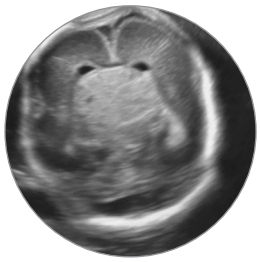

MRI in the Diagnosis of Fetal Intracranial Teratoma

Resonancia magnética en el diagnóstico del teratoma intracraneal fetal

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Summary

Fetal intracranial tumors are rare but, when they do occur they are associated with high fetal mortality. Many of the congenital brain tumors often result in intrauterine fetal death, limiting accurate assessment of tumor prevalence and incidence. The most frequent is teratoma, which is characterized by cells dependent on the three germ layers. These tumors are usually detected in the usual prenatal control ultrasounds, and are often so large that it is difficult to determine their origin. When ultrasound suspicion exists, a fetal MRI study is recommended. The case of a 20-week pregnant woman is presented, in which a fetal intracranial tumor was detected on a prenatal control ultrasound. It was decided to complete the study with a fetal MRI in which a pineal gland mass with anterior extension, compatible with teratoma, was visualized. Voluntary termination of pregnancy was decided, and the histological study after necropsy confirmed that it was an immature teratoma. MRI is the imaging test of choice after suspected diagnostic ultrasound, as its high resolution provides relevant information that allows better assessment of the internal architecture of the tumor, as well as its origin and extension. In addition, it also serves to evaluate the rest of the intracranial structures.

Resumen

Los tumores intracraneales fetales son poco comunes, pero cuando aparecen están asociados con una elevada mortalidad. Muchos de los tumores cerebrales congénitos a menudo provocan la muerte fetal intrauterina, lo que limita la evaluación precisa de su prevalencia e incidencia del tumor. El más frecuente es el teratoma, que se caracteriza por estar constituido por células dependientes de las tres capas germinales. Estos tumores suelen detectarse en las ecografías habituales de control prenatal y, frecuentemente, son tan grandes que se dificulta determinar su origen. Por ello, ante la sospecha ecográfica, se recomienda completar el estudio con una RM fetal. Se presenta un caso de una gestante de 20 semanas, en quien se detecta un tumor intracraneal fetal en una ecografía de control prenatal. Se decide completar el estudio con una RM fetal en la que se visualiza una masa dependiente de la glándula pineal con extensión anterior, compatible con teratoma. Se decidió la interrupción voluntaria del embarazo, y en el estudio histológico tras la necropsia se confirmó un teratoma inmaduro. La RM es la prueba de imagen de elección tras la sospecha diagnóstica ecográfica. Por su alta resolución aporta información relevante que permite valorar mejor la arquitectura interna del tumor, así como su origen y extensión. Además, sirve para evaluar el resto de estructuras intracraneales.

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Introduction

Congenital intracranial tumors are a rare entity, with a frequency of approximately 0.5-1.5% (1) of all pediatric brain tumors. Overall, they constitute only 10% of all prenatal tumors. The prognosis is generally poor, but depends on histology, location and size, and can cause cardiovascular involvement.

Fetal tumors have a different prevalence, location, histology and biological behavior than pediatric tumors. Early detection of prenatal neoplasia is crucial for fetal, maternal and neonatal care.

The most common tumor in fetuses is teratoma –which accounts for approximately half of all cases– followed by astrocytoma, lipoma, choroidal plexus papilloma, craniopharyngioma and embryonal tumors– among which the formerly called PNET have been included in the latest WHO classification of tumors. Most of them are diagnosed during prenatal ultrasound controls. In the case of suspected ultrasound, the test indicated is fetal magnetic resonance imaging (MRI), since it allows better characterization thanks to its high resolution. Sometimes, its large size makes it difficult to determine its origin (2).

The case of a 20-week pregnant woman is described, in whom a fetal brain tumor was detected in a routine

ultrasound, with MRI as a complementary study, whose findings were compatible with pineal teratoma. It was decided to legally terminate the pregnancy and perform a necropsy, which confirmed the diagnosis.

The objective of this work is to highlight the importance of fetal MRI for prenatal diagnosis of intracranial tumors, in order to characterize the lesion and to specify its location and extension.

Case presentation

35-year-old woman, pregnant for 20 weeks, no history of interest. A screening ultrasound was performed in the 20th week of pregnancy, in which a predominantly hyperechogenic, heterogeneous fetal intracranial mass was observed, with origin in the midline, extending into the thalamic region and third ventricle, with impression on the body and splenius of the corpus callosum and lateral ventricles, without hydrocephalus. In the study with Doppler it presented moderate vascularization. No other fetal abnormalities or polyhydramnios were observed (figure 1).

It was decided to complete the study with a fetal MRI performed at 20 weeks and 5 days, in which a solid mass with small cysts inside was identified, located in the pineal gland and extending in an anterior direction towards the third ventricle, both thalamus, columns of the fornix, splenius and corpus callosum. The lesion extends from the middle line, affecting both cerebral hemispheres. It pushes the medial wall of the lateral ventricles and the posterior margin of the cavum septum pellucidum. Caudally occupied partially the quadrigeminal cistern, in contact with the upper quadrigeminal tubers. Its signal was intermediate with respect to the cerebral parenchyma in T1 and T2 potentiated sequences, and high in diffusion sequences, with low signal in the ADC map, which indicates high cellularity of the lesion. No blood content was visualized in its interior, cerebral hematoma was ruled out (figure 2). These findings were compatible with pineal teratoma, confirmed by histological study.

Discussion

Congenital intracranial tumors are rare (3) and their physiopathology is different from other pediatric neoplasms. Normal embryological cells have a high mitotic potential, so it is believed that the origin of congenital tumors is due to an abnormal differentiation and maturation of these cells. Therefore, these lesions, whether benign or malignant, grow rapidly (2). In fetuses they usually appear at the supratentorial level, while in the rest of the pediatric population their origin is infratentorial (2, 4).

Intracranial teratoma is the most common fetal brain tumor, approximately half of the cases, followed by astrocytomas of different grades, lipomas, choroidal papillomas, craniopharyngiomas and embryonal tumors (2).

The most common ultrasound findings associated with brain tumor pathology are usually: polyhydramnios, produced by the alteration of hypothalamic function and fetal swallowing; hydrocephalus, by increased CSF production, as in choroid plexus papillomas, or by the mass effect of the tumor obstructing the ventricular system and macrocephalus, due to the tumor itself or secondary to hydrocephalus (2, 3).

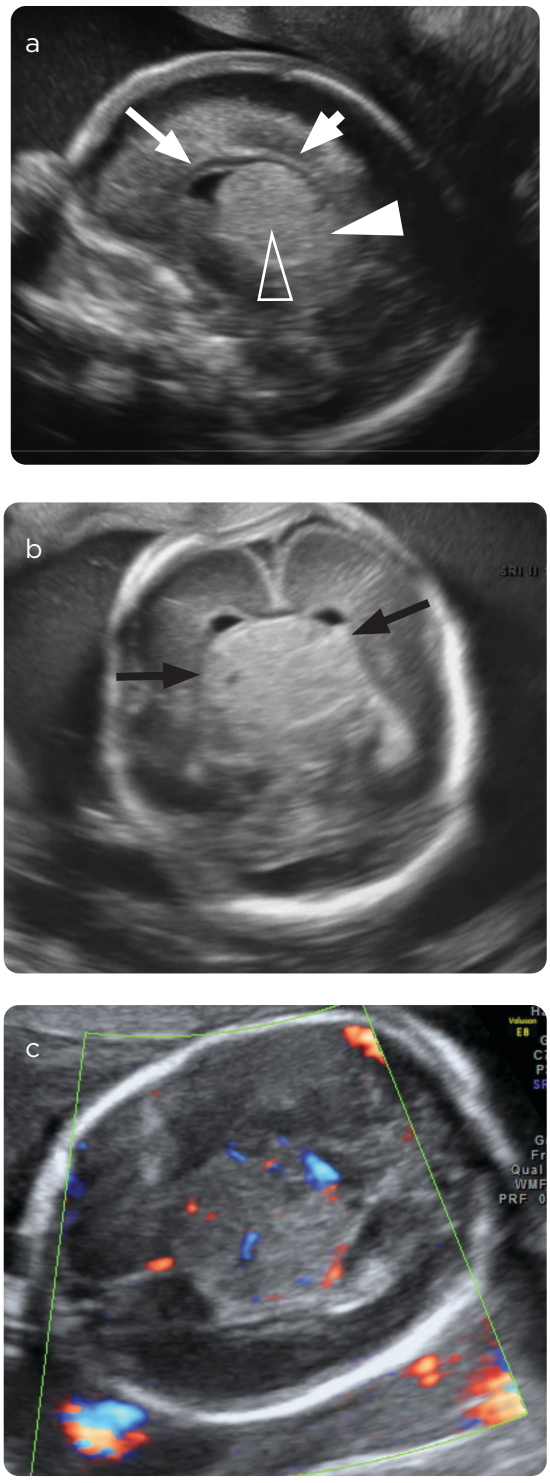

Figure 1. Immature teratoma of the pineal gland. Gestational age 20 weeks and 5 days. a) Neurosonography with mid-sagittal plane. Mass that originates in pineal region (solid white arrowhead) and extends in anterior direction to the third ventricle and thalamic region (empty white arrowhead), splenius and corpus callosum trunk (solid short white arrow) and reaches the posterior margin of cavum septum pellucidum (solid long white arrow). In caudal direction it reaches the quadrigeminal plate (empty long white arrowhead). b) Neurosonography with coronal plane. The lesion crosses the midline and affects both cerebral hemispheres with an impression on the medial wall of the lateral ventricles (black arrows). c) Eco-Doppler Neurosonography in axial plane. The mass presents moderate vascularization.

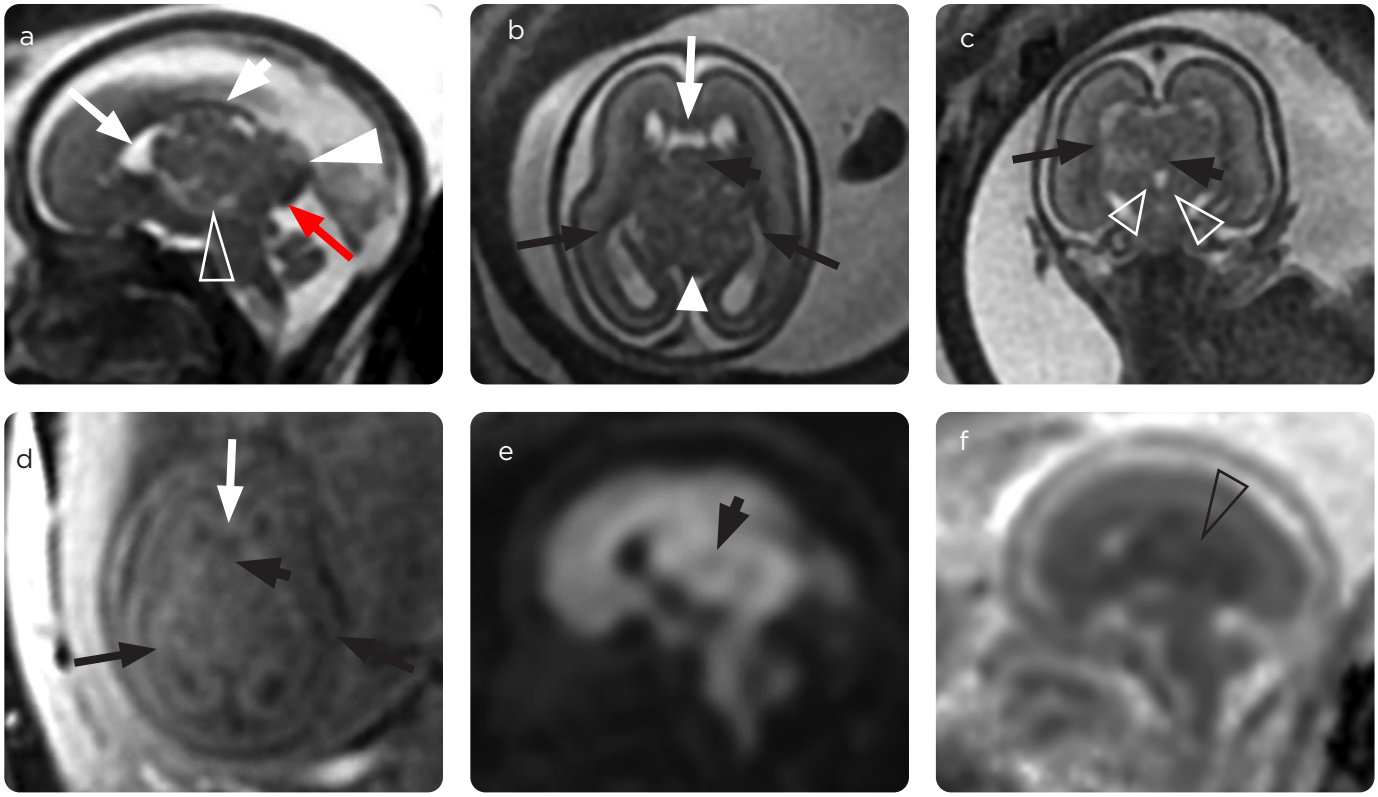

Figure 2. Teratoma of the pineal gland. Gestational age 20 weeks and 5 days. a) Sagittal SS FSE T2. b) Axial SSFSE T2. c) Coronal SS FSE T2. d) Axial 3D gradient T1 (LAVA). Mass that originates in pineal region (solid white arrowhead) and extends in previous direction to the third ventricle and thalamic region (empty white arrowhead), splenius and trunk of the corpus callosum (solid short white arrow) and reaches the posterior margin of the cavum septum pellucidum (solid long white arrow). In flow direction it reaches the upper quadrigeminal tubers (empty long white arrowhead). Intermediate signal in T1 and T2 sequences with respect to the parenchyma and crosses the midline (short black solid arrows) affects both cerebral hemispheres and produces impression on the medial wall of the lateral ventricles (long black solid arrows). e) Sagittal diffusion image. f) Sagittal diffusion sequence map The lesion is high signal in the diffusion image (solid black arrowhead) and low signal in the ADC map (empty black arrowhead) with low ADC values ranging from 1.05-1.2 ×10-3 mm2/s.

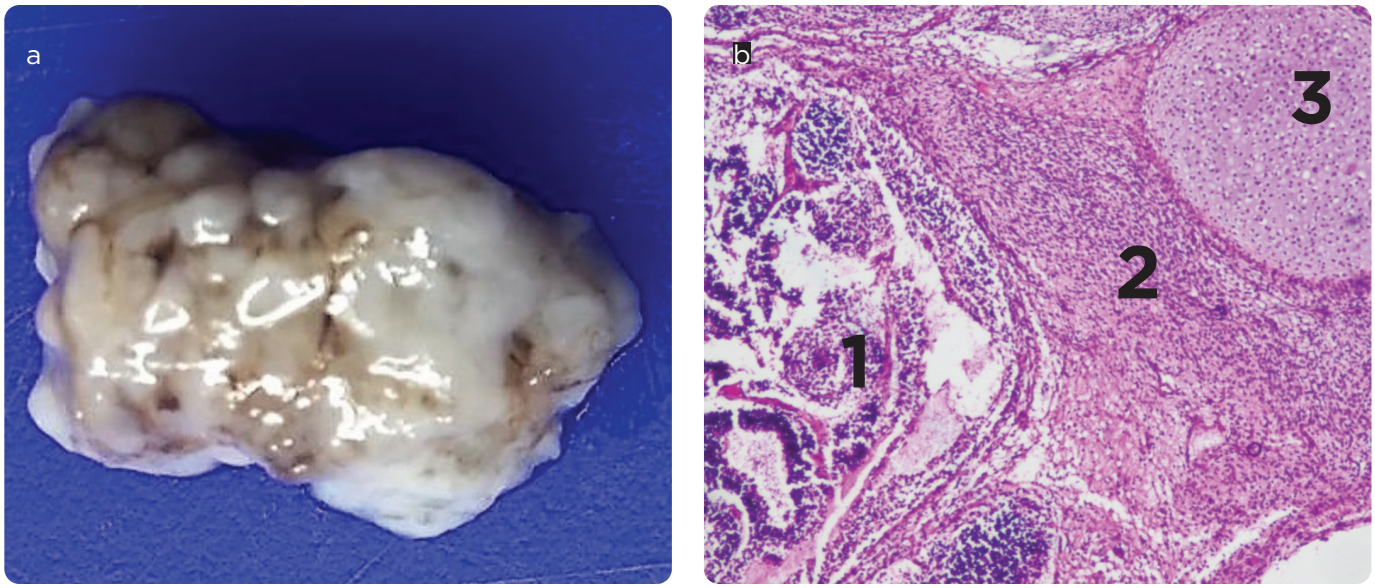

Figure 3. Immature teratoma. a) Necropsy, macroscopic part. Solid congestive multinodular brain tumor in the midline, 26×10 mm maximum diameter, well defined and easily enucleated. The solid tumor has a consistency similar to the cerebral parenchyma with small cystic areas. b) Histological study. 1: Neuronal tissue with predominance of immature forms from the ectodermal leaf forming neuroblastic rosettes, 2: with stromal tissue and 3: small islets of immature cartilage.

Teratomas represent the abnormal development of pluripotent cells of the three germ lines and immature neuroglial elements. They are usually located in the pineal region, cerebral hemispheres, hypothalamic area, suprasellar region and third ventricle (2, 4).

The exact etiology of teratoma is unknown. It is believed to be due to a failure of the primary germ cells to migrate. Histologically, they can be mature, immature or malignant, rare in fetuses (5). Mature teratomas contain multiple varieties of adult tissues, while immature teratomas contain fetal tissues. However, in both cases they contain tissues from all three germ layers: ectoderm, mesoderm, and endoderm —including muscle tissue, cartilage, bone, bronchial epithelium, intestinal epithelium, and neural tissue.

Fetal intracranial tumors are usually detected in prenatal ultrasounds, especially in the third trimester, around 60% after 30 weeks (2,4). In contrast, in this case the diagnosis was early —in the 20th week. They can exhibit rapid growth and be very large at diagnosis, since embryonic cells present a high mitotic potential.

Ultrasonically, they generally appear as complex solid-cystic masses, with or without calcification foci, and vascularization is observed in the Doppler study (3, 4, 6).

Generally, in the case of suspected ultrasound, the study is completed with MRI, which is the test of choice for the evaluation of fetal intracranial tumors and confirmation of the diagnosis, since it provides information on the internal architecture thanks to its spatial resolution, multiplanar capacity and tissue characterization. It also allows the identification of the site of origin, as well as the rest of the intracranial structures (4, 7-9).

The typical radiological findings of teratoma are mixed masses generally of great size, with solid-cystic component, with or without calcification focuses (3). In T1 sequence, they have a variable signal: high, if it presents fat or cystic component with protein content; intermediate, due to the solid component; low, due to calcium foci. In T2 sequence, a heterogeneous behavior is also observed, and in postnatal cases, where intravenous contrast medium can be administered, enhancement of the solid component (2, 10).

The main differential diagnosis is intracranial hematoma (1, 2, 5), which is ruled out because flow was demonstrated in the Doppler study –absent in hematoma– and in the MRI no high signal areas were observed in T1-weighted sequences suggesting bleeding inside. The differential diagnosis is also made with other congenital intracranial tumors (2, 5) that, in general, were ruled out because of the typical location of the teratoma in the pineal gland and because they are less frequent.

Astrocytoma normally presents as a solid-cystic mass with calcification foci originating in the thalamus or cerebral hemispheres (8), while craniopharyngiomas develop from squamous cell debris from the Rathke pocket, thus appearing as a heterogeneous suprasellar mass, frequently with calcifications (11). Their behavior is similar to that of teratomas, but they have different locations.

Choroid plexus papillomas originate in the lateral ventricles, third or fourth ventricle, have lobed morphology and are homogeneous, medium or high signal in T1 and low signal in T2. They usually originate uni or bilateral ventriculomegaly and usually present with hydrocephalus due to the production of CSF and an alteration in the reabsorption of the same (2, 11, 12).

Embryonic tumors are rare, very heterogeneous, of supratentorial location, normally in the cerebral hemispheres or within the lateral ventricles; in addition, they can present hemorrhage and the solid component is of intermediate signal in T1 and T2 to the cortical gray substance (2, 13). Therefore, due to the typical location in the midline and its behavior in the different sequences, the findings suggested the diagnosis of pineal teratoma. However, the definitive diagnosis was confirmed by histological study after necropsy.

In general, the prognosis of intracranial tumors is negative, with a high percentage of death in the intrauterine or neonatal period. When diagnosed before 30 weeks of gestation, the prognosis is particularly severe, with a mortality rate close to 96% (2, 7) and, therefore, voluntary interruption of pregnancy can be considered (1, 6, 14).

Teratomas represent the abnormal development of cells In the case described here, the parents decided to terminate the pregnancy when they were informed of the severity of the brain pathology. The necropsy confirmed a solid, multinodular midline tumor with well-defined cystic areas inside. Histologically, fetal congenital teratomas typically contain abundant immature tissue (15), as in this case, the brain tumor had neuronal tissue with predominance of immature forms from the ectodermal leaf and small islets of cartilage. These findings confirm that it was an immature teratoma dependent on the pineal gland (Figure 3).

Conclusion

Fetal brain tumors are rare, but are associated with high fetal and neonatal mortality (1). The most frequently detected in prenatal ultrasound are intracranial teratomas. In view of the suspicion, it is advisable to extend the study to fetal MRI, which provides valuable information about the tumor itself, its extension and origin (4). Although the definitive diagnosis is histological, the MRI is a useful test to establish a prognosis and fetal management (1-3, 5, 7, 8).

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