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CASE REPORT

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UNUSUAL RISK FACTORS ASSOCIATED WITH CONGENITAL HYDROCEPHALUS: CASE SERIES OF 3 PATIENTS

1) SYED ASKARI HASAN
2) SHAYAN UL HAQ ABBASI
3) SHOUKAT ALI BAIG
4) SYED MANSUR –UL –HAQ
5) SHAZIA JABBAR

1) 3rd year Student, Dow Medical College
2) 3rd year Student, Dow Medical College
3) 3rd year Student, Dow Medical College
4) 3rd year Student, Dow Medical College
5) Department of Gynaecology and Obstretics, Unit I, Civil Hospital Karachi.

Correspondence:
Syed Askari Hasan
Address: A-19 Al-Azam Appartments, Gulshane Iqbal 13-A, Karachi, Pakistan
Email: askari_hasan2004@hotmail.com
Phone: 9221 3343146540

ABSTRACT

Many risk factors has known to be associated with congenital hydrocephalus although some like advancing maternal age, maternal diseases e.g. diabetes and hypertension, and alcoholism have received more attention than others. Our case series shows that three factor that are not in such a limelight have been consistent in the three cases we present. They are namely multiparous pregnancies, anemia and a possibility of this condition being more common in male gender. The mothers in the three cases were multigravida 5, 7 and 8 respectively. Complete Blood Count (CBC) report of the mothers revealed anemia 8.9 g/dl, 9.7g/dl and 10.1 g/dl respectively. The sex of the neonates is male which brings out a possibility of thus condition being linked to male gender. This case series highlights the three factors which might be important in causing congenital hydrocephalus and therefore sets a platform for further research in this field.

Key words: congenital hydrocephalus, multiparous, multigravida.

INTRODUCTION

Hydrocephalus can lead to a variety of disabling symptoms that can have disabling effect on intelligent quotient, gait, cognition, balance and urinary incontinence. It is characterized by an abnormal increase in the volume of the cerebrospinal fluid within the skull, resulting in raised intracranial pressure.[1] Congenital hydrocephalous, in most cases, in the newborn is due to an obstruction of aqueduct of sylvius (adequectal stenosis).[2]

The most common clinical manifestation is size of head, with wide anterior fontanelle.[3] Apart from physical abnormality it also effects the mental debilitation as the Performance IQ is low and learning problems are common in children represented with this anomaly.[3]

A number of risk factors are involved. Maternal health is very critical to the developing fetus hence maternal diseases like diabetes, chronic hypertension and hypertension during gestation[4] are found to be associated with increased risk of fetus developing hydrocephalus. Alcoholism once considered the most common cause for mental deficiency in fetus is also responsible for development of CH in fetus [1, 4, 5]. Obesity also a possible factor needed to confirm whether maternal overweight is also implicated but further studies are needed to confirm the fact.[6] Oral contraceptives[7] and drugs[8] in the first trimester also lead to increased risk. Early marriages will results in multiparous pregnancies and a study confirms that this also related to increased risk of developing CH [4].Apart from physical factors ,there is also a familial trend to CH [4, 9, 10] and a possibility of being X linked.[14] Congenital hydrocephalus is amongst the top 12 congenital anomalies recognized in Philippines.[14]

CASE 1

A 30 Years old female patient with blood group B +ve was admitted in Gynecology and Obstetrics Unit I. Her obstetrics history revealed that her gravida was 5 with parity of 2+2. Family and personal histories were normal and did not reveal any significant information. Ultrasonography was performed twice during her pregnancy which progressively showed dilated ventricles as suspicious of hydrocephalus. The patient misplaced the report and now she presented at 32 weeks of pregnancy at our outpatient department.

She was not a book case from her first month. She got booked around seventh month of her gestation. She was not taking folic acid tabelets.

Blood and urine test were done which clearly shows anemia of Hb level of 8.9 gm/dl. Following tests were recommended CBC, RBS, Urine DR, LFTs, PT, APTT, UCE and these report no significant results. 4 pints of blood were transfused. All other systems were unremarkable.

The patient had emergency lower segmental caesarian section (EmLSCS) because of increase bi-parietal diameter which raises the suspicion that the baby will not deliver vaginally because of large head and small pelvis. The baby boy was born with enlarged size head, bulging anterior fontanelle. Meningo-mylocele and talipes equinovarus of both feet, head drain was performed in lithotomic position and he was immediately transferred to National Institute of Child Health. At birth, the hearth rate was 120 beats /min and respiratory rate is 48 / min APGAR score is 6.

Patient was then discharged as advised was given to get herself booked as soon as the pregnancy is confirmed. Post-operative medications were given that included tablet Flagyl, Postan Forte. Parhodel, Qalsan.

CASE 2

A 25 year old woman came to the hospital complain of labor pain. She was admitted to the gynecology department. Her ultrasound test showed that she was pregnant with a male hydrocephalic baby (larger than normal head).the baby BPD was 13.1 (suggestive of hydrocephalusas shown in fig 1).at the 30th week which increased to 13.8 at the 38th week.. The FL length was 13.6cm in 30th week and CRL was 38.6 in the 38th week. Her obstetrics history revealed that she was 7TH multigravida with parity 6+0.the last child was born two and a half years back.

Her hemoglobin level was 9.7g% (normal 11.5-16.4g %) indicating that she was anemic. Her blood group is A+. Glucose random taken at the 30th week showed a reading of 87mg/dl (70-200 normal). It was on the lower side (didn't get what was on the lower side). She was also suggested to take folic acid supplements. The urine test showed traces of albumin and neutrophils levels were raised indicating of some infection. The mother was advised not to deliver baby vaginally although the presentation is cephalic but lower segmental cesarean section is needed due to increase cephalo-pelvic disproportion (CPD). She gave birth to a baby boy of 4 kg at term. The APGAR score was 10; with heart rate 130 beats /min and respiratory rate of 26/min

The apgar score was 10. The baby was boy full-term with 4.0kg weight. 300cc of the fluid was drained after birth using spinal needle and for sent for spinal fluid examination. The report was unremarkable.



Fig.1:
BPD of 13.1 at 30th week which is suggestive of hydrocephalus.(case 2)

CASE 3

This case involves a 23yr old pregnant woman who was admitted through OPD to Gynecology ward because she was 37 (+2) gestational week gravida 8 Para 7. Her prenatal course was eventful, according to the patient she had normal menstrual cycle. Since her last menstrual period she developed nausea and vomiting. Her pregnancy was confirmed by urine pregnancy test. She felt fetal movement at 5th month of gestation. No signs of ATN, DM, P/V bleeding in 2nd and 3rd trimester. She visited Civil Hospital Karachi (CHK) at 38 weeks.

After admission, the patient underwent physical and obstetrical examinations. Her heart rate was 86/min, Blood Pressure 110/60mm of Hg, Respiration rate 18/min. Her appetite, sleep, micturition were normal and wasn't addicted to any drug or chemical abuse. She was diagnosed with polyhrdroamniosis. She was requested Ultrasound, Liver Function Tests, Hematology, Activated Partial Thromboplastintime, Prothrombin time (PT), Serum electrolyte test.

After Admission laboratory tests showed hemoglobin 10.10gm/dl (reference range 11.5 to 16gm/dl); RBC count 4.20Mil./µL (reference range 3.7 to 5.7µL); Mean corpuscular hemoglobin 24.30 Pg(reference range28 to 32Pg); Total Leucocyte count 10000/µL (reference range 4000 to 11000/µL); RBC morphology was hypochromic; Activated partial thromboplatin time was 31.0 sec (control 26 sec); PT was 10.0 sec (control 11sec); Total bilirubin, ALT, Urea, Creatinine and Random blood glucose level were all under normal ranges; Serum electrolytes were also found to be in normal range.

Next day after hospitalization she underwent assisted beach vaginal delivery (ABDV) after administering epidural anesthesia. The outcome was IUS female delivered by ABDV.

The baby was diagnosed as having Hydrocephalus. Postpartum prescription included Flagyl 500mg, Amikain 500mg, Cac Tablets.

DISCUSSION

According to a research carried out in the Department of Gynecology and Obstetrics, Lyari General Hospital, during the period of January 2000 to October 2005, neural tube defect (NTD) was found to be the commonest (65.8%) type of anomaly. and anencephaly.[13] Among the most frequent NTD was hydrocephalus. Therefore the need to have an insight into the possible causative agents of hydrocephalus is required. While analyzing the three cases in our OPD some factors appeared common to all three and a suspicion that considering our socioeconomic structure these could be an important cause in eliciting the disease. The cases brought to us were multigravida with 5 , 7 and 8 respectively. In Pakistan early marriages is a common practice in rural areas and woman brought even in very young ages are multigravida. A very detailed retrospective study for 10 years revealed strong evidence between multigravida uterus and a tendency of having congenital hydrocephalus.[4]
A unique finding in this study was Anemia. In all three Cases mothers were severely anemic hence this new finding needs further prospective study.
Another factor similar to 3 cases was anemia. This also relates to our socioeconomic structure as woman in our rural areas are normally malnourished and anemia is therefore common. Our case reports shows a possible link between these two anemia and congenital hydrocephalus
Another factor which is striking is that the infants given birth are males and have features typical of that of a hydrocephalus presentation: enlarged size head, bulging anterior fontanelle. This shows more tendency in male gender and of possibly being X linked. [3, 1, 12].

REFERENCES

1. Snell RS: Clinical neuroanatomy. 7th edition. Philadelphia: Wolters Kluwer Lippincott Williams & Wilkins; 2010.
2. Sadler TW, Langman J: Langman's medical embryology. 10th edition. Philadelphia: Lippincott Williams & Wilkins; 2006.
3. Rizvi R, Anjum Q: Hydrocephalus in children. J Pak Med Assoc 2005, 55:502-507.
4. Van Landingham M, Nguyen TV, Roberts A, Parent AD, Zhang J: Risk factors of congenital hydrocephalus: a 10 year retrospective study. J Neurol Neurosurg Psychiatry 2009, 80:213-217.
5. Zhang J, Williams MA, Rigamonti D: Genetics of human hydrocephalus. J Neurol 2006, 253:1255-1266.
6. Stothard KJ, Tennant PW, Bell R, Rankin J: Maternal overweight and obesity and the risk of congenital anomalies: a systematic review and meta-analysis. JAMA 2009, 301:636-650.
7. Stoll C, Alembik Y, Dott B, Roth MP: An epidemiologic study of environmental and genetic factors in congenital hydrocephalus. Eur J Epidemiol 1992, 8:797-803.
8. Angerpointner TA, Pockrandt L, Schroer K: Catamnestic investigations in children with congenital hydrocephalus. Z Kinderchir 1990, 45:151-155.
9. Haverkamp F, Wolfle J, Aretz M, Kramer A, Hohmann B, Fahnenstich H, Zerres K: Congenital hydrocephalus internus and aqueduct stenosis: aetiology and implications for genetic counselling. Eur J Pediatr 1999, 158:474-478.
10. Adams C, Johnston WP, Nevin NC: Family study of congenital hydrocephalus. Dev Med Child Neurol 1982, 24:493-498.
11. Zlotogora J, Sagi M, Cohen T: Familial hydrocephalus of prenatal onset. Am J Med Genet 1994, 49:202-204.
12. Halliday J, Chow CW, Wallace D, Danks DM: X linked hydrocephalus: a survey of a 20 year period in Victoria, Australia. J Med Genet 1986, 23:23-31.
13. Perveen F, Tyyab S: Frequency and pattern of distribution of congenital anomalies in the newborn and associated maternal risk factors. J Coll Physicians Surg Pak 2007, 17:340-343.
14. Mossey PA, Castilla E, World Health Organization. Human Genetics Programme., World Health Organization. Management of Noncommunicable Diseases.: Global registry and database on craniofacial anomalies : report of a WHO Registry Meeting on Craniofacial Anomalies : Baurú, Brazil, 4-6 December 2001. Geneva, Switzerland: Human Genetics Programme, Management of Noncommunicable Diseases, World Health Organization; 2003.
15. Sipek A, Gregor V, Horacek J, Masatova D: [Congenital hydrocephalus 1961-2000—incidence, prenatal diagnosis and prevalence based on maternal age]. Ceska Gynekol 2002, 67:360-364.
16. Ingalls TH, Pugh TF, Macmahon B: Incidence of anencephalus, spina bifida, and hydrocephalus related to birth rank and maternal age. Br J Prev Soc Med 1954, 8:17-23.