

CASE REPORT

Embryological, clinical and anatomopathological considerations on a very rare case of a bicephalous fetus (*dicephalus dipus dibrachius*)

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Abstract

In this paper, we present the very rare case of a 21 weeks pregnant woman, examined clinically and by ultrasound. We could observe the present of two heads and a common trunk and therefore we proceeded to the small caesarian of necessity. We examined the fetus from the clinical and pathological point of view and we could observe a rare case of pregnancy with a bicephalous malformed fetus. During the autopsy, we could reveal the common elements and the devised ones, which are described as it follows. The case is a very rare one and that probably explains the fact that until the 21 weeks examination, even if she was clinically and ultrasound examined there was the appreciation that it was a twin pregnancy with a normal evolution. In this case, it is necessary to underline that the difficulties concerning the diagnosis and the medical behavior are also because the patient was a primipara 34-year-old woman without any pathological or specific family history. This case analysis shows the necessity of a preconception genetic advice together with the extension of the analysis of the genetic risk in all mothers under 35-year-old, mandatory and supported by the medical system. At the same time, an ultrasound examination of great performance proves to be necessary in order not to prolong the development of a pathological pregnancy.

Keywords: pathological pregnancy, bicephalous fetus, *dicephalus dipus dibrachius*.

☐ Introduction

The case presented is very rare.

The particularity lies in this appearing in a pregnant primipara woman, clinically healthy without any history of obstetric pathology, with a pregnancy evolution that we considered as normal, without the intervention of risk factors, as history revealed. The patient is a nurse care with a high educational level and socio-economic environment.

We have to mention that the pregnant woman could not perform other tests such as Torch-test, Double-test or Triple-test, although they recommended, because these tests are too expensive for us and are not subsidized by the health insurance system of our country.

Another feature of the case is that being very rare, the first ultrasound examination performed on the patient at 12 weeks suggested twin pregnancy and was diagnosed as such.

The multiple or the twin pregnancy presents a fundamental characteristic in the majority of the mammals.

The conjoined twins represent a rare abnormality, one case in 50 000 births [1].

The cases of bicephalous fetus are even more rare, in the last decades being reported only a few worldwide. The survival until the adult age depend by the absence of some cardiopulmonary and intestinal malformations, which is hardly probable to happen, the malformations being usually complex in nature [2].

☐ Patient, Methods and Results

The case presented in our paper is a 34-year-old primipara, primigesta woman, who came to a specialist examination with a 21 weeks pregnancy accusing hypercontractility of the uterus.

The medical history showed that the patient had no significant family history, neither personal, physiological or pathological conditions. Menarche at 12 years, normal menstrual cycles of 30 days length, moderate menstrual flow, five days duration, without dysmenorrhea. The patient is primipara (IP) and primigesta (OG). The husband showed no pathological conditions or family

history. None of the partners suffered from any sexually transmitted diseases. Both partners reported no genetic disorders in the family. The pregnancy was achieved spontaneously without any treatment, at 34-year-old because the patient married later. The current pregnancy evolved normally without any subjective or objective events and the pregnant did not follow any treatments. The patient showed no exposure to hazards does not work or interact with any toxic substances or other pollutants.

There were no bacterial or viral infections during pregnancy. Previously and during pregnancy, she did not take any medication. No drinking, no smoking, no drugs, coffee occasionally.

The patient is a nurse in a pediatric ward of a county hospital, and although she may not make any correlation, we suspect a possible viral or bacterial infection contracted at work and that would have onset during the period of subclinical pregnancy.

The patient was previously clinically and with ultrasound examined and she was diagnosed with a twin pregnancy.

During pregnancy, she did not perform the TORCH Complex, its interpretation could have shown us previous vaccination or disease status for rubella, toxoplasmosis, herpes and cytomegalovirus, and even during the current hospitalization she refused performing it, affected by the discovery of the malformation.

She did not afford to perform the Double-test or Triple-test, though the tests recommended by her gynecologist.

The laboratory tests preceding the hospitalization are considered as normal, having the following values: Hemoglobin 12.8 g/dL, APTT 33.3%, glucose 73.4%, sterile urine cultures, Pap smear test type II, polymorphic vaginal flora.

During hospitalization, the clinical general examination revealed no pathological elements. The breast exam showed pregnancy changes, negative from an oncological point of view.

It should be mentioned that the patient was previously repeatedly clinically and ultrasound examined and she received the diagnosis of twin pregnancy with a normal evolution. She did not perform the double test, or the triple test or other genetic tests.

The laboratory tests she already had taken were within the normal values.

The anamnesis and the general clinical examination did not show any pathological signs. At the same time, there were no previous twins in the family.

At the clinical examination it was observed a shortened closed cervix, without any lesions, the patient did not lose any blood or amniotic liquid, she presented an increased uterus with an ovoid volume with the big longitudinal axis height of the uterus' upper part (IU) equal to 24 cm, contractile, with a slightly increased tonus, slightly sensitive, fetal heart beat (BCF) present – 164/minute.

The exploration of first intention was the conventional abdominal ultrasound, which showed the clear presence of two fetal heads with biparietal diameter 52 mm, and the other one with biparietal diameter 53 mm, a unique

thorax, only one bone pelvis, two superior members and two inferior members.

We proceeded to the 4D sonographic examination and we observed:

The cardiac image did not suggest the eventual presence of two hearts completely separated, but the presence of only one heart with a normal heart rate of 144 beats/minute, possibly malformed (Figure 1).

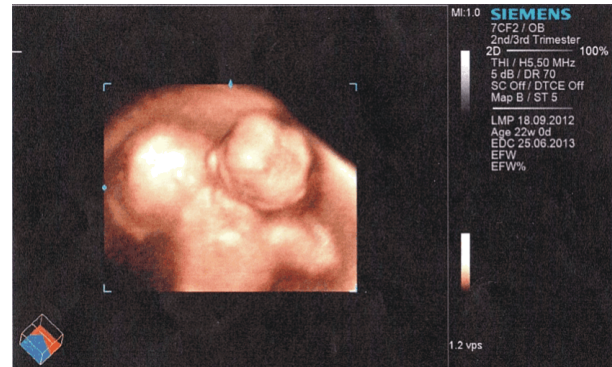


Figure 1 – 4D echographic ultrasound image of the bicephalous fetus.

The umbilical cord was a unique one, with a circular paracervical at the basis of the convergence of the two cervical columns united in a “V”.

The placenta is posteriorly positioned with a normal echo structure.

We decided to perform a C-section surgery in order to extract them after we proceeded to all the other tests and investigations: the hematologic, urinary ones, together with the anesthesiologist and the neonatologist consults.

We needed to perform a small C-section and we could observe the existence of one fetus, with only one trunk and one pair of superior and inferior members, two necks and two heads.

The child was a male, L=25 cm, W=780 g, alive in the moment of the extraction and dead five minutes later (Figure 2).



Figure 2 – Bicephalous fetus after extraction from the uterus.

The fetus presented one circular cord around each neck and two circulars in a scarf around the trunk. The cord was a unique one and it was centrally inserted in the placenta. The placenta was a unique one and from the macroscopic point of view we could observe an over dimension for the age of the pregnancy G=480 g. It was appreciated an extra quantity of the amniotic liquid.

The surgery and anesthesia intervention took place under normal circumstances.

The post surgery evolution of the patient was good, with a per-prima lesion healing.

The patient was ablactated. She left the hospital the 6th day, clinically and surgically cured with the usual recommendations, also to continue the genetic investigations for the couple.

We initially appreciated, after the first ultrasound and clinical examination and immediately after extraction of the fetus, we were dealing with a twin pregnancy with twin conjunctiva, which was invalidated by the pathological examination.

In the Department of Pathological Anatomy, they described a bicephalous (*dicephalus dipus dibrachius*) fetus meaning with two heads, two superior members and two legs.

Anterior wall of the thorax showed a normal macroscopic appearance (Figure 3), but the spines were double alongside their full length (Figure 4). There were four scapulas present, two on each side but with only two clavicles attached on a unique sternum. We did not observe any malformations for the skeletons of the heads and of the free superior and inferior members, the last ones being connected to a small unique pelvis.



Figure 3 – Bicephalous fetus: Dissection of the anterior wall of the thorax showed normal muscular and sternocostal plans.



Figure 4 – Bicephalous fetus: The vertebral columns were separated on their whole length.

Much more complex were the internal organs malformations. The lungs did not show any visible macroscopic malformations, there were two with normal

lobes: three lobes on the right and two lobes on the left while each of them was served separately by a trachea.

The heart had two pairs of ventricles but two common atriums. Consequently, we can say that there were no malformations present on the venous system while the aortic system was a double one until the L2 vertebra where they conjoined in a common abdominal aorta and then they separated in a normal way in two common iliac veins.

The pulmonary trunk began only in the right ventricle on the left side. The left pulmonary artery branched normally to the left lung, while to the right lung it branched only one with a small caliber. The left lung also received the blood from some pulmonary abnormal arteries, which came from the left descendent thoracic aorta.

The digestive tube was a double one almost until the ileocecal valve where the two small intestines conjoined into one dilated intestine on 1/1/0.5 cm. Surprisingly, the liver was a unique one, but bigger in volume, the left lobe went in the left hemithorax, due to a major left diaphragm hernia. The gallbladder was a double one, situated one into the other's proximity. This disposition in pair was kept for the small intestines too until their confluence described above. The urogenital system did not present malformations that could be seen from the macroscopic viewpoint.

Discussion

Taking into consideration all the results of the clinical and ultrasound examinations we decided upon the primipara, primigesta (IGOP) diagnosis of a 21 week pregnancy of Siamese twin brothers, possibly thoracopagous ones [3–5].

Although there could not be made a correlation with the exposure to pollutants, toxic materials or consumption of drugs, we still cannot postulate a viral infection, the patient working in a health care-a pediatric ward. The patient did not perform any specific genetic investigations during pregnancy and refused to perform some of them during hospitalization. She, however, agreed to perform a couple of tests discharged by the hospital [6, 7].

Taking into considerations the monozygotic division of the egg, there can appear different stages of the segmentation and cleavage, until the 14th day, when “the series cell head” start their development. The division is an asynchronous one and, at various times, it may involve the macromeres that can give birth to the embryo and the micromeres that will give birth to the trophoblast and fetal appendix.

When the division is produced after the 12th–14th day after the fecundation, in the stage of cleavage of the embryonic disk, the ovular covers being already present, there emerge double monsters or Siamese twins (the cleavage and the organogenesis not being completed).

The double monsters are linked fetuses in some region of the body, with loss of the total or segmentary individuality.

The incomplete duplication can be symmetrical or asymmetrical leading to two subjects linked equally or unequally. It is the situation of the thoracopagous twins that led to differential diagnoses problems in our case.

To be considered thoracopagus twins, it is necessary to have one pair of members for each fetus, which was not our case [8].

In our case, there was one fetus, with only one trunk and one pair of superior and inferior members, two necks and two heads. In Greek, *dicephalus dipus dibrachius* (*di* = double + *cephalus* = head, *di* + *pūs* = leg, *di* + *brachius* = arm] mean a person with two heads, two legs and two arms [9].

The ultrasound examination of great performance is necessary not to prolong the development of a pathological pregnancy [10–12].

The literature has described a case of a *dicephalus dibrachius* term newborn who survived for 11 days. The multiple tests performed (magnetic resonance, ultrasound X-ray scans) showed the presence of two spines, but lots of conjoined internal organs such as the heart, the liver, the pancreas, so the authors concluded that a surgical intervention could not have been performed in this case [1, 13].

The present case is very rare and the ultrasound diagnosis is established late due to a confusion with a normal twin pregnancy and ultrasound diagnostic difficulties [13, 14].

The incidence of Siamese twins is from 1 in 50 000 up to 1 in 100 000 births according to various authors [15–18]. Bicephalous are even more rare: since 1959 until 2010 there were reported only 48 cases [19].

Two mechanisms were taken into consideration: cleavage and fusion.

The cleavage may produce in different moments since fecundation. If it is produced four days since fecundation, when the trophoblast from embryoblast differentiation occurs (chorion), there will result monozygotic twins with separate covers (bichorial, biamniotic ones). If it takes place between the 4th and the 8th day since fecundation, the twins will be surrounded by a single chorion, but they will be each covered by a separate amnion (monochorial, biamniotic ones). Between the 8th and the 13th day since fecundation, the twins will share the same chorion and the same amnion (monochorial, monoamniotic ones). When the division produces after the 13th day since fecundation, within the cleavage stage of the embryonic disk, the ovular covers being already formed, there will result double monsters or Siamese twins (the cleavage and organogenesis are incomplete). Double monsters are fetuses connected in a certain part of the body by losing their total or segmentary individuality. In this mechanism, the moment and localization of cord knots cleavage are determinant [20].

The mechanism of the fusion involves a secondary connection of two embryonic disks, initially mono-ovular ones. This mechanism is supported by Rowena Spencer [21, 22] who enjoys the adhesion of most worldwide specialists [23–25].

The severity of malformations depends mainly on the craniocaudal separation of paleoaxes, namely the notochord [26]. This separation may be performed at various areas: from cranium basis, with two sides of the same cephalic extremity (diprosopus) [25], with two craniums occipitally fused [18], up to a complete separation of the spines [1].

According to some authors [27], gastrulation errors can be explained as errors in proliferation, migration and subsequent differentiation of the intra-embryonic mesoderm resulting in defective morphogenesis. We believe that the occurrence of malformations is the result of an internal and external complex factors (e.g. viral, microbial, vascular, nutritional, genetic, etc.) acting in the early stages of embryonic development.

Conclusions

The case presented in our paper is very rare and sometimes the diagnosis is established quite late. Although there could not be made a correlation with the exposure to pollutants, toxic materials or consumption of drugs, we still cannot postulate a viral infection, the patient working in a health care – a pediatric ward. In order to avoid any wrong interpretations, it is a necessity to proceed to a careful, accurate ultrasound investigation in all pregnancies in the first term of pregnancy. Analyzing this case highlights the need for preconception genetic counseling and the importance of mandatory free tests for genetic risk in mothers even under the age of 35-year-old.

Contribution Note

All authors have equal contribution for this article.

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