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A CASE REPORT OF HENOCH-SCHONLEIN PURPURA WITH TYPICAL SYMPTOMATOLOGY LATER COMPLICATED WITH NEPHRITIS.

Biochemistry

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ABSTRACT

Henoch- Schonlein Purpura(HSP) is one of the most common vasculitis in children commonly occurring in the first decade of life. Symptoms include a palpable skin purpura, arthritis, abdominal pain, and renal involvement. It is an auto-immune disorder where IgA immune complexes deposit in the vessel wall and renal mesangium predominantly. It is considered to be an acute self -limiting illness although renal involvement is the principal cause of morbidity and mortality.Supportive treatment is necessary as per the disease progression. We report the case of a 6year old female diagnosed as Henoch-Schonlein purpura with significant renal involvement (nephritis).

KEYWORDS

INTRODUCTION:-

Henoch - Schönlein Purpura (HSP) is an acute immunoglobulin A (IgA)– mediated disorder characterized by a generalized vasculitis involving the small vessels of the skin, the gastrointestinal (GI) tract, the kidneys, the joints, and rarely the lungs and the central nervous system (CNS). HSP is the most common vasculitis in childhood, the incidence decreasing with age.[1] HSP has typically been reported to be more common in males, with a male-to-female ratio ranging from 1.5-2:1, but some studies have found a more equal distribution between the sexes.[2,3] It has been reported that 6to 24 per 100 000 children younger than 17 years will develop HSP, depending on the ethnic background of the child. In Asia, the incidence is as high as approximately 70 cases/100 000 children per year.[4]. The prevalence of HSP peaks in children aged 3-10 years, with the mean age at presentation is 6 years, but the condition is also seen in adults.[5] The clinical features of palpable cutaneous purpura, arthritis, abdominal pain, and nephritis may develop over days to weeks and vary in their order of presentation. In one-half to two-thirds of children, upper respiratory tract infection (URTI) precedes the clinical onset of HSP by 1-3 weeks. HSP is typically an acute, self-limiting illness, and treatment is primarily supportive. Kidney involvement is the most important determinant of long-term morbidity. Up to 30-50% of children present with hematuria and/or proteinuria, or develop it within 4-6 weeks of the initial presentation. This is usually mild and self-limiting. However, approximately 7% of all HSP cases will develop either nephritis or nephrotic syndrome [6-11].

Case report:- We report the case of a 6year old female who with 2-3 episodes of passing high-coloured(cola colour) urine since 5days and generalized abdominal pain dull aching in nature and of moderate severity, not associated with vomiting or loose stools or any episode of hematemesis.

Palpable non-pruritic purpuric patches on the buttocks and extensor aspects of both the lower limbs were noticed which then extended to the extensor aspect of both the upper limbs and abdomen. Arthralgia was present in both the knee joints Pedal oedema (pitting type) which later progressed to generalised body oedema (anasarca)

Mother gave history of an episode of cold, cough and fever approximately 10 days prior to onset of the above symptoms. She received erythromycin and recovered well.

There is no significant family history and she was immunised to date; born to a non-consanguineous marriage.

On general examination vitals were stable .Generalised anasarca, bilateral pitting oedema of both lower limbs noted with no pallor. Abdominal examination, girth -64cms, soft on palpation with no organomegaly. On percussion, dull notes observed with positive fluid thrill and bowel sounds were heard.

The cardiac, nervous and respiratory system were normal on examination with no abnormality detected.

Laboratory investigations included the routine blood count, renal function tests, fasting lipid profile, serum electrolytes which were deranged significantly. Complete urine examination was significant for high proteinuria, presence of Rbc's and high urine protein-creatinine ratio (UPCR).

Immunology tests performed were complement C3&C4, ANA profile, Anti-CCP antibodies levels, RA factor ASO titre were all reported within the normal range.

An ultrasound-guided renal biopsy was performed which was reported as crescentic glomerulonephritis with underlying proliferative glomerulonephritis and co-relating with the clinical history and clinical features, and significant lab results Henoch- Schonlein nephritis (ISKDC class-IV) was the final diagnosis.

During her hospital stay, the patient received steroids (methyl prednisolone pulsatile doses *3) according to weight and age and immune suppressants(cyclophosphamide) were also given as part of the treatment regimen.

The patient stabilised and was discharged with oral steroids in tapering doses. On her follow-up visit, it was noticed that the UPCR ratio decreased whereas the renal function tests were not within the normal range .

Laboratory investigations:- Table 1(a-d) showing the results of various laboratory investigations :-

Table 1a :-complete blood picture, renal function tests, serum electrolytes, lipid profile, coagulation tests

Parameter	Value	Reference range
Haemoglobin*	10.1	12.6-17.4 g/dL
Rbc	4.20	4.70-6.10 M/uL
Mcv	78.6	80.0-94.0 fl
Mch	26.4	27.0-31.0 pg
Mchc	33.6	33.0-37.0 g/Dl
Wbc*	11.3	4.8-10.8 k/Ul
Neutrophils*	86.9	50.0-70.0 %
Lymphocytes	23.7	20.0-40.0 %
Monocytes	6.3	0.0-15.0 %
Eosinophils*	9.7	0.0-6.0 %
Basophils	0.4	0.0-2.0 %
Platelets*	4.34	130-450 k/uL
Hct	33	%

Bleeding time	1min 40sec	2-7 minutes
Clotting time	3min 20sec	8-15 minutes
Serum urea*	72.8	10.8-38.4 mg/dl
Serum creatinine*	3.52	0.5-1.2mg/Dl
Serum sodium	130.5	135-145 mEq/L
Serum potassium*	2.99	3.4-4.7 mEq/L
Serum chloride	115.9	90-110 mEq/L
Cholesterol*	421.8	<170 mg/dL
Triglycerides*	364	<150 mg/dL
Hdl	25	>45 mg/dL
Vldl	73	<120 mg/dl
Ldl	120	<110 mg/dl

Table 1b (complete urine examination)

Colour	Clear	Pale (straw)
Reaction	Acidic	Acidic
Specific gravity	1.014	1.005-1.030
Albumin*	++++	Nil
Sugar	Nil	Nil
Pus cells	Plenty	Nil
Epithelial cells	8-10	2-5/hpf
Rbc*	12-15	<2 /hpf

Table 1c (urine protein- creatinine ratio)UPCR:-

Proteins*	610	1-400 g/dl
Creatinine*	44	0.5-1.2 mg/dl
Ratio*	13.8	

Table 1d Immunology parameters and their values:-

Crp	11.6	<5 mg/dl
Aso titres	Negative	
RAfactor	4.3	<10-120 IU/mL
ANAprofile	3.6	<12
Anti-CCPantibodies	3.4	<12
Complement C3levels	97.9	90-180 mg/dl
Complement C4levels	27.4	10-40 mg/dl

*indicates values beyond the normal reference range
Ultrasound-guided renal biopsy:-

Renal biopsy was reported as crescentic glomerulonephritis and was classified as Henoch-schonlein nephritis (ISKDC IV).

DISCUSSION:-

Henoch-schonlein purpura serves as a first clue towards the probable diagnosis, as clearly seen in our patient. Routine laboratory investigations serve as an important tool that aids in the diagnosis and is also useful in the follow-up for assessing the progression of the disease. Henoch Schönlein Purpura (HSP), also named as IgA vasculitis according to Chapel Hill Consensus Conference (CHCC) 2012 nomenclature, is the most common systemic vasculitis in childhood [12,13]Although it is known that HSP is a small vessel leukocytoclastic vasculitis, its pathogenetic mechanisms are not yet fully identified. Researchers [14-17] have found elevations in the serum levels of immunoglobulin (Ig) A, IgA-containing circulating immune complexes and IgA rheumatoid factors in patients with HSP. IgAV is almost exclusively associated with abnormalities involving IgA1, rather than IgA2. IgA1 is phylogenetically younger and differs from IgA2 by insertion of a 13-17 amino acid sequence in the hinge region of the IgA1 molecule. [18] The predominance of IgA1 in IgAV may be a consequence of abnormal glycosylation of O-linked oligosaccharides unique to the IgA1 hinge region. Patients with IgAV and IgA nephritis express inherited galactose-deficient glycosylation of IgA1 molecules [19] IgAV-associated nephritis is characterized by an abnormal IgA1 glycosylation pattern with reduced galactosylation. The hinge region of IgA1 contains up to six major glycosylation sites at serine and threonine residues. The O-glycans include a core N-acetylgalactosamine (GalNAc), which is usually extended with galactose to form Galβ1,3GalNAc, which can bind to N-acetylneuraminic acid (Neu5Ac). Thus, each IgA1 O-glycan can have one of four short carbohydrate structures (types III, IV, V, and VI), leading to a mixture of IgA1 forms with varying degrees of galactosylation. Patients with IgAV-associated nephritis have a high prevalence of galactose-deficient (types I and II) IgA1.[20] The finding that galactose-deficient IgA1 molecules are only found in IgAV during episodes of nephritis lends support to the pathophysiological role of galactose-deficient IgA1 molecules in IgAV nephritis.[21]The typical clinical features at presentation may vary as a rash (95-100% of cases), especially involving the legs, abdominal pain and vomiting (35-85%), Joint pain (60-84%), especially involving the knees and ankles, Subcutaneous oedema (20-50%), Scrotal oedema (2-35%), Bloody stools. Renal involvement occurs in more than 75% of cases and, in the vast majority, this manifests as hematuria alone or hematuria and low-grade proteinuria. In 10%-20% of cases, there may be significant proteinuria, nephrotic syndrome, or acute glomerulonephritis. The diagnosis of HSP is based on the presence of any combination of the following four features: (1) purpuric rash on the buttocks and extensor surfaces of the hands and feet, (2) arthritis, (3) abdominal pain, and (4) renal involvement. The four features can occur in any sequence, which can complicate the recognition of the disease. The cutaneous manifestations are the most characteristic finding that facilitates the diagnosis. The renal involvement of HSP is marked by hematuria, proteinuria and, in severe cases, azotemia and hypertension. There is no routine laboratory test to identify HSP and it remains a clinical diagnosis. To confirm that a child has HSP nephritis requires measurement of blood pressure, microscopic examination of the urine, determination of urinary protein excretion, serum creatinine concentration, and calculation of glomerular filtration rate (GFR). A kidney biopsy specimen showing mesangial IgA deposition in a child with suggestive complaints and findings confirms the diagnosis.[22] .

The International Study of Kidney Disease in Children (ISKDC) classification is widely used for patients. According to this classification, HSPN is graded as follows: grade I: minimal glomerular abnormalities; grade II: mesangial proliferation without crescents or sclerosing lesions; grade III: focal segmental (IIIa) or diffuse (IIIb) mesangial proliferation with <50% crescents or sclerosing lesions; grade IV: mesangial proliferation with 50%-75% crescents or sclerosing lesions; grade V: mesangial proliferation with >75% crescents or sclerosing lesions; and grade VI: membranoproliferative-like lesions [9,23]. The Kidney Disease Improving Global Outcome (KDIGO) guidelines recommend that children with HSPN with persistent proteinuria, defined as >0.5-1 g/d per 1.73 m2, should be treated with angiotensin- converting enzyme inhibitors (ACE-I) or angiotensin receptor blockers (ARBs). It is recommended that after a trial of ACE-I or ARBs, those with persistent proteinuria, defined as >1 g/d per 1.73 m2, and glomerular filtration rate >50 mL/min per 1.73 m2, should be treated the same as patients with IgAN, with a 6-month course of corticosteroid therapy. According to the KDIGO guidelines, those with crescentic HSP with nephrotic syndrome and/or deteriorating kidney function should be treated the same as patients with crescentic IgAN [24]. For other patients, a wide variety of immunosuppressive agents (steroids, cyclophosphamide, azathioprine and calcineurin inhibitors) has been used for treatment. Some adjuvant therapies such as immunoglobulin, anticoagulants, antiplatelet drugs and vitamins have also been used in combination with immunosuppressive agents, but the reviews do not recommend these adjuvant treatments.[25]

CONCLUSION: -

To conclude, Henoch-schonlein purpura nephritis is diagnosed based on the clinical features supported by the laboratory investigations (routine and special). The incidence and etiology are not well defined and based on the pathophysiology it is also termed as IgA vasculitis, where there is a deficiency of glycosylated IgA1 molecules which primarily deposit in the skin and renal mesangium. Renal pathology is the major cause of morbidity and mortality. Treatment is mostly steroids and immunosuppressant's. Follow-up is necessary and should use a simple, routine laboratory test urine protein - creatinine ratio(UPCR) which helps in assessing the proteinuria and kidney function. It serves as a yardstick in grading of the disease severity and treatment modality .

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