

Henoch-Schönlein purpura

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Henoch-Schönlein purpura (HSP) is a common vasculitis of childhood with predominantly involvement of small vessels. It often follows an upper respiratory tract infection, or it can also be triggered by medication. Vasculitis mainly affects the vessels of the skin, digestive system, kidneys and joints. Immunofluorescent deposits of immunocomplexes with IgA are typical.^[1]

Features

The most common occurrence is in children from 3 to 15 years of age. The **incidence** is 12/100 000, more commonly affected boys than girls. The cause is unknown (indisputable role of intercurrent infection).

Leukocytoclastic vasculitis (in biopsy of skin lesions) is characteristic. IgA deposits appear in capillaries and venules, in the kidneys they are deposits of mesangioproliferative glomerulonephritis.^[2]

Clinical picture

The patient is affected by a skin exanthema, either as a confluent purpuric skin eruption over the extensors of the lower limbs, which may also appear on the upper limbs, or as hemorrhagic bullae). The rash does not itch and disappears within 2 weeks.

Up to **80 % of patients** also have joint pain, arthritis, which is transient, precedes the rash, but can affect any joint - especially knees and ankles, with arthritis comes swelling with pain and limited mobility.

In **50 % of patients**, abdominal pain of a colicky nature occurs, mainly in the navel area, positive occult bleeding may occur.

In **1/3 patients**^{[2][3][1]} kidney involvement of varying extent occurs - glomerulonephritis, which is microscopic hematuria and proteinuria, which rarely progresses to nephrotic syndrome)



Skin exanthema in more severe manifestations of HSP

Laboratory Finding

In the laboratory findings, there is an elevation of inflammatory parameters: sedimentation, CRP, leukocytosis^[3]. In contrast to thrombocytopenic purpura, the platelet count is normal or elevated^[3]. hematuria, proteinuria and blood in the stool^[3] may be present. Elevation of PAF^[2] occurs. Anemia^[2] occurs. Elevation of IgA persists, complement levels tend to be normal. In the case of **hemocoagulation** the capillary fragility test is pathological, the other parameters are normal. A kidney biopsy shows mesangioproliferative glomerulonephritis with IgA and complement deposits in the mesangium, and a skin biopsy shows IgA^[1] deposits.

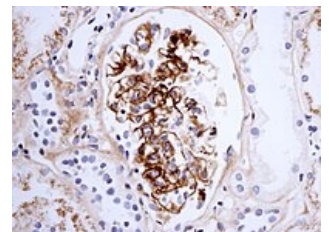


Image of IgA in the glomerulus in a patient with HSP glomerulonephritis

Diagnostic criteria

To be diagnosed with Henoch-Schönlein purpura, a patient must have 2 of the following 4 symptoms:

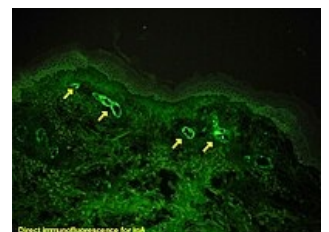
- purpura that does not disappear on palpation (in the absence of thrombocytopenia);
- abdominal pain (diffuse pain or intestinal ischemia);
- diagnostic biopsy (granulocytes in the wall of arterioles and venules);
- age under 20^[3].

These criteria have 87.1 % sensitivity and 87.7 % specificity^[3].

Differential diagnosis

- Other systemic vasculitides (Wegener's granulomatosis, polyarteritis nodosa, systemic lupus erythematosus, dermatomyositis, juvenile rheumatoid arthritis, Kawasaki disease etc.) ;
- thrombocytopenic purpura (idiopathic thrombocytopenic purpura, leukemia)^[3].

Therapy



Direct immunofluorescence of IgA in the glomerulus

Rest mode is the most important. So-called **symptomatic therapy** is proposed, which includes hydration and adjustment of electrolyte balance. **Non-steroidal anti-rheumatic drugs** are prescribed for acute arthritis. **Corticoids** are given to the patient for joint symptoms and abdominal pain. Methylprednisone is originally administered, but not as a prevention of glomerulonephritis, as meta-analyses have not shown a reduction in the risk of nephrotic or nephritic SY when used preventively. In the case of RPGN, a combination of steroid and azathioprine is suitable.

Prognosis

The long-term prognosis is favorable, but depends on the extent of renal involvement. *1-4 % of patients'* may develop chronic nephritis.^[1] The illness usually lasts 3-4 weeks^[2].

Links

Related Articles

- IgA nephropathy
- Vasculitides
- Manifestations of inflammatory rheumatic diseases on the musculoskeletal system and their surgical treatment

References

1. HRODEK, Otto – VAVŘINEC, Jan, et al. *Pediatric*. 1. edition. Praha : Galén, 2002. pp. 153-154. ISBN 80-7262-178-5.
2. DUNGL, P., et al. *Ortopedie*. 1. edition. Praha : Grada Publishing, 2005. ISBN 80-247-0550-8.
3. KLIEGMAN, Robert M. – MARCDANTE, Karen J. – JENSON, Hal B.. *Nelson Essentials of Pediatrics*. 5. edition. China : Elsevier Saunders, 2006. pp. 428-429. ISBN 978-0-8089-2325-1.