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Case Report

Parapharyngeal Neuroglial Heterotopia: A Case Report

Reza Saeidi[1], Mahmoud Hajipour[2], Ahmad Shah Farhat[3], Ashraf Mohammadzadeh[3], Fatemeh Serati[1*]

1. Neonatal Health Research Center, Research Institute for Children's Health, Shahid Beheshti University of Medical Sciences, Tehran, Iran
2. Pediatric Gastroenterology, Hepatology and Nutrition Research Center, Research Institute for Children's Health, Shahid Beheshti University of Medical Sciences, Tehran, Iran
3. Neonatal Research Center, Mashhad University of Medical Sciences, Mashhad, Iran

ABSTRACT

Background: Neonatal neck masses usually fall into three categories: developmental, inflammatory, and neoplastic. Malignant neck masses are rarely observed in neonates; nonetheless, If malignancy is suspected, the patient should be evaluated urgently. In this paper, we describe the case of a neonate with a very rare congenital neck tumor, nasal glial heterotopia, which was first described by Reid in 1852, and only 20 cases have been reported in the parapharyngeal space in an article published in 2005.

Case report: This case report presents a 3-month-old girl misdiagnosed with cystic hygroma at one month. She finally underwent surgery due to the progress of symptoms, airway obstruction, and mass neck enlargement. The pathological diagnosis confirmed a rare case of parapharyngeal neuroglial heterotopia.

Conclusion: One case of glial heterotopia of the parapharyngeal space was included and presented in the present study. Glial heterotopias are very rare in the parapharyngeal space. Histopathology and immunohistochemistry are essential to diagnose this lesion.

Keywords: Airway obstruction, Neck neoplasms, Neuroglia

Introduction

Neonatal neck masses usually fall into three categories: developmental, inflammatory, and neoplastic. Common congenital developmental masses in the neck include thyroglossal duct cysts, branchial cleft cysts, dermoid cysts, vascular malformations, and hemangiomas. Inflammatory neck masses can be the result of reactive lymphadenopathy, infectious lymphadenitis, or Kawasaki disease. Common benign neoplastic lesions include pilomatrixoma, lipomas, fibromas, neurofibromas, and salivary gland tumors. Malignant neck masses are rarely observed in neonates in the include lymphoma, rhabdo-myosarcoma, thyroid carcinoma, and metastatic nasopharyngeal carcinoma. If malignancy is suspected, the patient should be evaluated urgently. Heterotopic neuroglial tissue is defined as a mass composed of mature brain tissue isolated from the cranial cavity or spinal canal. The nose and the nasopharynx are the most common sites of location(1, 2).

Neuroglial heterotopia is a rare disease that was first described by Reid in 1852. Unlike encephalocele, neuroglial heterotopia is a benign ectopic neural tissue with no intracranial connection(3, 4). The proposed theories for the pathogenesis of this type of mass include descending brain tissue (encephalocele) through a skull defect, which finally closes to turn the encephalocele into neuroglial heterotopia(5). Another theory is the displacement of neuroectodermal cells or the attachment of the neuroectoderm to the surface ectoderm due to the failure of closure of the anterior neuropore(5, 6). Another proposed theory is olfactory bulb glial cells(4).

Neurons can still be observed in 10% of these cases, and an active choroid plexus can be observed

* *Corresponding author*: Fatemeh Serati, Neonatal Health Research Center, Research Institute for Children's Health, Shahid Beheshti University of Medical Sciences, Tehran, Iran. Tel: +982122061304; Email: saeedir@mums.ac.ir

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in pharyngeal neuroglial heterotopia(1,7). This describes the cystic component mechanism, which is indeed a cerebrospinal fluid produced by the active choroid plexus(8). Neuroglial heterotopia is a rare condition that has been diagnosed in only 200 cases mainly in the nasal cavity (mistakenly named nasal glioma)(8). Sometimes extra-nasal glial heterotopias occur in the scalp, orbit, palate, tongue, lips, middle ear, and pharynx(6). Neuroglial heterotopias in the parapharyngeal space are very rare, with fewer than 20 cases being reported in an article published in 2005(8). In this paper, we describe an intrauterine growth restriction neonate with nasal glial heterotopia who developed respiratory distress and neck mass.

Case report

The neonate was born via a cesarean section with a left-sided neck masse and respiratory distress on 2021 September 18 in Ahvaz, Iran. The patient was a term neonate with intrauterine growth retardation and a birth weight of 1,670 g. We were suspected of teratoma (cystic hygroma) and hemangioma. After complete mass resection on December 19, 2021, the patient respiratory distress continued. A repeated bronchoscopy demonstrated a multilobular mass resembling a nasopharyngeal lesion to the oropharynx with 90% obstruction, and severe laryngomalacia was diagnosed.

A computer tomography (CT) scan with contrast showed a large mass in the neck in the left submandibular space and the retropharyngeal space, causing nasopharyngeal and oropharyngeal stenosis. The lesion also caused carotid displacement and mandibular deformity. The facial artery provides the blood follow to this mass. The medial parts of the mass are displayed in the retropharynx of the cystic components. The most prevalent differential diagnoses in this age group are congenital causes, such as vascular abnormalities and teratomas. Nonetheless, other differential diagnoses are also considered, including deep neck infection, neural tube tumors, lymphoma, and other malignancies.

Video bronchoscopy revealed a multilobulated mass-like lesion from the nasopharynx to the oropharynx with 90% obstruction, shortness of aryepiglottic folds, and prolapse between TVC inspiration, normal vocal cords, and severe laryngomalacia. Microscopic pathology reporting pointed to an abnormal disorganized glial tissue with increased cellularity and aggregation of oligo-like cells with microcalcification. The final diagnosis was a submandibular mass, left, resection-neuroglial heterotopia.

Discussion

The literature review demonstrated very few cases of parapharyngeal neuroglial heterotopia across the globe. The majority of patients were girls, and their masses were located on the left side(5). Some cases had other abnormalities, such as heart defects, Pierre Robin sequence, and cleft palate(3). Clinical manifestations depend on the mass size and compressive effect on the adjacent structures. The patients typically present with respiratory distress, nutritional problems, stunting, and the presence or absence of neck masses. Sonography diagnosed one of the studied cases before birth who needed intrauterine therapy(5).

In the case of our patient, the first sign was respiratory distress. To diagnose this type of neck mass, radiological imaging was conducted. The CT scan was also carried out to obtain more details about the mass, as well as its size and location, and determine whether there was an intracranial connection. Diagnostic problems are often due to the normal prenatal history, lack of specific symptoms and signs, as well as the absence of confirmed pathognomonic features in radiology images. Lymphatic malformations are often the preferred differential diagnosis in these cases.

A characteristic observed in the majority of cases is bone deviation and displacement, particularly in the mandible and pterygoid plates, which often show parapharyngeal neuroglial heterotopia characteristics(4, 5, 9). In the majority of investigated cases with parapharyngeal neuroglial heterotopia, masses indicated both cystic and solid components, except in six cases that only showed a cystic mass(5, 10). The cystic component, the cerebrospinal fluid, is produced by the active choroid plexus in the neuroglial tissue(4). Neuroglial heterotopia is diagnosed only after the surgical removal of the mass(5).

At present, complete surgical excision is the standard disease management method (the current criterion standard treatment). The suitable time for surgery continues to remain challenging since it is more difficult when it comes to children. Nevertheless, delayed surgery may cause respiratory distress, nutritional problems, stunting, and cranial nerve palsy with mass enlargement. Our patient also underwent surgery due to respiratory distress, oxygen unsaturation during sleep, poor oral intake, and stunting(4, 5).

One of the subjects was monitored for six months; nonetheless, the mass became larger due

to an increase in the cystic component necessitating surgical intervention. Liquid drainage was also suggested as an option; however, it resulted in rapid fluid re-accumulation(10). A detailed follow-up of these patients is necessary due to the risk of recurrence, Tissue regrowth caused by recurrence is due to incomplete removal of the mass. Complete mass removal (as far as possible) is recommended; nevertheless, it is sometimes very difficult. Therefore, a stepwise removal is also recommended (7).

Conclusion

Neuroglial heterotopias is a rare differential diagnoses in congenital neck masses that may cause airway obstruction and respiratory distress.

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None

Conflicts of interest

None

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