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Parapharyngeal Neuroglial Heterotopia: A Case Report and Literature Review

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Patient: Female, 6-month-old
Final Diagnosis: Parapharyngeal neuroglial heterotopia
Symptoms: Facial swelling • failure to thrive • nasal obstruction • snoring
Medication: —
Clinical Procedure: —
Specialty: Otolaryngology • Pediatrics and Neonatology

Objective: Rare disease
Background: Pediatric neck masses have a wide differential diagnosis. Neuroglial heterotopia is a rare condition that was first described by Reid in 1852. The majority of neuroglial heterotopias are found in the nasal cavity (mistakenly termed as nasal glioma), but they can also occur in extra-nasal areas such as the scalp, orbit, palate, neck, and other areas. Only 20 cases of neuroglial heterotopia in the parapharyngeal space have been reported.
Case Report: In this case report, we present a 6-month-old girl who was misdiagnosed with unilateral choanal atresia at 1 month of age. As her symptoms progressed to airway obstruction and the size of her neck mass increased, she eventually required surgical management. The pathological diagnosis confirmed a rare case of parapharyngeal neuroglial heterotopia.
Conclusions: Neuroglial heterotopias is one of the differential diagnoses for masses causing airway obstruction in pediatric age groups. Preoperative diagnoses of parapharyngeal neuroglial heterotopias can be challenging, as they have no confirmed specific clinical or radiological features. This paper contributes to parapharyngeal neuroglial heterotopia research, which will ultimately enable clinicians to ascertain these tumors' characteristic features more promptly for earlier diagnoses.

MeSH Keywords: Airway Obstruction • Head and Neck Neoplasms • Nasal Obstruction • Neck • Neuroglia

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Background

Pediatric neck masses have a wide differential diagnosis. Neuroglial heterotopia is a rare condition that was first described by Reid in 1852 [1,2]. Neuroglial heterotopia is a benign ectopic neural tissue with no intracranial connection, unlike encephalocele.

Proposed theories for the pathogenesis of this type of mass include descending brain tissue (encephalocele) through a skull defect that eventually closes so the encephalocele evolves into neuroglial heterotopia [3]. Another theory is the displacement of neuroectodermal cells [1,2] or attachment of the neuroectoderm to the surface ectoderm due to failure of anterior neuroepithelial closure [3]. Another proposed theory is the entrapment of olfactory bulb glial cells [1,2].

Neurons can be still present in 10% of these cases and a functioning choroid plexus can be found in pharyngeal neuroglial heterotopia [2]. This suggests the mechanism of cystic component, which is in fact cerebrospinal fluid-like fluid [2] produced by the functioning choroid plexus [4].

Neuroglial heterotopias are unusual. Only 200 cases have been reported, and the majority presented in the nasal cavity (mistakenly termed as nasal glioma) [1–4]. Extra-nasal glial heterotopias occasionally occur in the scalp, orbit, palate, tongue, lips, middle ear, pterygopalatine fossa, and pharynx [5]. Neuroglial heterotopias in the parapharyngeal space are extremely rare; fewer than 20 cases were reported in an article published in 2005 [4].

Case Report

Our patient was a 6-month-old medically healthy girl born via spontaneous vaginal delivery after an uneventful pregnancy. She was full term with a birth weight of 3 kg. She was discharged home in good condition with her mother.

At 1 month of age, she presented to a small hospital with a fever and symptoms of upper respiratory tract infection with a nasal discharge more from the left side. On examination, a catheter could not be inserted into her left nostril; hence, unilateral choanal atresia was provisionally diagnosed with no radiological investigation at the time. Her symptoms improved after antibiotic treatment.

When she was 4 months old, her mother noticed swelling on the left side of her face along with decreased oral intake and fever. She was again treated with antibiotics, which improved her pain, overlying redness, and the amount of swelling, but the swelling did not resolve.

At 6 months of age, she presented to our hospital with left facial swelling for the prior 2 months associated with poor feeding, failure to thrive, and snoring. The size of the mass had minimally progressed, and the patient had no pain or shortness of breath. On examination, her growth parameters were below the fifth percentile, her vitals were stable, and she had no respiratory distress. A 4×3 cm mass was present over the left parotid area, but there were no overlying skin changes and it was not tender with palpation. A throat examination revealed a hard left palate bulge.

The patient was admitted. Her blood work was unremarkable. Her white blood cell (WBC) count was 15 (reference range 6–18), C-reactive protein (CRP) was 6.26 (reference range 0–3), and polymerase chain reaction (PCR) was negative for human immunodeficiency virus and hepatitis B and C. Computed tomography (CT) with contrast (Figure 1) showed a 6×4 cm enhanced cystic lesion in the left parapharyngeal space. The mass extended to the masticator space, retromandibular space, and skull base with erosion of the left sphenoid bone, subluxation of the left temporomandibular joint, airway narrowing caused by the mass, and no choanal atresia. Magnetic resonance imaging (MRI) (Figure 2) showed the same findings. The cystic mass was enhanced in T2 images with no intracranial connection. The final report suggested combined venolymphatic malformations. The most frequent differential diagnoses in this age group include congenital causes such as vascular anomalies and teratomas. However, other differential diagnoses, including deep neck space infection, nerve sheath tumors, lymphoma, and other malignancies, are also considered.

Ultrasound-guided fine-needle aspiration found clear fluid stained with blood, but no pus was aspirated. The patient's WBC count was 1278, red blood cell (RBC) count was 275, lymphocytes were 58%, and monocytes were 32%. The culture was positive for *Streptococcus pneumoniae*, and the cytology was negative for malignancy.

After discussion, the patient was planned for surgery as she had symptoms of airway obstruction (snoring and on and off oxygen desaturation during sleep) and poor oral intake with failure to thrive. Based on radiological imaging, lymphatic malformation was our working diagnosis.

During surgery, dissection was performed in layers until the parotid fascia was opened. The mass was identified. It had a hard consistency; hence, dissection was initiated around it. The facial nerve was identified and preserved, and part of the mass was below it. Superiorly, it reached the skull base. The mass adhered to the patient's pterygoid plates. Due to the mass extension and pterygoid plexus bleeding that was eventually controlled, these 2 parts (the part below the facial nerve and another part attached to the skull base) were left

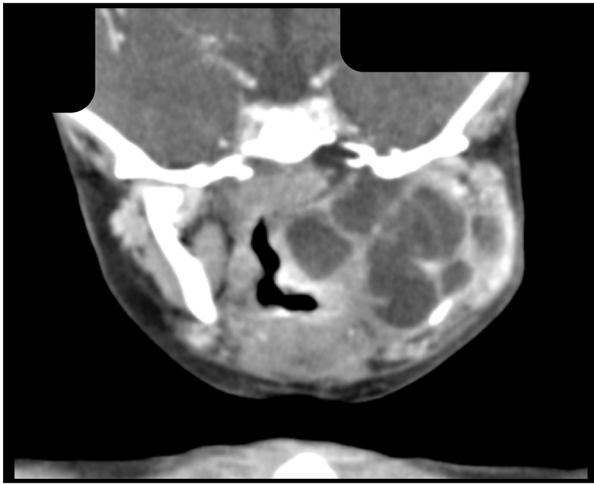


Figure 1. CT – coronal section showing the mass and the airway narrowing.

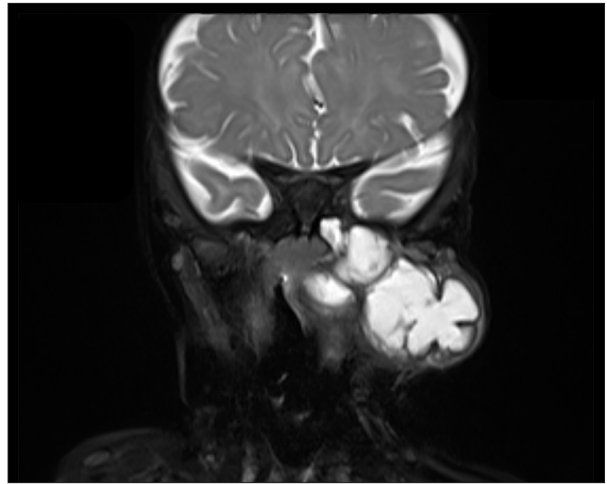


Figure 2. MRI – coronal section showing the T2 enhanced cystic lesion.

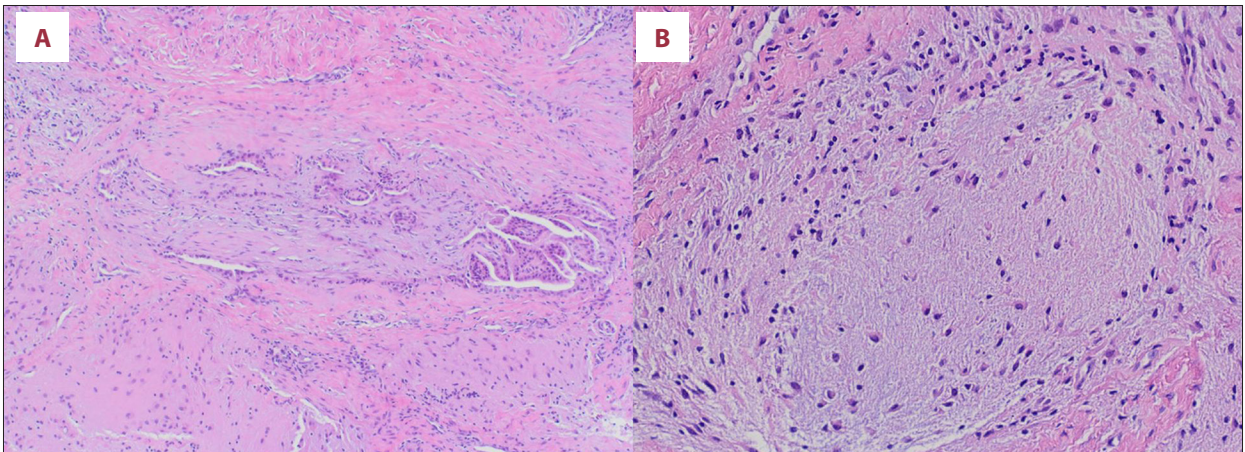


Figure 3. (A, B) H&E stain – glial tissue.

as the frozen section came back as a benign lesion. Closure was done. We waited for the permanent sample histopathology report to treat the patient accordingly. Postoperatively, the patient had grade IV facial palsy but was otherwise fine and tolerating oral intake.

The final histopathology report was positive for glial fibrillary acidic protein (GFAP) and S100 in the neuroglial tissue and cytokeratin 7 (CK7) in the choroid plexus (Figures 3, 4). The patient was diagnosed with mature glial tissue and neuroglial heterotopia.

A 2-week postoperative CT reported postoperative changes with incremental improvement of airway narrowing and no discrete collections. The patient was discharged home. Regular follow-ups and an MRI were scheduled for 6 months postoperatively. One month postoperatively, her facial nerve paralysis had improved to grade II.

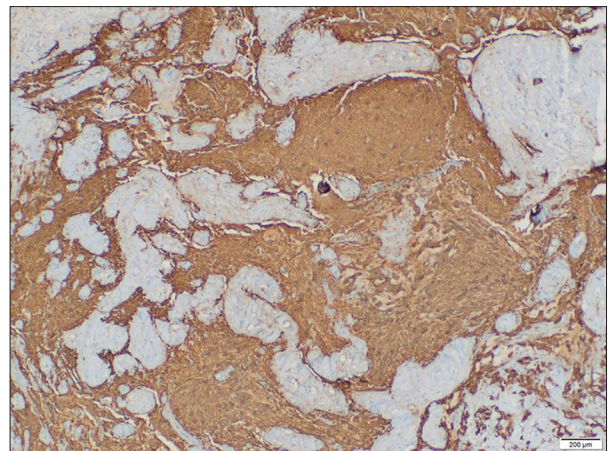


Figure 4. GFAP immunostain – glial tissue.

Discussion

Our literature review found very few parapharyngeal neuroglial heterotopia cases worldwide. Most patients were females, and the mass was on the left side [4,5]. Some cases were associated with other anomalies such as heart defects, Pierre Robin sequences, and cleft palates [1].

Clinical presentation of these cases depends on the mass size and the compression effect on adjacent structures. Patients usually present with respiratory distress, feeding difficulties, failure to thrive, and the presence or absence of neck masses. One of the cases reviewed was discovered through prenatal ultrasound and required ex utero intrapartum treatment (EXIT) [3]. In our case, the first presentation was left-sided nasal discharge that was mistakenly diagnosed as unilateral choanal atresia. To diagnose this type of neck mass, radiological imaging, CT, and MRI are preferred to further determine the mass details, location, and extent and ascertain whether there is any intracranial connection.

Diagnostic difficulty is often caused by an unremarkable prenatal history, no specific signs and symptoms, and no confirmed pathognomonic features on radiological investigations. Lymphatic malformation is usually the top differential diagnosis in these cases.

One feature observed in most cases is bone distraction and displacement, especially the mandible and pterygoid plates, which often suggest parapharyngeal neuroglial heterotopia features [2,6]. In our case, only mild erosion was noted in the left sphenoid bone.

In most of the reviewed parapharyngeal neuroglial heterotopia cases, the masses demonstrated both cystic and solid components, except for 5 cases that presented as cystic masses, similar to our patient [3,7]. The cystic component, which is in fact cerebrospinal fluid [2], is produced by a functioning choroid plexus [4] in the neuroglial tissue. Fine-needle aspiration of our patient reported 58% lymphocytes. Neuroglial heterotopia was diagnosed only after surgical excision, which was positive for GFAP and S100 in the neuroglial tissue and CK7 in the choroid plexus.

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At present, complete surgical excision is the criterion standard management. The timing of surgery remains controversial as neonatal procedures are more difficult due to the surgical plane. However, delaying surgery may cause respiratory distress, feeding difficulties, failure to thrive [2], and cranial nerve palsy [3] as the mass progresses in size. Surgical management was selected for our patient because she demonstrated snoring, oxygen desaturation during sleep, poor oral intake, and failure to thrive.

Observation was attempted in one of the reported cases for 6 months [3], but the mass progressed due to increased cystic components; therefore, surgical intervention was eventually necessary. Drainage was also proposed as an option, but it led to the rapid re-accumulation of fluid [6].

Careful follow-up is required in these patients as the risk of recurrence is up to 30% [7] (which is in fact tissue regrowth due to incomplete excision [5]). Complete resection as much as possible is recommended [2] but it is sometimes difficult to achieve; hence, staged resection is also proposed [5,7].

Conclusions

Neuroglial heterotopias should be considered as differential diagnoses for masses causing airway obstruction in pediatric age groups. Labelling patients with unilateral choanal atresia without investigation may delay diagnoses of these cases. Preoperatively diagnosing parapharyngeal neuroglial heterotopias is difficult, as there are no confirmed specific clinical or radiological features. This paper contributes to parapharyngeal neuroglial heterotopia research, which will ultimately enable clinicians to ascertain these tumors' characteristic features more promptly for earlier diagnosis.

Conflicts of interest

None.