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HENOCH-SCHONLEIN PURPURA IN CHILDHOOD

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Henoch-Schonlein purpura is the most common vasculitis in children. The process affects small blood vessels of the skin, joints, gastrointestinal tract, kidneys and the central nervous system.

The clinical manifestations in children with Henoch-Schonlein purpura were analyzed. The count of white blood cell, trombocytopenia, CRP, LDH, CPK, titer of antistreptolysin antibody (ASO) were analyzed. Urinalysis and urine culture tests were performed. The values of complements (C3 and C4) and immunoglobulins (IgG and IgM) for viruses HSV, EBV sand CMV (ELISA test) were analyzed as. Nasal and throat swabs were examined as well.

We examined 35 children, aged 5 to 17 years. The recorded clinical manifestations were: fever (47.21%), abdominal pain (28.61%), joint pain (25.33%), and muscle pain (18.03%). Changes such as purpura were present on hands (6.5%), forearm (12.3%), lower leg (81.2%), gluteal region (28.13%), and feet (21.35%). We also recorded: leukocytosis (11.37%), thrombocytopenia (7.21%), high level of CRP (6.21%) and high level of serum CPK and LDH (7.21%). ASO titer antibody was positive in 18.47% of children.

The majority of children had previous respiratory and urinary tract infection. Most of the children had an infection of viral origin. The prognosis for most children is good, with no pathological kidney damage with proteinuria. A small number of children required the use of corticoid in therapy. *Acta Medica Medianae 2015;54(4):32-36.*

Key words: *Henoch-Schonlain purpura, children, clinicall characteristics*

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Introduction

Henoch-Schönlein purpura is a clinical syndrome comprised of purpura, abdominal pain and arthralgia. Changes in the skin like purpura, abdominal pain and arthralgia are the most common symptoms in children. Henoch-Schonlein purpura (HSP) is defined as vasculitis of predominantly immune-complex deposits (IgA) in small blood vessels. It is typically localized in the skin, gut and glomeruli and is associated with arthralgia or arthritis (1, 2).

Henoch-Schonlein purpura is the most common vasculitis in the pediatric population. The average incidence of occurrence of Henoch-Schonlein purpura in EU countries is 13-15/100 000. The disease occurs in the pediatric popula-tion, in the age group 2-11 years. The average age of affected children is approximately 5.5 years, over 75% of affected children are under the age of 10. The disease is more common during winter months; in 50% of cases it is preceded by a respiratory infection of the upper or lower respiratory tract (3-5).

Skin changes like purpura are present on the extensor sides of the lower legs, gluteal region and upper legs; trunk and face are rarely affected. Symptoms on the skin may be in the form of palpable purpura and petechiae of the large ecchymosis. Clinical symptoms are edema of the face, hands, feet and scrotum. Arthralgia are mostly transient (6). Joint pain is often associated with pain in the muscles of the lower legs, all of which limit the mobility of patients, thus creating a picture of a seriously ill child. Abdominal pain is frequently of marked intensity and is associated with nausea and vomiting. Melena and hematemesis are rare symptoms. Symptoms of glomerulonephritis (hematuria, proteinuria) and nephrotic syndrome are characteristic of later stage of disease. Symptoms of disorder of the CNS are very rare.

The beginning of disease may be abrupt or gradual. Clinical symptoms may follow one another in a 2-3 weeks period. The most common infections of the respiratory or gastrointestinal tract precede the disease.

Inflammation of the endothelium of blood vessels is a basic part of pathogenetic process du-

ring the disease with different evolution and prognosis. Different etiological agents (bacteria, viruses, parasites, etc.) can be the triggers of the inflammatory process. Beta-hemolytic Streptococcus, Staphylococcus, Mycoplasma, Ebstein-Barr virus, varicella, adenovirus are very frequent etyologic agents. Sometimes, vaccination or intake of a drug can be the cause of the disease onset (7).

Disease has a good prognosis in most cases. Recurrences of the disease can be after several months or years. Patients should be monitored for the development of kidney disease. About 20% of affected children develop damage of the kidney 20 years after the outset of the disease (8).

Examinees and methods

Children with clinical symptoms (purpura, joint pain, abdominal pain) typical of Henoch-Schonlen purpura (HSP) were examined. The study was conducted at the Clinic for Children's Internal Medicine, Clinical Center Niš.

The analysis of history data (data about previous infection, treatment, use of drugs, risk factors in the prenatal period), clinical symptoms (abdominal pain, symptom on skin like purpura, muscle pain, feet joint pain, headaches, abnormal pain and symptoms on other organs and systems) and evolution of clinical disease (evolution of symptoms, hematuria, more intense symptoms, evolution of abdominal pain, application of corticosteroids, proteinuria) was performed.

Analyzed laboratory parameters: hematologic parameters (number of leukocytes, erythrocytes, platelets, monocytes, lymphocytes, hemoglobin), parameters of inflammation (CRP), urinalysis and urine culture tests, enzymes, cell suffering (CPK, LDH) values of complement (C3, C4), throat and nasal swabs, titers of antistreptolizin antibody (ASO), Elisa test (CMV, HSV and EBV).

Results

Analyzed patients with the diagnosis of HSP were treated at the Clinic for Children's Internal Medicine, Clinical Center Niš, in the period from April 2012 to June 2013. Thirty-five children, 15 of which were girls and 20 were boys aged 5 to 17 years, were examined.

The predominant clinical symptoms of examined children was fever (47.21%). Abdominal and joints pain (knees and ankles), characteristic symptoms for the diagnosis of HSP, were present in 28.61% and muscle pain in 18.03% of the children (Table 1) .

Table 1. Clinical symptoms

Fever	47,21%
Stomach pain	28,61%
Joint pains (knee, ankle)	25,33%
Muscle pains	18,03%

Symptoms on the skin like purpura were present on hands (dorsal side), upper arm, body and thigh (6.5%), forearms (12.3%), legs (81.21 %), gluteal region (28.13%), feet (21.35%) of the patients. (Table 2).

Table 2. Purpura of the skin

Legs	81,2%
Gluteal region	28,34%
Feet	21,35%
Forearm	12,3%
Hand (dorsal side), upper arm, face, whole body, thigh-high	6,5%

Hematologic parameters: leukocytosis in 11.37% and thrombocytopenia in 7.21% of the children. Higher values of CRP (C-reacting protein) were present in 6.21% of the children. Elevated enzymes of cell suffering CPK (creatine phosphokinase) and LDH (lactate dehydrogenase) were present in 7.21% of the children. Titer of antistreptolizin antibody (ASO) was positive in 18.47% of the childre (Table 3).

Table 3. Laboratory parameters

Leucocytosis	11,37%
Thrombocytopenia	7,21%
Crp	6,21%
Cpk	7,21%
Ldh	7,21%
Aso	18,47%

Analysis of the presence of IgG antibodies in the herpesvirus (HSV, CMV and EBV) shows the presence of IgG antibodies in all patients (ELISA) (Table 4).

Table 4. Positive antibody for viruses in serum (ELISA test)

CMV (igg+)	+
EBV (igg+)	+
HSV (igg+)	+

Urinanalysis in 12.3% of patients showed that the sediment had plenty of fresh RBCs, fading red cells, epithelial cells. Proteinuria was not recorded. Urine culture test showed the presence of E. coli > 100.000 in 7.2% of patients.

Values of complement C3 and C4 in the examined children were within the reference ranges.

The analysis of the throat swabs in 16.5% of patients showed the presence of Staphylococcus aureus; in 4.9% of patients Haemophilus parainfluenza and Escherichia coli were found.

X-ray of the heart and lungs in PA with 16.8% of the patients indicated the existence of the bronhial interstitial reaction.

Respiratory infections of the upper respira-

Table 5.Clinical manifestations

Respiratory infection of the upper respiratory tract	72.2%
Respiratory infection of the lower respiratorni tract	14.3%
ITU (infection of the urinary tract)	7.5%
Allergy to cow`s milk proteins	7.2%

tory tract (72,2%), respiratory infections of the lower respiratory tract (14.3%), urinary tract infection (7.5%) and allergy to cow's milk protein (7.8%) were recorded in the examined group of children.

Discussion

Clinical manifestations that define Henoch-Schönlein purpura in children are the subject of extensive research and analyses in order to establish criteria for the diagnosis of diseases, but above all to establish the etyopathogenic mechanisms responsible for the formation of pathological events, examine the prognosis of the disease and the application of corresponding therapy.

The majority of patients had characteristic respiratory or urinary infections. The same was observed in the test group of children (respiratory tract infection in 86.5% of patients and infections of the urinary tract with 7.2% of patients). The medical history indicated that the infections preceded HSP (approximately 10-14 days). The age of the children (between 5 and 17 years) corresponds to the findings of other authors (3).

One of the common symptoms of the examined children was fever 47.21%, which corresponds to the preceding infection, and that was the trigger for the onset of the disease. Serological analysis showed a positive IgG antibody titer to viruses (HSV, EBV and CMV), which can be considered as a probable etiological factor for infections that accompany the occurrence of HSP in examined children. The presence of urinary tract infection was also evident in patients (7.5%).

All children in the study had purpura on the skin, which is an important parameter for the diagnosis of HSP. Previous studies have pointed to purpura as one of the important clinical parameters for the diagnosis of HSP in children (9). Purpura was seen on the legs in 81.21% of patients.

Peru et al. described 254 children with HSP, in whom the following clinical changes were evident: purpura on the skin (100%), arthritis (66 %), gastrointestinal symptoms (56%), and symptoms of the kidney - proteinuria or hematuria (30%) (10). Purpura is the most common clinical symptom in 57-69% of patients and most notable on legs and gluteal region (10). Local angioedema can precede the occurrence of purpura on the skin. Arthritis (joints, feet and knees) is also an important clinical symptom (10). Examined children had the same incidence of clinical symptoms which coincides with the results of other findings.

The most common gastrointestinal symptom is abdominal pain, which is sometimes associated with vomiting or haematemesis (11). Arthralgia and abdominal pain were equally frequent clinical symptoms in examined children (29%). Intussusception is a serious acute surgical complication occurring in 0.7-13.6% of patients (11). Joint pain, swelling and limited mobility were present in 75% of patients with HSP in one study (14). Gastrointestinal bleeding may be present in about 5% of patients (12, 13). Microscopic hematuria is usually in the acute phase of disease, but nephrotic syndrome or glomerulonephritis can also develop of (15).

The prognosis of the disease is good in many children with HSP. In the UK, 1.6-3% of children with HSP develop nephritis (16). Patients with HSP must be monitored for at least two years after the normalization of urine sediment (17).

Antibiotic therapy was applied in children with the symptoms of infection during HSP. Corticosteroids were given to patients with intensive, recurrent purpura and abdominal pain. Anti-inflammatory therapy (NSAI-nonsteroid anti-inflammatory therapy) is applied with antihistamines. Severe form of HSP was not registered in our examined group of children.

Laboratory parameters indicate the presence of inflammation. Parameters of cell destruction (LDH, CPK) during the inflammation are very important for the prognosis and therapy of the disease.

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HENOH ŠONLEJNOVA PURPURA KOD DECE

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Henoh Šonlejnova purpura je najčešći vaskulitis u dečijoj populaciji. Proces zahvata male krvne sudove kože, zglobova, gastrointestinalnog trakta, bubrega i centralnog nervnog sistema.

Analizirane su kliničke manifestacije dece sa Henoh Šonlejnovaom purpurom. Analizirani su: broj leukocita, trombocita, vrednosti parametara: CRP, CPK, LDH, titar antistreptolizinskih antitela (ASO). Analiziran je urin: opšti pregled i urinokultura. Ispitivane su vrednosti komplemenata i vrednosti imunoglobulina (IgG i IgM) na viruse HSV, EBV i CMV (ELISA test), bris grla i nosa.

Ispitivano je 35-oro dece uzrasta od 5 do 17 godina. Evidentirane su kliničke manifestacije: povišena telesna temperatura (47,21%), bol u stomaku (28,61%), zglobovima (25,33%) i mišićima (18,03%). Promene po tipu purpure bile su zastupljene na šakama (6,5%), podlakticama (12,3%), potkolenice (81,2%), glutealna regija (28,13%), stopala (21,35%). Evidentirana je leukocitoza (11,37%), trombocitopenija (7,21%), povišene vrednosti CRP-a (6,21%) i povišene vrednosti CPK i LDH (7,21%). Titar ASO antitela bio je pozitivan kod 18,47% dece.

Kod većine ispitivane dece prethodila je infekcija respiratornog i urinarnog trakta. Većina dece imala je infekcije virusnog porekla. Prognoza kod većine je dobra, bez patoloških oštećenja bubrega sa proteinurijom. Mali broj dece je zahtevao primenu kortikoida u terapiji. *Acta Medica Medianae 2015;54(4):32-36.*

Ključne reči: *Henoh-Šonlejnova purpura, deca, kliničke karakteristike*