Prenatal diagnosis of fetal craniofacial teratoma: US, MRI findings, a rare case report

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ABSTRACT

Background: Craniofacial teratoma is a rare congenital malformation. We present a case of an intracranial expanding orofacial teratoma, where pathological examination revealed the findings of remarkable ultrasonographic signs and magnetic resonance imaging (MRI).

Case Presentation: In the 23-week ultrasound examination, a solid cystic mass lesion was revealed in the craniofacial region. MRI showed that the mass had an intracranial extension and spread to the fetal face, and also revealed a marked nasopharyngeal obstruction by the mass. There was a growing tumor protruding from the fetus's mouth. Slight polyhydramnios was also detected.

Conclusion: In detecting prenatal anomaly, although the ultrasound is the first scan method applied, MRI in prenatal diagnosis is very useful for evaluating the extent, nasopharyngeal obstruction degree, and extension into the intracranial region of craniofacial teratoma.

Keywords: Prenatal ultrasound, prenatal MRI, fetal teratoma, fetal abnormality, polyhydramnios.

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Background

Congenital tumors are very rare and teratomas are the most common congenital tumors [1]. The most common benign tumor in newborns is teratoma; however, it occurs in less than 1/250,000 in the craniofacial region. Teratomas are usually in the sacrococcygeal region and in order of frequency, they can be observed in gonads, anterior mediastinum, and retroperitoneum [1,2].

Intracranial and extracranial extensions of craniofacial teratomas are rarely seen. The cranial congenital teratomas are often fatal due to the mass effect, intracranial involvement, airway obstruction, and respiratory failure, and they have rapid growth potential [3]. The teratomas are usually benign and a malignant transformation is rare [1,4].

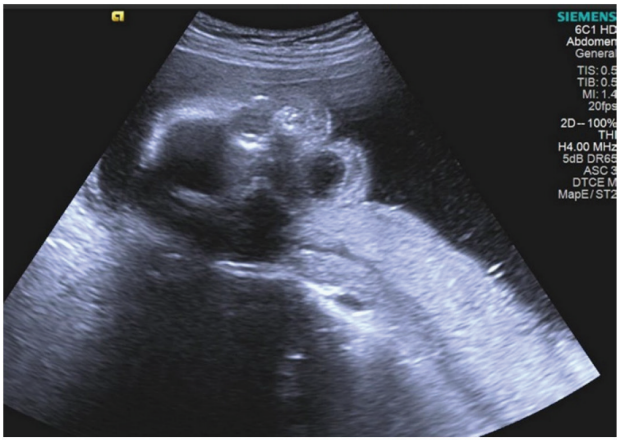
We aim to report a case of teratoma with an extension into the nasopharyngeal, facial, and intracranial regions, causing airway obstruction, which was detected prenatally via sonography (USG) and magnetic resonance imaging (MRI).

Case Presentation

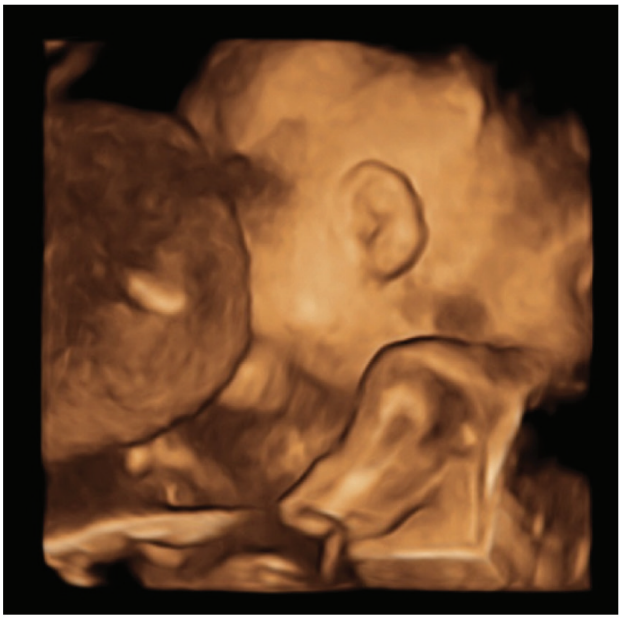
A healthy 25-year-old primigravida was referred to the fetal radiology unit of our hospital for the first prenatal obstetric examination at 23 weeks of gestation. The patient lived in the countryside, and a first trimester examination could not be obtained. The sonographic examination was carried out using the ACUSON S2000 USG system scanner equipped with a curved array transducer (Siemens Medical Solutions, Mountain View, CA). USG evaluation of the fetus revealed a complex mass in the midsagittal line originating from the mouth, nose, and interorbital space measuring 45 × 50 mm. The mass lesion contained solid parts and a cyst formation (Figure 1). The amniotic fluid amount had increased slightly; however, the rest of the examination was normal. Although teratoma was considered in the differential diagnosis, neuroblastoma and giant hemangioma were also assumed in one of our minds.

Considering the presence of polyhydramnios, fetal MRI examination was proposed to reveal the mass nasopharynx and oropharynx relationship. Sagittal T2-weighted MRIs revealed that a large, heterogeneous mass with cystic components was centered in the soft tissues of the midfacial line, extending outwardly over the inter-globe and nose, aggressively filling the oropharynx and nasal cavity (Figure 2). Also, there was intracranial extension of the mass toward the frontal lobe anterior.

Based on these findings, postpartum respiratory distress seemed inevitable and the presumptive diagnosis was a teratoma. Pregnancy was terminated with a normal delivery in the 25th week at the request of the family.



(a)



(b)

Figure 1. *Coronal USG images. (a) A solid cystic component mass lesion extending to the interorbital distance is observed by filling the lower half of the face. (b) Sagittal 3d USG image showing mass lesion appearance on the face.*

The presence of bone tissue, cartilage tissue, and mature squamous epithelium was revealed in the microscopic examination of the tumor (Figure 3a,b). Cellular atypia or significant mitotic activity was not noted. These findings are consistent with those of mature craniofacial teratoma.

Discussion

Teratomas are genuine congenital neoplasms consisting of tissues originating from three germinal layers and derived from totipotential/(pluripotent) cells. Congenital intracranial teratoma has been described as three forms: (1) massive intracranial teratoma; (2) smaller, more localized and less extensive, intracranial teratoma; (3) intracranial teratoma with extension into the extracranial structures, a solid version extending through the cranial base into the

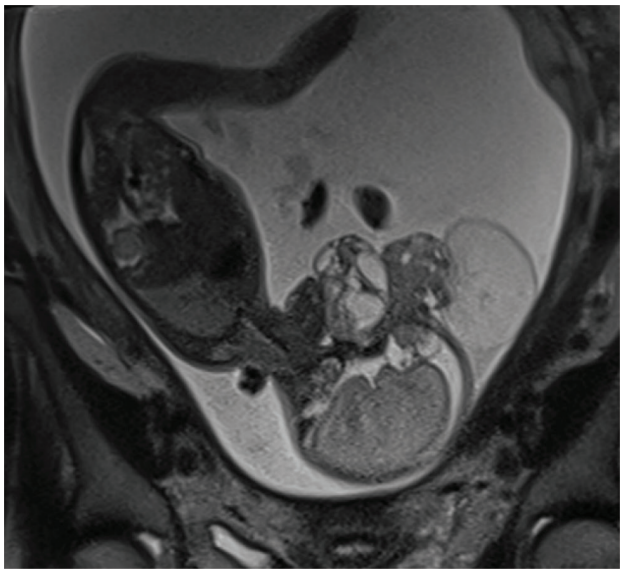
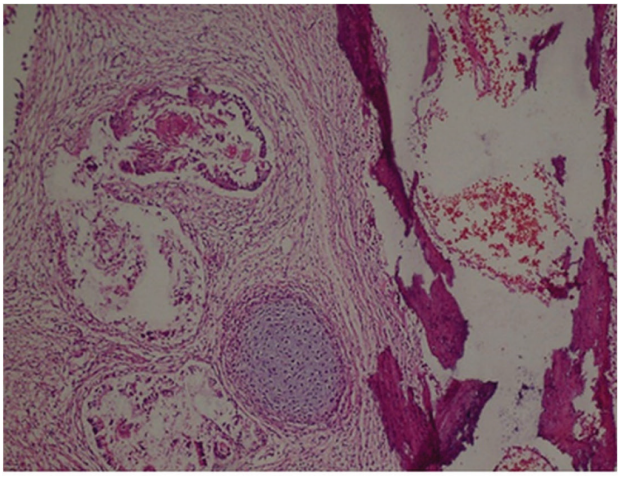


Figure 2. *Sagittal T2-weighted MRI. An outward protrusion of the mass lesion by obliterating the mouth and nasal cavity is observed. Also, this sequence revealed the postnatal prognosis of the mass with the brain extension and nasopharynx relationship.*

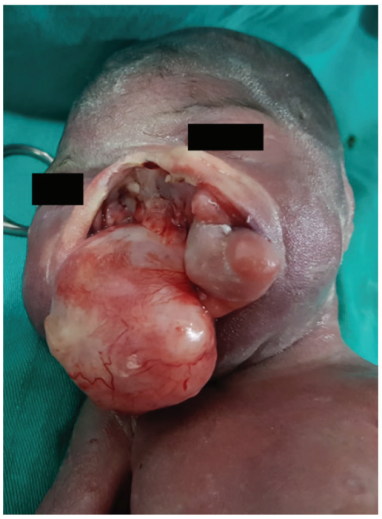
face and neck region (also known as epignatus) [5-7]. Our case is also an oropharyngeal teratoma with an extracranial component on the scalp and extension to the brain.

Teratomas of the face and neck may frequently present as great masses containing both cystic and solid elements and arise from the thyrocervical area, palate, or nasopharynx [8]. The oropharyngeal teratoma is also called epignathus and can be easily detected by the prenatal USG. The term "epignathus tumor" refers to a teratoma of the oropharyngeal cavity in the fetus whose origin is nonspecific [9]. At fetal teratomas' diagnosis, prenatal USG is the cornerstone. The USG reveals that large tumors can distort the normal appearance of the face, lips, nostrils, and nose [10,11]. USG diagnosis has proven to be successful in the very early stages of pregnancy, even at 15-17 weeks; however, the vast majority of lesions are diagnosed in the late second and third trimesters [10,11]. It is generally seen as a large solid and cystic mass extending toward the anterolateral direction of the fetal face and neck. The USG is noninvasive and harmless for anomaly screening during pregnancy. In prenatal USG applications, teratoma can be detected as a complex intracranial or extracranial mass with calcification. Only 50% of the cases' calcifications may show but this finding may not be obvious in the USG [12]. In our case, craniofacial teratoma did not include calcification.

The mass often has an intracranial extension, as in our case, and in such cases, the prognosis is not good [9]. The characteristic view of the teratoma is a giant, heterogeneous mass with cystic components in the T1- and T2-weighted images, with no obvious distinction between mature and immature teratomas [3]. In our case, especially the sagittal T2-weighted MRI demonstrated the intracranial extension of the mass and nasopharyngeal



(a)



(b)

Figure 3. *(a) Histopathological examination. Bone tissue, cartilage tissue, and glandular structures are seen in the section (hematoxylin and eosin ×100). (b) A mass lesion that protrudes outward, filling the mouth and nose and widening the interorbital space, in the photograph of the stillborn neonate.*

obstruction very well. MRI is a very useful imaging technique for describing the extent of such tumors in facial and cranial soft tissues. Fetal MRI can be particularly useful in determining whether a controlled delivery such as *ex utero* intrapartum treatment (EXIT) procedure is necessary [3,4]. The treatment of neck masses that cause significant airway obstruction, such as cervical or intrathoracic teratoma, usually involves the EXIT procedure [3,4,13]. In the EXIT procedure, the fetus is partially born by cesarean section and is subjected to intubation or surgical procedures, while the placenta and umbilical cord remain intact [13].

One-third of these tumors are associated with polyhydramnios due to difficulty in swallowing amniotic fluid. In our case, polyhydramnios had started to develop moderately [9]. Hydrocephalus and intracranial invasion, polyhydramnios, and hydrops, as well as along with tumor size >5 cm, are a sign of poor prognosis for the fetus [14]. Histopathologically, teratomas are classified as mature or immature [9]. This distinction is based on the differentiation of tumor cells [7]. Mature teratomas contain a variety of adult tissue types, while immature teratomas contain completely undifferentiated parts that are similar to fetal tissues. Both entities generally contain tissues from all three germ layers, such as skeletal muscle, cartilage, bone, bronchial epithelium, gut epithelium, and neural tissue. Our case has been included under the category of congenital teratoma with mature bone, cartilage, skin, and glandular elements.

Conclusion

Craniofacial teratoma is an uncommon status that must be handled by a specialized multidisciplinary team. Radiological imaging methods are a very accurate route for this team. This case demonstrates that radiological imaging methods, such as the USG and especially MRI, are important guides that will provide the fetus with the correct approach. Prenatal diagnosis has made better the perinatal management of these lesions.

List of Abbreviation

EXIT	Ex utero intrapartum treatment
MRI	Magnetic resonance imaging
USG	Ultrasound

Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this article.

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Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

Consent for publication

Written consent was obtained from the patient.

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Summary of the case

1	**Patient (gender, age)**	25-year-old Female
2	**Final diagnosis**	Kraniyofascial teratoma
3	**Symptoms**	Abnormal ultrasound findings at pregnancy
4	**Medications**	N/A
5	**Clinical procedure**	N/A
6	**Specialty**	Radiology