

Antenatal ultrasound diagnosis of dicephalus dipus dibrachius - and its correlation with autopsy

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Ind J Radiol Imag 2006 16:1:107-108

Keywords: Antenatal diagnosis, fetal anomalies, conjoined twins, dicephalus dipus dibrachius

INTRODUCTION

The antenatal diagnosis of dicephalus dipus dibrachius is extremely rare. Two cases of dicephalus dipus dibrachius diagnosed at different periods of gestation are illustrated here. These fetuses were unique for the absence of spinal duplication and the presence of a median facial proboscis in one. The ultrasonographic findings were confirmed after therapeutic abortion and ultrasonographic autopsy.

CASE REPORT

This report is aimed at highlighting the antenatal features of dicephalus dipus dibrachius conjoined twins. The ultrasonographic features of the two fetuses scanned by the US machine (Aloka 5000 Pro, Tokyo) with 3.5 to 10 MHz high density probes are given below.



Fig 1. The coronal US scan of the first fetus shows single spine, part of ribs and the fused heads (1- first head, 2- second head)

Fetus No. 1. A twenty two year old primipara was referred for a first trimester scanning at 14 weeks of gestation. She had no contributory personal or family history. The uterine size was corresponding to her period of amenorrhoea. The liquor and placental features were

normal. The fetus was appearing normal but for a slightly elongated skull contour on sagittal plane. A coronal section showed the two lateral projections representing the heads above the level of the neck (Fig. 1). The axial section of the head showed the fused skulls at the occipital regions. The brain in each calvaria was normal but for the fused occipital portions at their posterior aspects. The ventricles showed normal width and the choroid plexus were normal (Fig. 2). The facial details were technically poor even by an endovaginal scanning. The body was single and only a pair of upper and lower limbs was seen. The spine showed no duplication.



Fig 2. The ventricles showed normal width and normal choroid plexus (marked *). The occipital regions are fused.

Fetus No. 2. A twenty four year old primipara came for routine ultrasound scanning at 26 weeks of gestation. Her personal and family histories were also not contributory. The uterine size was normal for the period of gestation. Ultrasound scanning showed normal placental parameters and liquor volume. The skull bones showed an irregular contour. Two main bulges formed the head. The brain matter was totally abnormal with out delineation of the

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Received 7 April 2005; Accepted 12 September 2005

internal anatomy. A prominent cystic space was seen on each side, out lined by the brain matter (Fig. 3). An anterior mid-line proboscis was seen among the two heads. The nose was not definable. The orbits and mouth were poorly defined in one head. The trunk was single and there were only a pair of upper and lower limbs. The vertebral column showed no splitting up to the head.

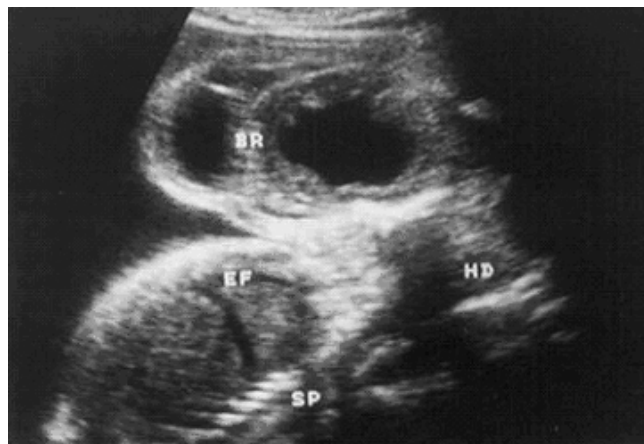


Fig 3. The second fetal head showed a prominent cystic space on each side of the fused heads, out lined by the brain matter (BR). The internal structures were disorganized (HD-head, EF- minimal pleural effusion, SP- spine).



Fig 4. The second fetus after abortion showed the fused heads and deformed facies. The median proboscis is seen between the heads.

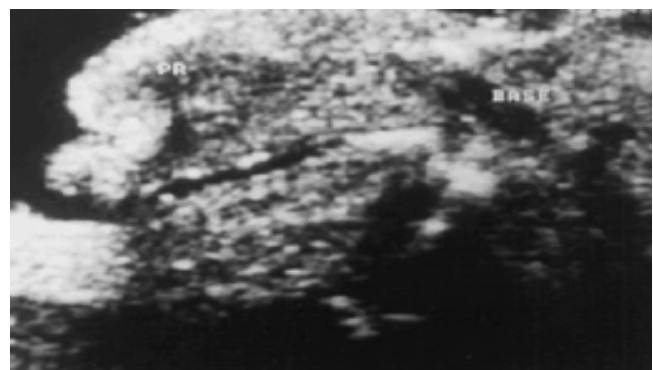


Fig 5. Ultrasonographic autopsy shows the fine details of the anterior median proboscis as an elongated soft tissue structure attached to the area between the two heads.

Both pregnancies were terminated after the confirmation of the diagnosis by repeated scanning. The abortus showed the features consistent with the antenatal ultrasound details. The examination of the first abortus could not be carried out because of degenerative changes. The second one (Fig. 4) was subjected to ultrasonographic autopsy using a 10 MHz high density probe after immersing in a tray of water. The anterior median proboscis was seen as an elongated soft tissue structure attached to the area between the two heads (Fig. 5).

DISCUSSION

Conjoined twinning results from incomplete division of the embryonal axis between 13 to 15 days after fertilisation. This is unique among the monochorionic monoamniotic twins (1 in 300 monozygotic twins). The incidence of conjoined twins is reported to be around 1 in 50,000 to 100,000 births [1,2,3]. They are classified according to the anatomical sites of incomplete splitting (Terata catadidyma, terata anadidyma, terata ancatadidyma). Among terata anadidyma, the subtype craniopagus is characterized by the incomplete splitting of the head. The trunks and the limbs are separate. This occurs in less than 2% of conjoined twins. Dicephalus falling under the group terata catadidyma is extremely rare characterized by the presence of two heads and one body.

The literature shows one report of antenatal ultrasound diagnosis of dicephalus at 12 weeks [4]. Another asymmetric parasitic dicephalus fused at the neck of the co-twin was reported at 17 weeks of gestation [5]. In dicephalus, though the body appears single externally, the spine may show varying extent of duplication. The number of the upper limbs may vary from 2 to 4 [6]. The dicephalus fetus with two heads, one trunk and two upper limbs is known as dicephalus dipus dibrachius. These two fetuses described above had typical features of this variety but the spine showed no duplication. The presence of a median proboscis in one fetus is reported for the first time. The available literature shows no description of proboscis in these fetuses. Surgery is not advocated for them as the prognosis is usually lethal [7].

CONCLUSION

Dicephalus dipus dibrachius is very rare among the conjoined twins. The morphological features vary extremely between them and demand autopsy for the correlation of ultrasonographic details.

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