

# Fetus amorphous acardious: report of a rare case and differential diagnosis from placental teratoma with review of the literature

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**Abstract.** – Fetus amorphous acardious is a rare fetal malformation, lacking a functional heart and bearing no resemblance to human embryos. The main differential diagnosis is with placental teratoma and is based on the degree of skeletal organization and umbilical cord formation. A 27-year old woman delivered a healthy newborn at 37 weeks' gestation. An amorphous mass, covered with healthy looking skin, was connected to the placenta with a short pedicle. X-ray examination of the mass revealed the presence of vertebral column associated with ribs and pelvic bones. Histopathologic examination demonstrated the presence of spinal tube inside the vertebral column. Microscopy of the pedicle was consistent with umbilical cord. Various other tissues were also discovered, such as adipose tissue, gastric and large.

#### Key Words:

Fetus amorphous acardious, Placental teratoma.

## Introduction

Fetus amorphous acardious is a rare fetal malformation, lacking a functional heart and bearing no resemblance to human embryos<sup>1,2</sup>. It is observed only in monozygotic, monochorionic, twin, rarely triplet<sup>3</sup>, gestations. Its incidence is estimated to be one in 35,000 deliveries<sup>4</sup>. Differential diagnosis from placental teratoma is problematic and depends on the degree of skeletal organization and umbilical cord formation<sup>5</sup>. The proposed by Fox and Butler-Manuel<sup>5</sup> criteria for the distinction of the two entities have received some criticism in the past<sup>6</sup>. These authors have

suggested that the presence of an umbilical cord, even a rudimentary one, and of craniocaudal skeletal organization favours the diagnosis of amorphous acardious<sup>5</sup>. Since then a few reports of fetus amorphous acardious have been published in the English literature<sup>1-3,7,8</sup>, albeit not all of them fit with the aforementioned criteria<sup>6,7</sup>. We present a case of fetus amorphous acardious and discuss the differential diagnosis from placental teratoma.

## Clinical History

A 27-year-old woman, gravida 1, para 1, presented in our hospital at 33 weeks' gestation due to ultra-sonographic discovery of a heterogeneous globular mass of 117 × 69 mm in the uterine cavity that lacked cardiac activity and was attached by a short pedicle to the placenta. A normal fetus was also present and no fetal distress was noted. Previous ultra-sound examination, at 23 weeks' gestation, was unremarkable. Two weeks later (35 week of gestation) the mass measured 150 × 110 and at 37 weeks' gestation the patient gave birth by vaginal delivery to a healthy boy weighing 3110 gr. An ovoid mass, covered by mature skin, was attached to the placenta via a short pedicle. The postpartum course was uneventful.

## Materials and Methods

The specimen was subjected to x-ray examination and then fixed in 10% formalin. Representative sections were embedded in paraffin and stained with hematoxylin-eosin (H & E).

## Results

An oval firm mass, associated with a short, 25 mm long, pedicle was delivered to the pathology department. The mass measured 200 × 119 × 73 mm and weighed 855 gr. It was covered with healthy-looking skin that was hairless apart from a small area at one pole of the mass that was covered with black hair (Figure 1A). The most prevailing diagnosis at this time was placental teratoma. However, x-ray examination of the specimen revealed the presence of a well developed vertebral column that consisted of 12 vertebrae and was connected with 9 ribs on each side. Three additional bones were noted at what was considered the caudal portion of the specimen and reminded of rudimentary pelvic bones (Figure 1B). These findings raised the possibility of a severely malformed fetus.

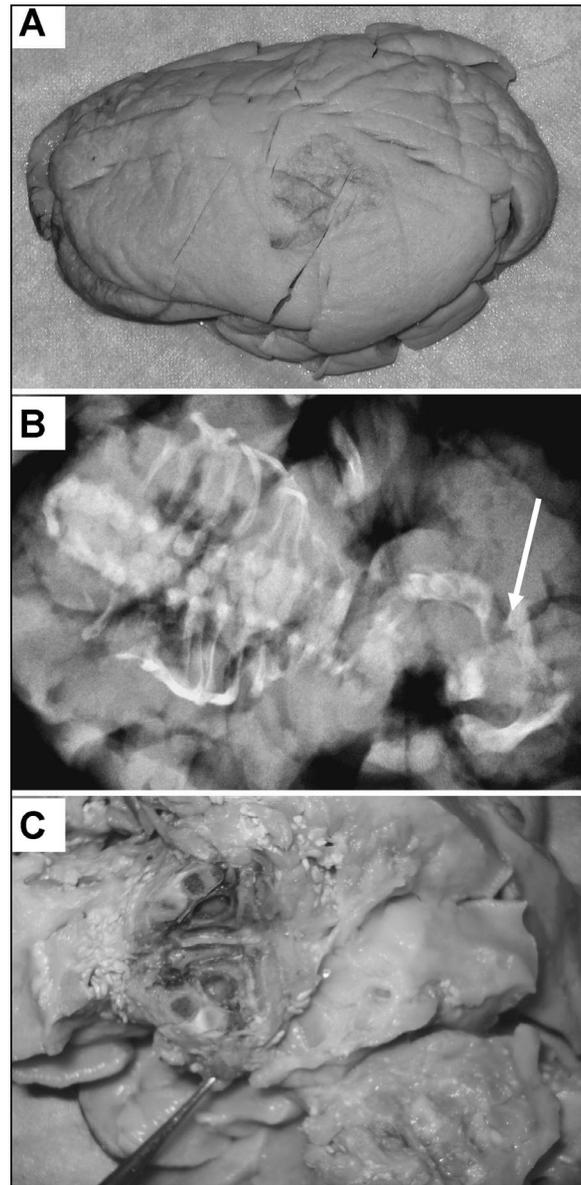
On cross section, adipose tissue comprised most of the specimen (Figure 1C). Three cystic areas were noted. One was located at the cranial part, respectively to the hairy skin and at the opposite pole of the rudimentary pelvic bones, and had reddish-tan fleshy tissue protruding in its cavity. Light microscopy proved the later to be choroid plexus overlying neuroglia (Figure 2A), thus confirming that this was the cranial part of the specimen. The other cystic areas, measuring 11 and 42 mm, were found in between the adipose tissue and were full of powdery white-tan material. Microscopically, they were composed of stomach and large bowel wall, respectively.

Interestingly, a spinal tube was found in the centre of the vertebral column, which contained a cylindrical, white-tan tissue, with soft consistency (Figure 1C). Histologic examination revealed that this tissue was composed of neural elements (neuroglia and sparse neurons) (Figure 2B), representing a residual or poorly developed spinal cord.

The pedicle that connected the mass to the placenta was comprised of two vessels (one artery and one vein) surrounded by mucomyxoid substance (Figure 2C), that was Wharton's jelly.

The placenta was delivered to the pathology department separately. It measured 18 × 15 × 3.5 cm, weighed 645 gr and was reddish tan in color. One amniotic cavity was present. No gross abnormalities were noted.

Based on the radiographic, gross and microscopic findings, a diagnosis of fetus amorphous acardious was made.

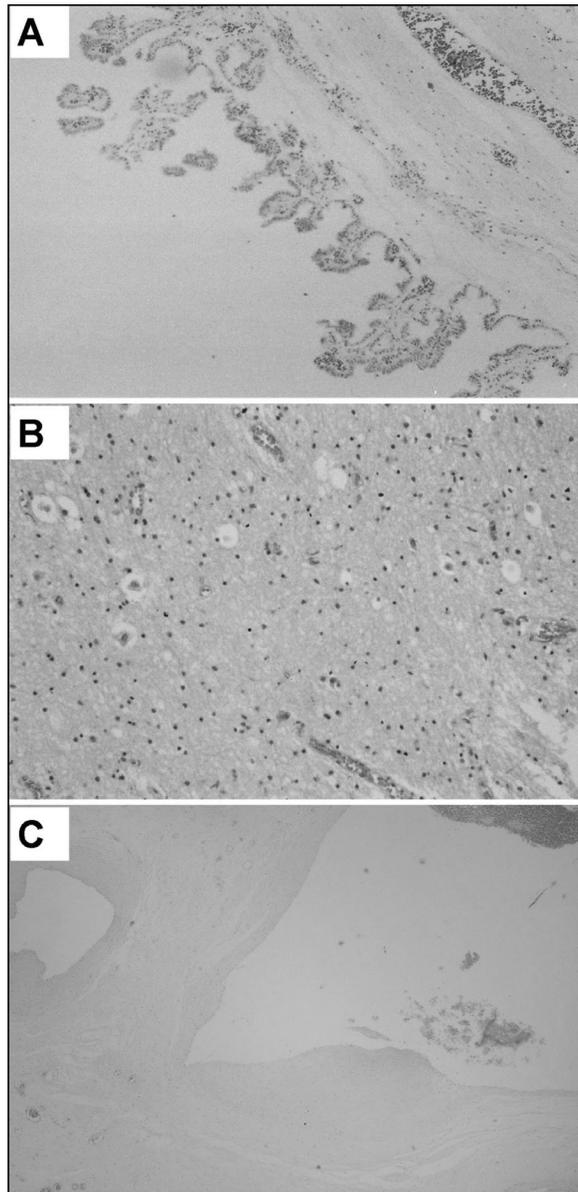


**Figure 1.** **A**, The mass was covered with healthy-looking skin. Note the black hair at one pole of the specimen. **B**, Radiographic examination revealed a well-developed vertebral column connected with ribs laterally and pelvis-like bones (arrow) caudally. **C**, Cross section of vertebral column unveiled a spinal tube containing a spinal cord-like tissue.

## Discussion

Fetus amorphous acardious is a rare fetal malformation, occurring in one in 35,000 deliveries<sup>4</sup>. It is characterized by total absence of heart and displays no resemblance to a human embryo<sup>1,9</sup>. It is always accompanied by the presence of a co-twin, since it needs the functioning heart of the

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**Figure 2.** **A**, Chorioid plexus overlying neuroglia was found at the cranial part of the mass (H & E,  $\times 200$ ). **B**, Nervous tissue that consisted of neuroglia and sparse neurons was found in the spinal tube (H & E,  $\times 200$ ). **C**, Light microscopy of the rudimentary umbilical cord showed that it contained one vein and one artery surrounded by Warton's jelly (H & E,  $\times 100$ ).

other embryo to provide circulation for both of them<sup>4</sup>. That is why it is always found in monozygotic, monochorionic twin<sup>1,4</sup> or rarely triplet<sup>3</sup>, pregnancies.

The pathogenesis of this condition is still ambiguous. It has been proposed that the malformed embryo has either a weak heart or no heart at all<sup>7</sup>.

This leads to a reversion of the blood flow from the normal embryo to the anomalous twin through anastomosing vessels between the two circulations<sup>1,7</sup>. According to this theory the deprived of nutrition and oxygen blood in the chorionic plate arteries results in hypoxia and malnutrition of the anomalous embryo, which can interfere with normal organ development. This theory has received a lot of criticism<sup>1</sup>. It has also been postulated that there is a primary failure in the development of many organs (including the heart) of the embryo<sup>1</sup>. However no chromosomal defect has been detected in these embryos<sup>8,10</sup>. The most prevailing theory seems to be that there is an existence of artery-to-artery anastomoses between the circulations of the twins, leading to reversal of the circulation and subsequent impairment in the development of the heart and other organs<sup>9</sup>.

Diagnosis of fetus amorphous acardious largely depends on the presence of an umbilical cord and of craniocaudal skeletal organization, as was proposed by Fox and Butler-Manuel<sup>5</sup>. Our case exhibited a clear-cut craniocaudal organization with a fairly developed vertebral column that contained a spinal cord and was associated with ribs and pelvis-like bones. There was also a cystic space at the cranial portion of the fetus with choroid plexus-like protrusions in its cavity that strongly resembled cerebral ventricles, although the possibility that it can represent cystic hygroma commonly seen in these fetuses<sup>9</sup> can not be excluded. A well formed umbilical cord was also noted. It is highly unusual for a teratoma to have such a high degree of organization<sup>11,12</sup>. The rarely encountered entity of fetiform teratoma, also called homunculus<sup>13-16</sup>, complexes the matter even more, but has never been described at the placenta. Fetiform teratoma is a mature ovarian teratoma containing a solid structure that bears a high degree of organization resembling an embryo and even contains vertebrae<sup>13-14</sup> or vertebrae-like bones<sup>16</sup>. However, a well developed vertebral column with a centrally located spinal cord, as was seen in our case, is an extremely rare feature of teratomas and has been encountered only once in the fetus-like structures of fetiform teratomas<sup>13</sup>.

It has been proposed<sup>6</sup> that the distinction between fetus amorphous acardious and teratoma is arbitrary, since they probably represent the two portions of a continuum. However, the two entities have a different origin (teratoma is a neoplasm whereas fetus amorphous acardious is a

fetal malformation) and different impact on the other embryo. Teratomas are rarely associated with fetal distress<sup>12</sup> while acardiac twinning can result in hydramnios, preterm delivery, congestive heart failure and even death of the co-twin<sup>4,9</sup>. Fortunately, our case had an uneventful course without any intervention. Alternatively, disruption of arterial supply to the anomalous embryo can be considered when the co-twin is at risk<sup>9,17-19</sup>.

Cytogenetic studies may aid the differential diagnosis in difficult cases<sup>16</sup>. Teratomas are homozygous for chromosome polymorphisms near the centromere, whereas normal tissues are heterozygous for these polymorphisms<sup>20</sup>. However, genetic analysis of fetus amorphous acardius has rarely been reported<sup>8,10</sup> and has not been performed in our case, as well. Furthermore, information about the polymorphism status of placental teratomas is not available, probably due to the rarity of the entity. Cytogenetic studies of placental teratomas and amorphous fetuses are needed to validate the use of genetical analysis in the differential diagnosis of placental neoplasms and fetal malformations.

In conclusion, distinction between the two entities, albeit difficult on some occasions, should always be attempted and largely depends on the radiographic, gross and microscopic features of the specimen.

### References

- 1) ALDERMAN B. Foetus acardius amorphus. *Postgraduate Med J* 1973; 49: 102-105.
- 2) CHI JG. Acardiac twins. *J Korean Med Sci* 1989; 4: 203-216.
- 3) DAHIYA P, AGARWAL U, SANGWAN K, SEN J. Antenatal diagnosis of twin-reversed arterial perfusion sequence (acardiac amorphous) in a triplet pregnancy: case report. *Arch Gynecol Obst* 2004; 269: 147-148.
- 4) MOORE TR, GALE S, BENIRSCHKE K. Perinatal outcome of forty-nine pregnancies complicated by acardiac twinning. *Am J Obstet Gynecol* 1990; 163: 907-912.
- 5) FOX H, BUTLER-MANUEL R. A teratoma of the placenta. *J Pathol Bacteriol* 1964; 88: 137-140.
- 6) STEPHENS TD, SPALL R, URFER AG, MARTIN R. Fetus amorphous or placental teratoma? *Teratology* 1989; 40: 1-10.
- 7) CALAME JJ, VAN DER HARTEN JJ. Placental teratoma or acardius amorphus with amniotic band syndrome. *Eur J Obstet Gynecol Reprod Biol* 1985; 20: 265-273.
- 8) CHEN CP, SHIH JC, CHANG TY, CHERN SR, LIN CY, WANG W, TZEN CY. Prenatal diagnosis of an acardiac amorphous twin and the anomalous co-twin in a monoamniotic twin pregnancy. *Prenat Diagn* 2005; 25: 618-620.
- 9) WONG AE, SEPULVEDA W. Acardiac anomaly: current issues in prenatal assessment and treatment. *Prenat Diagn* 2005; 25: 796-806.
- 10) LATTANZI W, DE VINCENZO RP, DE GIORGIO F, STIGLIANO E, CAPELLI A, ARENA V. An acephalus acardius amorphous fetus in a monochorionic pregnancy with sex discrepancy. *Twin Res Hum Genet* 2006; 9: 697-702.
- 11) ELAGOZ S, AKER H, CETIN A. Placental teratoma. A case report. *Eur J Obstet Gynecol Reprod Biol* 1998; 80: 263-265.
- 12) MEINHARD K, DIMITROV S, NICOLOV A, DIMITROVA V, VASSILEV N. Placental teratoma—a case report. *Pathol Res Pract* 1999; 195: 649-651.
- 13) ABBOTT TM, HERMANN WJ JR, SCULLY RE. Ovarian fetiform teratoma (homunculus) in a 9-year-old girl. *Int J Gynecol Pathol* 1984; 2: 392-402.
- 14) KUNO N, KADOMATSU K, NAKAMURA M, MIWA-FUKUCHI T, HIRABAYASHI N, ISHIZUKA T. Mature ovarian cystic teratoma with highly differentiated homunculus: a case report. *Birth Defects Res A Clin Mol Teratol* 2004; 70: 40-46.
- 15) LEE YH, KIM SG, CHOI SH, KIM IS, KIM SH. Ovarian mature cystic teratoma containing homunculus: a case report. *J Korean Med Sci* 2003; 18: 905-907.
- 16) WEISS JR, BURGESS JR, KAPLAN KJ. Fetiform teratoma (homunculus). *Arch Pathol Lab Med* 2006; 130: 1552-1556.
- 17) MCCURDY CM, CHILDERS JM, SEEDS JW. Ligation of the umbilical cord of an acardiac-acephalous twin with an endoscopic intrauterine technique. *Obstet Gynecol* 1993; 82: S708-711.
- 18) SEPULVEDA W, BOWER S, HASSAN J, FISK NM. Ablation of acardiac twin by alcohol injection into the intra-abdominal umbilical artery. *Obstet Gynecol* 1995; 86: 680-681.
- 19) TANAWATTANACHAROEN S, MANOTAYA S, WACHARA-PRECHANONT T, UERPAIROJKIT B, TANNIRANDORN Y, CHAROENVIDHYA D. The management of acardiac twins at King Chulalongkorn Memorial Hospital. *J Med Assoc Thai* 2004; 87: S284-287.
- 20) LINDER D, MCCAW BK, HECHT F. Pathogenetic origin of benign ovarian teratomas. *N Engl J Med* 1975; 292: 63-66.