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Prenatal diagnosis of concurrent facial and cerebral vascular malformation which caused congestive heart failure ☆

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ABSTRACT

Arteriovenous malformations (AVMs) are rarely reported antenatally. Most in utero diagnosis of vascular malformation is related to vein of Galen malformation (VGM). We describe a case of simultaneously diagnosed pial arteriovenous fistula (AVF) and facial vascular malformation in a 20 weeks old fetus. The dilated intracranial venous pouch appeared as a midline anechoic structure which was misdiagnosed as a VGM in her previous ultrasound exam. Another AVM was diagnosed in the same side of fetal face which fed by a branch of external carotid artery and communicated with the mentioned pial AVF. High output cardiac failure and hydrops were evident. To our knowledge this is the first report of prenatally detected combination of facial and cerebral vascular malformations at such as early pregnancy week.

1. Introduction

Arteriovenous malformations (AVMs) are rarely reported antenatally. Most in utero diagnosis of vascular malformation is related to vein of Galen malformation (VGM). We describe a case of simultaneously diagnosed pial arteriovenous fistula (AVF) and facial vascular malformation in a 20 weeks old fetus.

2. Case report

A 33 years old healthy female was referred to our hospital at 20 weeks of gestation with the diagnosis of VGM and hydrops fetalis.

Her last week sonography revealed an intracranial midline vascular lesion which was associated with skin edema over the skull, pericardial effusion, mild ascites and cardiomegaly. The parents had no remarkable past medical history and the first trimester sonography was normal.

The ultrasound exam in our institution revealed a midline dilated venous pouch lower than midbrain and posterior to the Willis circle. Using color Doppler several feeding vessels were identified. Findings were compatible with a pial AVF. This venous pouch draining was into the dilated superior petrosal sinus and then transverse sinus. Both jugular veins were prominent. Vein of Galen and straight sinus were normal (Fig. 1).

Such early presentation of hydrops fetalis was unusual for pial AVM, so looking for another probable reason we found an AVM at the same side of the face extending from squamosal portion of temporal bone down to the mandibular ramus. This AVM was communicating with the mentioned pial AVF via the probable dilated sphenopetrosal sinus. Also this AVM was receiving a feeding artery from ipsilateral external carotid (Fig. 2). No other brain abnormalities were detected.

Because of the associated high output cardiac failure (Fig. 3) and resultant hydrops the parents chose pregnancy termination. Parents precluded histopathologic evaluation of their fetus.

3. Discussion

AVM consists of disorganized vascular channels called nidus which directly connect arteries to the venous system. AVF is a vascular malformation similar to AVM but with no intervening nidus

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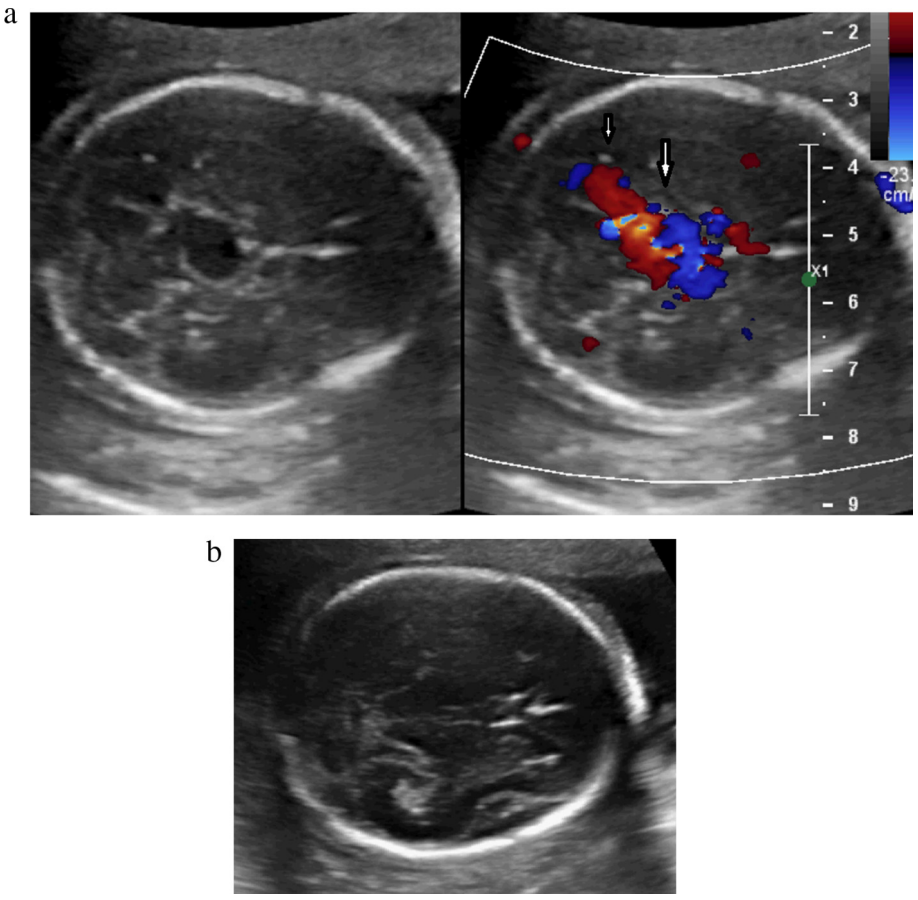

Fig. 1. a: Gray scale and color Doppler sonography show pial AVF as a midline dilated venous pouch at the level of willis circle which fed by multiple dilated arteries (larger arrow) and was draining into a dilated superior petrosal sinus (smaller arrow). b: Axial view at the level of midbrain. No pathology was found at the quadrigerminal plate cistern.

[1]. Their genesis seems to occur actually during 4th to 6th week [2,3]but often progress in late infancy and childhood period [4].

Most of the cerebral vascular malformation diagnosed at antenatal period is VGM [5–7]. Other types are rarer in fetal period. Pial AVF includes pial arteries directly open into a dilated draining vein. In dural sinus malformations (DSM) the fistula is located adjacent to the dural sinus with the sonographic appearance of a huge cystic lesion next to the dura matter. Both of them are distinguished from VGM by lack of median prosencephalic vein involvement [1,8,9].

Embryologically median prosencephalic vein is a precursor of vein of Galen which is supplied by internal cerebral veins. In VGM median prosencephalic vein does not involute and become dilated as a result of arteriovenous fistula. It is located at the velum interpositum and quadrigerminal plate cistern dorsal to the tectum and drain through the straight or falcine sinuses into the superior sagittal sinus [6].

In our case the dilated draining vein was located in the midline and misdiagnosed as a VGM but the distinguishing feature was its lower and more anterior position (posterior to the Willis circle and lower than midbrain, not in quadrigeminal plate cistern). Also it opened into a laterally located vein (superior petrosal sinus), not midsagittally located straight or falcine sinuses.

Garel et al reported three cases of pial AVF (one case near the midline and two laterally located) and mentioned the main distinguishing feature from VGM is the site of the vascular malformation [8].

VGMs and pial AVFs consistent 30% and 17% of cerebral malformations among pediatric group, respectively [6,8]

There are a few in utero detected cases of pial AVF. Although all of pial AVFs were presented in 3rd trimester [8,10,11] limited cases of DSMs and VGMs were detected around 23–25 weeks [9] The incidence and location of VGM may be the factors that facilitate their earlier sonographic diagnosis.

There is no other antenatally detected facial AVM in literature review. In head and neck, AVMs occur rarely in comparison to low flow vascular anomaly [3,12]s but their true incidence is not well known [3].They are commonly observed in the cheek and mid face [4]. Their behavior is locally aggressive usually progressing during puberty and adolescence, as an expansive mass with cosmetic disturbance [13].

Our facial AVM was fed by a branch of external carotid artery. Both jugular veins were dilated reflecting increased cardiac output. Mortality and morbidity often are related to cardiac failure which is associated with more complex and extensive lesions and earlier presenting cases [1,2,5,10].

Color Doppler sonography is the most available and the first line method in diagnosis of these types of vascular anomalies. Also this method provides information about important prognostic factors such as cardiac failure, hydrocephalus, brain ischemic lesions and ultimately on pregnancy outcome and postnatal therapeutic options [10,11].

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