CASE REPORT

Intracranial extracerebral neuroglial heterotopia with parapharyngeal extension: pre-natal and post-natal imaging findings

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ABSTRACT. We report an unusual case of intracranial extracerebral neuroglial heterotopia (IENH) presenting with parapharyngeal extension. Neuroglial heterotopias are rarely reported pre-natally and, to our knowledge, there are no reported cases presenting pre-natal imaging findings of IENH with parapharyngeal extension. Because pre- and post-natal imaging studies are essential to diagnose IENH and plan surgical treatment, radiologists should be familiar with this rare condition.

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Neuroglial heterotopias (NHs) are rare congenital lesions characterised by the presence of differentiated neuroectodermal tissue in locations where neuronal and glial cells do not typically occur [1]. These lesions usually involve the head and neck regions and are more commonly described in extracranial locations such as the nose and nasopharynx, oral cavity, oropharynx, palate, tongue, lips, scalp and orbit. Although intracranial extracerebral NH has been described more frequently with the increased availability of MRI, it remains a very rare condition [2].

Intracranial extracerebral neuroglial heterotopias (IENHs) with parapharyngeal or oropharyngeal extension have been presented in a limited number of reports [2, 3]. To our knowledge, pre-natal imaging findings of this uncommon combination have not been reported before. We report a case of IENH with parapharyngeal extension and describe its antenatal and post-natal imaging features.

Case report

A 28-year-old pregnant female (gravida 3, para 2) was referred to our institution after an obstetric ultrasound examination at 25 weeks' gestation identified a foetal intracranial mass with extension to the left side of the neck. A repeat obstetric ultrasound performed in our department showed an osseous malformation in the left frontotemporal area of the cephalic pole resulting from a solid, round, predominantly hypoechoic intracranial mass (4.8 × 4.2 cm) with an anechoic area inside. This mass appeared to be contiguous with a predominantly

cystic mass (6.7 × 5.9 cm) on the left side of the neck (Figure 1). A foetal MRI was performed at 27 weeks gestation; this showed a solid cystic mass lesion in the left anterolateral portion of the craniofacial region (Figure 2). Subsequent ultrasound evaluations showed that the mass had remained stable during pregnancy.

The female infant was delivered pre-term by the *ex utero* intrapartum treatment procedure (EXIT) at 36 weeks gestation because of the risk of airway compression. Apgar scores at 1 min and 5 min were 9 and 10, respectively. The patient, who weighed 3045 g at birth, was admitted into the neonatal intensive care unit on mechanical ventilation. On clinical examination, the neonate had a large, soft, compressible cervical mass involving the frontotemporal region of the head.

An MRI scan performed at the age of 1 day showed a large heterogeneous lesion with solid and cystic components. The solid component was an intracranial extracerebral lesion located in the left middle cranial fossa that presented a signal intensity similar to that of cerebral parenchyma, with clear-cut demarcation from the adjacent left temporal lobe. The left temporal lobe was hypoplastic and compressed by the lesion (Figure 3). Following the intravenous injection of paramagnetic contrast medium, the lesion showed a slightly more pronounced enhancement than the adjacent brain. The cystic component was extracranial, contiguous with the intracranial lesion, isointense with the intracranial cerebrospinal fluid (CSF) spaces and extended inferiorly into the neck region.

A CT scan demonstrated a remodelling of the left frontal and temporal bones as a result of the mass lesion. The skull base anatomy (notably the sphenoid bone) was also distorted by the lesion, which had extended inferiorly into the infratemporal fossa, displacing the upper airway to the right and causing orbital dysplasia (Figure 4).

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Figure 1. (a–c) Obstetric ultrasound at 25 weeks gestation shows an intracranial extra-axial hypoechoic mass lesion in the left frontotemporal region, seemingly contiguous with an anechoic lesion in the left cervical region. (d) Colour Doppler image reveals the flow within the mass.

The patient underwent complete excision of both cervical and temporal lesions when 29 days old. Initially, head and neck surgeons excised the cervicofacial component of the mass until they reached the base of the skull. Neurosurgeons then resected the intracranial portion. No connection to the adjacent brain was seen intra-operatively.

A subsequent histopathological examination revealed a mass of mature non-neoplastic neuroglial tissue. The neuropathological findings in combination with imaging data led to the diagnosis of NH. The post-operative recovery was uneventful. The patient was discharged 40 days after surgery and remains well and active 4 years later without any clinical signs of developmental delay.

Discussion

Since Reid first reported a case of NH in 1852 [4], several terms have been used to describe this type of lesion, including glioneural or neuroglial choristoma, glioneural hamartoma, brain heterotopia, heterotopic brain tissue and ectopic brain, among others [5]. The term choristoma is used synonymously with heterotopia

to define histologically normal tissue occurring in an abnormal location [5]. In 1999, Gyure et al [1] defined the term “intracranial extracerebral neuroglial heterotopia” to describe a case with involvement of the middle and anterior cranial fossae. It should be noted that because of its benign pathological and clinical nature, this type of lesion is considered to be a developmental malformation rather than a true neoplasm [1, 6].

IENHs are exceedingly rare lesions and, unlike meningoencephaloceles, they do not have any connection with the subarachnoid space [1]. The pathogenesis of NH remains unknown, although several theories have been proposed to describe possible mechanisms: (a) an early encephalocele through the chondrocranium might subsequently lose its connection to the central nervous system (CNS), thereby evolving to a NH [7]; (b) displacement of pluripotent neuroectodermal cells during early embryogenesis might later lead to a differentiation into neural tissue in an ectopic location [8]; (c) an entrapment of glial cells from the olfactory bulb outside the CNS might develop into a NH [7]; and (d) an extra third evagination occurring inferior to the telencephalic vesicles (which develop during the fifth week of embryogenesis) might subsequently be separated from

Figure 2. Foetal MRI at 27 weeks gestation. (a,b) T_2 weighted axial, (c) coronal and (d) sagittal images show a solid cystic mass lesion in the left anterolateral portion of the cranio-facial region, with an intracranial extra-axial component causing compression to the left frontal and temporal lobes.

the developing CNS, thereby evolving to a NH (or an "accessory brain", as termed by the authors of the theory) [9].

The clinical presentation of NH depends on the location (most commonly reported on the left side) and size of the mass. An enlargement of head circumference can be seen in cases of intracranial involvement; unilateral involvement can present with facial asymmetry [5], as observed in our case. On the other hand, patients with extracranial masses commonly present with respiratory distress and feeding difficulty [10]. Some developmental anomalies have been associated with NH; these include cleft palates, Pierre Robin syndrome, congenital heart disease, pectus excavatum and micrognathia [11, 12].

NH is more frequently diagnosed during infancy and in most cases a pre-natal diagnosis is not performed [10]. The pre-natal detection of a cervical mass allows the early establishment of therapeutic strategies for possible airway compression at birth; airway obstruction due to neck masses is associated with high morbidity and mortality [13]. In our case, the EXIT procedure was adopted and the anatomical details provided by the foetal MRI scan played a valuable role in the planning of this strategy [13].

For a complete radiographic assessment of the lesion, a post-natal MRI study and a complementary CT scan is recommended. The high soft tissue resolution of MRI

enables the specific characterisation of size and extent of the mass and any displacement of adjacent structures. In all pulse sequences, NH has a signal intensity that is similar to normal brain tissue [4]. The vascularity of the lesion may affect the degree of contrast enhancement noted. MRI can be helpful in discerning an intracranial connection through skull base defects, as in our case. In this context, MRI can assess whether there is any direct communication with the subarachnoid space. Cystic components are isointense with the intracranial CSF spaces and might represent CSF-like fluid-filled spaces [4]. A CT scan can demonstrate bony erosions, most commonly of the pterygoid plate, mandible and skull base, besides delineating the mass location and its relationship to the base of the skull.

The differential diagnosis of IENH comprises conditions characterised by the presence of mature brain tissue outside the CNS, such as teratoma and encephalocoele. Differential diagnosis of neck masses in a neonate most commonly includes lymphangioma, teratoma and encephalocoele [13, 14]. Histologically, the mass is composed of neurons and mature glial elements; choroid plexus-like structures can also be found [10]. NH can entwine with the surrounding soft-tissue structures, which can lead to an incorrect pathological diagnosis of a teratoma [1]. Therefore, it is essential that there is correlation between the surgical, radiological and pathological findings.

(a)

(b)

(c)

(d)

Figure 3. Post-natal MRI at 1 day old. (a,b) T_2 weighted axial and (c) coronal images demonstrate an intracranial extra-axial mass lesion located in the left middle cranial fossa, with a signal intensity similar to normal brain parenchyma, causing compression to the adjacent frontal and temporal lobes; an extracranial cystic component can be seen. (d) Post-contrast T_1 weighted axial image demonstrates a slight enhancement of the solid component of the mass compared with the adjacent brain.

(a)

(b)

(c)

(d)

Figure 4. Post-natal CT scan. (a,b) Post-contrast axial and (c,d) coronal images show a mass lesion inducing a remodelling of the left frontal and temporal bones and distortion of the skull base anatomy.

The definitive and curative treatment for NH is complete surgical resection. Recurrence is rare and it seems to be a result of incomplete resection [4]. As a result of the benign characteristics of the lesion, the long-term prognosis is excellent. Imaging studies are essential for the diagnosis and surgical planning of IENH and therefore radiologists should be familiar with this rare condition.

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