CHAPTER 19

HENOCH-SCHONLEIN PURPURA – IgA VASCULITIS (D69.0)

Elif ÇETİNKAYA[1]
Ayşe Nilhan ATSÜ[2]

REMEMBER

- Henoch-Schönlein purpura (HSP) is the most common form of systemic vasculitis in children. Average time to appear: around 6-7 years of age. Although rare, it can also be observed in adulthood.
- The annual incidence of the disease in childhood has been reported as 2.4-15 per 100,000. The incidence in adulthood is estimated at 1.3 per 100,000.
- The etiology of the disease is not known exactly. Vaccines, viral and bacterial infections, certain drugs and autoimmunity have been blamed.
- Major clinical signs (HSP disease tetrad):
  - Palpable purpura without thrombocytopenia or coagulopathy
  - Arthritis/arthralgia
  - Stomachache
  - Kidney diseases
- The disease has a seasonal predisposition: it is more common in autumn and winter.

[1] MD., Basaksehir Cam and Sakura City Hospital, Dermatology Clinic, elif.cetinkaya96@hotmail.com
[2] MD., Istanbul Kent University, Faculty of Health Sciences, nilhan.atsu@kent.edu.tr

- Pain, tenderness, and swelling in the testis/scrotum are typical clinical findings.
- Orchitis, epididymitis, testicular torsion and testicular hematoma may occur. All of the listed conditions above are urological emergencies, and should be consulted with a urologist.

Neurology

- HSP may also lead to cerebral vasculitis. As a result, ischemia, edema, bleeding and infarction may develop.
- Headache, ataxia, seizure, encephalopathy, myelopathy, focal neurological deficit, intracerebral hemorrhage and central/peripheral neuropathy may be observed in some of the patients.
- In the presence of the above-mentioned neurological symptoms/signs, the opinion of a pediatric neurologist should be sought.

Pulmonary diseases

- Since impaired lung diffusion capacity, and mild interstitial changes in chest imaging have been reported in patients with IgA vasculitis, evaluation by a pulmonologist is required.

Cardiology

- Cardiac involvement is more common than expected, and occurs 2-4 weeks after the onset of the disease.
- Palpitations, bradycardia, and chest pain may occur.
- Conduction anomalies, myocarditis, endocardial vasculitis and congestive heart failure may develop. Cardiac examinations of patients should be performed routinely, and evaluated by a cardiologist.

Ophthalmology

- Henoch-Schönlein purpura may also lead to keratitis, and uveitis. Cases having retinal artery vasculitis, episcleritis, and retinal artery occlusion have been reported. In clinical doubt, an ophthalmologist should be consulted urgently.

REFERENCES

Gardner-Medwin JMM, Dolezalova P, Cummins C, Southwood TR. Incidence of Henoch-Schönlein purpura, Kawasaki disease, and rare vasculitides in children of different ethnic origins. Lancet. 2002;360(9341):1197–202.

Hetland LE, Susrud KS, Lindahl KH, Bygum A. Henoch-schönlein purpura: A literature review. Acta Derm Venereol. 2017;97(10):1160–6.

Kang Y, Park J su, Ha YJ, Kang M il, Park HJ, Lee SW, et al. Differences in clinical manifestations and outcomes between adult and child patients with henoch-schönlein purpura. J Korean Med Sci. 2014;29(2):198–203.

Calvo-Río V, Loricera J, Mata C, Martín L, Ortiz-Sanjuán F, Alvarez L, et al. Henoch-schönlein purpura in Northern Spain: Clinical spectrum of the disease in 417 patients from a single center. Med (United States). 2014;93(2):106–13.

Yang YH, Hung CF, Hsu CR, Wang LC, Chuang YH, Lin YT, et al. A nationwide survey on epidemiological characteristics of childhood Henoch-Schönlein purpura in Taiwan. Rheumatology. 2005;44(5):618–22.

Alexander K C Leung, Benjamin Barankin KFL. Henoch-Schönlein Purpura in Children: An Updated Review. Curr Pediatr Rev . 2020;16(4):265–76.

Trapani S, Micheli A, Grisolia F, Resti M, Chiappini E, Falcini F, et al. Henoch Schonlein purpura in childhood: Epidemiological and clinical analysis of 150 cases over a 5-year period and review of literature. Semin Arthritis Rheum. 2005;35(3):143–53.

Chang WL, Yang YH, Lin YT, Chiang BL. Gastrointestinal manifestations in Henoch-Schönlein purpura: A review of 261 patients. Acta Paediatr Int J Paediatr. 2004;93(11):1427–31.

Çakıcı EK, Gür G, Yazılıtaş F, Eroğlu FK, Güngör T, Arda N, et al. A retrospective analysis of children with Henoch–Schonlein purpura and re-evaluation of renal pathologies using Oxford classification. Clin Exp Nephrol [Internet]. 2019;0(0):0. Available from: http://dx.doi.org/10.1007/s10157-019-01726-5

Ghrahani R, Ledika MA, Sapartini G, Setiabudiawan B. Age of onset as a risk factor of renal involvement in Henoch-Schönlein purpura. Asia Pac Allergy. 2014;4(1):42.

Dalpiaz A, Schwamb R, Miao Y, Gonka J, Walzter W, Khan SA. Urological manifestations of henoch-schonlein purpura: A review. Curr Urol. 2014;8(2):66–73.

Bulun A, Topaloglu R, Duzova A, Saatci I, Besbas N, Bakkaloglu A. Ataxia and peripheral neuropathy: Rare manifestations in Henoch-Schönlein purpura. Pediatr Nephrol. 2001;16(12):1139–41.

Stefek B, Beck M, Ioffreda M, Gardner L, Stefanski M. Henoch-Schönlein purpura with posterior reversible encephalopathy syndrome. J Pediatr. 2015;167(5):1152–4.

Chaussain M, de Boissieu D, Kalifa G, Epelbaum S, Niaudet P, Badoual J, et al. Impairment of lung diffusion capacity in Schönlein-Henoch purpura. J Pediatr. 1992;121(1):12–6.

Bérubé MD, Blais N, Lanthier S. Neurologic manifestations of Henoch-Schönlein purpura. Handb Clin Neurol. 2014;120:1101–11.

Du L, Wang P, Liu C, Li S, Yue S, Yang Y. Multisystemic manifestations of IgA vasculitis. Clin Rheumatol. 2021;40(1):43–52.

Aristodemou P, Stanford M. Therapy insight: The recognition and treatment of retinal manifestations of systemic vasculitis. Nat Clin Pract Rheumatol. 2006;2(8):443–51.

Min Seung Kang, Han Jo Kwon JEL. Retinal Capillary Reperfusion from Ischemic Retinal Vasculitis in Henoch-Schönlein Purpura: A Case Report. Ocul Immunol Inflamm . 2021;17(1).

BÖLÜM 18

HENOCH-SCHÖNLEİN PURPURASI–IgA VASKÜLİTİ (D69.0)

Elif ÇETİNKAYA[1]
Ayşe Nilhan ATSÜ[2]

HATIRLA

- Henoch-Schönlein purpurası (HSP) çocuklarda sistemik vaskülitin en sık gözlenen formudur. Ortalama görülme zamanı 6-7 yaş civarıdır. Nadir olmakla birlikte erişkin yaşta da ortaya çıkabilir.
- Hastalığın çocukluk çağında yıllık insidansı 100.000'de 2.4-15 olarak bildirilmiştir. Yetişkin dönemdeki insidans ise 100.000'de 1.3 olarak tahmin edilmektedir.
- Hastalık etyolojisi tam olarak bilinmemektedir. Aşılar, viral ve bakteriyel enfeksiyonlar, bazı ilaçlar ve otoimmünite patogenezde suçlanmıştır.
- Major klinik bulgular (HSP hastalık tetradı):
  - Trombositopeni veya koagülopati olmaksızın gözlenen palpabl purpura
  - Artrit/artralji
  - Karın ağrısı
  - Böbrek hastalığı
- Hastalık mevsimsel yatkınlık gösterir: sonbahar ve kış aylarında daha sık izlenir. Bu nedenle enfeksiyöz tetikleyicilerin hastalıkta rol oynayabileceği üzerinde çok durulmuştur.

[1] Dr., TC Sağlık Bakanlığı Çam ve Sakura Şehir Hastanesi, Dermatoloji Kliniği, elif.cetinkaya96@hotmail.com
[2] Dr., İstanbul Kent Üniversitesi, Sağlık Bilimleri Fakültesi, nilhan.atsu@kent.edu.tr

- Orşit, epididimit, testis torsiyonu ve testis hematomu görülebilir. Bu durumların ürolojik aciller arasındadır ve üroloji uzmanına konsülte edilmelidir.

Nöroloji

- HSP serebral vaskülite de neden olabilir. Buna bağlı olarak iskemi, ödem, kanama ve enfarkt gelişebilir.
- Hastalarda baş ağrısı, ataksi, nöbet, ensefalopati, myelopati, fokal nörolojik defisit, intraserebral kanama ve santral/periferal nöropati gözlenebilir.
- Hastalar bu yönlerden de takip edilmeli ve yukarıda bahseilen nörolojik semptom/bulgular varlığında nöroloji uzmanının görüşü alınmalıdır.

Göğüs Hastalıkları

- IgA vasküliti olan hastalarda bozulmuş akciğer diffüzyon kapasitesi, akciğer görüntülemelerinde hafif düzeyde interstisyel değişiklikler raporlanmış olduğundan gerekli durumlarda göğüs hastalıkları görüşü alınabilir.

Kardiyoloji

- Kardiyak tutulum sanıldığından daha sık olarak görülür ve hastalık başlangıcından 2-4 hafta sonra ortaya çıkar.
- Çarpıntı, bradikardi ve göğüs ağrısı görülebilir.
- İletim anomalileri, miyokardit, endokardiyal vaskülit ve konjestif kalp yetmezliği gelişebilir. Hastaların kardiyak muayeneleri rutin olarak yapılmalı ve kardiyoloji uzmanınca değerlendirilmelidir.

Göz Hastalıkları

- Henoch-Schönlein purpurası keratit ve üveite de neden olabilir. Retinal arter vasküliti, episklerit ve retinal arter oklüzyonu gelişen vakalar bildirilmiştir. Bu durumlarda acil olarak göz hastalıkları uzmanından görüş alınmalıdır.

KAYNAKLAR

Gardner-Medwin JMM, Dolezalova P, Cummins C, Southwood TR. Incidence of Henoch-Schönlein purpura, Kawasaki disease, and rare vasculitides in children of different ethnic origins. Lancet. 2002;360(9341):1197–202.

Hetland LE, Susrud KS, Lindahl KH, Bygum A. Henoch-schönlein purpura: A literature review. Acta Derm Venereol. 2017;97(10):1160–6.

Kang Y, Park J su, Ha YJ, Kang M il, Park HJ, Lee SW, et al. Differences in clinical manifestations and outcomes between adult and child patients with henoch-schönlein purpura. J Korean Med Sci. 2014;29(2):198–203.

Calvo-Río V, Loricera J, Mata C, Martín L, Ortiz-Sanjuán F, Alvarez L, et al. Henoch-schönlein purpura in Northern Spain: Clinical spectrum of the disease in 417 patients from a single center. Med (United States). 2014;93(2):106–13.

Yang YH, Hung CF, Hsu CR, Wang LC, Chuang YH, Lin YT, et al. A nationwide survey on epidemiological characteristics of childhood Henoch-Schönlein purpura in Taiwan. Rheumatology. 2005;44(5):618–22.

Alexander K C Leung, Benjamin Barankin KFL. Henoch-Schönlein Purpura in Children: An Updated Review. Curr Pediatr Rev . 2020;16(4):265–76.

Trapani S, Micheli A, Grisolia F, Resti M, Chiappini E, Falcini F, et al. Henoch Schonlein purpura in childhood: Epidemiological and clinical analysis of 150 cases over a 5-year period and review of literature. Semin Arthritis Rheum. 2005;35(3):143–53.

Chang WL, Yang YH, Lin YT, Chiang BL. Gastrointestinal manifestations in Henoch-Schönlein purpura: A review of 261 patients. Acta Paediatr Int J Paediatr. 2004;93(11):1427–31.

Çakıcı EK, Gür G, Yazılıtaş F, Eroğlu FK, Güngör T, Arda N, et al. A retrospective analysis of children with Henoch–Schonlein purpura and re-evaluation of renal pathologies using Oxford classification. Clin Exp Nephrol [Internet]. 2019;0(0):0. Available from: http://dx.doi.org/10.1007/s10157-019-01726-5

Ghrahani R, Ledika MA, Sapartini G, Setiabudiawan B. Age of onset as a risk factor of renal involvement in Henoch-Schönlein purpura. Asia Pac Allergy. 2014;4(1):42.

Dalpiaz A, Schwamb R, Miao Y, Gonka J, Walzter W, Khan SA. Urological manifestations of henoch-schonlein purpura: A review. Curr Urol. 2014;8(2):66–73.

Bulun A, Topaloglu R, Duzova A, Saatci I, Besbas N, Bakkaloglu A. Ataxia and peripheral neuropathy: Rare manifestations in Henoch-Schönlein purpura. Pediatr Nephrol. 2001;16(12):1139–41.

Stefek B, Beck M, Ioffreda M, Gardner L, Stefanski M. Henoch-Schönlein purpura with posterior reversible encephalopathy syndrome. J Pediatr. 2015;167(5):1152–4.

Chaussain M, de Boissieu D, Kalifa G, Epelbaum S, Niaudet P, Badoual J, et al. Impairment of lung diffusion capacity in Schönlein-Henoch purpura. J Pediatr. 1992;121(1):12–6.

Bérubé MD, Blais N, Lanthier S. Neurologic manifestations of Henoch-Schönlein purpura. Handb Clin Neurol. 2014;120:1101–11.

Du L, Wang P, Liu C, Li S, Yue S, Yang Y. Multisystemic manifestations of IgA vasculitis. Clin Rheumatol. 2021;40(1):43–52.

Aristodemou P, Stanford M. Therapy insight: The recognition and treatment of retinal manifestations of systemic vasculitis. Nat Clin Pract Rheumatol. 2006;2(8):443–51.

Min Seung Kang, Han Jo Kwon JEL. Retinal Capillary Reperfusion from Ischemic Retinal Vasculitis in Henoch-Schönlein Purpura: A Case Report. Ocul Immunol Inflamm . 2021;17(1).