JOURNAL OF THE ANATOMICAL SOCIETY OF INDIA 63 (2014) S39–S42

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Case Report

# Non-immune hydrops foetalis in combination with hydrocephalus and epidural haematoma: A rare togetherness

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ARTICLE INFO

*Article history:*
Received 27 January 2014
Accepted 9 May 2014
Available online 3 June 2014

*Keywords:*
Non-immune hydrops foetalis
Hydrocephalus
Epidural haematoma
Foetal anaemia

ABSTRACT

Non-immune hydrops foetalis is a serious disorder characterized by abnormal fluid accumulation in two or more foetal serous compartments together with generalized soft tissue oedema. It is a consequence of aberrant fluid homeostasis and prenatal cardiac failure, usually resulting from foetal anaemia caused by a variety of factors. We report one such case of a male infant aborted at 23 weeks of gestation which showed classical signs of hydrops scalp oedema, pleural effusion, ascites. In addition, hydrocephalus (enlarged head) and epidural haematomas were also observed. Hypertelorism and low set ears were the facial abnormalities noted. To the best of our knowledge, this is the first case of its kind to be reported with all the three conditions (non-immune hydrops foetalis, hydrocephalus, epidural haematoma) associated together. This coexistence is best explained by the common pathway of intracranial haemorrhage and the development of anaemia.



## 1. Introduction

Non-immune hydrops foetalis is a serious clinical condition characterized by accumulation of excessive abnormal fluid in two or more foetal compartments – skin/foetal subcutaneous tissue, pleura (pleural effusion), pericardium (pericardial effusion), peritoneum (ascites), amniotic fluid (hydrops amnion); frequently resulting in death either in-utero or shortly after birth. It represents the prenatal form of terminal stage of cardiac failure and aberrant fluid homeostasis resulting from a variety of maternal, foetal and placental causes.[1,2]

Apart from idiopathic causes, most of the cases are attributed to the following reasons: Cardiovascular pathologies inclusive of congenital cardiac defects and functional

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http://dx.doi.org/10.1016/j.jasi.2014.05.003

cardio-pulmonary anomalies (30%); Chromosomal anomalies (20%) – turner's syndrome, trisomy 21, etc.; Anaemias (15%); Malformation syndromes (10%) involving pulmonary (congenital cystic adenoid malformation- most common), renal, gastrointestinal, neurological or skeletal systems; Infections (10%) – toxoplasmosis, rubella, cytomegalovirus, herpes simplex, syphilis, parvovirus B19; Inborn errors of metabolism (5%) – glycogen storage disorders, mucopolysaccharidosis, Niemann–Pick's disease, etc.; Liver diseases (5%) and Others (5%) such as placental chorangioma, twin to twin transfusion in foetus, etc.[3–5]

The pathophysiologic mechanism responsible in majority of the cases is development of aberrant fluid homeostasis, which usually stems from foetal anaemia leading to hypoxia and high output cardiac failure. The severity of hydrops is related to the level of anaemia and the degree of reduction in serum albumin (oncotic pressure), which is partly due to hepatic dysfunction. Alternatively, heart failure may decrease right heart pressure, with subsequent development of oedema and ascites. Failure to initiate spontaneous effective ventilation because of pulmonary oedema or bilateral pleural effusions results in birth asphyxia or severe respiratory distress.[1,4]

## 2. Case report

An unbooked patient, a 25-year-old woman with 23 weeks of gestation (Gravida 4, Para 2, Abortion 1, Living 1) was admitted with complaints of bleeding per vagina. The obstetric history revealed nothing significant and the patient denied any medicinal intake or radiation exposure. There was no history of diabetes, pre-eclampsia or consanguinity (of any degree) in marriage. Also, there was no family history suggestive of any metabolic disorder. The important findings of ultrasound examination were: absence of foetal heart rate and movements, pleural effusion, ascites, umbilical herniation and positive spalding sign (overriding of foetal cranial bones). The blood group of the mother was O-positive. Condition of the foetus was explained to the mother and pregnancy was terminated with spontaneous expulsion of abortus and complete removal of placenta and membranes. The chromosomal analysis tests were refused by the parents due to financial constraints. After taking parental consent, the male foetus weighing about 450 g was sent for autopsy to the department of Anatomy, Pt. B.D. Sharma Postgraduate Institute of Medical Sciences, Rohtak. Gross abnormalities in foetus were recorded and photographs were taken. The foetus had a large head and distended abdomen. The skin and scalp were oedematous. Hypertelorism and low set ears were the facial abnormalities noted. There were no obvious signs of any skeletal deformities. No abnormalities like brachydactyly, clenched fists, club foot, contracture deformities, etc. were recorded in the limbs. There was no evidence of any neural tube defect like spina bifida or myelomeningocoele (Fig. 1).

Dissection of the foetus was done with a midline incision. The right lung was shrunken when compared to the left lung. Bilateral pleural effusion was present and the effusion on the left side was haemorrhagic in nature. The heart, great vessels and pericardium were normal. Pericardial effusion was not seen. The diaphragm was normal. All the viscera i.e. stomach, liver, pancreas, spleen, large intestine, small intestine, kidney, urinary bladder and testis were normal in shape, size and anatomical location. There was presence of free fluid in the peritoneal cavity (ascites) (Fig. 2).

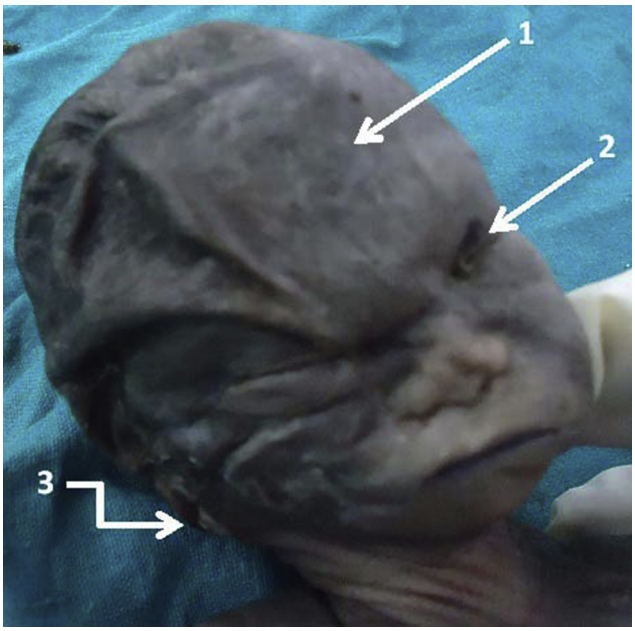


**Fig. 1 – Facial dysmorphic features of the foetus. 1: Enlarged head (hydrocephalus), 2: Hypertelorism, 3: Low set ears.**

The skull was opened using a mid-sagittal incision. Bilateral epidural haematomas (collections of blood between dura

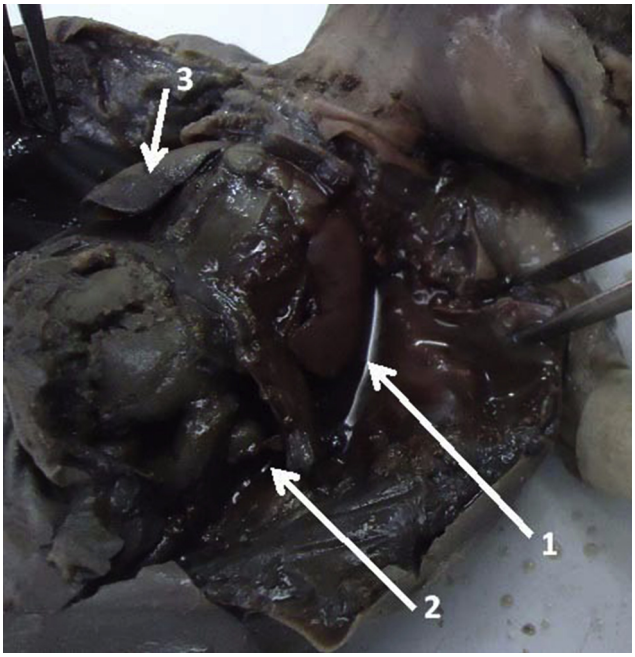


**Fig. 2 – Findings on dissection of the foetus using an anterior midline incision. 1: Haemorrhagic pleural effusion, 2: Ascites, 3: Shrunken right lung.**

mater and skull) were found. No skull fracture was seen. The extents, attachments and folds of dura mater were normal. Mid-sagittal hemisection of the brain showed no gross abnormality in the ventricles or any other part of the brain. No signs of intracranial haemorrhage were present (Fig. 3).

## 3. Discussion

Non-immune hydrops foetalis presenting before 24 weeks is usually due to chromosomal aberrations while that presenting after this is mostly attributed to structural anomalies (such as cardiac and pulmonary).[4] Autopsy plays an important role in determining a definite cause in majority of cases facilitating genetic counseling and prediction of recurrence. It also highlights important associated anomalies, which can aid in better management of the cases. Likewise in the case reported here by us, hydrocephalus and epidural haematoma were the associated anomalies observed.

The epidural bleed or haematoma is derived either from branches of middle meningeal artery or from dural venous sinuses.[6] The hydrocephalus developed during foetal life which manifests subsequently is termed as congenital hydrocephalus. Hydrocephalus resulting from obliteration of the subarachnoid cisterns in brain or malfunction of the arachnoid villi is known as communicating or non-obstructive hydrocephalus, the most common cause being subarachnoid haemorrhage. It can also result from meningitis, malignancies, arterio-venous malformations, etc. Whereas, non-communicating or obstructive hydrocephalus results from obstruction within the ventricular system of brain. Its multifactorial etiology includes aqueductal stenosis (mostly due to infections or intracranial haemorrhage), Arnold–Chiari malformation, Klippel–Fiel syndrome, Dandy–Walker syndrome, tumours, etc.[7]

The case reported here by us is of non-immune hydrops and not that of immune hydrops because the mother's blood group was Rh-positive, hence no chance of development of anti-D antibodies. The most suitable explanation for the coexistence of hydrops and hydrocephalus is an intracranial haemorrhage leading to anaemia. The development of anaemia explains the symptoms of hydrops (pleural effusion, ascites) whereas intracranial haemorrhage explains the presence of hydrocephalus. Cases illustrating the coexistence of non-immune hydrops foetalis with other associated anomalies have been scarcely reported in literature. Caulson CC et al[8] reported a case with non-immune hydrops and hydrocephalus secondary to foetal intracranial haemorrhage and cited development of anaemia as the common etiology, similar to ours. Among the other causes which can simultaneously lead to both the conditions, the most likely ones are chromosomal abnormalities. A case of antenatally diagnosed Turner's syndrome reported by Kalpatthi et al[9] was associated with hydrocephalus, hydrops foetalis, cystic hygroma and hypoplastic left heart syndrome. A similar case was also reported by Othman ARM et al[10] in which ultrasonographic examinations of the 28 week old foetus showed multiple congenital anomalies: severe hydrocephalus, scalp oedema, pleural effusion, bilateral clenched fists with overlapping fingers, bilateral clubfoot, markedly decreased amniotic fluid and facial dysmorphic feature such as low set ears, micrognathia, hypertelorism and downslanting palpebral fissures. Since the parents did not agree with karyotyping the foetus, the differential diagnosis of the authors included trisomy 18, arthrogryposis and akinesia sequence. But in our case, the gross examination of the foetus did not reveal the typical features of such abnormalities. Only hypertelorism and low set ears were noted, which are not adequate to label it as a case of any specific chromosomal abnormality. Moreover, there was neither any history of risk factors like increased maternal or paternal ages, radiation or drug exposures, consanguinity in marriage; nor any family history of inherited genetic or chromosomal disorders. Further confirmation using karyotyping or chromosomal analysis for gene mutations could not be done as the parents refused for these tests due to financial constraints. Intrauterine infections, especially "TORCHS" group i.e. toxoplasmosis, rubella, cytomegalovirus, herpes simplex, syphilis; also figure in the common etiology list of non-immune hydrops and hydrocephalus. Many authors like Ergunay et al[11] have even reported Parvovirus B19 as an important causative agent of non-immune hydrops. Bonnefoy et al[12] reported a case of extradural haematoma diagnosed in-utero associated with hydrocephalus, but no signs of intracranial haemorrhage or hydrops foetalis were present. The blood sample of the mother tested positive for pertussis infection. In the present case, there was no history of any antenatal check-up or maternal vaccination, thereby increasing the chances of intrauterine infection. Macdermot et al[13] have reported a case of hydrops foetalis associated with communicating hydrocephalus, osteopenia and abnormal dentition. The facial abnormalities observed in the infant were hypertelorism, orbital hypoplasia without proptosis, brachydactyly, frontal and temporal bossing of skull. The signs of connective tissue disorder manifested as pathological fracture, yellowish discolouration of teeth, blue sclera and easy bruising. However, facts such as non-consanguineous marriage, insignificant family history and autopsy findings like absence of hepatosplenomegaly, renal anomalies, contracture deformities, skeletal dysplasias etc. minimize the possibility of metabolic disorders as an etiology in the case reported by us.

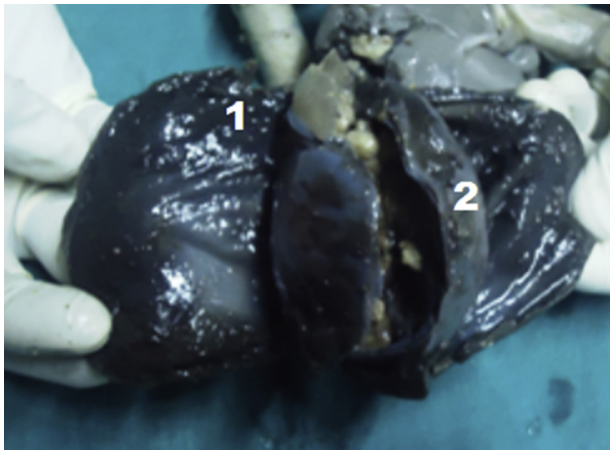


**Fig. 3 – Findings on dissection of scalp and brain. 1: Epidural haematoma, 2: Dura mater.**

The presence of epidural haematoma in a hydropic foetus with hydrocephalus as in our case is extremely rare and has not been reported yet in literature, according to the best of our knowledge. The differential diagnosis regarding the cause of the disease in the present case accounting best for the coexistence of these three clinical conditions together, includes intrauterine infections and development of anaemia secondary to intracranial haemorrhage.

## Conflicts of interest

All authors have none to declare.

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