Prenatal Diagnosis of Nasal Glioma Associated with Metopic Craniosynostosis: Case Report and Review of the Literature

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ABSTRACT

Nasal gliomas (nasal glial heterotopia) are rare benign congenital frontonasal lesions occurring in approximately 1: 20.000 - 40,000 live births. The diagnosis is rarely reported prenatally. Nasal gliomas are typically isolated lesions, with syndromic association being exceedingly rare. Metopic craniosynostosis can occur as an isolated abnormality or in association with multiple syndromes. This case is the first reported case of nasal glioma in association with craniosynostosis in the published literature.

CASE REPORT

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Imaging Findings

A 24-year-old primigravida with no significant past medical history underwent a routine anatomy ultrasound at 19 weeks gestation. Fetal sonography revealed a solid mass of the left face between the nose and medial aspect of the left orbit (Figure 1a-d). No extension to the orbit or intracranial involvement was identified. The intracranial structures and bony structures of the head and face were normal and no additional abnormalities were identified. Prenatal fetal MRI was performed at 21 weeks gestation. Fetal MRI demonstrated a circumscribed 8 x 7 x 11 mm frontonasal mass at the nasal root extending to left of midline with isointense signal intensity to brain parenchyma on T2-weighted images (Figure 2a & b). The intracranial structures were normal and there was no calvarial defect or direct intracranial connection. The parents were informed that the most likely diagnosis was nasal glioma however alternative diagnoses such as teratoma or subtle encephalocele were difficult to excluded prenatally.

A female fetus was born uneventfully by spontaneous vaginal delivery at 39 weeks gestation with a birth weight of 2875 grams (small for gestational age) and Apgar scores of 9 at 1 and 5 minutes. Examination revealed a 3cm soft, round mass at the nasal bridge with patent nasal fossae. Otherwise, she was clinically well with an intact neurological examination. Neonatal MRI was performed at 2 days of age, at which time, the mass demonstrated isointense signal intensity on T1- and T2-weighted images to brain parenchyma with no

pathologic enhancement (Figure 3a-d). The absence of intracranial extension was confirmed, excluding the diagnosis of frontonasal encephalocele. Additionally, trigonocephaly was present (Figure 3e). Surgical resection was planned for several months following birth.

Management

The patient was referred for head CT at 2 months of age for evaluation of metopic craniosynostosis and surgical planning. CT demonstrated a subtle midline defect of the left nasal bone with extension of the nasal glioma into the nasal cavity (Figure 4a & b). Metopic craniosynostosis with trigonocephaly was confirmed (Figure 4c & d). At 4 months of age the patient underwent uneventful excision of nasal glioma, nasal bone reconstruction, and endoscopic treatment of metopic craniosynostosis by metopic suturectomy. Pathological examination confirmed the diagnosis of nasal glioma (glial heterotopia) (Figure 5).

DISCUSSION

Etiology & Demographics

Nasal gliomas are rare congenital frontonasal lesions estimated to occur in 1: 20,000 - 40,000 live births with a male:female ratio of 3:2 [1]. Approximately 60% of nasal gliomas are extranasal, 30% are intranasal, and 10% are extranasal with intranasal extension.

Nasal gliomas are non-neoplastic masses composed of heterotopic neuroglial tissue and thus these lesions are also referred to as nasal glial heterotopia. During early gestation the nasal bone is separated from the frontal bone by a small fontanelle called the fonticulus frontalis. The fonticulus frontalis closes as the frontal and nasal bones fuse, forming the frontonasal suture. Extranasal gliomas are formed when brain tissue has herniated through the fonticulus frontalis and becomes trapped over the bridge of the nose following its closure. The slightly less common intranasal gliomas are formed when brain tissue herniates through the foramen cecum into the prenasal space and becomes disconnected from the brain during closure of the frontal and nasal bones. Nasal gliomas lack a direct fluid-filled connection to the intracranial subarachnoid space, differentiating them from encephaloceles.

Craniosynostosis is the pathologic premature closure or ossification of a cranial suture. The metopic suture, which separates the frontal bones, is the first suture to close, typically between 3 to 9 months of age [2]. Pathologic premature closure of the metopic suture is the second most common type of craniosynostosis (incidence 1:5,200) [3]. Metopic craniosynostosis results in trigonocephaly, or a triangular head shape. The pathogenesis of craniosynostosis is poorly understood and is likely multifactorial including fetal constraint in utero and hormonal and genetic factors. Trigonocephaly typically occurs as an isolated abnormality.

Clinical & Imaging Findings

Nasal gliomas present on ultrasound as a solid frontonasal mass with characteristic end-diastolic low arterial flow velocity on Doppler imaging [4]. On MRI, nasal gliomas have intermediate signal intensity on T1-weighted images and intermediate to high signal intensity on T2-weighted images due to gliosis. Occasionally, a tract of fibrous tissue can be seen extending from the nasal glioma to the frontonasal region. However, the most important observation to make on MRI is the lack of a distinct connection to the intracranial structures, thereby differentiating nasal glioma from encephalocele.

Nasal gliomas are typically isolated lesions occurring in an otherwise normal newborn. Syndromic occurrence is exceedingly rare with only one such case in the reported literature in which a nasal glioma occurred concurrently with PHACE syndrome [5]. Since nasal gliomas occur due to anomalous development of the fonticulus frontalis, which is contiguous with the metopic suture, it is possible that anomalous development of the fonticulus frontalis and the metopic suture might occur concurrently. Additionally, as intrauterine constraint has been proposed as a possible factor in the etiology of craniosynostosis, it is possible that the presence of a nasal glioma may result in constraint of the fetal calvarium, thereby resulting in craniosynostosis. However, this is the first reported case of this association in the medical literature.

Treatment & Prognosis

Follow diagnosis of a nasal glioma, surgical resection early in life is the treatment of choice to reduce complications including nasal deformity and visual disturbances. Surgical resection is typically curative.

Differential Diagnoses

Prenatal diagnosis of nasal glioma is uncommon with only 4 such cases in the published literature [6-9]. Prenatally, the differential diagnosis of nasal glioma includes frontonasal encephalocele, orbital dermoid, dacryocystocele, and hemangioma. The primary differential diagnosis in most cases of nasal glioma is a frontonasal encephalocele. Frontonasal encephalocele is a mass of heterotopic neuroglial tissue in a similar location to nasal glioma (near the bridge of the nose or within the nasal cavity). Nasal glioma and frontonasal encephalocele have a similar appearance on imaging including a solid mass on ultrasound, T1 isointensity and T2 iso- to hyperintensity, and no significant enhancement. Frontonasal encephaloceles have a direct fluid connection to the intracranial subarachnoid space, which is the primary differentiating feature on imaging. Orbital dermoid is a developmental ectodermal inclusion cyst, which typically presents as a painless subcutaneous nodule near the orbital rim in childhood. Orbital dermoids have a characteristic heterogeneous appearance on both sonography and MRI with internal fat contents and a thin rim of peripheral enhancement. Dacryocystocele is a cystic orbital mass located inferiomedially to the globe, which typically presents in the neonatal period and resolves spontaneously. Dacrocystoceles are typically located slightly more lateral than nasal gliomas and frontonasal encephaloceles, which are typically paramidline. Dacrocystoceles present on imaging as a round cystic lesion, which is anechoic on ultrasound and follows fluid signal intensity on all sequences on MRI. Minimal peripheral enhancement can be seen. Thickened, enhancing walls suggest superimposed infection. Hemangiomas can

present as a growing reddish soft tissue lesion of the scalp or face of neonates or infants. While the majority of these lesions regress spontaneously, large hemangiomas or those with high vascular flow can cause high output cardiac failure. Hemangiomas have a variety of imaging appearances including solid to mixed solid and cystic with a heterogeneous appearance on ultrasound and MRI. Intense enhancement is characteristic of hemangiomas.

TEACHING POINT

Nasal gliomas present as a solid frontonasal mass on prenatal ultrasound and as an intermediate T1, intermediate to high T2 frontonasal mass on MRI without direct fluid connection to the subarachnoid space.

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FIGURES

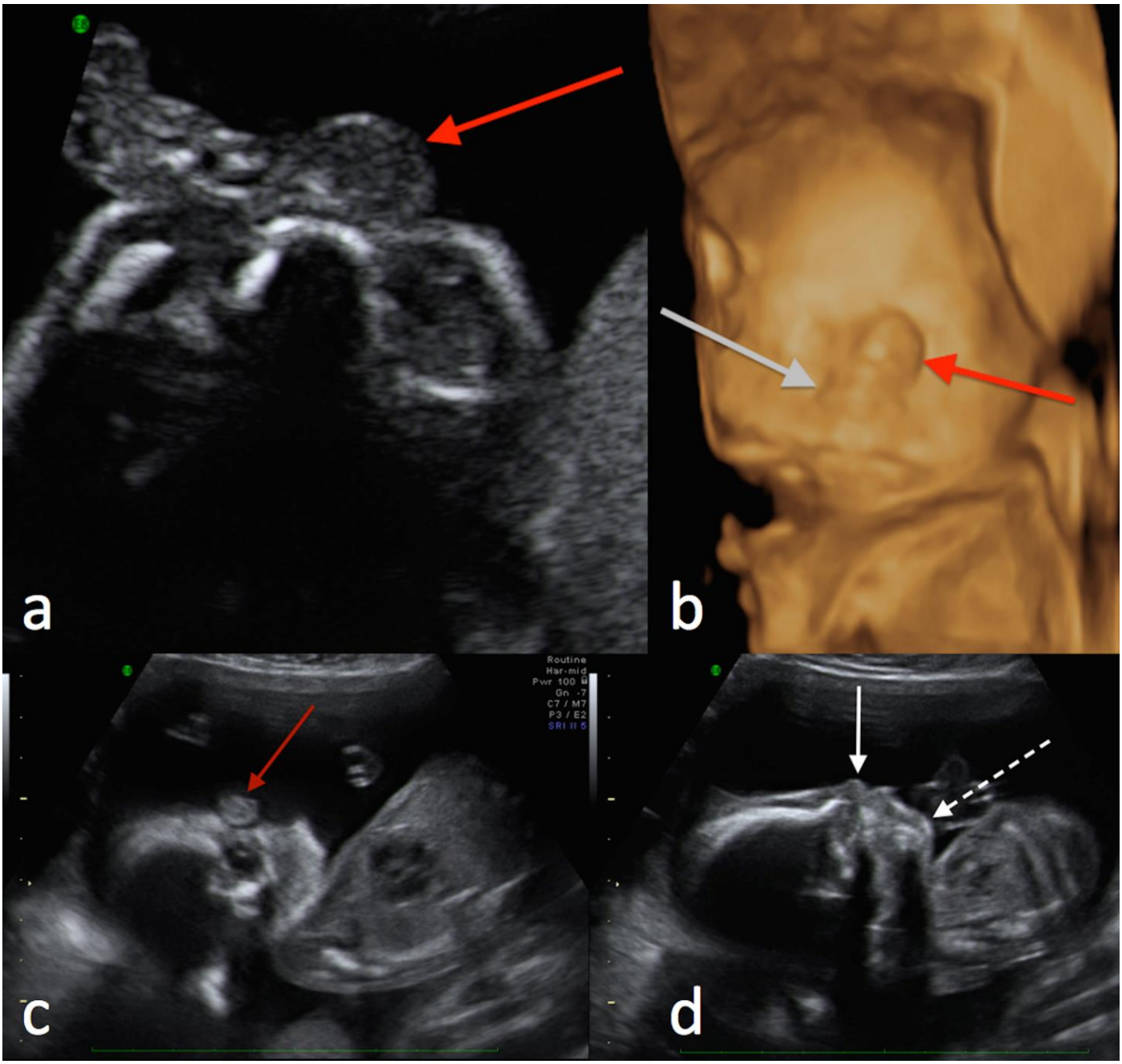

Figure 1: Female fetus at 19 weeks gestation with nasal glioma. Transverse (a) and 3-dimensional frontal (b) images from prenatal ultrasound demonstrate a frontonasal soft tissue mass (red arrows) superior and to the left of the nose (white arrow). Off-midline saggital prenatal ultrasound image (c) demonstrates a rounded frontonasal mass (red arrow). Midline sagittal ultrasound image (d) demonstrates the location of the nose (solid white arrow) and the chin (dashed white arrow) in relation to the nasal glioma seen in image 1c. Technique: Voluson 730, GE Healthcare, USA. RAB4-8-D wide band convex volume transducer (2-8 MHz).

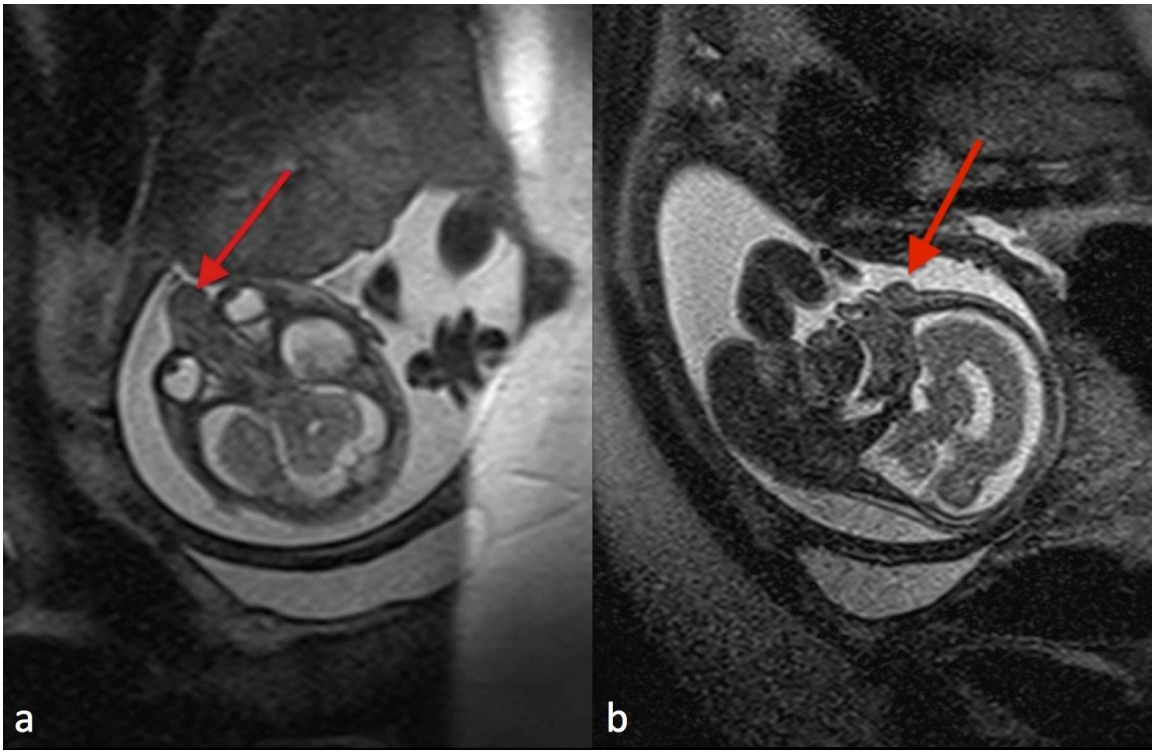

Figure 2: Female fetus at 21 weeks gestation with nasal glioma. Axial (a) and sagittal (b) T2-weighted MRI demonstrates a frontonasal mass (red arrow) that is isointense to brain parenchyma with no definite connection to the intracranial structures. Technique: 1.5 T magnet strength, Avanto MR, Siemens, Germany, axial T2 sequence 1100 TR, 105 TE, 3.0 mm slice thickness; sagittal T2 sequence 1100 TR, 108 TE, 2 mm slice thickness.

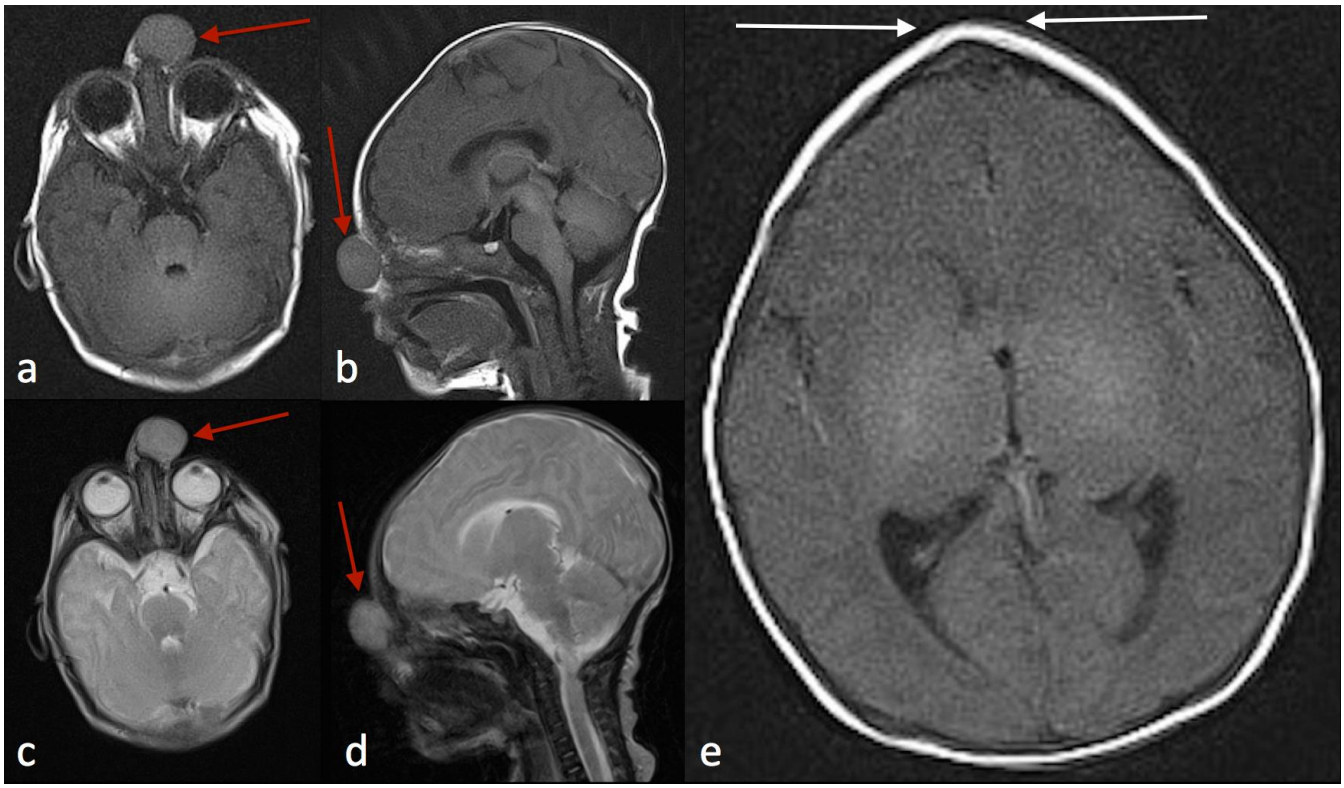

Figure 3: Two-day-old female neonate with nasal glioma. Axial and midline sagittal T1 Blade MR images (a & b), axial T2 Blade MR image (c), and sagittal T2 tse image (d) demonstrate a 1.8 x 1.5 x 2.0 cm left paramidline frontonasal mass immediately superior to the nasal bridge that is isointense to brain parenchyma on all sequences. The lack of connection to the intracranial structures differentiates this nasal glioma from a frontonasal encephalocele. An additional axial T1 Blade image (e) demonstrates trigonocephaly (white arrows). Technique: 1.5 T magnet strength, Avanto MR, Siemens, Germany, axial T1 blade sequence 1800 TR, 55 TE, 4.0 mm slice thickness; sagittal T1 blade sequence 1940 TR, 55 TE, 4.0 mm slice thickness; axial T2 blade sequence 4000 TR, 99 TE, 4.0 mm thickness; sagittal T2 weighted image 5120 TR, 115 TE, 4.0 mm slice thickness.

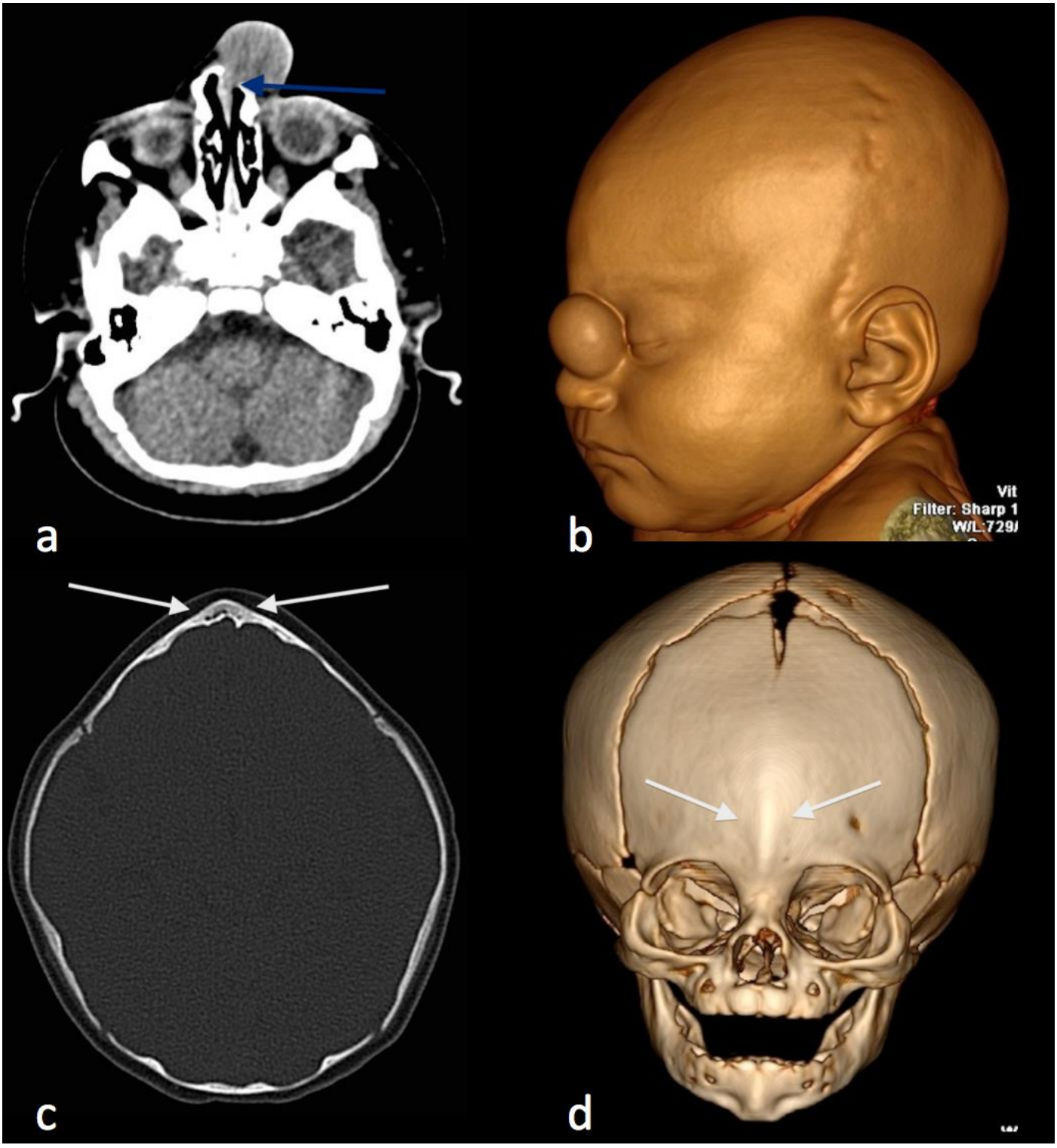

Figure 4: 2 month old female with nasal glioma and trigonocephaly. Axial head CT in soft tissue windows (a) demonstrates subtle extension of the nasal glioma into the nasal cavity (blue arrow). 3-dimensional soft tissue (b) demonstrates the paramidline, supra-nasal location of the nasal glioma. Axial CT in bone windows (c) and 3-dimensional frontal image (d) demonstrate premature closure of the metopic suture with trigonocephaly. Technique: 64-row multidector sensation CT, Siemens, Germany, 240 mas, 100kVP, 1.0 mm reformation. 3D reconstructions using Vitrea, Toshiba Corporation, Japan.

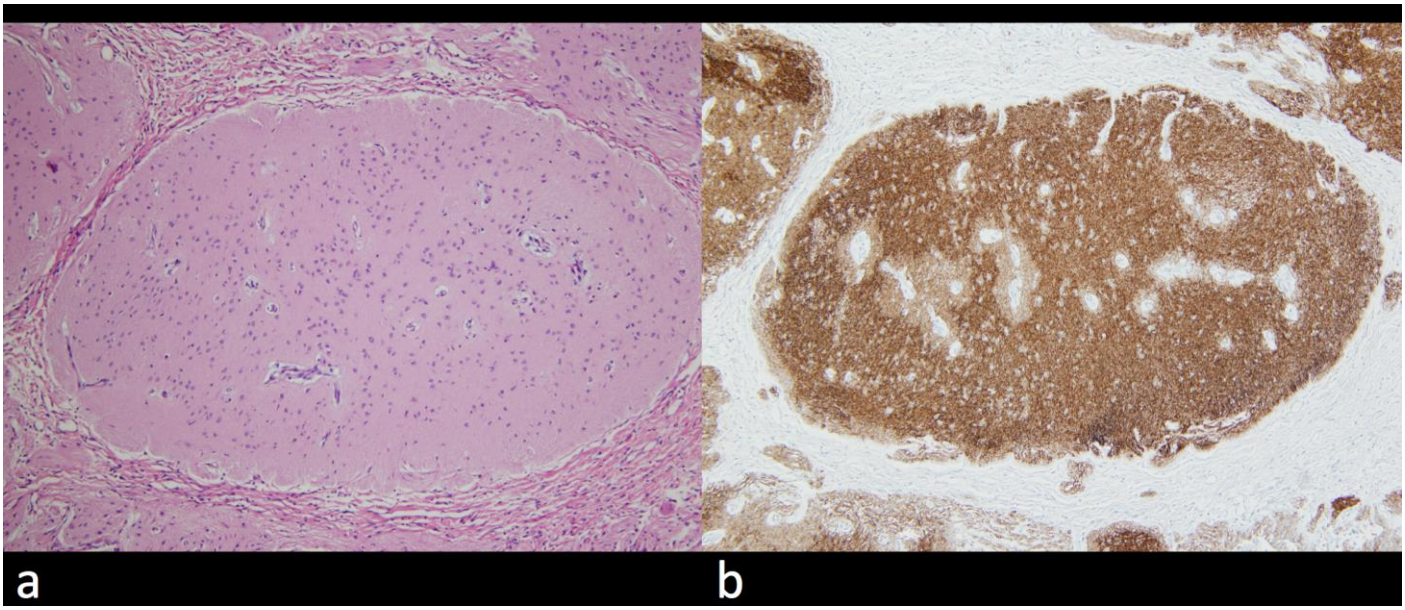

Figure 5: Surgical pathology specimen of nasal glioma (nasal glial heterotopia). Hematoxylin and eosin stain, 100x magnification (a) demonstrates a focus of neuroglial tissue showing the small, bland nuclei of numerous astrocytes present in a background of finely fibrillar eosinophilic neuropil. Immunohistochemical stain for glial fibrillary acidic protein (GFAP), 100x magnification (b) demonstrates strong, diffuse expression of GFAP, an intermediate filament protein found in the cytoplasm of astrocytes, within this focus of neuroglial tissue.

Etiology	• Benign, sporadic, developmental lesion of heterotopic neurogenic tissue. • Arises during closure of the fetal frontal and nasal bones. • Rarely associated with other congenital malformations.
Incidence	• 1 : 20,000 – 40,000 live births.
Gender ratio	• Male:female ratio of 3:2.
Age predilection	• Congenital lesion typically diagnosed at birth or during infancy.
Classification	• Extranasal – 60%: Located off midline at the nasal bridge. Present clinically as a red-blue colored mass without pulsations. • Intranasal – 30%: Located medial to the middle turbinate bone within the nasal passage. May present clinically with nasal congestion or obstruction, obstruction of the nasolacrimal duct, meningitis, or epistaxis. • Mixed – 10%
Risk factors	• No risk factors have been described.
Treatment	• Surgical resection is the treatment of choice, ideally performed early to prevent secondary visual disturbances or nasal deformity.
Prognosis	• Complete surgical resection is usually curative. • 10% recurrence rate with incomplete resection. • Good prognosis due to typical lack of other congenital malformations.
Findings on imaging	• US: Solid frontonasal mass with a characteristic low arterial velocity during end-diastole on Doppler. • MRI: T1 isointense to brain parenchyma. Typically no enhancement. T2 isointense to hyperintense due to gliosis. No direct fluid connection to the intracranial subarachnoid space.

Table 1: Summary table of key information of nasal glioma.

	Clinical History	US	MRI	Pattern of enhancement
Nasal Glioma	• Congenital mass of heterotopic neuroglial tissue near the bridge of the nose or in the nasal cavity. • Slow growth compared to adjacent tissues. • No change in size with Valsalva.	• Solid frontonasal mass. • Doppler: Classic end-diastole low arterial velocity.	• T1: Isointense to brain parenchyma. • T2: Isointense to hyperintense to normal brain due to gliosis. • No direct fluid connection to the intracranial subarachnoid space.	• No enhancement within soft tissue.
Frontonasal Encephalocele	• Congenital mass near the bridge of the nose or in the nasal cavity. • Changes in size with crying or Valsalva.	• Solid frontonasal mass. • Associated with widened interorbital distance.	• T1: Isointense to brain parenchyma. • T2: May be hyperintense due to gliosis. Hyperintense CSF surrounds herniated soft tissue. • Contiguity with intracranial parenchyma through an osseous defect.	• No enhancement within soft tissue.
Orbital Dermoid	• Ectodermal inclusion cyst. • Typically presents in childhood and teenage years. • Firm, painless subcutaneous nodule near the orbital rim.	• Complex lesion with variable attenuation.	• T1: Strongly hyperintense components due to fatty tissue. • T2: Isointense or mildly hypointense with heterogeneous debris.	• Typically has a thin rim of enhancement. • May have avid enhancement if complicated by rupture or inflammation.
Dacryocystocele	• Round cystic orbital mass medial to the globe in a neonate. • Located more lateral than nasal glioma or encephalocele. • Most resolve without surgery.	• Round, cystic anechoic mass. • Resembles an extra eye.	• T1: Hypointense, well-circumscribed mass at the medial canthus. • T2: Hyperintense due to fluid. • May have variable signal intensity if complicated by infection.	• Minimal enhancement of the cyst wall. • Thickened enhancing wall can be seen if infected.
Hemangioma	• Growing reddish soft tissue mass in the neonate or infant. • Most regress spontaneously. • Large lesions or those with high vascular flow can cause high output cardiac failure if not treated.	• Heterogenous and hyperechoic. • Solid or mixed solid and cystic.	• T1: Heterogenous hypo- to isointense. • T2: Heterogenous hyperintense signal. • Flow voids are commonly present.	• Intense enhancement.

Table 2: Differential diagnosis table for frontonasal mass in a fetus or neonate.

ABBREVIATIONS

CT: Computed tomography
MRI: Magnetic resonance imaging
PHACE syndrome: Posterior fossa abnormalities, hemangioma, arterial lesions, cardiac anomalies, eye abnormalities

KEYWORDS

Nasal glioma; Trigonocephaly; Fetal MRI