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Huge oro-naso pharyngeal neuroglial choristoma: a case report and review of literature

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Abstract:

Choristoma is a general term referring to normal tissue found in an abnormal location. Glial choristoma is a mass that consists of mature, normal brain tissue, isolated from the cranial cavity or spinal canal. The involvement of an extracranial non-midline site is exceptionally rare. Here we report, a case of newly born baby girl with oro-naso-pharangeal mass. The patient presented with air-way obstruction that had lasted for 6 days, with the radiological studies revealing a mass-like lesion with soft tissue density in the oral cavity. The patient had no previous history of head trauma during vaginal delivery, and no evidence of central nervous system connection was noted on the radiological or operative findings. The patient underwent debulking of the mass. Histologically, the mass was composed of disorganized but mature, normal glial tissue with significant choroid plexus component. The clinicopathologic findings of the case are presented and the relevant literature including the aetiopathogenetic hypothesis of choristoma are discussed.

Keywords: Choristoma, neuroglial, heterotopia

1 Introduction:

Heterotopia, or choristoma, represents mass consisting of normally formed tissues that are present in abnormal anatomical sites. For instance, the presence of gastric mucosa in the upper third of the esophagus, pancreatic tissue in the gastrointestinal wall, or a parathyroid gland within the thymus. Glial choristoma or heterotopic glial tissue represents a mass lesion that consists of mature, normal brain tissue that is separated from the cranial cavity or spinal canal. Neuroglial choristomas usually involves the midline extracranial structures of the head and neck, such as the nose, nasopharynx, oropharynx, palate, tongue, and tonsil. Nasal cavity represents the most common site[1, 2]. The ateopathogenesis of the neuroglial choristomas is related to the failures in the normal course of embryonic development of the neural tube. This can result in a great variety of defects in the form of choristomas. Choristomas are benign tumor like growth that should be separated from neoplasms. They are different from hamartomas which are tumor like malformations composed of a focal overgrowth of mature normal cells located where they are normally found [3] [4-8].

2 Case Report:

Here we report a case of choristoma in a one day-old female infant (body weight: 1800-gram) who presented with manifestations of air-way obstruction during vaginal delivery. General examination was unremarkable. The cranium and vertebral column were intact. No craniofacial defects were observed. There was a mass lesion protruding from the mouth, encroaching on the oral cavity and causing airway obstruction.

The anaesthetist immediately did endotracheal intubation to save the airways. CT showed soft tissue mass lesion filing the left side of the oral cavity protruding anteriorly through the mouth. The mass was extending into the left parapharyngeal space and displacing the left masticator space (including the left mandibular ramus). The mass was extending to the left infratemporal fossa and the nasopharynx. The mass had heterogenous appearance being formed mainly of solid elements, fatty tissue and cystic areas (the cystic components mainly in the nasopharyngeal region).

The differential diagnosis included tumors and neuroglial choristoma.The differential diagnosis of in our case include glioma, teratoma, dermoid cyst, hemangioma, cystic hygroma, neurofibroma, and sarcoma. Debulking was performed through intraoral approach and the patient was clinically stable. Grossly, the lesion consisted of 11X6X3 cm mass with solid and cystic grayish white areas and soft consistency. Histologically, the mass was composed of mature neuroglial and choroid plexus tissue. The glial tissues were composed of cells with one or more nucleus, basophilic fibrillary cytoplasm resembling astrocytes. The glial cells were supported by a fibrillar homogenous eosinophilic stroma. No other ectodermal, mesodermal or endodermal derivatives were observed.

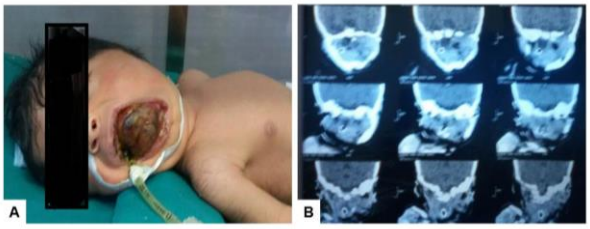

Figure 1: Clinical and radiological features of neuroglial choristoma. Clinically, there is mass protruding from the mouth. CT shows soft tissue mass lesion filing the left side of the oral cavity and extending into the parapharyngeal space.

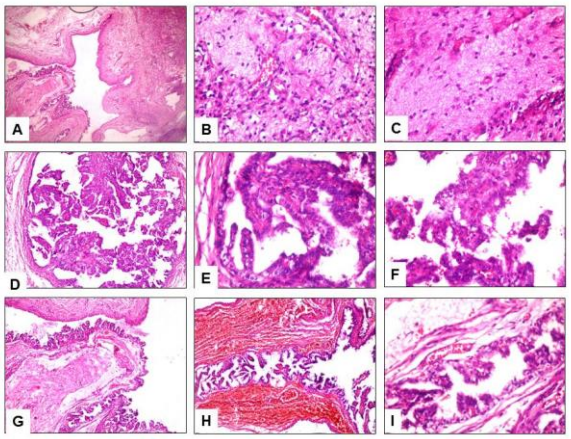

Figure 2: Histological features of neuroglial choristoma. The mass consists of mature neuroglial and choroid plexus tissue. The glial tissues is composed of cells with one or more nucleus, basophilic fibrillary cytoplasm resembling astrocytes. The glial cells are supported by a fibrillar homogenous eosinophilic stroma (A-C). The choroid plexus tissue is composed of papillary formations. The latter consists of epithelial layers around delicate stroma.

3 Discussion:

Choristomas are defined as the presence of histologically normal tissues that are located at abnormal locations [1, 2]. Neuroglial choristoma are rare developmental anomalies that can be divided into extraneuraxial and paraneuraxial groups [5]. Extraneuraxial neuroglial heterotopia (choristomas) are more common and usually involves the nasal cavity

or the superficial soft tissue of the head and neck. On histology, they are characterized by a disorganized mixture of neuroglial and mesenchymal tissues. It is usually obvious at birth but may remain asymptomatic until late childhood or even adulthood. Complete surgical excision is curative in most cases [3] [4] [9]. Paraneuraxial neuroglial heterotopia (choristomas) are rare and may be seen in the paracranial or paraspinal spaces. It consists of organized brain tissue and is usually diagnosed shortly after delivery or in early childhood [5-8] . Histologically, oral glial choristomas have disparate histopathologic features. Neuroglial choristoma is composed of a variety of elements of the central nervous system (ectodermal elements alone), such as astrocytes, oligodendroglia and neurons, ependyma, retinal components and choroid plexus. The cells are mitotically inactive, with fibrovascular stroma that are poorly encapsulated and adhere to surrounding tissues.

The ateopathogenesis of neuroglial heterotopia is poorly understood. Several pathogenetic hypotheses of extraneuraxial neuroglial heterotopia include (i) herniation of neuroectodermal tissue through a primary bony defect [3] [4] ; (ii) separation and detachment of cerebral precursors from the brain primordium during early embryogenesis [3] [7]; (iii) aberrant migration of pluripotential embryonic tissue with subsequent neuroglial differentiation[3] ; (iv) retention of neuroectoderma remnants [10]; and (v) teratoma formation with exclusive neuroglial elements [3] . Paraneuraxial neuroglial heterotopia is closely related to neural tube defects, such as encephalocele and myelomeningocele [5] [7] [8]. A connection between the heterotopic neuroglial tissue and the intervertebral region is found in some cases, [5] [6] . Accordingly, the “herniation and sequestration” hypothesis seems plausible to explain the pathogenesis of paraneuraxial neuroglial heterotopia.

To conclude, here we report a case of neuroglaial choristoma of the oro-naso pharyngeal region. Neuroglial choristoma should be a consideration in newborns with air-way obstruction. Surgical treatment is usually curative.

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