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Discordant Anencephaly in a Set of Triplets

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Abstract. A 38-year-old woman, gravida 5, para 3, with a triplet prgnancy discordant for anencephaly, is reported. The diagnosis was made prenatally by ultrasound. Autopsy of the anencephalic member revealed small adrenal glands with absent fetal cortex, and a coincidental tetralogy of Fallot. Studies of the histocompatible lymphocyte antigens of both parents were performed and they failed to demonstrate HLA B-27. The literature on anencephaly in triplets is summarized. The association of congenital heart disease and HLA B-27 with anencephaly is discussed.

Key words: Triplets, Anencephaly, Adrenal hypoplasia, Tetralogy of Fallot, HLA B-27

INTRODUCTION

Neural tube defects are the second most common congenital anomaly in the United States, the most common being congenital heart disease. The incidence of triplets is approximately 1 in 7000 births. Therefore, the expected incidence of anencephaly in a triplet gestation is extremely rare. This is the fourth case report of anencephaly in a set of triplets.

CASE REPORT

The patient was a 38-year-old white woman gravida 5, para 3, abortus 1. She had three term normal deliveries and one elective termination after genetic amniocentesis revealed a Klinefelter's syndrome. She had taken no medications except prenatal vitamins. She had a strong family history of insulin-dependent diabetes, but no history of congenital anomalies. Ultrasonic assessment at 8 weeks gestation in her referring doctor's office revealed two definite gestational sacs and a possible third. A repeat level-II study done at 24 weeks gestation showed triplets with one anencephalic fetus. Her pregnancy proceeded unevent-

fully until 32 weeks. There was no associated polyhydramnios, pregnancy induced hypertension or anemia. At 32 weeks, she was admitted to our hospital in premature labor. Cervical dilatation was 5 cm on arrival and no attempt was made to stop labor. Ultrasound demonstrated that triplets I and II were vertex and triplet III (the anencephalic member) was breech. Triplets I and II were born spontaneously 3 minutes apart. They were both female, weighed 1660 and 960 g, respectively, and Apgar scores were both 9. The anencephalic triplet III, also female, was delivered by breech extraction and weighed 1022 g. Two separate placentas were released spontaneously.

Monozygosity of triplets II and III was supported by placental examination. This demonstrated a separate placenta for triplet I, weighing 330 g, with a dichorionic dividing membrane between it and the fused placenta for triplets II and III. This was a diamniotic monochorionic placenta that weighed 600 g and presented minor vascular anastomoses. Bloodgroup examination also supported strong evidence for monozygosity of triplets II and III.

Autopsy of the anencephalic member demonstrated the classic features of anencephaly with absence of the cranial vault and protruding eyes. The combined weights of the adrenal glands was 0.5 g (normal being 5 g) and microscopic examination revealed absence of fetal cortex. In addition, there was a major congenital heart anomaly, tetralogy of Fallot. The lungs were hypoplastic and, on microscopy, demonstrated marked intraalveolar hemorrhage.

The mother's post partum course was uneventful. Because of the strong family history of diabetes, hemoglobin A_1C was drawn but was in the normal range. The patient and her husband had HLA typing and both failed to demonstrate a HLA B-27 locus. Antigens could not be determined from the anencephalic member due to extremely poor cell viability from samples of lymph nodes and spleen taken 16 hours post mortem. Unfortunately, a heparinized sample of cord blood suitable for HLA determination was not obtained.

DISCUSSION

There are numerous case reports of anencephaly in twins [5, 7]. In contrast, anencephaly in triplets has only been reported three times. In 1966, Scott and Paterson [10] reported a case of concordant anencephalic monozygotic triplets. Discordant anencephaly in triplets was described by Kohler and MacDonald in 1972 [8] and another case was reported in a Pergonal-induced triplet pregnancy by Greenberg et al in 1981 [6].

Kohler and MacDonald [8] reported that the adrenal glands of the anencephalic triplet had a combined weight of 1.8 g well in excess of the usual in anencephalics and microscopy demonstrated an appreciable fetal zone. It was postulated that this may have been caused by ACTH stimulation received from the normal synchorionic cotriplet. Our case did not demonstrate any fetal zone.

Congenital malformations associated with anencephaly have been well described. David and Nixon [3] reviewed 294 cases of anencephaly and found that 41% of cases had other malformations. Seventeen of the 294 cases had cardiovascular system defects, 3 with tetralogy of Fallot.

Attempts have been made to correlate the histocompatibility locus with neural tube defects. Results are conflicting. Pietrzyk and Turowski [9] have shown a relationship between HLA B-27 and spina bifida, and the study of Amos et al [1] is in agreement with this. Other investigators [2, 4] have failed to demonstrate a linkage between HLA type

and spina bifida. In a report of twins concordant for anencephaly [5], HLA typing of both parents failed to demonstrate HLA B-27 locus in either. This was also our experience. At the present time, it seems unlikely that HLA typing will be recommended to identify high-risk families for genetic counseling.

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