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Case Report

# Case series of congenital heterotopic neuroglial tissue in the parapharyngeal space

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ABSTRACT

Cases of congenital heterotopic tissue presenting in the head and neck are frequent in the pediatric otolaryngology literature. Heterotopic glioneuronal tissue is rare and fewer than 20 cases of heterotopic glioneuronal tissue in the parapharyngeal space have been reported. We present two cases of infant children who were seen at the Children's Hospital of Pittsburgh in 2013 with glioneuronal heterotopic masses in the parapharyngeal space.

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## 1. Introduction

Congenital heterotopic tissues found in the head and neck have been reported in the literature. Heterotopic thyroid, salivary gland, and gastric tissue have been described in locations such as scalp, neck, nasopharynx, palate and tongue [1]. There have only been 200 cases of heterotopic glioneuronal tissue reported and most are within the nasal cavity [2]. Other locations of the head and neck such as the orbit, middle ear, tongue, palate, pharynx, scalp or parapharyngeal space have also been described [1–10]. Here we present two cases of glioneuronal heterotopia arising in the parapharyngeal space. The first case was discovered on prenatal ultrasound and excised surgically. The second patient presented with mild airway obstruction and was managed conservatively. We discuss the clinical presentations, diagnostic features, and management of these lesions.

## 2. Case 1

A 6-day-old male born at 36 weeks gestation presented with a left-sided neck mass first visualized on prenatal ultrasound. A prenatal magnetic resonance image (MRI) at 36 weeks revealed a $3 \times 3 \times 2$ cm complex cystic mass involving the left parotid space and buccal space with medial extension into the pharynx and retropharyngeal spaces. MRI characteristics were interpreted as consistent with a venolymphatic malformation, and the location of the mass was concerning for potential upper airway obstruction.

An ex utero intrapartum treatment (EXIT) procedure was performed due to postnatal airway concerns. The infant was noted to have subtle facial asymmetry with the left mandible and submandibular region slightly larger than the right. A pre- and post-contrast $T_1$, $T_2$, and diffusion-weighted MRI of brain and neck revealed a $3.7 \times 3.8 \times 3.1$ cm mass with both solid and cystic components. The mass was centered within the left parapharyngeal space and extended into the left palatine tonsil region and left parotid gland, with bowing of the left lateral oropharyngeal wall, medial nasopharyngeal wall, and body and ramus of the mandible. The left carotid sheath contents were displaced posteriorly without changes in course or caliber of the major vessels.

One week later the patient underwent direct laryngoscopy with biopsy of the mass. The pathology demonstrated strips of soft tissue partially lined by respiratory-type epithelium with admixed glial and neuronal tissue, a mixed inflammatory response, and fibrosis (Fig. 1A and B). The decision was made to conservatively manage the mass with follow up MRI after six months. In the interim, the patient remained asymptomatic with no signs of dysphagia, stridor, or cranial nerve deficits. Six months later an MRI showed an increase in the size of the mass to $5.4 \times 4.5 \times 3.3$ cm with a more prominent cystic component (Fig. 2).

Based on the significant growth and risk of airway compromise, the decision was made to proceed with surgical excision. A combined transparotid–transcervical approach was used to expose the mass. The mass was carefully dissected off the facial nerve, carotid artery, and jugular vein. Final surgical pathology revealed predominantly glial tissue with cystic cavities lined with choroid plexus

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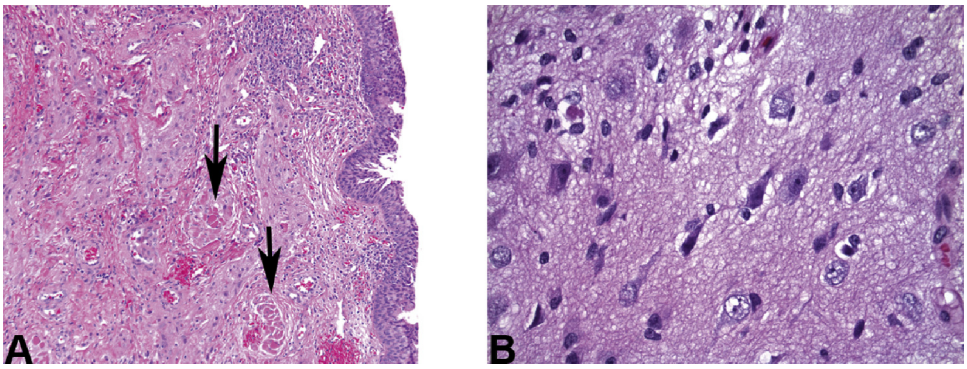


**Fig. 1.** (A) Medium magnification view of parapharyngeal mass showing layer of respiratory-type epithelium (right) overlying a moderately cellular lesion arranged in vague lobules and fascicles. Small entrapped regions of striated muscle are present (arrows) (HE, 100×). (B) High magnification view of lesion showing a mix of cells with a neuronal phenotype showing large round nuclei with single small nucleoli and amphophilic cytoplasm and smaller nuclei resembling astrocytes embedded in a neuropil background (HE, 400×).

epithelium (Fig. 3A–D). The patient's post-operative course was uncomplicated. He underwent an esophagram on post-operative day four, which showed no leak. Subsequently he was started on an oral diet and discharged home. The patient will be followed with subsequent MRIs.

## 3. Case 2

A 4-week-old girl presented with stertor and stridor since birth which were exacerbated with oral intake. There was no associated cyanosis or dyspnea. Prenatal history was significant for maternal methadone maintenance during pregnancy and subsequent morphine treatment as a newborn for neonatal abstinence. Physical exam revealed a large, bulging, firm mass on the left posterior pharyngeal wall and palate. The mass occluded 50–60% of the nasopharynx, with the tonsil and palate displaced anteriorly and rightward uvular deviation. A computed tomography (CT) scan revealed a $1.5 \times 2.2 \times 1.6$ cm heterogeneous solid and cystic mass in the left parapharyngeal space. With concern for airway obstruction and compression of neurovascular structures, the patient was intubated for MRI, direct laryngoscopy, and endoscopic biopsy.

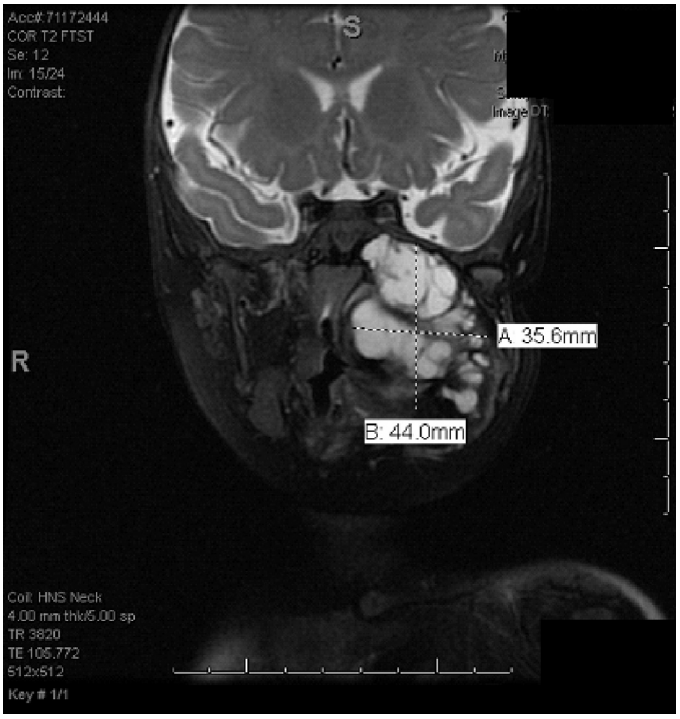


**Fig. 2.** $T_2$-weighted coronal slice MR image showing left parapharyngeal cystic mass. Bowing of the left oropharynx is evident. No communication with brain or CNS was visualized.

Pre- and post-contrast $T_1$, $T_2$, and diffusion-weighted MR images showed a well-demarcated left parapharyngeal space lesion which was inhomogeneously enhancing, with central hyperintensity and peripheral hypointensity on $T_2$ (Fig. 4). Fluid was noted in the fascial planes of the left masticator space, as well as stranding of the superficial soft tissue of the left cheek. No connection to the central nervous system was seen.

The patient underwent direct laryngoscopy and excisional biopsy of the mass. The pathology demonstrated a polypoid mass covered by stratified squamous epithelium with underlying infiltration by glioneuronal tissue (Fig. 5A–C). After the procedure she was extubated and developed mild inspiratory stridor with occasional subcostal retractions. Subsequent flexible fiberoptic nasolaryngoscopy revealed glottic and supraglottic edema and erythema with questionable left vocal cord paralysis. She was put on nasal CPAP and her breathing improved over the course of the week. After this week, patient began oral feeds which were tolerated well. The decision was made to forgo surgical excision of the mass in light of the patient's improved respiration and feeding tolerance. At two week follow up, patient was doing well with resolution of breathing symptoms, and no issues with feeding or gaining weight. Patient will be followed with subsequent MRIs.

## 4. Discussion

Congenital glioneuronal heterotopia is a rare entity in the parapharyngeal space. Fewer than 20 cases of heterotopic neuroglial tissue in the parapharyngeal space have been reported in the literature [9-14] (Table 1). The typical presentation in these cases is a neonate with clinical symptoms of airway obstruction or difficulty feeding with or without an accompanying palpable neck mass. Most reported cases are female with masses presenting on the left side. Associated congenital anomalies include cleft palate, Pierre-Robin syndrome, and heart defects. In a minority of cases, heterotopic glioneuronal tissue presents as a cystic lesion, likely due to the presence of functioning choroid plexus epithelium, as seen in the first case presented in this report. These cystic lesions can expand rapidly and are more likely to cause airway obstruction or acute respiratory distress. In the first case presented, the mass was large and fast-growing but entirely asymptomatic. This mass was heterogeneously solid and cystic, with choroid plexus epithelium. In the second case,