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A CASE OF GLIAL HETEROTOPIA INVOLVING ROOT OF NOSE- A RARITY

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ABSTRACT Heterotopic neuroglial tissue represents normal brain tissue in an abnormal location away from the CNS. It is a rare congenital anomaly and the majority of these lesions are diagnosed at birth. We present this case report to highlight the occurrence of heterotopic glial tissue in the nose in children. It being a rare disease, very few articles in the literature have documented the occurrence of heterotopic glial tissue in extracranial sites. The entity put the surgeons in dilemma as to the diagnosis and management.

KEYWORDS : Glial Heterotopia, Nose, Nasal Glioma

Introduction-

Glial heterotopias are rare congenital displacements of cerebral tissue in extracranial sites. The incidence of congenital nasal masses is reportedly 1 in 20,000-40,000 live births (1,2,3) , and nasal glial heterotopia accounts for approximately 5% of them(4). The term was first described by Scmidt(5). The most common areas of occurrence include root of nose and nasal cavity (6). However, occurences in other areas like the tongue(6), skin and palate (6) have also been reported.

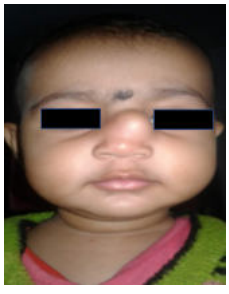
The development of heterotopic glial tissue has been a matter of interest for head neck surgeons. It is usually said that the embryological remnant of neuroglial tissue gets detached from the arachnoid connection and gets sequestered in the extracranial site (7,8). It is essentially non neoplastic and mimics dermoid, encephalocoele and meningoencephalocele.

Case Report-

A female patient aged 3 years presented to us with slow growing painless swelling over her root of nose. The swelling was present since birth and was initially of the size of lentil and then increased to the size of a pea.

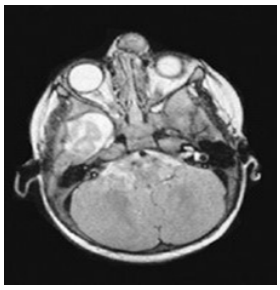
The swelling did not increase in size during crying. She did not have any history of respiratory distress. She did not have any complaints of nasal obstruction or discharge from nose. The antenatal and postnatal history were unremarkable.

On examination, a nodular swelling was seen over the root of the nose. The swelling had a smooth surface, regular and well defined margins. It was 2cm X 2 cm in dimension. It was non- tender, mobile, firm in consistency and was non compressible. Transillumination test was negative.



Preoperative clinical picture

MRI was done which showed a regular homogenous mass with well defined margins over the root of nose with no connection to cranial cavity. The mass was hypointense on T1W image and was hyperintense on T2W image

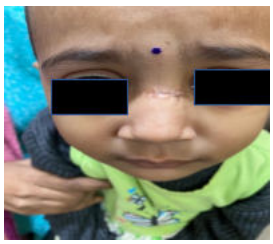


T1W MRI image of the mass

Surgical excision was planned under general anaesthesia. The mass was found to be lying in the subcutaneous plane and was well separated from the surrounding soft tissue.

It was sent for histopathological examination which showed the presence of astrocytes with vesicular nuclei suggestive of glial tissue.

During follow up, she exhibited a recurrence of the mass over her nose at 6 months after surgery. The radiological characteristics were same. The mass was excised with few millimetre cuff of surrounding tissue and sent for Histopathological examination. It came out to be a glial tissue again. We performed an immunohistochemistry of the tissue and found it to be positive for S100.



2 months post operative status after revision surgery.

Discussion-

Heterotopias represent isolated rests of nervous tissue without attachment to the nervous system. Many theories have been proposed as to the mechanism behind the development of these rare tumors. These include an encephalocele that loses its intracranial connection resulting in heterotopic neuroglial tissue (9,10). Another mechanism is that of glial cells migrating from the olfactory bulb [10]. Another mechanism is displacement of neuroectodermal cells during early embryogenesis, which later develops into mature glial tissue [11]. These tumors are rare and are hence misdiagnosed. They are essentially slow growing and non neoplastic (12). Although cosmesis

was the primary concern for the parents in our case, heterotopias in other areas, like tongue and palate can lead to difficulty in feeding and respiratory distress (if mass in the posterior part of tongue encroaches upon airway) . The other probable lesions can be Dermoid, Lymphovascular Malformation, Encephalocoele, Lipoma, Meningoencephalocoele. Preoperative differentiation between these lesions is difficult, however, encephalocoeles will exhibit definite connection to the brain or spinal cord. Also, encephalocoeles show heterogeneity on imaging compared to dermoids(8). Histopathological examination, supported by immunohistochemical analysis will aid in diagnosis.

Conclusion-

Heterotopic glioma is a rare clinical condition and hence diagnosing this disease is difficult. Improper preoperative diagnosis led us to a surprise at the histopathological report. Subsequent recurrence made us more cautious while removing the mass. Although imaging investigatons are available, they may not give us entire data for correct diagnosis. With limited number of case reports all over the world, this entity is documented by us.

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