

Ultrasound Diagnosis of Fetal Eye Mass

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Introduction

Fetal/neonatal orbital mass is extremely uncommon. We report a rare case of a fetal orbital mass noted on prenatal ultrasound with postnatal pathologic diagnosis of conjunctivoid variant of a dermoid cyst.

Case Report

A 23-year-old G2P1001 noted to have fetal left orbital mass on prenatal ultrasound at 31 weeks gestation measuring 3.5 x 2.6 x 3.4 cm after a normal anatomy scan at 20 weeks. The mass was clearly separate from the globe and was complex with septations and no vascularity. At delivery, the female infant had severe left eye proptosis and was unable to close the eyelid over the globe. MRI of the orbit and brain revealed a macrocystic fluid-filled mass of the left orbit most consistent with lymphangioma. Oculoplastics recommended urgent drainage of the lesion due to concern for exposure keratopathy. Left eye orbitotomy with partial resection of the cyst was performed; pathology revealed a benign cyst with non-keratinizing squamous epithelium and focally cuboidal epithelium, likely representing a conjunctivoid variant of a dermoid cyst.

Discussion

The differential diagnosis of a fetal orbital cyst includes congenital cystic eye, microphthalmos with cyst, cephalocele, hemangioma, lymphangioma, orbital teratoma, conjunctival epithelial cyst, and dermoid cyst (Shields 2004). Congenital cystic eye occurs when the optic vesicle fails to invaginate in the 2-7mm stage of fetal development. Microphthalmos with cyst is caused by failure of the embryonic fissure to close at the 7-14mm stage of fetal development. It presents on imaging as a cyst connected to a small globe. Cephalocele occurs when a portion of the brain herniates through an osseous defect. Hemangiomas are benign tumors composed of blood vessels. Orbital teratomas, conjunctival epithelial cysts, and dermoid cysts can be differentiated based on histopathological examination. True orbital teratomas contain tissues derived from all 3 germ layers. They appear as multiloculated complex cysts with tissues of varying echogenicity on ultrasound. Conjunctival epithelial cysts and conjunctival dermoid cysts are both composed of nonkeratinizing stratified squamous epithelium. The presence of dermal appendages distinguishes a dermoid cyst from a conjunctival epithelial cyst. Cases of conjunctival epithelial cysts presenting on prenatal ultrasound have been reported (Yen 2001, A. Singh 2009). To our knowledge, this is the first conjunctival dermoid cyst identified in utero on prenatal ultrasound. This variant comprises a small fraction of orbital dermoid cysts and typically presents later in childhood rather than at the time of delivery or on prenatal ultrasound.