

Case Report

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Congenital extranasal glial heterotopia in infancy: a rare case report

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ABSTRACT

A seven-month-old female child has been presented with extra nasal mass on the dorsum of the nasal bridge since birth. On clinical examination a 1.6x1.4 cm globular, firm, non-compressible, non-pulsatile mass was noted over the nasal dorsum, slightly left of midline. Radiological studies were suggestive of extranasal glioma without intranasal or intracranial extension. Histopathological examination revealed a non-encapsulated, ill-defined lesion composed of sheets and nests of benign and mature glial tissue. Glial heterotopia is defined as the mass of mature brain tissue isolated from the cranial cavity or spinal canal. Nasal glial heterotopias are congenital tumors. The nose and nasopharynx are the most common sites of location. Nasal glial heterotopias are rare benign lesions that can be challenging to diagnose clinically and radiologically due to their uncommon presentation. In this case report; we discuss a rare case of extranasal glial heterotopia, highlighting its clinical presentation, diagnostic challenges, radiological and histopathological features.

Keywords: Nasal glioma, Heterotopia, Glial tissue, Congenital nasal swelling

INTRODUCTION

Glial heterotopia is a rare non hereditary, benign congenital malformation composed of mature glial tissue isolated from cranial cavity and spinal cord. The incidence of Glial heterotopia is 1 in 20000-40000 live births with female preponderance.¹ Most structures involve midline structures, including the nose and nasopharynx as well as oropharynx, palate, tongue, tonsils and lips.² The term “glioma” is a misnomer, as these lesions are not neoplastic.³ “Neurological heterotopia” is a more accurate term. A total of 264 cases has been reported in world literature since the first described by Reid in 1852.⁴ We diagnosed the present case as extranasal glial heterotopia on histopathology.

CASE REPORT

A seven-month-old female child presented with a firm mass over the nasal bridge present since birth. On clinical examination a 1.6x1.4 cm globular, firm, non-compressible, non-pulsatile mass was noted over the nasal dorsum, slightly left of midline. The overlying skin was normal. There was no transillumination or increase in size with crying. Clinically no other congenital anomaly was present and was diagnosed as nasal dermoid cyst. Further radiological evaluation by Computed Tomography showed a well-circumscribed, non-enhancing nasal soft tissue mass without intra-cranial and intra-nasal communication suggestive of extra nasal glioma. Also, cranial ultrasound and Magnetic resonance imaging was suggestive of extra nasal glioma. Complete surgical excision was performed.

Grossly the mass was skin covered, globular with tan, solid homogenous and firm appearance on cut surface. Histopathological examination revealed skin with underlying dermis showing a non-encapsulated ill-defined lesion composed of variable size sheets and nests of benign and mature glial tissue. The astrocytes appeared dispersed in neurofibrillary matrix in the background of vascularised connective tissue. There was no evidence of atypia, mitosis or necrosis. This confirmed the diagnosis of extranasal glial heterotopia on microscopy. After 2 months of complete surgical excision, the patient is in good health without any signs of recurrence.



Figure 1: Mass over nasal septum.

DISCUSSION

The heterotopia is applied to microscopically normal cell or tissues that are present in abnormal location. Heterotopic neuroglial tissue is defined as a mass composed of mature brain tissue isolated from the cranial cavity or spinal canal. The differential diagnosis includes congenital nasal dermoid cysts, nasal glioma, nasal encephalocele, hemangioma, fibrous dysplasia, as well as malignant tumor such as rhabdomyosarcoma, neuroblastoma.⁵

The pathogenesis of these lesions remains uncertain, and several hypotheses have been proposed. Firstly, they may represent a sequestered encephalocele. Secondly, they could result from separation of primitive embryonic neural tissue from the developing brain owing to altered timing of chondrocranial fusion. Thirdly, they may arise from displacement of isolated pluripotent cells during early embryogenesis with subsequent differentiation into mature neural tissue. Finally, some authors suggest entrapment of abnormal glial tissue from the olfactory bulb. The most widely accepted theory considers heterotopic neural tissue

to be a variant of encephalocele in which the central nervous system connection is either absent or regresses to a vestigial remnant.^{2,3}

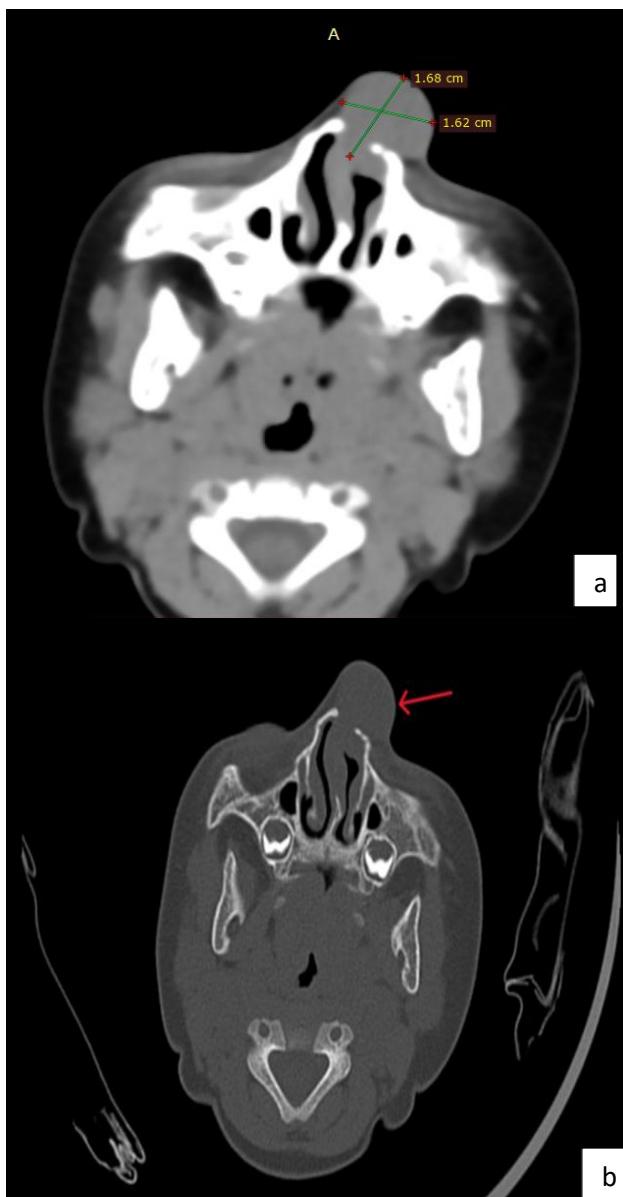


Figure 2 (a and b): CT reveals a well- circumscribed, non-enhancing nasal soft tissue mass without bony erosion or intracranial communication.

Neuroglial heterotopias can be classified according to their anatomical location. They include intraparenchymal central nervous system lesions, dural and leptomeningeal lesions, intracranial but extracerebral lesions, midline nasal glial heterotopia thought to originate as encephaloceles, and non-midline lesions of the head and neck.² Encephaloceles and glial heterotopia have a similar embryologic origin. The encephalocele is a herniation of cranial contents through a defect in the cranium with meninges, whereas glial heterotopia is thought to be an encephalocele that has lost the intracranial connection.^{6,7}

Nasal glial heterotopia are congenital tumors arising from the normal neurectodermal tissue which is entrapped during the closure of the anterior neuropore.⁸ These are locally aggressive lesions which are usually present at birth and 90% cases are usually diagnosed before the age of 2 years.⁹ They often manifest as extranasal (60%), intranasal (20%) and mixed (10%).¹⁰

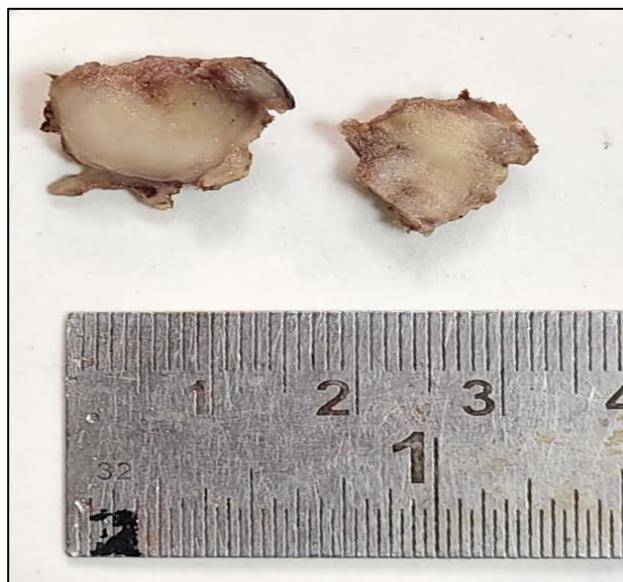


Figure 3: Gross examination homogenous, tan, firm cut surface.

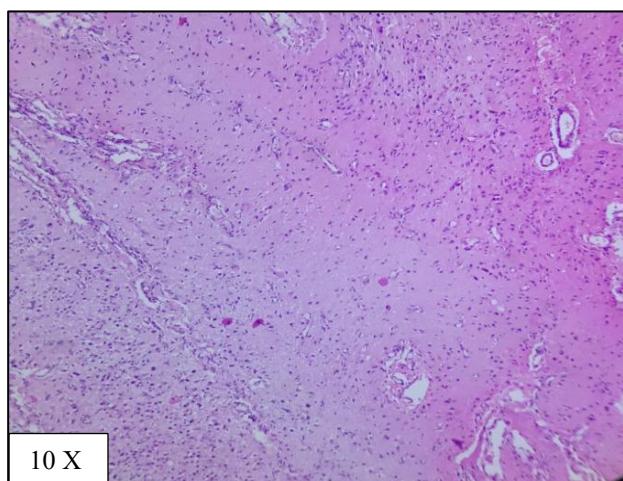


Figure 4: Glial tissue arranged in sheet.

Clinically, extranasal glial heterotopia present as masses that do not transilluminate and whose volume remains stable despite crying and efforts. These are benign without any potential for malignant transformation and may result in distortion of nasal septum resulting in facial deformity.⁴

Evaluation of congenital swelling is mostly done by magnetic resonance imaging to rule out intracranial and intranasal connection. Biopsy is contraindicated because of the increased risk of meningitis or perhaps the removal

of functional brain matter. However histopathological examination of excised specimen is mandatory for establishing diagnosis. Histologically, nasal glial heterotopia is characterized by varying size of neurons and glia, few showing gemistocytic astrocytes and varying degrees of fibrosis. A large amount of fibrotic tissue can undermine that glial tissue in H and E sections or glial tissue may mimic fibrotic tissue, as in our case.⁵ The treatment of choice is complete surgical excision with 4-10% recurrence rate.¹¹

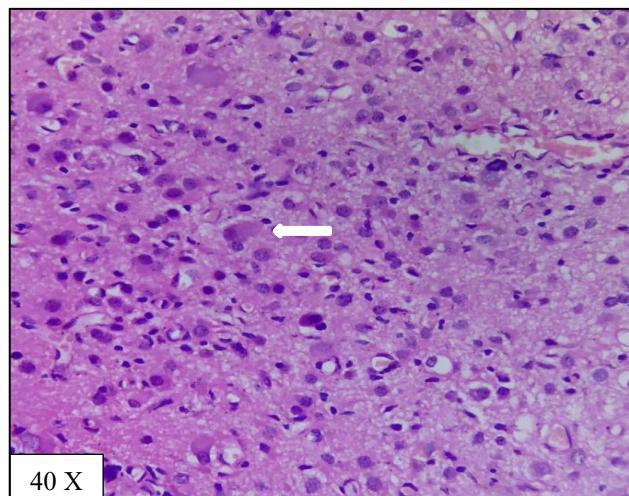


Figure 5: Astrocytes in neurofibrillary background.

CONCLUSION

In summary, nasal glial heterotopia are rare benign lesions that can be challenging to diagnose clinically and radiologically due to their uncommon presentation. This highlights the essential role of histopathology examination for their diagnosis and to rule out other pediatric malignant tumors. A thorough histopathological workup along with clinico-radiological correlation is mandatory for early diagnosis and treatment.

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