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RESEARCH ARTICLE
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HETEROTOPIC NEUROGLIAL TISSUE LEADING TO AIRWAY OBSTRUCTION IN NEWBORNS: A CASE REPORT

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ABSTRACT
Heterotopic neuroglial tissue which comprises of well differentiated neuroectodermal cells represents developmental heterotopia of neuroglial tissue rather than true neoplasm. Fewer than 30 cases have been reported in the medical literature[1]. Herein, we present a patient with submandibular heterotopic neuroglial tissue who presented to the emergency department with respiratory distress and feeding difficulty in the first week of her neonatal period. The lesion was resected and post operatively confirmed histologically as heterotopic neuroglial tissue.

INTRODUCTION
Heterotopia is the presence of a particular tissue type (normal in morphology) at a non-physiological; ectopic site usually co-existing with original tissue in its correct anatomical location[2]. The neuroglial cells are a collective group of cells with the main function to protect and maintain the optimum functioning of the nervous system[3]. The glial cells, or neuroglia, can be broadly categorised into two variants[3]:
Microglia: The microglia are phagocytic cells. They serve a defensive role within the nervous system. Furthermore, they have flexible shapes and are present throughout the brain and spinal cord.

Macroglia: The macroglia are the larger neuroglia present in the nervous system. Moreover, they may be further divided into 7 variants. Based on their function and location:

a) Astrocytes- Astrocytes are the star-shaped cells found in the Central Nervous System (CNS). They provide structural integrity by filling up the spaces between neurons. They have perivascular end-feet which wind around the blood vessels and thus constitute the blood-brain barrier. Further, the astrocytes also conduct the metabolite exchange between the neurons and blood vessels.

b) Oligodendrocytes- The oligodendrocytes are glial cells present in the CNS that help in the making of the myelin sheath.

c) Ependymal Cells- The ependymal cells are of three variants: ependymocytes, tanycytes, and choroidal epithelial cells.These cells are a part of the CNS and they deal with aspects of the cerebrospinal fluid (CSF).

The ependymocytes promote the free movement of molecules between the neurons and the cerebrospinal fluid. The tanycytes respond to alterations in the blood-derived hormonal levels. The choroidal epithelial cells control the chemical composition of the cerebrospinal fluid.

d) Radial Glial Cells- The radial glial cells are found in the CNS. They provide a temporary support structure for new neurons to latch on to.

e) Schwann Cells- The Schwann Cells are a counterpart to the oligodendrocytes, in the Peripheral nervous system(PNS). Schwann cells help in the making the myelin sheath. These cells are also phagocytic in nature

f) Satellite Cells- The satellite cells are found in the PNS surrounding the nerve cells of the sensory system and the autonomic system. The main function of the satellite cells is to maintain a stable chemical environment around the neurons.

g) Enteric Glia Cells- They are a part of the PNS, found in the gastrointestinal tract. Thus, they help in digestion and also in homeostasis.

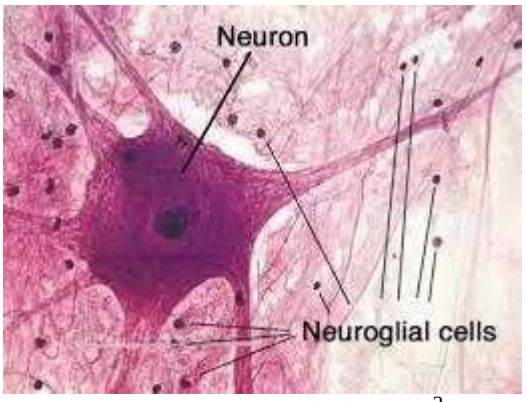


Figure 1: Neuroglial tissue[3]

INCIDENCE: Fewer than 30 cases have been reported in the medical literature so far.

CASE DETAILS

We present a case of a 7day old neonate who presented with complaints of labored breathing, snorting, nasal flaring and with features suggestive of pneumonia. She was born full term by normal vaginal delivery, weighing 2.6 kg to a multiparous woman who received antenatal care in an institutional setting. Baby cried immediately after birth and is immunized till date.

On examination, the baby had a firm swelling in the left side of neck measuring approximately 4*5 cm , the baby was irritable, crying with labored breathing and showed early signs of dehydration. No craniofacial abnormalities were evident, however, there was a firm mass on left side of the neck . The mass was non-compressible, non-trans illuminant, painless, not fixed to the skin with mobility present in both vertical and horizontal planes. There was no redness or local calor associated with the mass and it didn't move with crying or deglutition. Baby was nursed in prone position, which seemed to reduce the respiratory distress.

MANAGEMENT

An ultrasound of the neck was done in preparation for surgical resection which revealed a heterogenous mass causing compression over the trachea, with solid as well as cystic components taking up minimal vascularity on colour doppler.

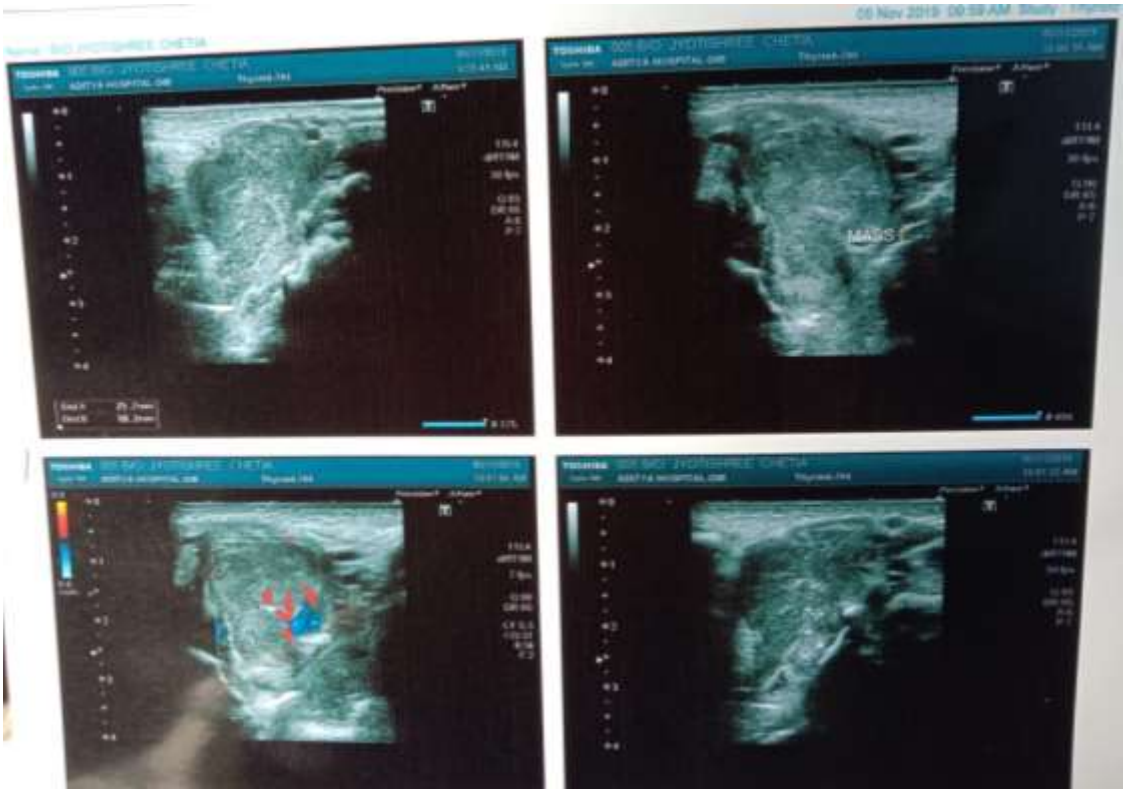


Figure 2: Ultrasonography neck revealing the mass

After resuscitation in the form of fluid administration and oxygenation, the patient was taken up for surgical intervention.
Transverse incision was given over the swelling giving ample visualization of the operative field. Precise resection of the mass was done and the sample was sent for biopsy.

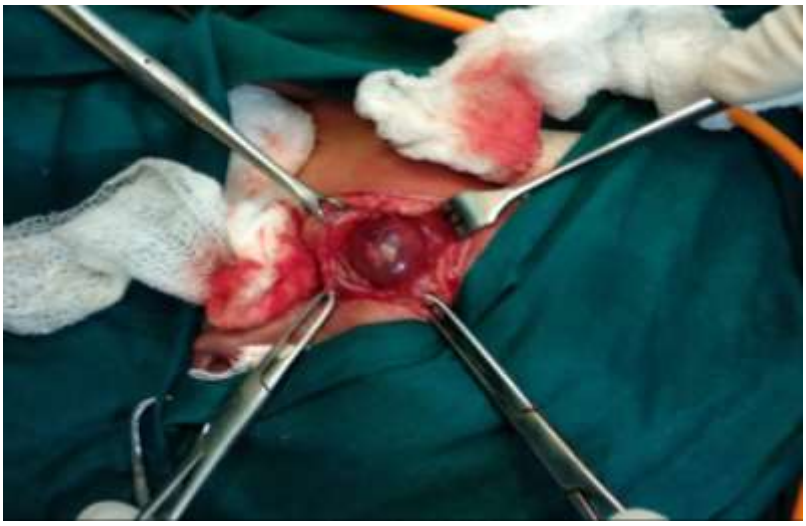


Figure 3: Intraoperative picture showing the mass

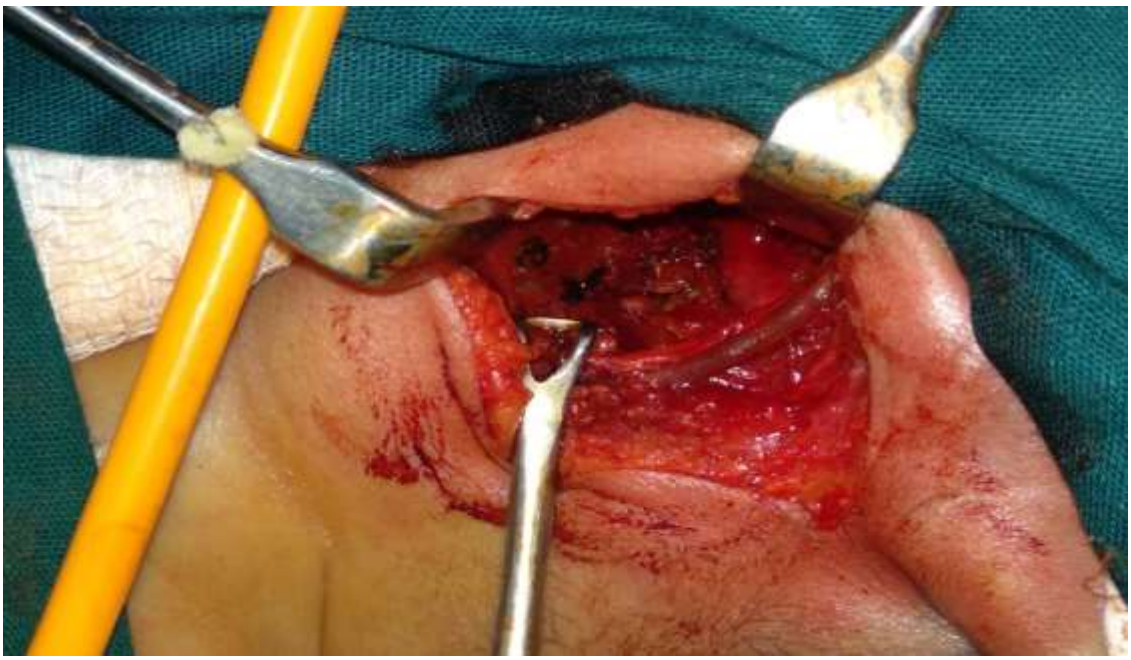
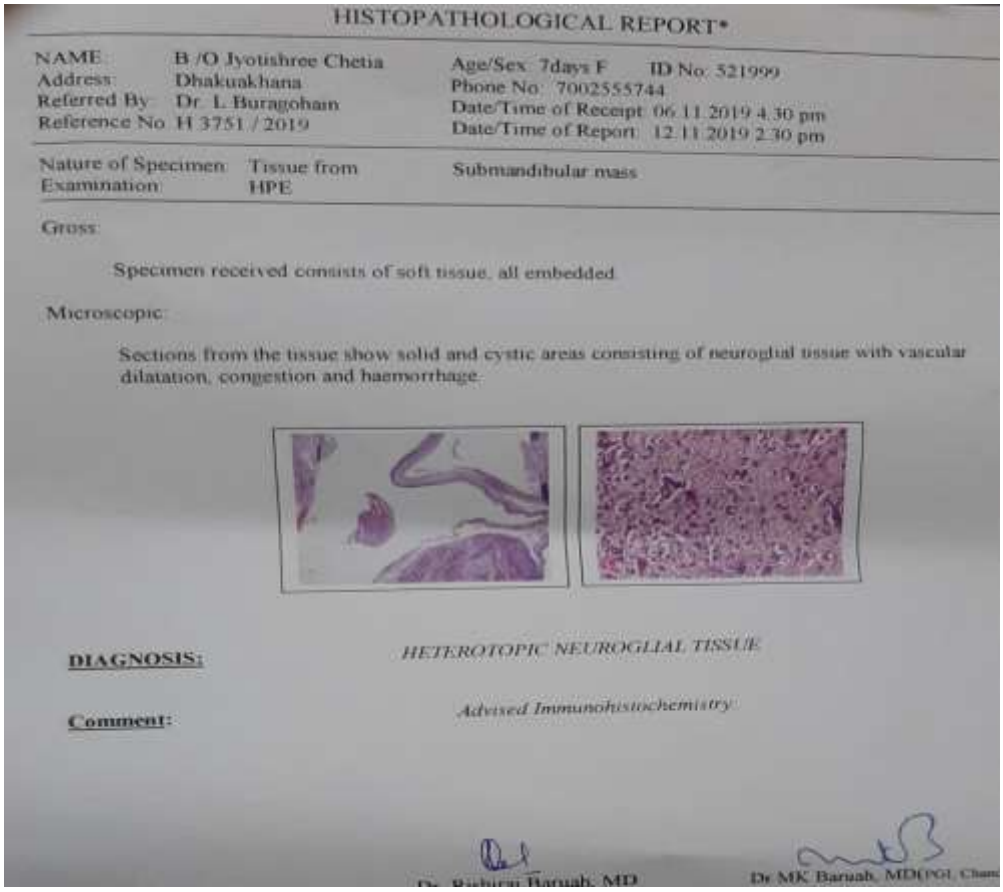


Figure 4: Intraoperative picture of left side of the neck , after mass resection

Immediate post operative period was uneventful with no signs of breathlessness and the patient started taking frequent, adequate feeds after 24 hours of surgery and was subsequently discharged on post operative day 5 on antibiotics and analgesics.

Patient came for follow up after 2 weeks with the following HPE reports and the diagnosis of Heterotopic Neuroglial tissue was made.



HISTOPATHOLOGICAL REPORT*

NAME: B /O Jyotishree Chetia
Address: Dhakuakhana
Referred By: Dr. L Buragohain
Reference No: H 3751 / 2019

Age/Sex: 7days F ID No: 521999
Phone No: 7002555744
Date/Time of Receipt: 06.11.2019 4.30 pm
Date/Time of Report: 12.11.2019 2.30 pm

Nature of Specimen Examination: Tissue from HPE — Submandibular mass

Gross:

Specimen received consists of soft tissue, all embedded

Microscopic:

Sections from the tissue show solid and cystic areas consisting of neuroglial tissue with vascular dilatation, congestion and haemorrhage

DIAGNOSIS: *HETEROTOPIC NEUROGLIAL TISSUE*

Comment: *Advised Immunohistochemistry*

Dr. Rishiraj Baruah, MD — Dr MK Baruah, MD(PGI, Chan

Figure 5: Histopathological report of the resected mass

Patient was followed up again after 6 months and there were no complaints of recurrence

DISCUSSION

EMBRYOLOGY AND PATHOGENESIS: The pathogenesis of heterotopic neuroglial tissue remains unclear but some of the proposed mechanisms are stated below.

- Brain tissue protruding through the chondrocranium (early skull base) at 12 weeks' gestation might result in a frontal encephalocele and that encephalocele losing its intracranial connection might result in heterotopic neuroglial tissue[4–6].
- A change in the timing of fusion of the chondrocranium might result in the separation of a segment of primitive neural tissue from the main portion of the developing brain[5].
- During embryogenesis, displacement of totipotent neuroectodermal cells occur which later develop into mature neural tissue[1,4].
- Abnormal migration of glial cells from the olfactory bulbs[6].

PATHOLOGY: The differential diagnosis of masses causing airway obstruction in the newborn includes glioma, teratoma, cystic hygroma, hemangioma, neurofibroma, ectopic thyroid, brachial anomaly and heterotopic brain.

Brain heterotopias are composed of nests of neural tissue, without mitoses, embedded within varying amounts of fibrovascular stroma[6]. Neurons can be present in up to 10% of cases. Focal calcifications might be present. Reactive changes, a paucity of neurons, and focal calcifications are changes usually observed in poorly perfused neural tissue[6].

Unlike nasal glioma, heterotopic pharyngeal neuroglial tissue might contain neurons and astrocytes as well as more complex central nervous system elements such as ependymal-lined structures, a functioning choroid plexus, and pigmented cells of retinal differentiation[1,3,6]. The absence of these more complex structures from nasal glioma might indicate that they are indeed separate entities[1].

Heterotopic neuroglial tissue is composed solely of ectodermal elements[7], which distinguishes it from teratoma, which is composed of all 3 germ layers.

Grossly, heterotopic neuroglial tissue is solid, firm, and dark brown or red. It might have cystic components containing cerebrospinal fluid–like clear fluid. The tissue is relatively avascular and poorly encapsulated and adherent to surrounding soft tissues.

RADIOLOGY: Axial and coronal CT images of the head, neck, and brain delineate the location of the tumor and its relationship to the skull base. Displacement and distortion of the mandible and pterygoid plates at birth is commonly noted and seems to be a differentiating feature of these tumors[7]. In addition, erosion of the floor of the middle cranial fossa is characteristically associated with brain heterotopias in the parapharyngeal space. These CT attributes might help differentiate brain heterotopias from more common congenital lesions such as cystic hygroma[8].

Magnetic resonance characteristics of heterotopic neuroglial tissue resemble normal brain tissue in all pulse sequences[1]. Cystic elements might be present and represent cerebrospinal fluid–like fluid-filled spaces. A CT cisternogram might give additional information regarding connection with the subarachnoid space.

TREATMENT: Surgical intervention is necessary in patients with heterotopic neuroglial tissue that causes airway distress, dysphagia, or failure to thrive. Tumor resection in the newborn period might allow early oral feeding and avoidance of tracheotomy. The surgical approach should be similar to that for cystic hygroma. Resection should be as complete as possible without sacrificing vital structures or compromising function. Multiple surgical resections might be necessary to accomplish this task.

The timing of the surgery is controversial. Early surgical intervention in the newborn period seems to be beneficial for 2 reasons. First, further growth of neuroglial heterotopias might cause distortion and erosion of bone resulting in facial deformity requiring future correction. Second, delay in resection might preclude normal development of the swallowing function and pharyngeal

coordination. Proponents of delayed resection believe that resection might be safer in the older child, in whom vital neurovascular structures are more easily salvageable and blood volume is greater[9]. However, delayed resection of heterotopic neuroglial tissue frequently requires airway control and alternate routes for feeding. The increased morbidity and mortality rates associated with tracheotomy and gastrostomy feeding must be considered. In addition, further distortion of surrounding soft tissue and bone of the facial skeleton might ensue.

Surgical excision can be performed through a transcervical, lateral pharyngotomy approach with primary closure of the pharynx. This allows early identification and preservation of vital neurovascular structures. Preservation of pharyngeal mucosa with meticulous dissection of the submucosal portion of the mass might improve functional outcome. A transoral and/or trans-palatal approach can be combined with the external approach[5]. Rigorous postoperative care includes attention to nutritional status and speech and swallowing therapy.

Tracheotomy and gastrostomy might be necessary in patients with co-morbidities precluding early surgical exploration (eg: complex congenital heart disease). A nasopharyngeal airway might be an alternative to tracheotomy in select newborns with a prominent nasopharyngeal component and minimal involvement of the oropharynx and hypopharynx.

MANAGEMENT APPROACH: If a mass is identified in the vicinity of the skull base, CT (with and without cisternography) and MRI can be used to exclude an intracranial connection. Next, an incisional biopsy and airway endoscopy should be performed. If the biopsy specimen contains mature neural tissue, the differential diagnosis is limited to 3 entities: teratoma, encephalocele, and heterotopic neuroglial tissue. Differentiation of these tumors can be made by clinicopathologic and radiographic correlations. Surgical resection can be proceeded, if the general health of the child permits. A nasopharyngeal tube might be useful for airway management in some patients to avoid tracheostomy. Patients with airway compromise, failure to thrive, and other significant comorbidities might require delayed surgical resection, tracheostomy, or gastrostomy tube insertion.

CONCLUSION

Heterotopic neuroglial tissue must be considered a differential diagnosis of airway obstruction in the newborn. Management is surgical resection, with attention to vital structures. More than one surgical procedures might be needed in some of these patients[8].

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