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Case Report

Nasal heterotopy in a Newborn Infant: A Case Report

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ABSTRACT
Glial heterotopias are rare, congenital, benign, midline, non-teratomatous extracranial glial tissues which are mostly present in the nose and may masquerade as encephalocele or dermoid cyst. These masses appear to share a similar embryogenic origin. Herein, we present a neonatal nasal glioma on the nasal root and glabella area. Although rare, because of their potential to connect to the central nervous system, these disorders are clinically important.
Keywords: Glial tissue, heterotopy, newborn

Introduction

Glial heterotopy is a midline, congenital mass located in the nose, which contains mature glial tissue. Although the incidence is not fully known, taking reported cases into consideration, it has been reported as 1:20.000-40.000 (1,2). Depending on the location, it may be extra nasal, nasal or mixed type. Other rare locations of heterotopic brain tissue may be the lips, tongue, within the scalp, nasopharynx or oropharynx. Encephalocele from midline defects and embryonic origin nasal glioma are similar and are related to the abnormal partition of the ectoderm and neuroectoderm during the development of the nose (2). Diagnosis is made through histopathological evaluation by the removal of the mass with excisional biopsy. Differentiation of nasal glioma from encephalocele cannot be made with histopathological examination. The most important feature of glial heterotopy is that there is no intracranial connection of the mass. It is important to make a clear diagnosis because of the risk of the mass involving functional brain tissue. Although midline located masses of the nose are rarely seen, we presented the case of a newborn infant with nasal glioma to emphasize the significant details of diagnosis and treatment.

Case Report

A female infant weighing 1120 gram, of 26 weeks six days gestation according to the last menstrual cycle was delivered to a 31-year old mother as the 3rd live birth of 5 pregnancies. No fetal anomaly was detected during the prenatal follow-up, and no nasal mass was reported on prenatal ultrasound. Her Apgar score was 7-8 at 1st and 10th minutes respectively. The infant had findings of respiratory distress, so was admitted to the neonatal intensive care unit. In the physical examination, a non-pulsatile mass was seen of polypoid structure with regular borders, located on the midline towards the left with a wide base adjacent to the nasal root (Figure 1a, b). No accompanying malformation was determined. Laboratory investigations revealed no hypothalamic dysfunction. On radiological examination, cranial tomography showed a mass of soft tissue density with no accompanying bone defect, and no extension to the intracranial or the orbital region, and not creating an obstruction in the nasal cavity. On contrast magnetic resonance imaging, a heterogeneous, hypointense appearance was observed on T1 and T2-weighted images and was evaluated as a mass with a mild level of heterogeneous contrast (Figure 2). The material removed with excisional

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biopsy was identified as a piece of cutaneous with hair and subcutaneous tissue of polypoid structure 4x3x2.4 cm on a base 2.5x2.8 cm. Immunohistochemically it was evaluated as glial heterotopia with (+) glial fibrillary acidic protein. The postoperative appearance of the case is seen in Figure 3. The patient was followed-up in neonatal intensive care unit because of being 3 months premature. Informed consent was obtained from the patient's parents.

Discussion

Glial heterotopy is defined as the congenital abnormal location of mature glial tissue in an area outside the central nervous system without intercranial extension. This terminology is not appropriate as it does not include neoplasm or tumoral tissue. By definition, it is a special type of choristoma. It is not a teratoma structure containing ectoderm, endoderm, and mesoderm layers. The dura, pia and arachnoid of the brain tissue and/or leptomeninges that are defined as a bone defect with herniation continuing to the cranial cavity are clearly separated from the surrounding encephalocele (1). There may be erosion and deformity in the adjacent bone and frequently accompanying hypertelorism. Clinically, lesions may be externally adjacent to the nose (60%), in the nasal cavity (30%) or both areas (10%). Typically it is congenital or develops within the first two years (3,4). Therefore, as in the current case, diagnosis is often made in the neonatal period, but occasionally, the diagnosis of those with intranasal location may be made in adulthood. Other rare locations for heterotopic brain tissue are the lips, the tongue, inside the scalp, the nasopharynx, and the oropharynx. Although rare, they are important because of their potential relationship with the central nervous system (5). The actual incidence is not known, but on the basis of reported cases, it has been stated to be 1:20.000-40.000 (1,2). Family history has not been defined. There have been reports of cases with the combination of corpus callosum agenesis and cleft palate (6,7). Patients present in the early stage of life (often at birth or in the first months) with a subcutaneous nodule in the nose or polypoid mass in the nasal cavity. In patients with an intranasal mass, non-specific findings of nasal obstruction, chronic rhinosinusitis, otitis or allergic symptoms may be seen. If there is accompanying leakage of cerebrospinal fluid, there should be focus on encephalocele (5). A reddish colour in the physical examination of those with extranasal location could be telangiectasia, and those that are non-pulsatile with a covering of skin are often slow-growing polypoid structure masses. In lesions with insufficient blood supply, reactive changes, local calcifications, and ependymal type cystic degeneration may be seen. The masses with intranasal location are on the lateral wall of the nose. Before diagnosing a nasal polyp in unilateral intranasal polypoid masses in particular, detailed tests must be applied. In the current patient, the lesion grew within days, and necrotic changes were seen on the surface. Histologically it was fibrovascular soft tissue containing mature glial cells (astrocytes and oligodendrocytes). Multinuclear giant cells are often seen. In 10-60% of case series, neuron mass is seen. A low level of oxygen in the mass and insufficient neuroectoderm

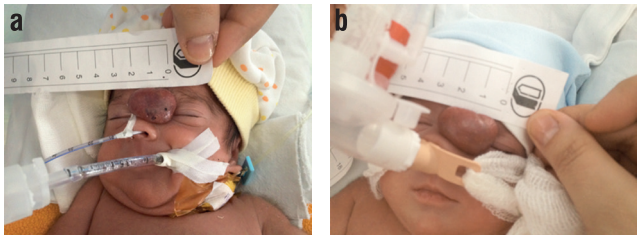

Figure 1. a) The pre-operative apperance of glial heterotopy; a polypoid structure with regular borders, located on the midline mass, b) The pre-operative apperance of glial heterotopy; a polypoid structure with regular borders, located on the midline mass

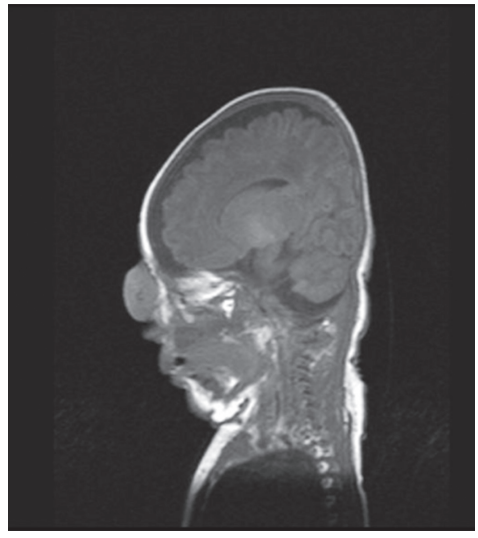

Figure 2. Heterogeneous, hypointense lesion on T1 and T2-weighted magnetic resonance imaging image

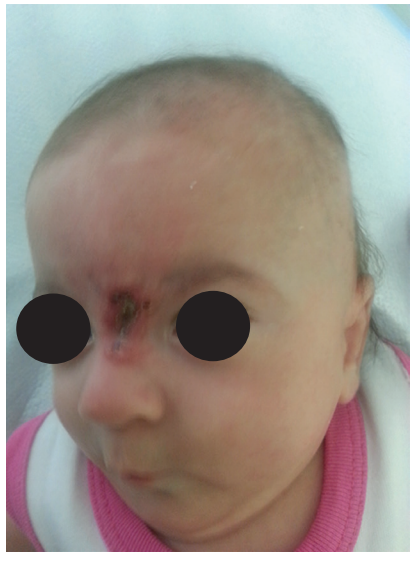

Figure 3. Post-operative apperance of the patient

development cause a low neuron content. Sections prepared for pathological examination may not contain the glial component, and staining with only haematoxylin eosin may not yield information. In suspected cases of glial heterotopy, special staining and immunohistochemical evaluation is necessary. S-100 protein together with Masson Trichrome staining and glial fibril acid staining are important techniques in revealing neurological tissue in fibrotic tissue in particular, and in confirming the diagnosis. In the current case, the diagnosis was confirmed by neuronal tissue showing in glial fibrillary acid staining in the removed mass. Neuron-specific enolase is another stain that can be used (8). Differentiation of glial heterotopy and encephalocele cannot be made with pathological examination. No connection of glial heterotopy with the central nervous system is the most important feature in differentiating it from encephalocele (9). A connection with the central nervous system can be revealed with imaging methods or during surgical procedures. Microscopic invasion, mitotic findings or metastases have not been reported (10,11). Recurrence occurs when the primary lesion could not be fully excised and this has been determined in 4-10% of cases (12). Unlike surgical approaches, evaluation with imaging methods is necessary for the differentiation before the excision procedure of the mass. While tomography gives information related to the defect in the bone structure in particular, magnetic resonance imaging shows soft tissue and intracranial connection. The biopsy and aspiration of pediatric nasal masses are contra-indicated because of the high risk of meningitis and potential damage to functional brain tissue associated with encephalocele. Complete surgical excision is a curative treatment for glial heterotopy (3).

In conclusion, as nasal glioma has the same embryonic root origin, it may be confused with encephalocele and dermoid cysts. Nasal glioma must be carefully evaluated, showing that there is no intracranial connection and that it is ectopic tissue, not herniation tissue. Patients with cerebrospinal fluid leakage during surgical procedures must be re-evaluated. The case presented here is of a rarely seen nasal glioma as a midline defect of the nose which could be confused with encephalocele.

Ethics

Informed Consent: Informed consent was obtained from the patient's parents.

Peer-review: External and internal peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: A.A.K., Concept: A.A.K., T.B.K., Design: A.N.T., Data Collection or Processing: A.A.K., Analysis or Interpretation: A.N.T., Literature Search: T.B.K., Writing: T.B.K.

Conflict of Interest: No conflict of interest was declared by the authors.

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