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Case Report On 12-Year-Old Boy With Hemolytic Uremic Syndrome And Thalassemia Trait

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Abstract:
Haemolytic Uremic Syndrome (HUS) is an uncommon disease that mostly affects children under ten. Damage to the lining of blood channel walls and the destruction of red blood cells are common causes of kidney failure. Because injured red blood cells and other factors might clog the tiny blood capillaries in the kidneys and induce scarring, the child may generate little urine. The kidneys have to work harder to eliminate wastes and excess fluid from the bloodstream as a result of this. High blood pressure, swelling of the hands and feet, and fluid buildup can all be caused by the body's inability to eliminate excess fluid and waste (oedema). A 12-year-old male came in Paediatric department at tertiary care HospitalWardha, with a chief complaints of Headache, giddiness and scaling of skin, breathlessness, vomiting 3-4 episodes, as narrated by her mother. Now visit to tertiary care hospital for further treatment. After historycollection and physical examination and all necessary investigations were done, The doctor identified a case of Hemolytic Uremic Syndrome With Chronic Kidney Disease with a known case of beta Thalassemia Trait With Hypertension.inj. Meropenem 420 mg , ,Inj. Colistin, inj. Piptaz 2 g ,Inj. Fluconazole 150 mg ,inj. Dexa 5 mg iv,inj. Erythropoietin, inj. Iron sucrose 100, tab. Amlodipine 5 mg bd , tab. Clovidine 0.1 mg, tab. labetalol 100mg, Tab. Prazosin 5 mg, TDS, tab. Spironolactone 25 mg bd was given.Patient responded to both medicine and physician counselling. His general condition was improved.

Keywords: Haemolytic uremic syndrome, Chronic Kidney Disease, Thalassemia Trait, Hypertension, dialysis.

Introduction

Hemolytic uremic syndrome (HUS) is a disorder caused by damaged and inflamed tiny blood vessels in the kidneys. [1] Clot can form in the vessels as a result of this. The clots restrict the kidneys' filtration system, resulting in renal failure, which can be fatal.[2] In children, it is the most common cause of acute renal damage. Approximately 5% of children with HUS die during the acute phase of the disease, with early studies indicating a death rate of up to 21%.[3]The most common cause of acute kidney injury in children is a haemolytic uremic syndrome.[4]

Hypertension is a common complication of HUS and can be challenging to manage. Local renin-angiotensin activation is thought to have a role in thrombotic microangiopathy, which results in a vicious cycle of increasing renal damage and uncontrollable hypertension. [5]Beta thalassemia is a blood condition in which the amount of haemoglobin produced is reduced. The iron-containing protein haemoglobin is found in red blood cells and is responsible for transporting oxygen to all of the body's cells. Low levels of haemoglobin cause an oxygen shortage in numerous regions of the body in patients with beta-thalassemia.[6]

The type of thalassemia a person has is determined by the number and type of thalassemia characteristics they inherited from their parents. If a person inherits a beta-thalassemia trait from both his father and mother, he will develop a beta-thalassemia major.[7]
In the general population, HUS is predicted to affect 1-3 per 100,000 people.[8] The overall incidence of HUS in the United States is estimated to be 2.1 instances per 100,000 people per year, with a peak incidence between the ages of six months and four years.[9] The mortality rate in affluent countries is 2-4 percent. Acute renal failure associated with HUS causes only a small percentage of patients to die. [10]There are an estimated 270 million people worldwide who have defective haemoglobins and thalassemias, with 80 million of them carrying the -thalassemia gene. According to recent surveys, between 300,000 and 400,000 new-borns are born each year with a significant haemoglobin disease (23,300 with thalassemia major), with up to 90% of these births taking place in low- and middle-income countries.[11]

Patient-specific information:
A Case ofa 12-year-old male admitted in paediatric ICU attertiary care Hospital with the chief complaints of Headache, giddiness and scaling of skin since 15 days, breathlessness since 12 days, vomiting 3-4 episodes 11 days back narrated by his mother.

The primary concern and symptoms:
As narrated by mother, child was apparentlyalright, then child developed headache and giddiness and he was taken outside hospital and blood was transfused as per investigation low haemoglobin and it started emplacing of breathlessness and had 2-3 episodes of non-projective and non-foul swelling vomiting, and then the child was taken to Nagpur, and it was diagnosed with chronic kidney disease with ARDS. Hb 8.1 gm. %, WBC 10000, Platelet 0.79 Lack, Sodium 143, Potassium – 3.49, Urea 20, Creatine 6.60, 2 point of PRC was transfusion, 6 hours of dialysis was done. PS was positive for schistocytes with? Haemolytic uremic syndrome and hypertension were diagnosed and the child's parents were taken DAMA and brought here for further management of it.

Medical family and psychosocial history:
The child was a known case of Beta-thalassemia Trait since birth. His father is former, and her family belong to a middle-class family. His parents have a consanguineous marriage. His father and mother have also known cases of Beta-thalassemia Trait from birth, and his father also has a history of hypoplastic kidney. He maintains a good interpersonal relationship with his parents.
Birth History: The child was born with Full term normal vaginal delivery with birth weight was 2.5 kg, no history of any NICU tray, a started all milestones and immunization till date as per age.

Clinical findings:
On physical examination, the child was conscious, co-operative and well oriented to time, place and person.He looks anxious, depressed, dehydrated and febrile, heart rate were 110 beats /min, respiration were 22 breaths /min & blood pressure was 150/120 mmHg,oxygen saturation was 98%and thin body built, hygiene wasnot maintained properly. His weight was 28kg, and his height was 1.20m. abdominal girth were 65 cm , Facial puffiness present, scaling of the skin with xerosis present, abdomen examination – soft -abdominal distension and pain present umbilical eversion present, shifting dullness test positive and S1 and s2 present bilateral lung clear, breathlessness present.
Timeline:A child was admitted in hospital with the complaints ofHeadache, giddiness for 15 days, breathlessness since 12 days, vomiting 3-4 episodes 11 days back and high blood pressure.
Day 1: on admission Facial puffiness present, scaling of the skin with xerosis present for that doctor advised Imulac Moisturizing Lotion bd for 2 weeks,abdominal distension present and hypertension, cough with blood-tinged sputum present for that inj. Emset 3mg iv sos, inj. Pan 25vmg iv 24 hourly, inj. Ceftriaxone1.2 gm iv 12 hourly, tab. Amlodipine 5 mg bd, syp. Shelcal 5 ml bd, tab. Soda mint500 mg .qid ,syp. Bevon 5ml bd, syp. Sacral o 10 ml bd, syp. Mucaine gel 10ml TDS, inj. Lasix 5mg iv stat , tab.cloridine 1 +1/4 tab bd.
Day 4: Cough with blood-tinged sputum present, persistent hypertension BP 160/100mmhg
Day 5: 3rd cycle of haemodialysis done, hypertension present 160/130 mmHg for that doctor tab. Clovidive 0.1 mg 2 TB(15 mcg/kg/ day)bp reading between 90th- 95th centile, respiratory distension present with urine output on the lower side.
Day 17: Abdominal distension were present, tolerated haemodialysis well, BP 160/120mmhg.

Day 20: Cough present, Periorbital oedema present, tolerated Haemodialysis well, hypertension BP -172/140 mmHg, abdominal girth 71 cm,
Day 21: Abdominal pain present, abdominal distension, ascitic fluid cumm. S/O, TLC-110 cells.
Day 28: Bleed in ET, abdominal distension, BP -170/120 MMHG for that doctor sodium Nitroprusside 2-5 ml in 7.5 ml D_5 0.8 ML/hour, W/H vitamin D3 sacket 60,000iu 1/week and tab. Prednisolone 20 mg 1 tab. BD, deflate ET tube cuff 6 hourly X 10 min.
Day 30: Tolerated Haemodialysis well.
Day 32: Complaint of fever spikes, respiratory distress present, cough, periorbital oedema present, maintaining saturation on O_2 by CPAP, BP -146/110 MMHG

Diagnostic Assessment:
Complement component 3 123 mg/dl, C4 62.4 mg/dl , chloride 111 mmol/l increased, **KFT** - urea 289 mg/dl increased, creatinine 11.4 mg/dl increased , sodium 135 decreased , potassium 6 mmol/l ,LDH 465 U/L increased, **LFT** – ALT 66 U/L Increased , AST 39 U/L Increased, Total Protein 5.6 g/dl ,phosphorus 8.6 mg/dl increased,**urinary protein (random)** 430 mg/dl, **urinary protein 24 hrs** -990 mg/day increased ,haemoglobin 9gm.% decreased , RDW 20.6% Increased , HCT 31% decreased ,PLATELET 0.79 Lack,**Microbiology report urin** no RBC, 1-2 pus cell/HPF, organism seen**. Urin**e **Culture report**: growth of enterococci species. **Blood culture report** report show growth of pseudomonas aeruginosa , **urin examination-**pus cells 5-6cells/hpf, RBC 1-5 RBCs/Hpf ,epithelial cell 2-3 cells/HPF , urine albumin present ++, color **Doppler study of renal** : increase RI in both renal parenchyma secondary to Renal parenchyma disease ,increase echotexture of both kidney with loss of CMD, right kidney 6.5 X2.5 CM, left kidney 6.1 X 2.6 CM , **USG impression** – grade III RPD, Gross ascites, minimal left-sided pleural effusion hyper echoic kidney with CMD.
Diagnostic Challenges: No diagnostic Challenges were faced

Diagnosis: Haemolytic uremic syndrome with chronic kidney disease with hypertension with known caseof Beta-thalassemia Trait.

Prognosis: This child's prognosis was poor. The majority of children with HUS recover completely. However, some people will get long-term kidney impairment.

Therapeutic Intervention*:*
Medical treatment were provided to child, such as O2 by CPAP, oral liquids allowed inj. Meropenem 420 g in 20 ml NS over 2 Hours iv 12 hourly, (D8), Inj. Colistin is iv in 20 ml ns over 2 hours iv 12 hourly (D7), inj. Piptaz 2 g in20 ml D5 over 30 min iv 12 hourly (D12),Inj. Fluconazole 150 mg iv 48 hourly (D28) inj. Dexa 5 mg iv 8 hourly iv ,inj. Erythropoietin 5000unit in 2 times/week, inj. Iron sucrose 100 mg in 100 ml Normal saline over 2 hours IV 2 times/week, ta. Amlodipine 5 mg 1 tab bd , tab. Clonidine 0.1 mg 3.5 tans bd (2-2) , tab. labetalol 100mg 1/4th tab. QID, Tab. Prazosin 5 mg in 5 ml NS, give 2 ml TDS (7-3-11), tab. Spironolactone 25 mg 2 tab, in 5 ml NS give 4 ml bd , maintain intake and output chart, nebulization with adrenaline 4 hourly,strict BP Monitoring, PRC transfusion was given, platelet transfusion given., iv fluid was given, catheter care was given.

Challenges in therapeutic intervention:
The child has CKD and requires continuously dialysis every tri-weekly. He had a total of 12 dialysis and was intubated due to pulmonary oedema. The child needs moredialysis for a further stay. A Child condition improved slowly but his blood pressure was notunder controlled.

Discussion:
A 12-year-old male came in PediatricOPD in our tertiary care Hospital with a chief complaints of headache, giddiness and scaling of skin for 15 days, breathlessness since 12 days, vomiting 3-4 episodes 11 days back narrated by her mother. The outside hospital diagnosed HUS with CKD, Hypertension, and he was a known case of beta-thalassemia traitcame for our hospital for further management.[12-15]
HUS is best treated with supportive treatment such as blood transfusions and dialysis. Anyone can acquire HUS, although it is more frequent in children under five.[12]HUS is frequently caused by infection with particular strains of the bacterium Escherichia coli (E. coli). [13]Other infections, drugs, or situations such as pregnancy, cancer, or

autoimmune illness can all cause HUS.[14]the majority of children with acute renal damage only require dialysis for a short period of time.
In some analysis, the prevalence of hypertension was comparable to that found in the CKiD group. 5, S8 In contrast to the CKiD study, ours found that hypertension at the time of enrollment was not a significant risk factor for CKD development. Children with uncontrolled hypertension, on the other hand, had a higher risk of CKD progression over time. [13-16]
The ESCAPE study found that strict blood pressure management was linked to a lower risk of CKD development, highlighting the importance of blood pressure control in juvenile CKD. [16-23]

Conclusion:
Low red blood cells, acute kidney failure, and low platelets counts are all symptoms of HUS, a category of blood disorders. Bloody diarrhoea, fever, vomiting, and weakness are the most common early symptoms. Children are more likely to be harmed, but most children recover without lasting health consequences, while some children may experience serious and sometimes life-threatening issues. In this case, a child has chronic kidney disease with Haemolytic uremic syndrome with a known case of Beta-thalassemia Trait with hypertension, and after taking treatment patient's condition was silently improved but his blood pressure was not controlled.

Informed consent: Before taking this case, information was given to the child, and their mother and informed consent was obtained from the child as well as the mother.
Conflict of Interest: No conflict of Interest
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