [9], sub-mandibular region [10], tonsillar fossa [11], orbit [12], scalp [6], lung [13] and middle ear. Extracranial congenital malformations of CNS include encephalocele, gliomas and heterotopias [14]. The difference between these partly

depends on the presence or absence of communication with the brain. Encephaloceles are continuous with the spinal cord and brain, whereas gliomas (heterotopias) retain their connection in only 30% of cases. In our case report of the child with nasal polyp, imaging study did not show any connection with the brain.

Most reviews of ectopic neural tissue have speculated on the possible pathogenetic mechanisms of heterotopia [15, 16]. These include an encephalocele that loses its intracranial connection resulting in heterotopic neuroglial tissue [17, 18]. Another mechanism is displacement of neuroectodermal cells during early embryogenesis, which later develops into mature glial tissue [18, 19]. Glial cells migrating from the olfactory bulb is yet another mechanism [17]. Though many possible mechanisms may exist, in our case displacement of totipotent neuroectodermal cells was the possible mechanism to explain the location, as the tongue and nose are ventral structures in development. Heterotopias exhibit the biologic behaviour of a slow growing benign lesion [14]. Recurrences have been reported only in 4–10% of cases and they have been treated by re-excision.

The description of heterotopic neuroglial tissue in classical references on head and neck pathology is sparse, at best meriting only a brief mention. They are composed of nests of neural tissue without mitosis, embedded within varying amounts of fibrovascular stroma [17]. Heterotopias of the tongue described earlier have been found to include ependymal lined structures, choroid plexus and pigmented cells of retinal differentiation apart from the dominant component of astrocytes, neurons and oligodendrocytes [17, 19]. Grossly the tissue is relatively avascular, solid, firm, poorly encapsulated and adherent to the surrounding soft tissues [20]. The removed specimen from the tongue showed similar features and the diagnosis was confirmed with immunohistochemical staining for S-100 protein and GFAP. However, no neuronal or other elements like choroid, ependyma or retinal tissue was noted in either case.

Clinical examination and preoperative imaging with CT and MRI scans cannot distinguish a glial choristoma from other midline swellings of head and neck. Benign lesions at this location include teratoma, haemangioma, lipoma, rhabdomyoma, leiomyoma, dermoid cyst, thyroglossal duct cyst and lymphatic malformations [21]. These benign lesions can be distinguished only by histopathological and immunohistochemical examination. However, CT and MRI images are helpful in delineating the extent of the lesion and its relation to surrounding structures. Surgical intervention is indicated for lesions causing airway distress, dysphagia or failure to thrive. The tongue lesion in the first case was approached by the less morbid intraoral corridor and in case of difficulty, suprahyoid route was contemplated. In the second case the mass was removed by

endoscopic removal without any consequential morbidity. Both patients did well after surgery.

Conclusion

Heterotopic neuroglial tissue of the tongue in a two and a half year old boy and glial tissue of nasal cavity in a three-year-old boy are presented for its rarity. The tongue swelling caused dysphagia and could have caused dysarthria and respiratory distress, if not excised. Complete excision of the mass was achieved by the intra-oral route, with minimal morbidity. Recurrence is unlikely as the lesion was completely removed, however, the child is under close follow up. The child in the second case with nasal glioma in the left nostril underwent endoscopic removal. Endoscopy is the key for diagnosis and management. The child was symptomatically rid of nasal block but continued to have watery rhinorrhoea. CT and MRI images did not contribute in the diagnosis. Histopathology and immunochemistry helped in clinching the diagnosis in these cases. Hence these cases merit documentation.

Key Messages

- Heterotopic glial tissue in the tongue is a rare presentation. Occurrence of this lesion in a two and a half year old child is unique; so far all case reports in literature were in neonates.
- The diagnosis was uncertain until the final pathological report.
- Complete excision was possible by intra oral approach.
- Special stains—S 100 protein and GFAP were positive, confirming lingual glial heterotopia.

References

1. Schmidt MB (1900) Ueber seltene spaltbildungen im Bereiche des mittleren Stirn fortsatzes. *Arch (Pathol Anat)* 162:365

2. Bratton AB, Robinson SHG (1946) Gliomata of the nose and oral cavity. A report of two cases. *J Pathol Bacteriol* 58:643
3. Potter EL (1953) Pathology of the foetus and the new born. Yearbook publishers, Chicago
4. Mosadomi A (1975) Neurilemmoma of the tongue. *J Oral Med* 30:44
5. Ofodile FA, Aghadiuno PU, Oyemade O (1982) Adebajo T: heterotopic brain in the tongue. *Plastic and Reconstr Surg* 69:120–124
6. Orkin M, Fischer I (1966) Heterotopic brain tissue (heterotopic neural rest). *Arch Dermatol* 94:699–707
7. Ruff T, Diaz JA (1986) Heterotopic brain in the nasopharynx. *Arch Otolaryngol Head Neck Surg* 94:254–256
8. Shapiro MJ (1968) Mix BS: heterotopic brain tissue of the palate. *Arch Otolaryngol Head Neck Surg* 87:522–526
9. Genut AA, Miranda FG, Garcia JH (1976) Organized cerebral heterotopia in the ethmoid sinus. *J Neurol Sci* 28:339–344
10. Okeda R (1978) Heterotopic brain tissue in the submandibular region and lung—report of two cases and comments about pathogenesis. *Acta Neuropathol* 43:217–220
11. Feldman BA, Schwartz RH, Chandra R et al (1982) Heterotopic brain simulating a neonatal tonsil. *Clin Pediatr* 21:428–430
12. Smith KR, Schwartz HG, Luse SA et al (1963) Nasal gliomas—A report of five cases with electron microscopy of one. *J Neurosurg* 20:968–982
13. Fuller C, Gibbs A (1989) Heterotopic brain tissue in the lung causing acute respiratory distress in an infant. *Thorax* 44: 1045–1046
14. Knox R, Pratt M, Garvin AJ, White B (1989) Heterotopic lingual brain in the newborn. *Arch Otolaryngol Head Neck Surg* 115:630–632
15. Bratton AB, Robinson SHG (1948) Gliomata of the nose and oral cavity: a report of two cases. *J Pathol* 58:643–648
16. Batsakis JG (1979) Other neuroectodermal tumors and related lesions of the head and neck in tumors of the head and neck, 2nd edn. Williams & Wilkins, Baltimore, pp 334–337
17. Patterson K, Kapur S, Chandra RS (1986) Nasal gliomas and related brain heterotopias: a pathologist's perspective. *Pediatr Pathol* 5:353–361
18. Cohen AH, Abt AB (1970) An unusual case of neonatal respiratory obstruction—heterotopic pharyngeal brain tissue. *J Pediatr* 76:119–122
19. Kallman JE, Loevner LA, Yousem DM et al (1997) Heterotopic brain in the Pterygopalatine fossa. *Am J Neuroradiol* 18:176–179
20. Behar PM, Muller S, Gerber ME, Todd NW (2001) Heterotopic neuroglial tissue causing airway obstruction in the new born. *Arch Otolaryngol Head Neck Surg* 127:997–1002
21. Takamizawa S, Inoue T, Ono Y, Satoh S, Nishijima E, Muraji T et al (2006) A case report of glial choristoma of the tongue. *J Pediatr Surg* 41:E13–E15