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Ischiopagus Which is the Rarest Form of Heteropagus Along With Ambiguous Genitalia and Kidney Agenesis: A Case Report

Ambiguus Genitalya ve Böbrek Agenezisi ile Birlikte Görülen Heterofagusun en Nadir Formu Olan İskiofagus Vakası

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Abstract

Parasitic twin (heteropagus) is a term to describe an incomplete fetus which is partially resorbed and located on a normal fetus. Ischiopagus which defines the parasitic twins connected from ileum is the rarest form of heteropagus abnormalities. In this case, a thirteen week old fetus was diagnosed with ischiopagus, ambiguous genitalia, and left kidney agenesis during the first trimester ultrasonography screening. Termination was applied with the family decision. These cases should be evaluated together with accompanying multisystem anomalies. Informing the family with details about the viability of the fetus is important to prevent unnecessary termination.

Keywords: Heteropagus, kidney agenesis, ambiguous genitalia

Öz

Parazit iki kiz parsiyel olarak rezorbe olmuş ve normal fetus üzerinde yerleşmiş inkomplet fetüsü tariflemek için kullanılan bir terimdir. İskiofagus normal fetuse ileumdan bağlanmış parazitik fetüsü ifade edip parazit ikiz vakalarının en nadir görülen formudur. Bu vakada on üç haftalık fetus birinci trimester ultrasonografik incelemesinde iskiofagus, ambiguus genitalya ve böbrek agenezisi tanısı almıştır. Ailenin kararı sonucu terminasyon uygulanmıştır. Parazitik ikiz vakaları eşlik edebilecek multisistemik anomaliler açısından dikkatli incelenmelidir. Aileyi fetüsün durumunun yaşamla bağdaşırlığı açısından bilgilendirmek gereksiz terminasyonları önlemek açısından önemlidir.

Anahtar Kelimeler: Parazitikikiz, böbrek agenezisi, ambiguus genitalya

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Introduction

Human extremities start to be formed four weeks after fertilization. Lower extremity buds begin to grow two days later than the upper extremity buds. Each bud has the ectodermal cells which provide the proximal to the distal growth. Parasitic twin (heteropagus) is a term to describe an incomplete fetus which is partially resorbed and located on a normal fetus [1].

Heteropagus is a rare abnormality which is seen one in 1.000.000 live births [2]. It was firstly described in 16th century by French surgeon Ambroise Pare as connection of the abdominal area to any part of the body. The most comprehensive case report was done in 2001 by Spencer; one hundred and fifty-seven cases were presented after literature review of over one hundred and twenty-five year data [1]. Spencer described eight anatomic locations according to the heteropagus connection sites: omphalopagus, thoracopagus, cephalopagus, craniopagus, ischiopagus, parapagus, pygopagus, rachipagus [1]. Thirty-nine cases were reported by Sharma et al. after 2001; only five of them (12%) were ischiopagus which is extremely rare to be seen [2].

Heteropagus is a rare developmental disorder of unknown etiology. Although there are some suspected ethiopathogenetic mechanisms, more studies are needed to explain exact pathogenesis. There is not any evidence about the effect of maternal age on heteropagus improvement.

There is not any heteropagus cases reported in mothers who uses alcohol, cigarettes, or any narcotic drugs. However, teratogen drug use, dietary factors, and maternal diabetes are theoretically likely in the etiology of heteropagus. In addition to them, there are some complex theories were discussed regarding the etiology; one of them says division or bifurcation of extremity buds was the pathology in heteropagus. Another one supports a fault in specification of extremity buds causes extremities more than normal number.

Management of heteropagus is based on the maternal and fetal conditions. Heteropagus is not a termination indication by itself, however, the fetus should be reassessed for the termination in the presence of additional fetal abnormality[2].

In this case report, it is aimed to describe the fetus with ischiopagus, ambiguous genitalia, and unilateral kidney agenesis.

Case report

The ethical approval was obtained from the local ethical committee. The case was twenty-eight years old multigravida mother (G2P1) at 13th gestational week without any chronic diseases. The mother neither had a family history of heteropagus nor a teratogen drug use, smoking, alcohol, narcotic drug use history. She had one year old son who was diagnosed as tetralogy of Fallot in antenatal period and had delivered with cesarean section due to presentation anomaly. Parents did not have endogamy. The first child was not diagnosed with any other congenital abnormality in neonatal visits; he referred to cardiovascular surgery department to be operated.

In the current pregnancy, a second left lower extremity belonged to the parasitic twin was described during the first trimester screening ultrasonography at 13th gestational week*(Figure 1)*. This extra lower extremity which was originated from sacral region was connected to the iliac wing with loose and ossified association. In addition, this extremity contained femur, tibia, and fibula, however, finger sequence was irregular and polydactyly was diagnosed*(Figure 2c)*. Left kidney agenesis and ambiguous genitalia were seen additionally*(figure 2d)*. Adrenal gland was seen in the left kidney area.

The family was informed about the chromosomal diseases, screening tests, and amniocentesis. After the perinatology council, termination of pregnancy was introduced to the family. Since the family requested to terminate the pregnancy, informed consent was obtained and the pregnancy terminated. Termination was carried out by labor induction with 100 mg sublingual misoprostol administration every three hours for twenty-four hours. Bumm uterine curette was used to empty the uterine cavity at the end of twenty-four hours. The patient was asked for the permission of this case report publication and usage of the figures, she approved the publication.

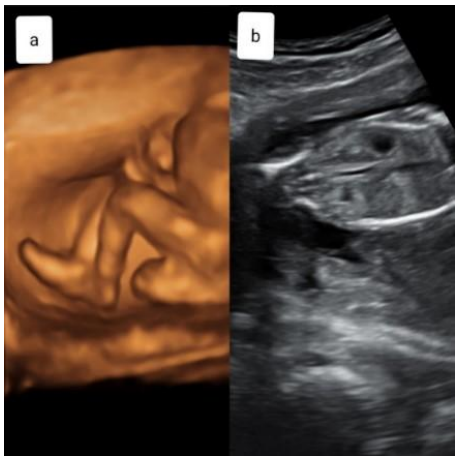

Figure 1: (a) *parasitic lower extremity 3D ultrasonographic picture* (b) *parasitic lower extremity (femur) ultrasonographic picture*

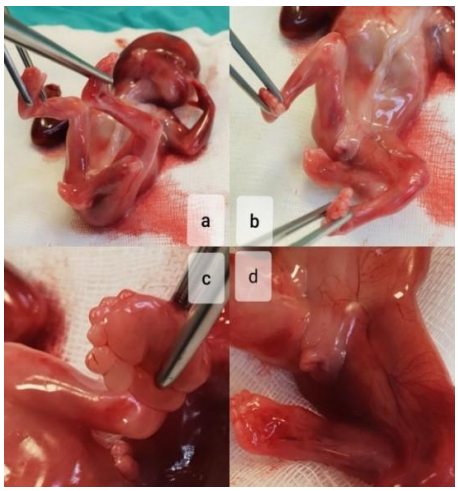

Figure 2: (a, b) *parasiticthird lower extremity of abortion material,* (c) *polydactyly of abortion material,* (d) *ambiguous genitalia of abortion material*

Discussion

Early antenatal diagnosis of heteropagus become easier with the developed imaging techniques. Therefore, family counseling, decisions of prenatal and postnatal management, and surgery planning can be done more safely.

Although the number is not a lot, there were some cases published in the past years about heteropagus. Norman published the case of ischiopagus which had duplicated leg and polydactyly in 1964 [3]. In our case, genital and urinary system abnormalities such as unilateral renal agenesis and ambiguous genitalia were accompanied. Additional system abnormalities were found in 2/3 of the cases [3]. Commonly seen additional system abnormalities are urinary and gastrointestinal system abnormalities and rarer than these, it is possible to see outer genital area malformations, abdominal wall deformities, and neural tube defects [3, 4, and 5].

A seventeen years old ischiopagus case which was reported by Gokcen in 2015 was operated successfully [5]. The success of the surgery were due to absence of accompanying gastrointestinal or genitourinary system abnormalities. It is important to keep in mind that this patient population requires multidisciplinary approach.

In our case, because of the early diagnosis and presented additional congenital abnormalities, family decided to terminate the pregnancy. It is thought that congenital cardiac abnormality in the previous pregnancy and psychological atmosphere due to the abnormalities in the current pregnancy effected the decision of the family.

Knowledge and experience are limited regarding the heteropagus management due to limited number of case reports. More radical and brave management techniques may be described with increasing new cases.

References

1. Spencer R. Parasitic conjoined twins: external, internal (fetuses in fetu and teratomas), and detached (acardiacs). ClinAnat2001;14:428–444
2. Sharma G, Mobin SS, Lypka M, Urata M. Heteropagus (parasitic) twins:a review. J PediatrSurg 2010; 45:2454–2463.
3. Temesgen M. Tail-likeCongenital Duplication of Lower Extremity (Extra Legor Vestigial Parasitic Twin). Ethiop J HealthSci.2017;28(1):103.
4. StahrN, GuggenbergerR, Kellenberger CJ, Wisser J, and Subotic U. In utero and postnatal imaging finding of parasitic conjoined twins (ischiopagusparasiticustetrapus). Pediatric. Radiol. 2015;45:767–770.
5. Gokcen EC, and Wamisho BL. Delayed presentation of a heteropagus (parasitic) twin: a case report of a 17-year-old patient. J. Pediatr. Orthop. 2015;B 24:567–572.