CASE REPORT

Fetus Amorphous Acardious – A Case Report

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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 26 year old woman delivered a healthy newborn at 38 weeks of gestation by caesarian section. 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 and axial skeleton. Histopathological examination demonstrates the presence of cartilage, bone, adipose tissue, skin with adnexal structures and neural tissue.

Keywords: Teratoma, Amorphous Fetus, Axial Skeleton

Introduction:

Fetus amorphous acardious is a rare fetal malformation lacking a functional heart and bearing no resemblance to human embryos [1, 2]. It is observed only in monozygotic, mono-chorionic twin, rarely triplet [3], gestations. Its incidence is estimated to be 1 in 35,000 deliveries [4]. Differential diagnosis from placental teratoma is problematic and depending on the degree of skeletal organization and umbilical cord formation [5]. The proposed criteria by Fox and Butler Manuel [5] for the distinction of the two entities have received some criticism in the past [6]. But they have suggested that the presence of an umbilical cord even a rudimentary one and craniocaudal skeletal organization favors the diagnosis of amorphous acardious [5]. Since then a few reports of fetus amorphous acardious have been published in the literature [1, 3, 7, 8]. Albeit not all of them fit with the aforementioned criteria [6, 7]. We present a case of fetus amorphous acrdious and discuss the differential diagnosis from placental teratoma.

Clinical History:

A 26 year old Christian woman Gravida$_3$, Para$_2$, Living$_2$ ($G_3P_2L_2$) with 38 week 2 days gestation with previous caesarian section (G_1) and VBAL with twin gestation IInd twin IUD (G_2).

In the present pregnancy ultrasonography on 29th Oct 2014 revealed twin gestation with 1st twin 34 weeks 5 days healthy and 2nd twin 25 weeks without cardiac activity. On 30th Oct 2014, single live fetus of 37(SLIUG) weeks 3 days ill defined fetus like structure anterior to 1st fetus, head and body could not be differentiated (AC-27 weeks 5 days). Emergency caesarian section was done on 4th Nov 2014. Liquor clear, placenta was diamniotic monochorionoc, congenital anomaly of 2nd twin- Amorphous fetus with single limb bud. Facial muscles ill developed and ill defined. 1st twin is male child of 2.5 kg.

Received amorphous fetus with placenta was weighing about 1.1 kg. External examination shows hair at one end 3 cm below the some process which is 1cm in length and 3cm below the process, there is umbilical cord attachment. At lower end there is limb bud measuring 3cm.

Weight of placenta-450gm measuring 15 x 12 x 5cms (Fig 1). Cut section of placenta is unremarkable. The specimen was subjected for X-ray examination and then fixed in10% formalin. A longitudinal section is taken and revealed gray brown soft mass with areas of white and yellow areas. Posterior most areas of the mass were bony with vertebral column (Fig. 2). Multiple sections were taken and stained with hematoxylin and eosin stains.

Microscopy revealed skin with adnexal structures (Fig. 4 and 6), adipose tissue, bone, cartilage (Fig. 5) and neural tissue (Fig. 7). **In view of radiological appearance of the mass which revealed axial skeleton, pelvic bones, vertebral column with ribs (Fig. 3), a diagnosis of fetus amorphous acardious was made.**

Fig. 2- Cut Section showing Heterologous Elements with Hair

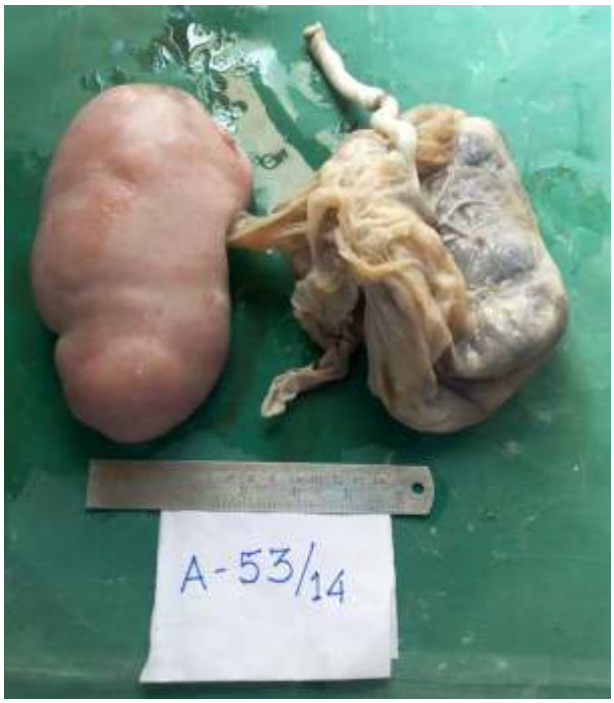

Fig. 1: Gross Picture Showing Fetus Amorphous with Placenta and Attached Umblical Cords For Twins

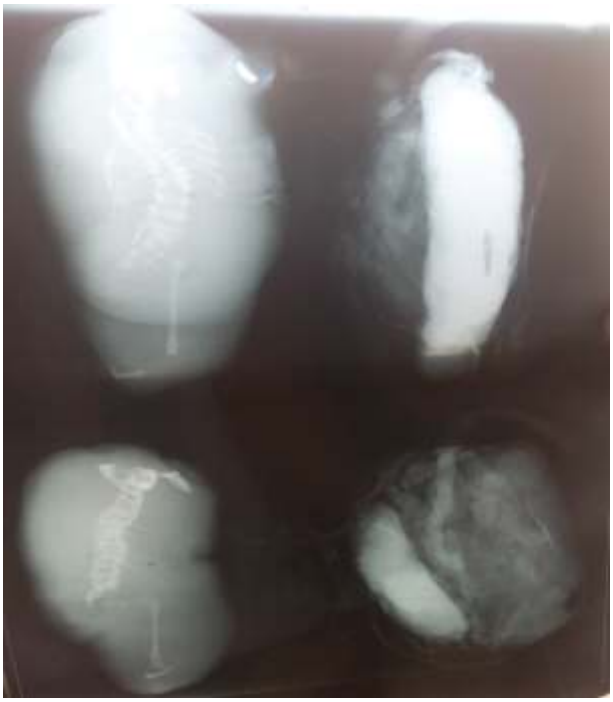

Fig. 3: X-ray of Amorphous Fetus Showing Axial Skeleton with Limd Bud

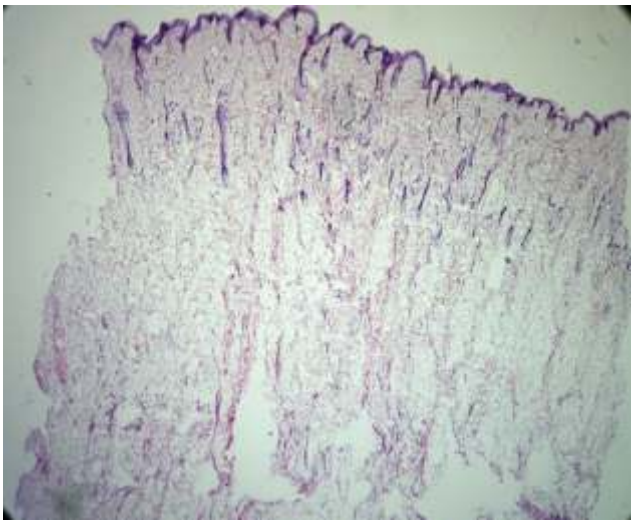

Fig 4: Skin showing Adnexal Structures

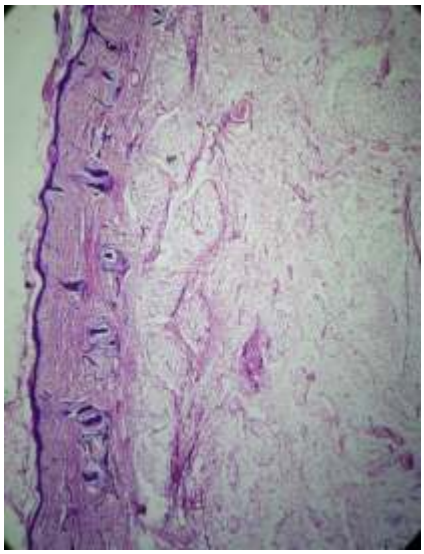

Fig. 6: Showing Skin with Adnexal Structures, Fat and Msenchymal Elements

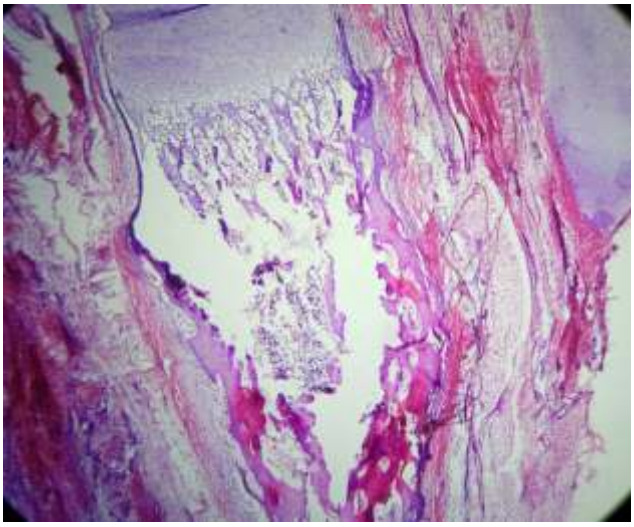

Fig 5: Showing Bone with Bone Marrow, Cartilage and Soft Tissue

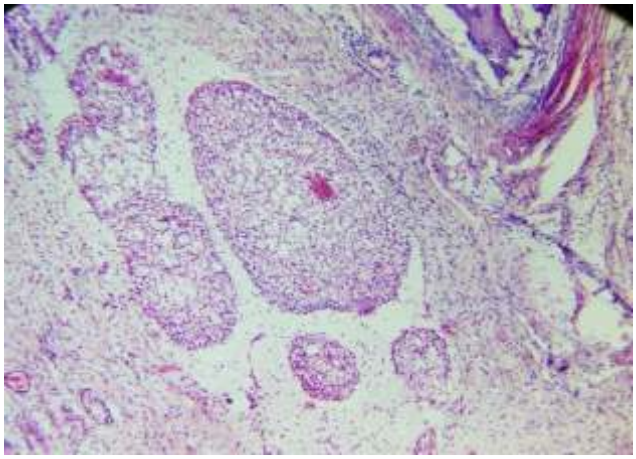

Fig. 7: Showing Bone with Neural Structures

Discussion:

Fetus amorphous acardious is a rare fetal malformation occurring in 1:35000 deliveries [4]. It is characterized by total absence of heart and displays no resemblance to human embryos [1,9]. It is always accompanied by the presence of a co-twin, since it needs the functioning heart of the other embryo to provide circulation for both of them [4]. That is why it is always found in monozygotic monochorionic twin [1, 4] or rarely triplet [3] 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 [7]. This leads to a reversion of the blood flow from the normal embryo to the anomalous twin through anastomosing vessels between the two circulations [1, 7]. According to this theory the deprival of nutrition and oxygenated 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. It has also been postulated that there is primary failure in the development of many organs of the embryo [1] (including heart). However no chromosomal defect has been detected in these embryos [8, 10]. The most prevailing theory seems to be that there is an existence of artery-to-artery anastomosis between the circulation of the twins leading to reversal of the circulation and subsequent impairment in the development of the heart and other organs [9].

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 [5]. Our case exhibited craniocaudal skeletal organization with vertebral column, ribs, pelvic bones and axial skeleton without any organs. There may be presence of brain, spinal cord stomach and large bowel wall. It is highly unusual for a teratoma to have such a high degree of organization. The rarely encountered entity fetiform teratoma, also called homunculus, complexes that matter even more, but has 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 or vertebrae like bones.

It has been proposed [6] that the distinction between fetus amorphous acrdious and teratoma is arbitrary, since they probably represent the portion of a continuum. However the two entities have a different origin (Teratoma is a neoplasm whereas fetus amorphous is a fetal malformation) and different impact on the other embryo. Teratomas are rarely associated with fetal disorders while cardiac twinning can result in hydramnios, preterm delivery, congestive heart failure and even death of the co-twin [4, 9]. 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 [9].

Cytogenetic studies may aid in the differential diagnosis in difficult cases. Teratomas are homozygous for chromosome polymorphisms near the centromere whereas normal tissues are heterozygous for these polymorphisms. However genetic analysis of fetus amorphous acardious has rarely been reported [8, 10] 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 rarity of the entity. Cytogenetic studies of the placental teratomas and amorphous fetuses are needed to validate the use of genetic analysis in

the differential diagnosis of placental neoplasms and fetal malformations.

Conclusion:

The distinction between the two entities, the placental teratomas and amorphous fetuses, albeit was difficult. In some occasions should always be attempted and largely depends on the radiographic, gross and microscopic features of the specimen.

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