

Glial Heterotopia of the Base of Tongue: A Case Report

Dil Tabanının Glial Heterotopisi: Bir Olgu Sunumu

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ABSTRACT

Glial heterotopia of oropharynx is a congenital anomaly, whereby ectopic mature glial tissue is found around oropharynx isolated from the brain and spinal cord. Herein we report a rare presentation of a mass at the base of tongue in a neonate. In addition, to underscore the rarity of oropharyngeal glial heterotopia, we discuss the dilemma in approaching its diagnosis and management in a neonate.

Keywords: Glial heterotopia, base of tongue, congenital

ÖZ

Orofarinksin glial heterotopisi, ektopik matür glial dokunun orofarinks çevresinde beyin ve omurilikten izole olarak ayrı bir şekilde bulunduğu bir konjenital anomalidir. Burada bir yenidoğanda dil tabanındaki bir kitlenin nadir bir prezentasyonunu sunmaktayız. Buna ek olarak orofarinksin glial heterotopisinin enderliğine vurgu yapmak üzere yenidoğanda tanısı ve tedavi yönetimindeki ikilemi tartışmaktayız.

Anahtar kelimeler: Glial heterotopi, dil tabanı, konjenital

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INTRODUCTION

Congenital mass at the base of tongue is uncommon and usually benign in nature. It can be due to inflammation, neoplasm or structural abnormalities developing during embryological development. The diversity of its etiology has proven to be a challenging issue in making the correct diagnosis. Glial cells are normally located in the central nervous system and serve as physiological support for the neural cells¹. Glial heterotopias at the base of tongue are rare developmental lesions that are located at the extracranial midline structure². They are potentially life-threatening when the airway or the swallowing function is impaired. Surgical intervention to excise the lesion is mandatory in the event of airway compromise.

CASE PRESENTATION

A two-day-old infant was referred to the otolaryngology department for evaluation of the upper aerodigestive tract when she developed stridor after birth. She was born full term preceding an uncomplicated pregnancy with a satisfactory birthweight of 2940 grams and a good Apgar score of eight. The stridor was accompanied by chest recession at two hours after birth. Her airway was adequately maintained with a nasal continuous positive airway pressure ventilation for a day before she deteriorated and required oral intubation using a size 3.5 Fr endotracheal tube. Physical examination revealed neither craniofacial abnormalities nor hemangiomas on the face and extremities. Intraoral examination at the base of tongue revealed a solitary midline submucosal mass with a broad base and covered with normal tongue tissue. The mass felt firm and non-pulsatile with a size of approximately 2 cm x 2 cm. Upon flexible nasopharyngoscopy, the mass occluded the retropalatal space completely. A direct laryngoscopy with telescopic view revealed a non-friable mass and was encroaching the vallecula. There was no obstruction below the level of the mass.

Ultrasonographic examination showed a well-defined hypoechoic lesion at the base of tongue with minimal internal vascularity, and confirmed the presence of a normal thyroid gland in the anterior neck. Contrast-enhanced computed tomography (CT) scan revealed a non-infiltrative, predominantly hypodense mass measuring 2.3 cm (width) x 1.6 cm (height) x 1.8 cm (length) occupying the oropharynx, of which origin was likely to be at the base of tongue. Any communication with



Figure 1. (*) Base of tongue mass, (A) endotracheal tube.

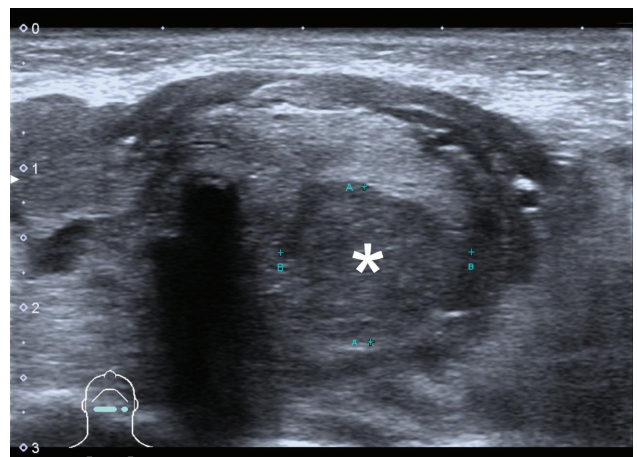


Figure 2. (*) Oblique axial ultrasound of the neck showing a homogeneous hypo-echoic solid mass on the base of tongue.

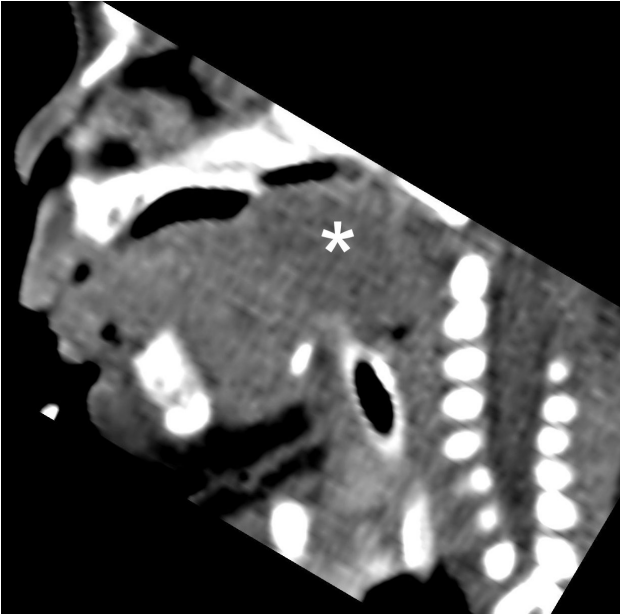


Figure 3. (*) Contrast - enhanced CT in oblique section demonstrating a hypodense mass arising from the base of tongue and occupying the oropharynx with well-defined margins.

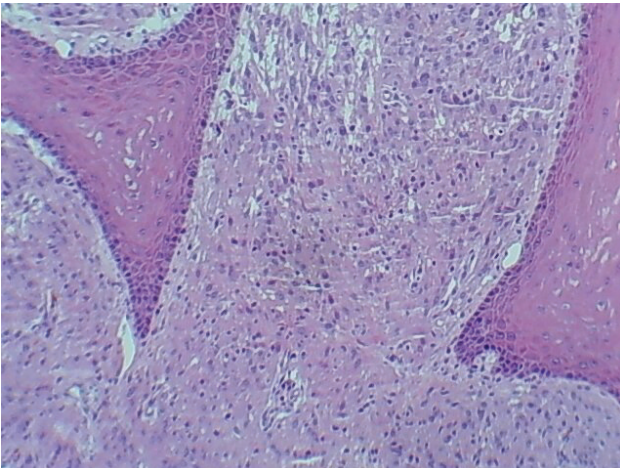


Figure 4. Glial tissue with overlying epithelium.

the cranial cavity or with the normally-positioned thyroid gland could not be demonstrated. The absence of high-density iodine content essentially ruled out ectopic thyroid tissue in this case. In addition, presence of a cystic area, calcification or fat could not be demonstrated.

The tumour along with an additional 1 cm diameter of healthy tissue surrounding the lesion was excised under general anaesthesia with electro-

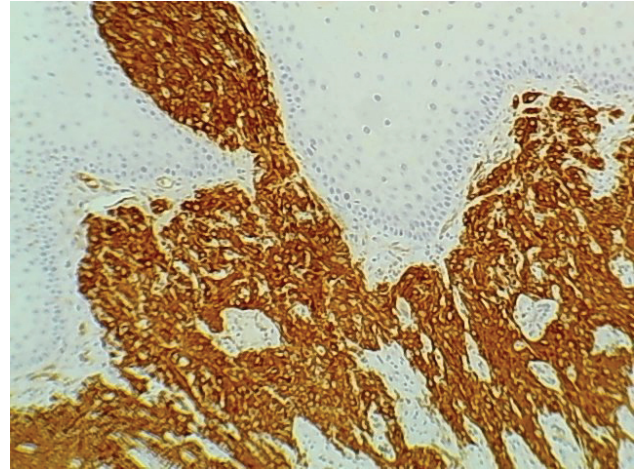


Figure 5. Glial tissue showed immunoreactive towards GFAP stain (x400).

cautery. Histopathological examination showed clumps of brain tissue (mature glial tissue composed of astrocytes, gliosis, and variable stromal fibrosis) overlined by stratified squamous cell which was diffusely immunoreactive to glial fibrillar acidic protein (GFAP). The child recovered well postoperatively with no recurrence seen during follow-up.

DISCUSSION

Glial heterotopia involving the dorsal surface of cervical spinal cord was first described by Wolbach in 1907²⁻⁴. Subsequently, few cases were reported worldwide. The locations that have been described include oral cavity, middle ear, tonsillar fossa, orbit, lung and skin and soft tissue^{2,5}. The nose and nasopharynx area are the most reported site of occurrence². Glial heterotopia, glial choristoma, and gliomatous teratoma are interchangeable terms that are used to describe a mere collection of normal mature glial tissue rather than true neoplasm, which is located at an abnormal anatomical site⁶. This rare congenital anomaly is reported to have a slight male predominance with a ratio of 3: 1⁷. Its estimated occurrence is 1 in 20.000 to 40.000 births⁷.

The pathogenesis of glial heterotopias remains unclear with various hypotheses being postulat-

ed. Harris et al proposed that the development of such lesion may be attributed to separation of an accessory third evagination of the neural tube from the developing central nervous system, which is located at the base of the telencephalic vesicles^{3,8}. On the other hand, Birrell et al, suggested the possibility of a displaced neural tissue through a persistent communication between cranial end of foregut (Seessel's pouch) and the base of occiput portion of the foetal head. An alternative theory suggested formation of a sequestered encephalocele following the formation of Ratke's pouch when a portion of craniopharyngeal canal persisted⁹. It is nevertheless being agreed upon that glial heterotopia represents a non-neoplastic developmental anomaly, even though cases of malignant transformation have been reported^{1,2}. Based on its location and putative pathological mechanism, Gyure et al further classified glial heterotopia into five categories which include intraparenchymal lesions, dural and leptomeningeal lesions, intracranial extracerebral lesions, distal lesions of the lung and uterus and last but not least, midline nasal glioma with related head and neck lesions^{3,4}.

The atypical location of a glial heterotopia at the base of tongue tend to mimic other common congenital lesions involving base of tongue such as lingual thyroid, teratoma, and thyroglossal duct cyst, thereby clouding the diagnosis making process^{5,6}. Imaging is helpful to provide guidance and support diagnosis. Ultrasonographic evaluation is deemed necessary to evaluate the thyroid gland, to determine the characteristic of the lesion as well as to assess the vascularity. CT and magnetic resonance imaging (MRI) are used to delineate the extension of the lesion and the involvement of the surrounding structures. However, they are unable to differentiate glial heterotopia from other midline base of tongue lesions⁵. MRI may play a superior role in evaluating the soft tissue of the surrounding oropharynx and oral cavity. In our case, the infant was intubated prior to performing any imaging modality. As a

result of the long duration of the scanning process with continuous interference from positive airway pressure ventilation, the images of an attempted MRI were, unfortunately useless because of motion artefacts.

The diagnosis of a glial heterotopia can only be confirmed through histopathological examination. Under the naked eye, it appears as a solid, and firm mass adherent to the surrounding soft tissue⁴. Morphologically, the lesion consists of mature astrocytes and glial fibres embedded in a fibrovascular stroma^{5,10}. Unusual components such as neurons, choroid plexus, and oligodendrocytes have also been found¹⁰. The lesion is immunoreactive to glial fibrillary acidic protein (GFAP), S100-protein, vimentin, CD 57, neuron-specific enolase (NSE) and neurofilament (NF) stains¹⁰. Surgical excision is the treatment of choice with a reported low rate of recurrence ranging from 4% to 10%^{2,5}. Rapid growth of a glial heterotopia has been described¹. Therefore, an early intervention is generally advocated regardless of its location to attain functional improvement and prevent complications.

In conclusion, glial heterotopia represents a rare entity and should be included as one of the differential diagnosis of a congenital lesion of the tongue base. Histopathological examination is the only avenue of confirming its diagnosis. Given its close to the airway, early intervention of any congenital tongue base mass must be carried out to avert potentially disastrous consequences.

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