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Case report

Henoch-Schonlein purpura (HSP) in a rare novel complicated case: A case report

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Hcnoch-Schonlein purpura is an IgA mediated small-vessel vasculitis and it is so common in children. The classic term of signs and symbols include polpuble purpura, arthralgia/arthritis, abdominal pain and also renal disease. The incidence of HSP in older age of the entire gastrointestinal tract (gastric, small intestine and large intestine) without arthritis/arthralgia and kidney involvement is rare. In patients with HSP, in older age people, kidney problems are more common than the children, which no kidney problem in this case. A 57-year-old man who was admitted to the Razi Hospital, Rasht, Iran, with Perumblical colicky abdominal pain. During the outpatient surveys, Spiral CT Abdominal-Pelvic with IV oral contrast was determined by abdominal-Pelvic Ultrasonography through small bowel follow, ileum and jejunum involvement. After performing CT, Palpable purpura skin lesions appeared in the upper and lower extremities which was characterized by a low ALB outbreak, S / E (OB +), high fecal calprotectin, high ESR and normal leucocyte. In this way, at Razi Medical Education Center, the endoscopy showed stomach and duodenum involvement and colonoscopy shows the ascending, transverse, and descending colon. Skin sampling shown IgA sediment in Direct Immunofluorescence (DIF). HSP, although it occurs at an early age, has been reported in the patient due to the appearance of skin and abdominal pain and the absence of involvement of other basic organs such as the kidney, lung, and the negative effects of other vasculitis diseases on the basis of skin pathology.

Keywords: Henoch-Schonlein Purpura (HSP), Vasculitis, Abdominal pain

Henoch-Schönlein Purpura (HSP) is an immune complex-mediated disease that affects small vessel vasculitis of childhood with the prevalence of 6.1 to 20.4 per 100,000 children per year (1, 2). HSP is an acute immunoglobulin A (IgA) mediated disorder and most commonly occurs in the pediatric age. It involves small vessels caused by the storage of IgA in them. It is characterized by the classic tetrad of palpable purpura, arthralgia, abdominal pain and acute renal compromise, however, some of them might not appear (3). The confirmed diagnosis is based on the presence of palpable purpura with one of the following symptoms: diffuse abdominal pain, skin biopsy with IgA deposits, arthritis or arthralgia, and/or renal compromise (hematuria and/or proteinuria) (4). Characteristic cutaneous manifestations contain of palpable purpura (80 to 100% of the cases) and edema, which can be followed by urticarial exanthema or maculopapular exanthema. Palpable purpura demonstrates a symmetric distribution and it is localized mostly in lower limbs and gluteus, but

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it can also affect the being frequent in adults, cutaneous face, trunk, and upper limbs (5, 6). Although, blisters are very infrequent in children, with a prevalence rate lower than 2%. Remarkably, HSP is significantly more common in the pediatric population, failure to diagnosis and treat adults can have serious ramifications. In this account, we report a case of HSP in a 57-year-old patient with a past medical history who initially presented to the clinic with Palpable purpura skin lesions in the upper and lower extremities and without kidney problem.

Case Report

A 57-year-old man with a severe abdominal pain (pain score8 / 10) periumbilical pain was referred to the Razi Hospital in Rasht. The pain of the patient is aggravated by eating, decreases with bowel movement and does not spread to the other organ. One set of feces (loose, bitterness, stinky) and a significant of 10kg weight loss over the past month indicates the Family history of duodenal cancer in his father. He was hospitalized at the center of neurology one year ago due to paresthesia in the upper left limb, which was discharged with the diagnosis of small vessels disease. In the history of the drug, Atorvastatine, ASA 80 mg/daily, Allupurinol 100mg/daily and Ibuprofen 400mg/PRN for reducing pain in the last month were prescribed. The patient was examined as an outpatient abdominal pain that, on day 17 (2 days after CT, with contrast), begins with symptoms of



Figure 1. Palpable purpura skin lesions on the lower limbs.



Figure 2. Small bowell involment.

Palpable purpura skin lesions in the upper and lower extremities (Fig 1). According to the skin lesions of the patient, the candidate for examination is vasculitis (Table 1) and remarkably, the previous NCV-EMG bias indicated the normal result. In imaging study (CT and Barium), intra luminal involvement was detected in the small intestine in the areas of ileum and jejunum (Fig 2). The patient was referred to the adult gastroenterology and liver center, Razi Hospital for examination of the digestive system. Endoscopy of the erosion of the stomach anterior, bulb, duodenum and sub epithelial peteche, and colonoscopy erythema and loss of vascularity was revealed in the ascending, transverse and descending colon (Fig 3). In the sampling of new skin lesions, the IgA sediment was determined in DIF and with the diagnosis of HSP, corticosteroid therapy started for the patient.

Clinical intervention

A patient with a 50-year history of familial duodenal cancers with abdominal pain and weight



Figure 3. Superfecial erosions in bulb.

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Table 1. Summary Laboratory reviews

Hematology		**Biochemistry**			
WBC	15400	BUN	7	ALB	2.6
Neutrophils	80%	Cr	1.1	Protein	6
Lymphocytes/10	14%	AST(SGOT)	18	Na	139
Monocyte	2%	ALT(SGPT)	16	K	9
Eosinophil	3%	Alkaline phosphatase(ALP)	135	Fe	34
Hb	11.5	amylase	46	TIBC	195
MCV	93	lipase	45	Ferritin	286
RDW	14.3	25(OH) Vit D	31	CRP	31
Plt	534				

Other				**Urin 24hrs Collection**	
ESR	70 mm/h	Hbs Ag	Negative	Volume	2000cc/24hrs
PT	13.9	Anti-PR3		Creatinine	1100
PTT	31	Cryo-globulin		Protein	20
INR	1.2	Fecal Calprotectin Ag	>1000		
C3,C4,CHAO	Normal				

U/A				**S/E**	
Color	yellow	Protein	Negative	WBC	Negative
Appearance	clear	Bilirubin		Occult blood (OB^+)	
Specific gravity	1.030	Ketone		RBC	
PH	6.5	Urobilingen		Ova of parasite	
WBC	2	Nitrite		Protozoa cyst	
RBC	1	Bacteria		Strongyloides Ab IgG	
Epithelial cells	2	Cast			

loss in an outpatient examination and imaging study was required to be investigated in the early stages of the patient's gastrointestinal tract. A test and imaging study was required. In experiments conducted with high ESR, low ALB, normal U/A, S/E (OB^+) and high Fecal calprotectin should be considered as gastrointestinal malignancy and the cause of low ALB with normal U/A, protein losing enteropathy or ALB drop can be consider, as the negative phase reactante. With the emergence of dermal purpura, intravenous contrast is a newer diagnosis of vasculitis, paraneoplastic manifestations of gastrointestinal malignancies. In this way, drug induced vasculitis (in the field of Alluporinol, Atorvastatin), response to intravenous contrast, which was not aided by a laboratory finding. So, it was better to sample the new skin lesions at the same time. After referral to the adult gastroenterology and liver center with regard to the possibility of malignancy in the digestive system, the probability of IBD with digestive problems, skin manifestations was investigated and considerably, diagnosis of stomach - duodenal colon was determined. In this way, a new skin lesion was sampled and carried out immunofluorescence in order to diagnose the sediment IgA.

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Discussion

HSP is characterized by the association of skin, joint, and gastrointestinal manifestations that may occur in consecutive episode (7). In addition to these manifestations, renal involvement is common, and the long-term prognosis depends on its severity is less common in adults. Although, HSP has been comprehensively studied in children, much less is known about its natural history in adults (8, 9). In adults, however, the incidence of HSP and the severity of its clinical manifestations appear not to be the same as in children. The incidence of renal involvement in adults varies from 45 to 85% of cases, depending on the data for patients and the definition of renal involvement. Among cases of glomerulonephritis, HSP is only responsible for 0.6 to 2% of adult nephropathies. The risk of progression to renal insufficiency, which ranges from 5% to 15% in children, seems to be much higher in adults, approximately 30% (0 to 50%). These differences are probably due to the small size of the series, the presence or absence of renal involvement, and the differences in the duration of follow-up (7, 10). In this patient, laboratory studies specified high ESR, low ALB, normal U/A, S/E (OB^+) and high fecal calprotectin and all these quantification indicated gastrointestinal malignancy. Conspicuously cause of low ALB with normal U/A, protein losing enteropathy or ALB drop is considered as the negative phase reactant. The differential diagnosis of HSP includes conditions such as vasculitis, paranoplacental manifestations of gastrointestinal malignancies, drug induced vasculitis (in the field of Alluporinol, Atorvastatin), malignancy in the digestive system and IBD with digestive problems. In this patient, whose purpura located on the upper and lower extremities, renal functions were normal at follow-up. Since the IgA plays an important role in HSP and is elevated in serum, the IgA level was high in this case. Also, it was sampled from new skin lesions which were diagnosed with IgA sedimentation immunofluorescence. Notably, One reasonable explanation is that many serologic studies revealed increased serum levels of IgA in only 50% of patients with HSP during the acute stage (11). Consequently, with the diagnosis of HSP, corticosteroid therapy stated for the patient. The use of early glucocorticosteroids (GCSs) may shorten the duration of abdominal pain, decrease the risks of intussusceptions and surgical intervention (11).

Conclusion

HSP, although it occurs at an early age, it has been reported in the patient due to the appearance of skin and abdominal pain and the absence of involvement of other basic organs such as the kidney, lung, and the negative effects of other vasculitis diseases on the basis of skin pathology and was treated with 30 mg prednisolone.

Author contributions

FMG did the conceptualization and supervision the project **FJ ISA, ASH, MB**, **MM** collected and recorded the patient's data and also examined the patients at the hospital. **TZ** was involved in some sections of writing the manuscript. **AAS** accomplished the data quality control, investigated and optimized the informatics database, accomplished the statistical analyses and data processing, wrote the paper and evaluated it. All authors revised the paper comprehensively. All authors read the manuscript carefully and confirmed the final edited version of the paper.

Ethical issues

There are no ethical problems for this project.

Informed consent

This project was done with the agreement and informed consent of all the patients.

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Conflict of interest

The authors declared no conflict of interest.

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