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Case Report

Acephaly, an Extremely Rare Malformation: First Case in Morocco

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Abstract

Introduction: The total absence of a head, or acephaly, is one of the severe prenatal abnormalities that cannot support life.

Decapitation by the amniotic bands has often been suggested as the most likely cause.

Presentation of Case: In this uncommon case study, a 26-year-old Moroccan woman gives birth to a headless male fetus at 20 weeks of pregnancy due to acephaly, an incredibly rare congenital condition.

Discussion: This case study explores many theories on the causes of acephaly, such as the function of amniotic bands and other variables. Even though this syndrome is rare, managing serious fetal malformations in the early stages of discovery depends on knowing its cause.

Conclusion: Comprehensive prenatal screening and diagnosis tools are crucial, and the report emphasizes the need for additional research to clarify the underlying etiology of acephaly and related abnormalities.

Keywords: *Acephaly; Decapitation; Amniotic Bands*

Introduction

Central nervous system (CNS) malformations constitute a wide range of congenital anomalies. Almost all of these malformations result in severe and irreversible neurological defects, and many cannot be kept alive [1].

Two possible mechanisms have been suggested for the total absence of the fetal head: reverse arterial perfusion of twins in monozygotic twin pregnancies and amniotic bands in singleton pregnancies. To date, 8 cases of acephaly in singletons - without other malformations - have been reported [2].

We report the case of a 26-year-old female patient who gave birth at 20 weeks of amenorrhea to a male fetus without a head, i.e. acephaly. The objectives of this case report were to present a new case of acephaly, the first case to our knowledge in Morocco, to discuss the role of amniotic bands and alternative mechanisms in the light of our results and those of the published literature, and to discuss a possible sequence of events that leads to this very rare type of perturbation. The work has been reported in line with the SCARE criteria [3].

Case Report

The case in question represented the second pregnancy of a 26-year-old woman who had a healthy boy who was operated on by caesarean section for the indication of a surgical pelvis. Her family history did not reveal any malformations. The couple were not consanguineous and the current pregnancy was unremarkable, with the exception of spotting in the 08th week and anaemia, for which iron was recommended. 13-week transabdominal ultrasound (US) showed a live foetus with normal femur length and abdominal circumference. Its biparietal diameter could not be measured and anencephaly was suspected. On a second ultrasound two weeks later, the cephalic pole could not be visualised. The amniotic fluid was normal. A third ultrasound at 18 weeks confirmed the presence of a live foetus with a complete absence of the head, which, despite a thorough search of the amniotic cavity, could not be detected.

At 20 weeks of amenorrhoea, the patient was consulted for dark bleeding with abdominal pain of the type of contracture. In view of the persistence of bleeding with thrombocytopenia 56000, the patient was operated on for suspicion of retro placental hematoma at birth. The head and part of the neck were absent, and the neck showed a clear scar at the cephalic end, covered by a thin translucent membrane and surrounded by skin (Figure 1).

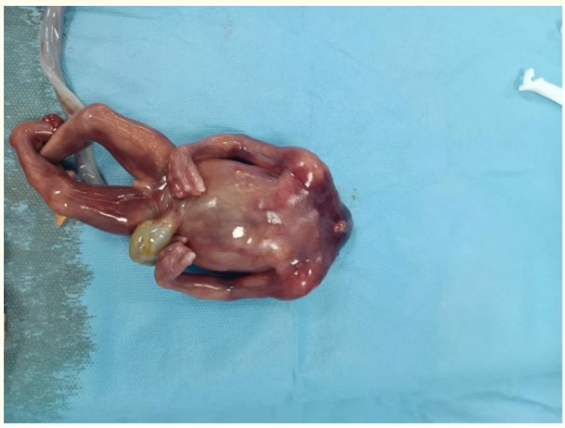

Figure 1: *Male foetus with normal upper limbs and body. The head and part of the neck are missing.*

Discussion

Congenital malformations of the central nervous system (CNS) are among the most common abnormalities, however data on the incidence of CNS malformations in autopsy populations is scarce. Pinar., *et al*. reviewed 4122 autopsies performed between 1958 and 1995. They found 363 cases (8.8%) of CNS malformations; 3% consisted of acephaly in four cases of twin reverse arterial perfusion (TRAP) [1].

Despite the fact that amniotic bands were only seen in one of the reported cases, decapitation by amniotic bands was considered to be the most probable mechanism leading to this type of acephaly. On the other hand, club feet were seen in all foetuses, including our own.

Cloven feet are often found in foetuses with amniotic band malformations and other conditions characterised by restriction of foetal movement, such as Moebius syndrome, which is caused by damage to the brainstem [4].

Acephaly, an extremely rare anomaly, is thought to occur mainly in complicated monochorionic-monoamniotic twin pregnancies, in which the twins' reversed arterial perfusion sequence (TRAP) occurs in the presence of acardia. In such cases, one twin is usually normal

and, with the help of in utero interventions, may survive [5]. However, the other twin presents with acephaly and acardia. Our case differs from the others in that it was a single pregnancy. In addition, the absence of chromosomal disorders in the parents and other foetal anomalies on post-mortem examination led us to suspect that environmental factors, rather than genetic problems, were the cause.

Amniotic band syndrome (ABS), which can cause many malformations of the head, trunk and extremities of the foetus due to mechanical pressure, is the primary etiological factor to be considered in the development of acephaly [6].

Amniotic band syndrome, which generally occurs sporadically, is rarely the consequence of exposure to teratogens or hereditary connective tissue disorders (Ehlers-Danlos type IV syndrome, osteogenesis imperfecta, etc.) [7].

US is more useful in assessing fetal anomalies due to ABS than ABS itself. The literature shows that it is not always possible to detect amniotic bands in cases of ABS [8]. US scans for fetal anomalies are usually performed in the second trimester, between the 18th and 22nd gestational weeks. In order to detect serious anomalies earlier and interrupt the pregnancy, if necessary [9].

At US at 13 weeks, in some cases [10,11] the head had already been decapitated and, when this was noted, the cervical organs were normal except in one case, indicating that the decapitation had occurred after the embryonic period. It can therefore be presumed that the event began at 8 weeks gestation and that the time taken to amputate the head was 1 to 2 weeks. In the most cases, there was no trace of the foetal head, and although it may have been overlooked, this indicates that it resolved over a period of between 1 and 2 weeks.

	Swinburne [12]				**Shipp and colleagues [13]**	**Unsal and Colleagues [10]**	**Haider and Colleagues [11]**	**Mazzitelli and colleagues [2]**	**Present case**
	1	**2**	**3**	**4**					
Status at delivery	Alive for 20 minutes	Prenatal death	Alive for 10 minutes	Prenatal death	Termination	Termination	Termination	Termination	Retroplacental hematoma
Gestational age, weeks	37	40	NS	30	15	13	13	21	20
Weight, g	950	2000	200	750	NS	NS	NS	290	350
Level of decapitation	Above C4	Above C1	Above C2	Above C2	Several cervical vertebrae visible	Above C2	Above C3	Above C1	Above C3
Placenta, cord, membranes	NS	NS	NS	Ragged membranes; placenta with small, scarred areas	NS	NS	NS	Short and undercoiled umbilical cord	Short umbilical cord
Fetal head identified	No	No	No	No	Yes, after delivery	No	Yes, on US	No	No

Table 1: *Characteristics of fetuses with acephaly.*

While most people think that amniotic band amputation is the most likely cause of acephaly, there is little evidence to support such a straightforward process. Diagnoses of "amniotic band sequence" (ABS) that, despite careful examination of the fetus and membranes, show no discernible evidence of bands are not unusual. There have been arguments that the bands are not always visible and that the placenta should be submerged in water to enable the bands to float and become visible, in accordance with Torpin's experiment [14].

Conclusion

In conclusion, several theories regarding a shared underlying cause that simultaneously causes amniotic rupture and fetal malformations-with or without band formation-have been proposed. It appears beyond question that acephaly results from a disruptive event. To establish the underlying etiopathogenesis of amniotic bands and demonstrate their causal role in this and other amputation-related disorders, more research is necessary.

Fetal anomaly scanning emphasizes the importance of a first trimester US examination between the 11th and 14th weeks for double test in order to identify severe anomalies early and end the pregnancy when necessary.

Conflict of Interest

The authors declare no conflict of interest.

Author Contributions

All authors contributed to the conduct of this work. All authors also declare that they have read and approved the final version of the manuscript.

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