Article published online: 2024-08-16

Fetal Ganglioneuroblastoma: A Rare Entity with Antenatal Diagnosis and Postnatal Follow Up

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J Fetal Med 2024;11:185–187.

Abstract

Fetal abdominal masses can be challenging to counsel due to uncertain diagnosis and outcomes. We report a case of a midline upper retroperitoneal mass found incidentally during a routine third trimester ultrasound. Despite not being a suprarenal mass, ultrasound and magnetic resonance imaging indicated neuroblastoma prenatally. With the generally favorable prognosis for prenatally diagnosed neuroblastomas, expectant counseling was given. Postnatal imaging and biopsy confirmed ganglioneuroblastoma, a rare subtype. A conservative approach was taken, and after initial growth, the mass spontaneously regressed 5 months after birth, nearly resolving by 8 months.

Keywords
- antenatal
- fetal
- ganglioneuroblastoma
- neuroblastoma
- tumor

Introduction

Neuroblastoma, ganglioneuroblastoma, and ganglioneuroma form a spectrum of neuroblastic tumors originating from the neural crest derived progenitor cells or neuroblasts, the precursor for ganglion cells in the sympathetic nervous system.[1,2] These tumors exhibit diverse outcomes, ranging from spontaneous regression to fatal disease.[3,4]

Neuroblastomas, though common in the neonatal period, are rarely diagnosed prenatally. Sixty to 90% of fetal neuroblastomas are situated in the adrenal glands; hence, the presence of a fetal suprarenal solid or cystic mass is a strong indicator of neuroblastoma.[5]

Case Report

A 30 year old primigravida with no history of significant medical issues or hypertension underwent a routine growth scan at 33 weeks of pregnancy. Her first trimester nuchal translucency scan and the anomaly scan at 20 weeks showed normal results.

Ultrasound examination showed a well margined hypoechoic lesion in the midline retroperitoneum, displacing the inferior vena cava to the right. On grayscale imaging, the lesion appeared solid, without any apparent cystic component or large calcifications, although few faint internal calcifications were detected within the mass (►**Fig. 1A, B**). Doppler imaging did not reveal significant internal vascularization or feeder vessels from the descending aorta (►**Fig. 1C**). A preliminary diagnosis of a fetal abdominal tumor was made, with neuroblastoma and extralobar pulmonary sequestration in the list of potential differential diagnoses.

Further assessment with fetal magnetic resonance imaging (MRI) confirmed a well defined T2 intermediate signal intensity lesion in the midline retroperitoneum at the level of the kidneys (►**Fig. 2A, B**). It displaced the aorta posteriorly and the inferior vena cava to the right. No intraspinal extension was observed. The calcific foci in the lesion and the absence of systemic feeder vessels favored fetal neuroblastic tumor over extralobar pulmonary sequestration.

article published online
August 16, 2024

DOI https://doi.org/10.1055/s-0044-1788925.
ISSN 2348-1153.

Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

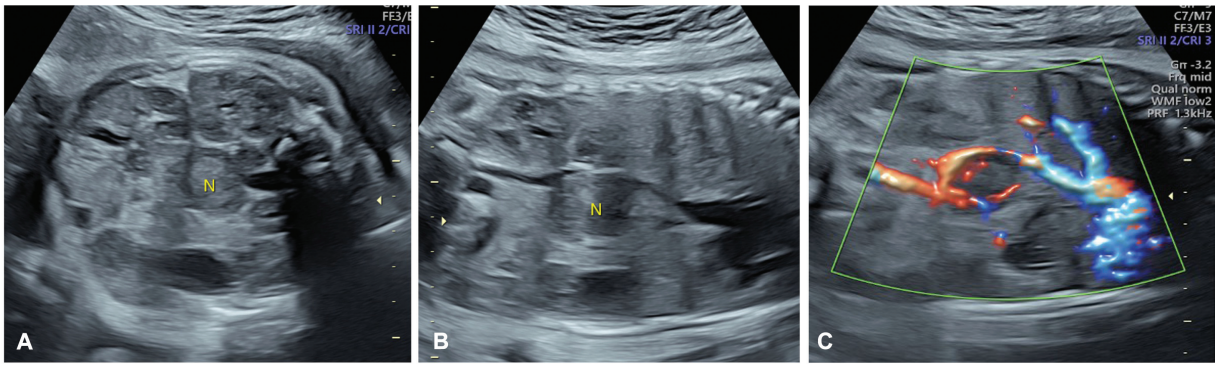

Fig. 1 (**A**, **B**) Ultrasound of fetal abdomen in axial and coronal planes, identifying a hypoechoic mass in the midline in retroperitoneum (N) at 33 weeks of gestation. (**C**) Color Doppler image shows no obvious blood flow within the lesion or any feeder vessel from the aorta.

A cesarean section was performed at 38 weeks for an obstetric indication, delivering a male child. The neonate exhibited no abdominal distension, organomegaly, additional swellings, or feeding difficulties. At 1 month, an ultrasound revealed a heteroechoic mass in the right paramedian retroperitoneum, larger compared to the antenatally detected lesion (►**Fig. 3A**). A trucut biopsy from the mass showed two morphological components; neuroblasts with monomorphic round cells with uniform nuclei and salt pepper chromatin, as well as ganglion cells with abundant granular eosinophilic cytoplasm and distinct cell boundaries. On immunohistochemistry, the neuroblasts were positive for synaptophysin and chromogranin while the ganglion cells were positive for S100 and glial fibrillary acidic protein. This confirmed the diagnosis of ganglioneuroblastoma. Considering the potential for spontaneous regression of antenatally detected neuroblastic tumors, a conservative approach without surgery or chemotherapy was opted for.

At 5 months, follow up ultrasound showed a decrease in the mass size accompanied by normalized urinary vanillylmandelic acid and homovanillic acid levels (►**Fig. 3B**). By 8 months, the retroperitoneal mass was barely discernible on ultrasound, and the infant remained asymptomatic and active during the follow up period.

Diagnostic Parameters

Fetal tumors are rare and present challenges due to various possible diagnoses and unpredictable course during pregnancy and infancy. Prognosis depends on factors like tumor type, location, and size, with most having poor outcomes. However, some exceptions like fetal neuroblastic tumors have a better prognosis.[3] Neuroblastoma, the most common childhood extracranial solid tumor, can also be detected incidentally in late pregnancy. Typically, they present as a well defined solid suprarenal mass on ultrasound, though some may be cystic. Microcalcifications and intratumoral hemorrhage are also seen.[6]

MRI can be a complementary tool to help confirm the anatomic location and to exclude adrenal hemorrhage. It is also useful for staging and evaluating metastases, which most frequently involve the liver and the placenta.[7]

Differential diagnoses include subdiaphragmatic extralobar pulmonary sequestration, teratoma, adrenal hemorrhage, and renal and pancreatic tumors.[1,5,8]

Subdiaphragmatic extralobar pulmonary sequestration is more common than neuroblastoma, typically appearing echogenic, left sided, and identifiable in the second trimester. In contrast, neuroblastic tumors can be either solid or

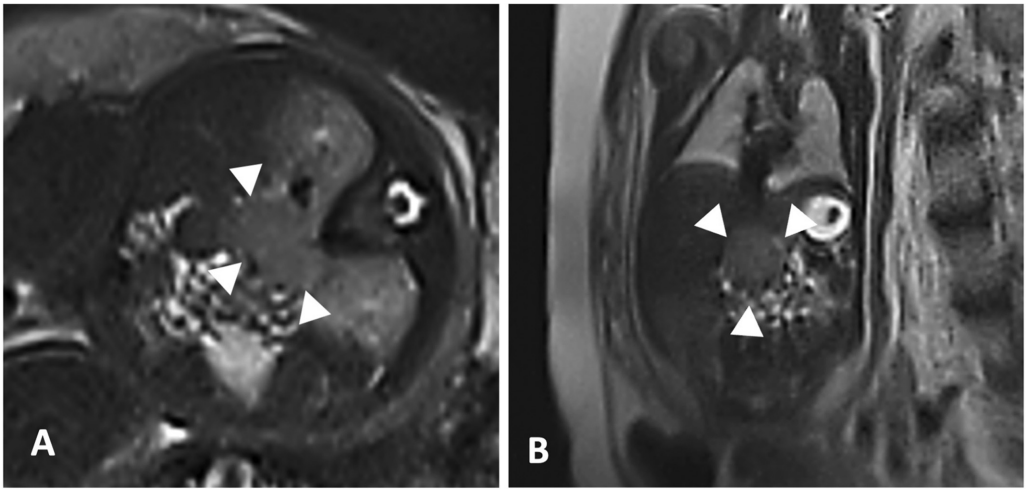

Fig. 2 (**A**, **B**) Fetal magnetic resonance imaging (MRI) T2-weighted (axial and coronal views), showing an intermediate signal intensity lesion (arrowheads) in the retroperitoneum at the level of the fetal kidneys.

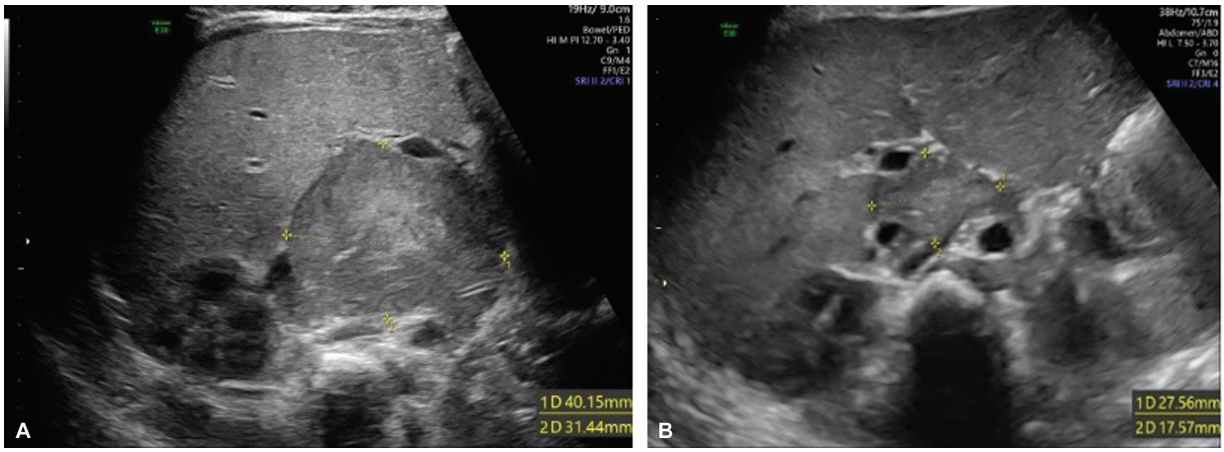

Fig. 3 (**A**) Postnatal ultrasound at 1 month, showing a larger heteroechoic mass in the right paramedian retroperitoneum. (**B**) Follow up postnatal ultrasound at 5 months, showing a decrease in the size of mass.

cystic, often appearing on the right side, and are typically identified in the third trimester. Neuroblastic tumors may show calcifications, unlike pulmonary sequestration. Doppler flow studies may help distinguish between them as neuroblastic tumors lack a single feeding vessel.[9]

Adrenal hemorrhage shows variable appearance, ranging from an echogenic lesion to a cystic suprarenal mass, typically changing appearance on follow up.[9] Teratoma has a more heterogeneous appearance with irregular cystic areas, large irregular calcifications, and, occasionally, identifiable bony structures.[9]

The overall prognosis of neuroblastoma detected during the fetal stage is excellent, with a 90 to 100% survival rate.[10] Spontaneous regression and maturation to more differentiated forms of neuroblastic tumors have been described and take on average 6 to 12 months. The mechanism of regression is unknown.[10]

Conclusion

We present a case of a retroperitoneal neuroblastic tumor, tracing its progression from prenatal diagnosis during the third trimester of pregnancy until its regression at eight months of age. This case study highlights the favorable prognosis for neuroblastic tumors identified during pregnancy due to their potential for resolution without surgical interventions or chemotherapy. Furthermore, it underscores the diagnostic and prognostic importance of ultrasound and MRI imaging in fetal tumors.

Conflict of Interest
None declared.

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