

Current problems in the diagnosis of Henoch-Schönlein purpura according to the ACR or EULAR/PReS criteria

Dear Editor,

We read with interest the article 'A patient with abdominal pain and a rash' by van Laarhoven *et al.*¹ They made the diagnosis of Henoch-Schönlein purpura (HSP) in their patient using the American College of Rheumatology (ACR) criteria.² Recently, however, the diagnostic criteria of HSP were modified by EULAR/PReS (the European League against Rheumatism/ Paediatric Rheumatology European Society)³ to overcome the weak points of the ACR criteria. In these new criteria, the age criterion was deleted, 'predominant IgA deposition' was included in the definition of the criterion describing 'biopsy', and arthritis and renal involvement were added to the group of criteria. Nevertheless, there are some problems in the correct diagnosis of HSP.³

Firstly, the ACR or EULAR/PReS criteria cannot detect the atypical presentation (e.g. delayed appearance of purpura) of HSP,⁴ leading to unnecessary procedures, such as appendectomy, or unfavorable outcome, such as death. Therefore, Kaneko *et al.* reported that measurement of plasma factor XIII (fibrin stabilising factor) might be useful for the early diagnosis of HSP, because it is decreased even without the purpuric rash, although not a diagnostic criterion of HSP.⁵

Secondly, skin biopsy cannot exactly differentiate HSP from hypersensitivity vasculitis, because IgA deposits may not be stained according to the biopsy site or various factors (e.g. phagocytosis with time) even in HSP.⁶

Thirdly, polyangiitis overlap syndrome can be missed even though a patient presenting with the features of HSP shows IgA deposits on biopsy specimens.⁷ Therefore, if a patient has an atypical presentation or a progressive course, the overlap of other vasculitis

should also be examined to prevent unexpected fatal situations.

In the future, further studies should be performed to elucidate the epidemiology and natural course of HSP based on the new EULAR/PReS criteria as well as the usefulness of the criteria.

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