Pediatrics & Therapeutics
ISSN: 2161-0665

Kumar et al., Pediat Therapeut 2013, 3:5
DOI: 10.4172/2161-0665.1000176

Case Report Open Access

Atypical HUS with Dysphagia and Palatal Palsy Responding to IVIG

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Abstract

D negative/Atypical HUS (Hemolytic Uremic Syndrome) is a difficult condition to diagnose and treat. It has been shown to be a disease of complement dysregulation. Treatment is difficult and revolves around plasma exchange. We report one case of an Atypical HUS in a toddler girl who developed multi organ dysfunction associated with never reported dysphagia and palatal paralysis which responded to IVIG (Intravenous Immunoglobulin) given over five days

Keywords: Atypical HUS; Dysphagia; Palatal palsy; IVIG

Introduction

The classical triad of microangiopathic hemolytic anemia, renal failure and thrombocytopenia seen in Typical HUS may not present acutely and simultaneously in an atypical form. Atypical HUS may involve multi organ system. CNS manifestations comprising of irritability, encephalopathy and thrombotic stroke is seen in 20-50% of the cases [1]. We present one case of an Atypical HUS in a toddler girl with multi organ dysfunction and never reported manifestation of dysphagia and palatal paralysis. Atypical HUS main line of treatment is plasma exchange. Due to non availability of plasma exchange therapy our patient was managed with FFP (Fresh Frozen Plasma) and supportive treatment while her dysphagia and palatal paralysis responded to IVIG.

Case Report

2 yr old female toddler was admitted with a history of (a) high grade intermittent fever without chills and rigors 15 days duration, (b) swelling over whole body starting over face and periorbital area 7 days duration (c) decreased urine output 1 day duration. On admission patient weight was 10.5 kg (85% of 50 centile) and height was 85 cm (92% of 50 centile NCHS standard). She was irritable, febrile-100°F, Pulse 120/minute regular normal volume, Blood Pressure 120/98 mm Hg (more than 99 centile), Respiratory rate 30/minute. Pallor and edema (periorbital and limb) was present. Systemic examination revealed firm hepatomegaly with ascites. Patient was started on restricted iv fluids (insensible loss and urine output), intravenous antibiotics (cefoperazone @ 150 mg/kg/day in three divided doses), diuretics, 5% albumin (single dose @ 1 gm/kg) and antihypertensives (nifedipine @ 1.5 mg/kg/day, atenolol 0.5 mg/kg/day and finally prazosin @ 0.1 mg/kg/day) and furosemide @ 2 mg/kg/day. Initial investigations revealed hb 7.9 gm%, Total counts 10200/cmm Differential count polymorphs 60% Lymphocytes 35% eosinophils 4% monocytes 5% platelets 3.1 lac/cumm, urine routine/microscopic 7-8 pus cells/hpf RBC 15-17/HPF protein 2+ blood urea/serum creatinine 196/3.2 mg%, serum sodium /potassium 136/4.4 meq/dl, serum albumin 2.8 gm%, serum bilirubin 0.8 mg% SGOT/SGPT 192/103 IU/L, serum cholesterol 170 mg%, C3, C4 was normal, serum amylase was normal, ANA, dsDNA, ANCA, DCT, HbsAg, HIV and anti HCV were negative. USG abdomen showed bilateral medical renal disease and ascites while Chest X-ray was normal. At this stage of the illness possibilities were AKI (Acute Kidney Injury) secondary to (a). RPGN (Rapidly Progressive Glomerulonephritis) (b). D negative HUS (Diarrhea negative Hemolytic Uremic Syndrome) (c). Interstitial Nephritis.

She improved in next three days so we started her on oral feeds. On day four of admission she developed hyperglycemia (486 mg%) with ketonuria 4+. For this she was managed with intravenous fluids, insulin and potassium. This transient DKA (Diabetic Ketoacidosis) state recovered in next 48 hours. Next three days her edema and renal function improved. But on day 10 of admission she developed tachycardia with increased pallor. Her hemoglobin had dropped to 5.4 gm% while PBS (Peripheral Blood Smear) showed features of intravascular hemolysis with thrombocytopenia (Table 1). Her LDH (Lactate Dehydrogenase) was 2000IU/L. She was transfused PRBC (Packed Red Blood Cell) @ 25 ml/kg over next three days. She was also given Fresh frozen plasma @ 10 ml/kg for the next three days, due to non availability of plasma exchange, guided by serum LDH level. Her condition stabilised and urine output improved. On Day 14 of admission she had hypokalemic (serum potassium 1.9 meq/dl) flaccid quadriparesis with bulbar paralysis which was managed with iV potassium followed by oral potassium supplements (Figure 1). Her paralysis recovered in limbs with normalisation of serum potassium but dysphagia, nasopharyngeal regurgitation and palatal palsy persisted. MRI brain was normal. CH50, factor H and I were normal. Serum and urine amino acid chromatography for cobalamin metabolism was normal. She was given IVIG @400 mg/kg for five day, with this she started showing improvement in next three days and her palsy completely recovered in ten days (Table 2). During this period she was kept on tube feeds her kidney function and liver enzymes normalised but her hypertension persisted. On follow up at one month there was no edema and blood pressure was between 90-95 centile with three antihypertensives. Renal function was normal. Pediatric nephrologist consultation was taken and it was advised not to do a renal biopsy at this stage. ADAMTS13 and complement gene mutations studies could not be done due to non availability.

Discussion

Central nervous system manifestations in the form of irritability,

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Received July 12, 2013; **Accepted** November 04, 2013; **Published** November 06, 2013

Citation: Kumar V, Sodhi K, Kanitkar M, Deepak, Anupama, et al. (2013) Atypical HUS with Dysphagia and Palatal Palsy Responding to IVIG. Pediat Therapeut 3: 176. doi:10.4172/2161-0665.1000176

	Day 16 of illness	Day 20	Day 26	Day 29
Hb gm%	7.9, no evidence of hemolysis	9.1	5.4, intravascular hemolysis present	8.2
TLC /cmm	10,200	16,300	6,800	7000
Platelets	3.1 lac/cu mm	2 lac/cu mm	40,000/cu mm	1 lac/cu mm
Blood Urea mg%	196	106	50	30
Serum creat mg%	3.2	0.9	0.8	0.5
Serum sodium				
meq/dl	136	134	130	124
Serum potassium meq/dl	4.4	4.9	3.9	1.9
Serum. Albumin gm%	2.8	3.2	3.5	3.4
Serum. Bilirubin mg%	0.8	0.9	0.8	0.7
SGOT/SGPT IU/L	192/103	203/86	67/45	45/40
Urine RE/ME	Pus cells 7-8/HPF 15RBC/HPF Urine Protein 2+	Urine protein 1+	NAD	Urine protein1+
RBC-15-17/HPF, Protein 2+	Urine protein 1+	NAD	Urine protein1+	
Lactate Dehydrogenase IU/L			2000	800
Direct coombs test (DCT)			Negative	
PBS(Peripheral Blood Smear)	Normocytic Normochromic		Schistocytes, burr cells	
present				
Blood culture		Sterile		

Table 1: Showing details of investigations: features of intravascular hemolysis with thrombocytopenia.

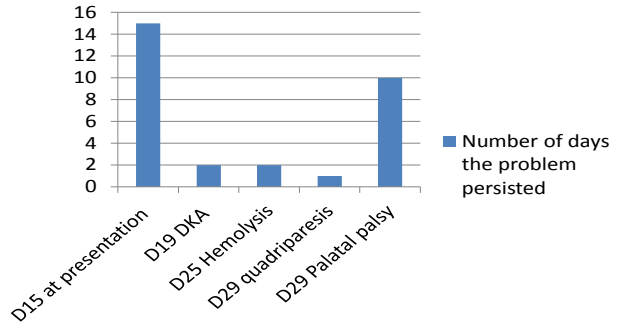

Figure 1: Bar chart depicting clinical Problems with duration.

1.	**Initial Presentation**	Restricted fluids, multiple antihypertensives, diuretics and supportive treatment
2.	**DKA**	Insulin, intravenous fluids and potassium
3	**Intravascular Hemolysis**	PRBC and FFP
4	**Hypokalemia/quadriparesis**	IV Potassium followed by oral potassium
5	**Dysphagia and palatal palsy**	IVIG

Table 2: Depicting various interventions: Improvement and recovery of Palsy.

drowsiness, seizures, cortical blindness, hemiparesis and coma are known extra renal complications of Atypical or D negative HUS [2]. These manifestations could be due to microangiopathic lesions or RPLS (Reversible Posterior Leukoencephalopathy Syndrome) secondary to hypertension. Both could be documented in MRI Brain. Complications of dysphagia and palatal palsy with a normal MRI brain are reported for the first time to the best of our knowledge. These complications getting cured using IVIG itself is a novel finding.

A typical HUS has shown to be due to alternative complement pathway dysregulation with 50% cases involving complement regulatory genes, factor H, factor I and MCP (Membrane Cofactor Protein) [3]. Plasma exchange therapy though cumbersome and invasive still forms the main modality of treatment. However Watt et al. had reported the only case of Atypical HUS responding to a combination of steroids and IVIG [4]. Our case who had developed a never reported finding of dysphagia and palatal palsy following Hypokalemia responded dramatically to five day therapy of IVIG (total 2 gm) alone. This could be a good alternative to plasma therapy apart from very promising and upcoming Eculizumab (monoclonal antibody against complement factor) [5].

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