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SHORT COMMUNICATION

Prenatal ultrasound and histological diagnosis of fetal nasal glioma (heterotopic central nervous system tissue): report of a new case and review of the literature

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Abstract
Introduction Nasal glioma is a rare, benign congenital midline facial lesion.
Materials and methods Prenatal ultrasound diagnosis performed at 2nd trimester of pregnancy revealed a right-sided mass at the level of the fetal face extending from the right internal canthus to the nasal bridge.
Conclusion Differential diagnosis of facial mass in the fetus represents a critical issue because is essential in guiding the prenatal counselling of the couple and in guiding the prenatal and/or postnatal management. Alternative diagnoses such as dacryocystocele, dermoid cyst, retinoblastoma or teratoma, hemangioma, and encephalocele that can not completely be excluded prenatally are discussed. Embryology, pathology, prenatal ultrasound diagnostic clusters of the lesion as well as MR imaging findings are discussed together with review of the literature.

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Keywords Nasal glioma · Nasal cerebral heterotopia · Congenital anomalies · Prenatal diagnosis · Ultrasound · Necropsy

Introduction

Nasal gliomas also known as cerebral nasal heterotopia are rare, congenital midline facial defect with an estimated incidence of only 1:20,000–1:40,000 live births and with a female to male ratio of 3:2 [1]. These lesions can be located extranasal in 60% of cases, 30% intranasal or be found in both sides. Nasal gliomas or heterotopic central nervous system tissue (HCNST) are considered a non-neoplastic glial heterotopia in an extracranial site [2]. Some theories have been advanced to explain the presence of extracranial nervous tissue. First of all, it is mainly regarded as a variant of encephalocele which has lost communication to the central nervous system. Another theory suggests that the presence of displaced pluripotential stem cells able to differentiate in mature nervous system [3].

Case report

A 38-year-old gravida was referred for 2nd level ultrasound due to right-sided mass at the level of the fetal face at 2nd trimester scan. Ultrasound examination was carried out using an Accuson 128XP/10 ART® (Universal Diagnostic Solutions, Ocean Drive, CA, USA) ultrasound machine equipped with a transabdominal 3.0–5.0 MHz probe. Ultrasound examination revealed a moderately hypoechoic mass extending from the right internal canthus to the nasal bridge (Fig. 1), with bright external contour probably representing the skin (Fig. 2). Doppler analysis was performed and

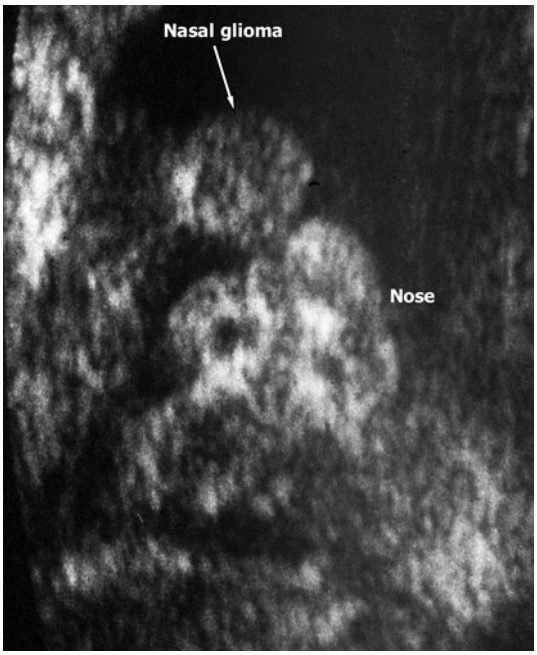

Fig. 1 Transabdominal 2D scan performed at 20 weeks of pregnancy showing a hypoechogenic, rounded mass on the right side of the fetal face extending from the right internal canthus to the nasal bridge

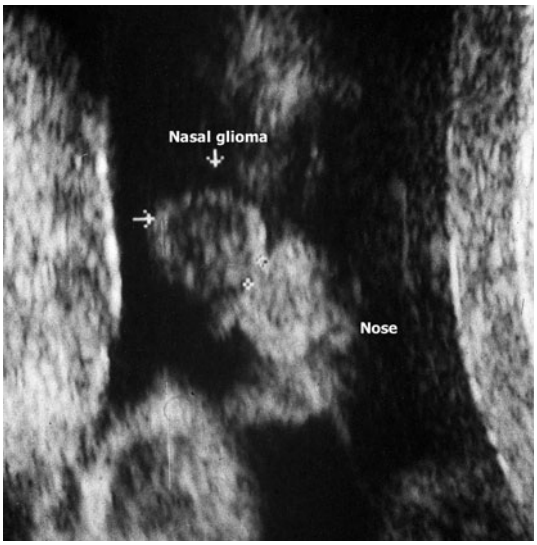

Fig. 2 Detailed of the lesion: note the bright external contour probably representing the skin

showed absent of vascular pattern within the lesion (Fig. 3). Uneventful amniocentesis was performed after signed informed consent and showed a 46,XX karyotype with elevated α-FP levels (75,000 ng/ml). Extensive and detailed discussion about presumptive and differential diagnosis, probable postnatal therapy and prognosis was undertaken by a multispecialists team.

To this regard, antenatal MR imaging was also advised in order to enhance the prenatal, ultrasound diagnostic accuracy. However, based on ultrasound findings consistent

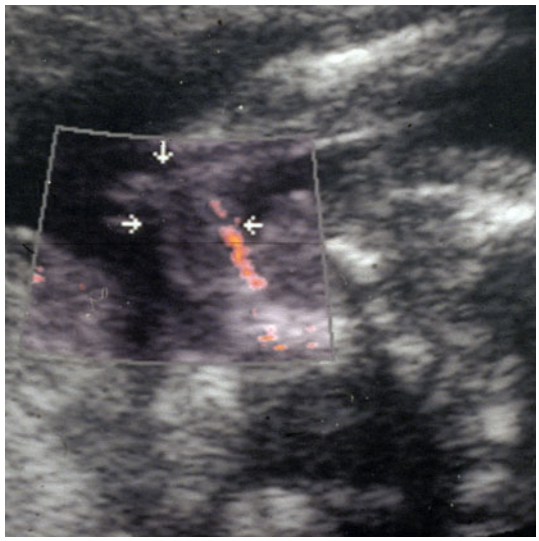

Fig. 3 Doppler analysis demonstrated absent flow within the lesion

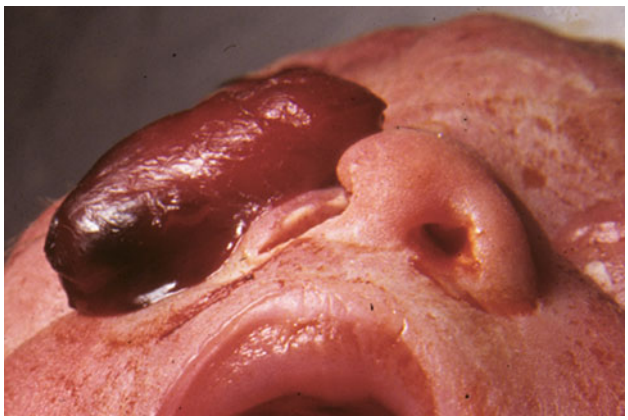

Fig. 4 Necropsy confirmed the presence of a purple, non-compressible mass of 18 × 11 × 10 mm

of possible fetal malformation, the couple refused complementary prenatal diagnostic investigations. A psychiatric certification demonstrating severe psychophysical disturbances associated with a high risk of serious life-threatening damages in the mother was provided. Legal termination of pregnancy (TOP) was at this stage achieved, after admission to hospital, by vaginal administration of prostaglandin E (Cervidil®, Serono, Latina, Italy), according to the Art.6, Art. 7 of the Italian Law 194/78.

Necropsy was performed and on gross examination the right nostril showed a superficial damage through which a purple, non-compressible mass of 18 × 11 × 10 mm protruded (Fig. 4). Histologically, the lesion was composed of a cellular proliferation similar to the white matter (Fig. 5).

There were astrocytes and neuroglial fibers intermixed with a fibrovascular connective tissue (Fig. 6). No neurons, mitoses or glomeruloid vascular proliferation were

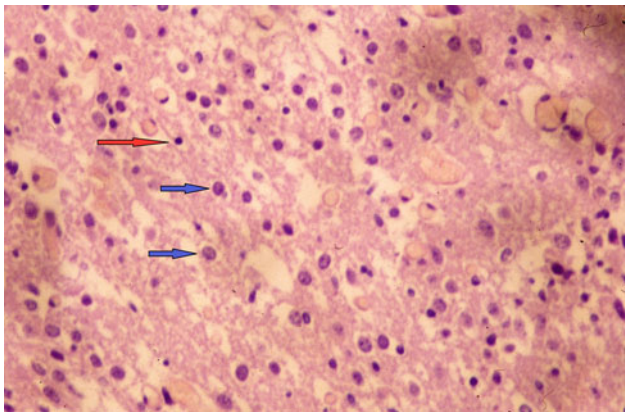

Fig. 5 Cellular proliferation similar to normal white matter with astrocytes (vesicular nuclei, *blue arrows*) and smaller darker oligodendrocytes (*red arrow*)

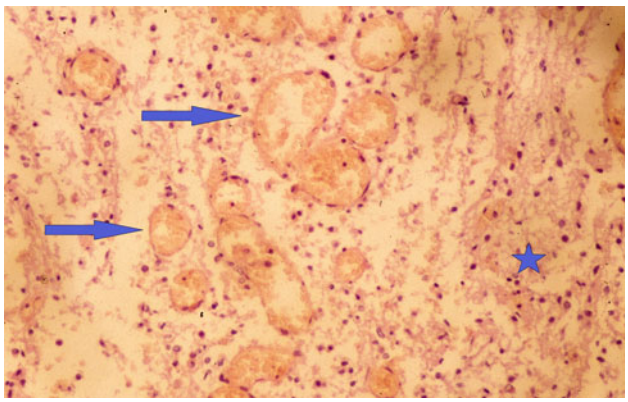

Fig. 6 Glial fibers (*star*) intermixed with a rich vascular network (*arrows*)

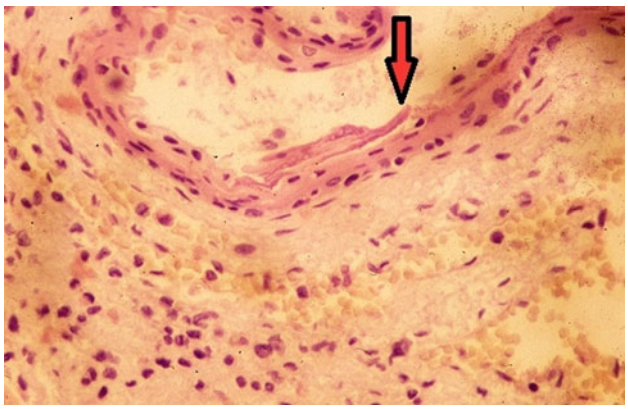

Fig. 7 Squamous epithelium (*arrow*) as an external cover of the polypoid mass

identified. Immunohistochemically, there was a strong reactivity for glial fibrillar acid protein (GFAP). Furthermore, the lesion was externally covered by squamous epithelium (Fig. 7).

Discussion

Nasal glioma is a rare, benign congenital midline tumor, with the potential for intracranial extension, that has been diagnosed prenatally in only two cases at 2nd trimester [4, 5] and in one case at 3rd trimester scan (31 weeks of gestation) [6]. The most known embryological theory was described by Grümwald in 1910 and is called the "prenasal space" theory. This theory is very attractive because of the embryopathogenic continuum proposed among dermoids, gliomas, and encephaloceles. However, while this theory can explain this continuum, it cannot explain the occurrence of craniofacial dermoids of the same topography. The differential diagnosis includes "prenasal space" developmental impairment, which are nasoethmoidal meningoencephaloceles, nasal dermoids, and epidermoid cysts [7]. Differential diagnosis is a critical issue because it is essential in guiding the prenatal counseling, planning the prenatal and/or postnatal management and explaining the related clinical prognosis. Typical location, Doppler findings, absent or presence of associated anomalies may aid the prenatal diagnosis although patients must be informed of the possible alternative diagnosis such as dacryocystocele, dermoid cyst, retinoblastoma or teratoma, haemangioma, and encephalocele that cannot completely be excluded prenatally [4, 8, 9].

Chmait et al. [6] referred in their case reporting on a nasal glioma diagnosed using 3D ultrasound that, although excellent visualization of the facial mass was obtained, the precise diagnosis remained uncertain until after delivery and further postnatal evaluation. Chmait et al. [6] confirmed the value of 3D ultrasound in the evaluation of such masses but underscores the difficulty in achieving a precise diagnosis prenatally and parents should be informed about uncertainties of the prenatal diagnosis [4] and of differential diagnosis. With exclusion of nasal glioma, the commonest forms of congenital midline masses are dermoid cysts, encephaloceles, and congenital haemangiomas (CHs) [10]. Dacryocistoceles, the commonest lesions of this group, are smooth-walled, hypoechoic cyst of the lacrimal duct that abut the orbits medially and be sometime bilaterally [11, 12]. Dermoid cyst usually occurs on the tip of the nose [12, 13] and can be thus easily excluded compared to liposarcoma and rhabdoid tumor that cannot be completely excluded prenatally [14]. Retinoblastoma may appear as a heterogeneous mass that arises directly from the eye and has an irregular echogenic structure surrounded by a sonolucent area and are covered by a membrane [9]. Congenital haemangiomas are heterogeneous mass that protrudes from and typically forms an obtuse angle to the skull that exhibit significant blood flow within the lesion at color Doppler analysis [15]. Within CHs, two clinical courses have been identified: rapidly involuting congenital hemangiomas

(RICH) and noninvoluting congenital hemangiomas (NICH) and only RICH has been reported prenatally [16]. Nasal teratoma usually has an outward direction that involved the tip of the nose without extension into the nose, nasal passages, or nasopharynx [13]. Nasal glioma must also be differentiated with frontal encephaloceles that usually involve the space directly above the nose [13] and antenatal MR imaging may be of value in such cases by ruling out underlying bony defects [4, 5].

In case of nasal glioma, T1-weighted imaging demonstrates the mass to be isointense to grey matter with moderate enhancement after contrast-enhanced T1-w image [5]. Similar MRI findings can be seen in case of CHs, with hypointense lesion on T2-weighted imaging and low resistance arterial flow on Doppler imaging [17]. Nasal glioma mimicking encephalocele may be also a challenging diagnosis in infancy due to its clinical presentation and endoscopic approach after failure to excise it through a craniotomy has been recently reported [18]. Endoscopic surgery is thus considered appropriate for the removal of intranasal glioma without intracranial extension [19] and radical excision is essential as nasal glioma or nasal glial heterotopia may recur following incomplete surgical removal [20]. Grzegorczyk et al. [5] in a very recent report suggests that pre- and postnatal MRI should routinely be performed to exclude intracranial connections and/or abnormalities and plan the surgical approach, when radiologic resources and competence may be available. Postnatal MRI and/or CT (computed tomography) are of particular interest in evaluating the intranasal extension of nasal glioma in order to tailor surgical treatment and reduce the risk of incomplete resection [4]. Huisman et al. [21] studying 11 patients (mean age of 4.5 years) with developmental nasal midline masses (nine dermoid sinus cysts, one meningocele, and one nasal glioma) demonstrated that intracranial extension was equally detected by CT and MRI using indirect imaging signs. When direct signs were evaluated, MRI suspected intracranial components in two patients that were undetected on CT. Moreover, MRI and CT matched the surgical findings in 9 out of 11 patients.

The conclusions were that neuroimaging is essential to characterize these lesions and the lack of radiation in a region of interest usually centered on the radiosensitive eye lenses favor the use of MRI. Furthermore, MRI did not show any false-negative results [21].

Form analysis of the medical literature and that of the reported cases, few clinical considerations can be drawn: (a) the prenatal diagnosis of nasal glioma can be made by ultrasound; (b) although nasal glioma is a benign fetal tumor, it has the potential for intracranial extension; (c) Doppler ultrasound may aids the prenatal sonographic diagnosis and be useful in differentiating nasal glioma from haemangioma; (d) MRI enhances the diagnostic accuracy either by indirect or direct imaging signs of intracranially located lesions and can exclude, better than prenatal ultrasound, an anterior meningoencephalocele and/or other associated cerebral abnormalities; (e) MRI and CT are essential to delineate the exact location, limits, and extension of the lesions when tailoring surgical approach; (f) radical surgical excision is recommended to prevent relapse of heterotopic neuroglial tissue while endoscopic approach is appropriate in case of intranasal extension of the tumor; and (g) good prognosis is expected when the prenatal and postnatal presumptive diagnosis are confirmed following histologic examination.

Conflict of interest None.

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