Published online: 2019-08-13

Case Report

Glial heterotopia of the lip: A rare presentation

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ABSTRACT

Glial heterotopia represents collections of normal glial tissue in an abnormal location distant to the central nervous system or spinal canal with no intracranial connectivity. Nasal gliomas are non-neoplastic midline tumours, with limited growth potential and no similarity to the central nervous system gliomas. The nose and the nasopharynx are the most common sites of location. Existence of glial heterotopia in the lip region is a rare developmental disorder. We report a case of large glial heterotopia in the upper lip region in a full-term female newborn which had intracranial extension with a fibrotic band. After the surgery, there was no recurrence in the follow-up period of 3 years. When glial heterotopia, which is a rare midline anomaly, is suspected, possible intracranial connection and properties of the mass should be evaluated by magnetic resonance imaging. By this way, lower complication rate and better aesthetic results can be achieved with early diagnosis and proper surgery.

KEY WORDS

Fibrotic band; glial heterotopia; lip

INTRODUCTION

Glial heterotopia represents collections of normal glial tissue in an abnormal location distant to the central nervous system or spinal canal with no intracranial connectivity. Nasal gliomas are non-neoplastic midline tumours, with limited growth potential and no similarity to the central nervous system gliomas. They are found in both sexes, are not familial, and most are diagnosed in newborns or infants. The nose and the nasopharynx are the most common sites of location. Nasal glial heterotopia or nasal glioma is used to describe these lesions. Extranasal sites of glial heterotopia are still rarer. Glial heterotopia can be located in scalp, orbit, tongue and palate. Clinically, nasal and nasopharyngeal gliomas present as a nasal obstruction and upper airway obstruction. Extranasal gliomas found at the nose or anywhere of the face cause a cosmetic deformity. These are solid, non-pulsatile, grey or pink coloured lesions, which can be confused with other midline lesions such as dermoid cyst, haemangioma and encephalocele. Confirmatory diagnosis of glial heterotopia

Access this article online	
Quick Response Code:	Website: www.ijps.org
	DOI: 10.4103/0970-0358.182244

How to cite this article: Dadaci M, Bayram FC, Ince B, Bilgen F. Glial heterotopia of the lip: A rare presentation. Indian J Plast Surg 2016;49:112-5.

can be done with of imaging and histopathological findings supported by the immunohistochemical presence of glial fibrillary acidic protein and S100 protein. Surgical excision is the mainstay of treatment for nasal gliomas.[1-3]

Existence of glial heterotopia in the lip region is a rare developmental disorder. We report a case of large glial heterotopia in the upper lip region in a full-term female newborn.

CASE REPORT

A 4-month-old female baby was brought to our clinic with the complaint of mass in the upper lip which existed since birth [Figure 1]. The mass filled midline and left upper lip totally and extended to the right upper lip in a small area. The mass deformed columella and caused superior deviation of the left alar. With palpation, the mass was mobile, had medium consistency, smooth borders, and a size of approximately 2 cm × 2 cm. There was no history of respiratory problems. The lesion did not transilluminate and there was no change in size when the child cried. There were no other pathologies in the head, neck and systemic examination of the child. Ultrasonography and echo–Doppler scanning revealed solid mass with no proliferation of vessels. We planned magnetic resonance imaging (MRI) to make the differential diagnosis from other congenital midline lesions and to evaluate the cranial connection of the mass. The MRI of the brain was normal. However, the MRI of the face showed a mass of 21 mm × 17 mm, which occupied midline and left upper lip, with no erosion of the bony walls and had an intracranial extension with a fibrotic band [Figure 2]. The patient was operated under general anaesthesia. The inner mucosa of the upper lip was incised and the mass was explored. It was seen that the mass continued as fibrotic band in the posterior and the fibrotic band was excised through nasopharynx [Figure 3]. After the removal of the mass, the excess skin in the lip area was excised with an incision from midline to the left alar rim. There was no destruction of the bony structures. The patient had no problems in the post-operative period. The definite pathological result of the mass was glial heterotopia.

In the early post-operative period, the elevation of the alar rim was obvious, but this elevation was indefinite in the post-operative 3rd year. There was no recurrence in the follow-up period of 3 years [Figure 4].

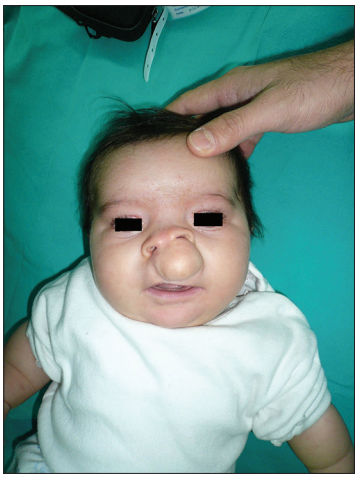

Figure 1: Pre-operative view of a 4-month-old female

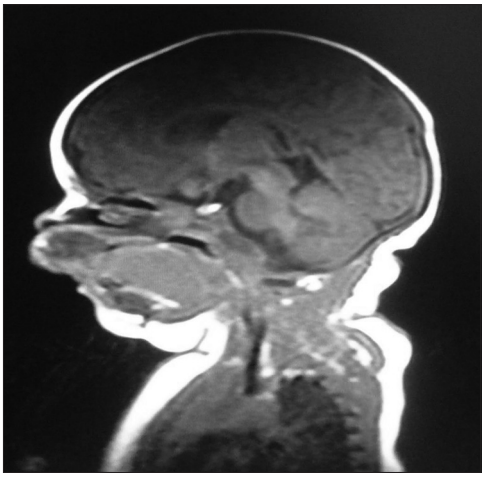

Figure 2: Pre-operative magnetic resonance imaging

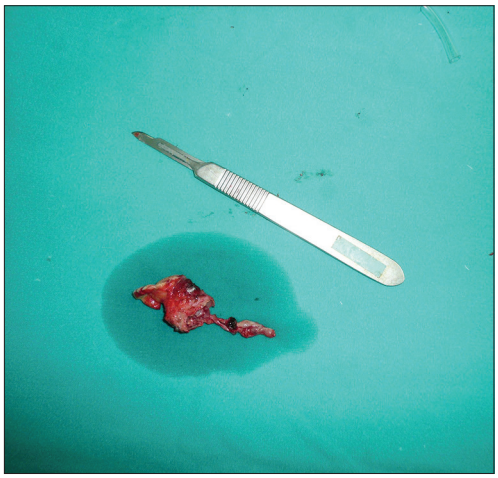

Figure 3: Appearance of mass and fibrotic band

DISCUSSION

Neuroglial heterotopia is a rare congenital midline lesion which originates from benign heterotopic neural tissue.

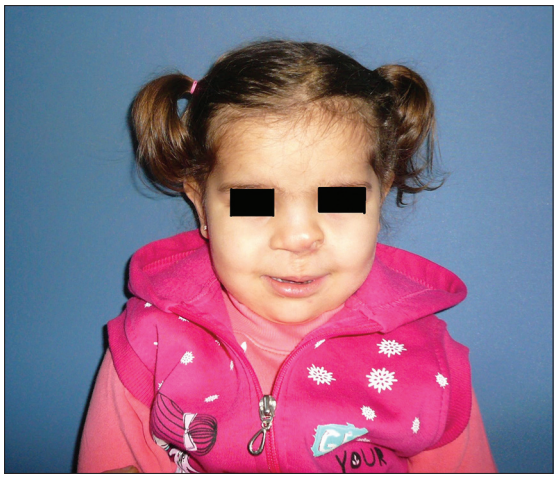

Figure 4: Appearance at post-operative year 3

Although neuroglial heterotopies can locate in nose, nasopharynx, orbit, palate, lip and scalp; they are most commonly located in the nose. Regarding the location of the nasal gliomas, 60% are located extranasally, 30% are intranasal and 10% are mixed.[1-3] In their series of 10 patients, which is the largest in the literature, Penner and Thompson reported that all lesions were located in the nose, either extranasally or intranasally.[1] Isolated lip location is very rare.

The exact aetiology of the nasal gliomas is not known. These lesions are not considered as tumours, rather they are extracranial presence of ectopic glial tissue as a result of abnormal closure of nasal and frontal bones. Nasal glial heterotopias usually have no connection with the central nervous system. However, 15–20% of the cases had intracranial connection with a fibrous band. In the cases with intracranial connection, cerebrospinal fluid leakage and recurrent attacks of meningitis can be seen.[1-5] In their series of 10 patients, Penner and Thompson reported intracranial connection with a fibrous band in one patient.[1] In our case, there was intracranial extension with a fibrotic band, and we detected it by MRI before surgery. We dissected the mass *en bloc* until the posterior part of the nasopharynx and excised. Pathologic examination revealed only fibrous tissue in this band.

Dermoids, dermoid cysts, gliomas, encephaloceles and haemangiomas are among the congenital midline lesions. Encephalocele is the most commonly encountered congenital midline lesion. Nasal gliomas are most commonly confused with encephaloceles. Although gliomas can be connected with cranial cavity with a fibrous band, encephalocele is defined as a neuroectodermal lesion with direct connection with the brain which can contain dura, brain tissue and cerebrospinal fluid. In addition, encephaloceles transilluminate. The most important method to determine the properties and intracranial connection of the masses is by MRI.[5-8]

The treatment of nasal gliomas is total surgical excision. Early surgery is mandatory in these cases. Although only cosmetic problems are present in cases located extranasally, severe complications such as meningitis, brain abscess can be seen in cases with intranasal location.[1,5] Total cure was achieved in our case with surgery but elevation of left alar rim and scar tissue in the lip persisted. Alar rim rotation and redirection and scar revision were planned in later ages.

When glial heterotopia, which is a rare midline anomaly, is suspected, possible intracranial connection and properties of the mass should be evaluated by MRI. By this way, lower complication rate and better aesthetic results can be achieved with early diagnosis and proper surgery.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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