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Henoch-Schönlein Purpura Presenting as Mononeuritis Multiplex

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Dear Editor,

Mononeuritis multiplex (MM) is a syndrome of the peripheral nervous system (PNS) involving progressive multifocal peripheral nerve lesions. Its distribution is typically asymmetric, and the course of the disease varies with the underlying etiology. MM is associated with several medical conditions, including vasculitis, immune-mediated diseases, diabetes, infections, neoplasms, infiltrative diseases, and drugs. Henoch-Schönlein purpura (HSP) is an IgA-mediated small-vessel vasculitis, and also a rare cause of MM.[1] The prevalence of HSP is lower in adults than in children, and no cases of MM with HSP in Korea have been reported previously. Here we report a case of MM with HSP in a Korean patient.

A 57-year-old female patient visited the emergency room due to weakness of her right ankle and right hand. She had been diagnosed with rheumatoid arthritis (RA) 10 years previously due to multiple arthralgia. Her RA was well controlled with medication, and she had no arthralgia at the present visit. One day before her presentation she had experienced shock-like pain that developed along her right upper arm and in her right ankle. A neurologic examination revealed weakness of wrist flexion, thumb abduction, and ankle dorsiflexion, and hypesthesia of the right finger tips and foot dorsum.

Purpuric skin lesions had developed on both feet 1 year before the current presentation, and the patient reported diffuse nonspecific abdominal pain (Supplementary Fig. 1 in the online-only Data Supplement). She had undergone a skin biopsy 1 month previously, which revealed leukocytoclastic vasculitis and IgA deposition within the purpuric lesion. The patient was diagnosed with HSP based on IgA vasculitis, purpuric skin lesions, and diffuse abdominal pain.

A nerve conduction study revealed multifocal axonal neuropathy (Table 1), leading to a diagnosis of MM. Laboratory tests revealed mild leukocytosis, elevated C-reactive protein and increased erythrocyte sedimentation rate (60.3 mg/L and 45 mm/hr, respectively). The findings of serologic tests were negative for antineutrophil cytoplasmic antibody, anti-SS-A antibody, anti-SS-B antibody, antinuclear ribonucleoprotein antibody, anti-Smith antibody, antihistone antibody, anti-double-stranded DNA antibody, cold agglutinin, and cryoglobulin. She was positive for antinuclear antibody at a titer of 1:640 and with a nucleolar pattern, and her serum rheumatoid factor was elevated (to >120 IU/L).

The patient underwent methylprednisolone pulse therapy (1 g per day for 5 days) without significant complications, and she was maintained on low-dose oral steroid. Her ankle and wrist weakness had improved at the 1-year follow-up, but the sensory loss persisted.

Among various etiologies, the most common cause of MM is vasculitic process, while HSP is rare.[2] HSP is an immune complex vasculitis that predominantly affects small vessels, and is characterized by nonthrombocytopenic palpable purpura, abdominal involvement, arthralgia, and renal involvement. Its pathophysiology remains unclear, but IgA is thought to play an essential role.[3] HSP mainly occurs in children and is uncommon in adults. Further-

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Table 1. Nerve conduction study revealed low compound motor action potentials in the right median and bilateral peroneal nerves, and low sensory nerve action potentials and slow sensory conduction velocities in the right superficial peroneal nerve

Site	Right			Left		
	Onset (ms)	Amp (mV)	Vel (m/s)	Onset (ms)	Amp (mV)	Vel (m/s)
Motor						
Median						
Wrist	2.50	0.5		2.42	12.8	
Elbow	6.41	0.5	54	5.78	12.1	61
Axilla	8.05	0.5	64	7.97	11.2	70
Ulnar						
Wrist	2.27	9.3		2.42	6.8	
B. elbow	6.02	8.8	60	5.78	6.8	62
A. elbow	7.73	8.4	60	7.58	6.8	57
Axilla	8.83	7.7	66	8.59	6.6	67
Peroneal						
Ankle	3.98	0.5		4.84	0.9	
B Fib	11.17	0.3	43	11.56	0.8	44
Tibial						
Ankle	3.59	17.0		4.45	13.6	
Knee	11.09	13.8	43	12.34	13.1	43
	Peak (ms)	Amp (μV)	Vel (m/s)	Peak (ms)	Amp (μV)	Vel (m/s)
Sensory						
Median						
F-W	2.59	41.3	52	2.38	64.1	53
W-E	3.41	38.2	58	3.78	31.4	52
E-A	2.06	101.9	58	1.94	130.2	58
Ulnar						
F-W	2.50	13.0	50	2.31	39.0	50
W-E	3.69	47.9	53	3.72	62.0	52
E-A	2.72	22.1	57	2.09	45.7	63
Sup peroneal	2.84	4.3	30	2.28	12.2	39
Sural	2.47	14.9	35	2.31	17.2	42
	Latency (ms)			Latency (ms)		
H-reflex						
Tibial	32.43			32.48		

E-A: elbow-axilla, F-W: finger-wrist, W-E: wrist-elbow.

more, PNS involvement in HSP is extremely rare. There have been a few reports regarding MM in HSP, but most were in children, with only rare cases in adults.

The clinical course of HSP in childhood is usually benign, but its clinical manifestation and prognosis are different in adults. Adult-onset HSP more commonly involves the kidneys, and severe renal involvement is more common in adults.[4] HSP is often treated conservatively in mild cases, but colchicine, dapsone, antileukotriene agents, corticosteroids, and other immunomodulating agents can be administered in severe cases. Although no well-organized trial has investigated immunomodulating agents, several agents have been used in combination with corticosteroids.[3] As mentioned above, due to its low prevalence, not much as is known about MM with HSP as other vasculitic MMs. Hence, the effectiveness of immunomodulating therapies in MM with HSP remains to be validated.

Our patient had MM with a diagnosis of HSP upon hospitalization. The etiologic evaluation for MM revealed no other findings. Although she had a prior history of RA, no symptoms of RA were evident at the time of MM. Though she exhibited positivity for antinuclear antibody and elevation of rheumatoid factor, these findings are not specific for RA and can be detected in several autoimmune diseases. Furthermore, the biopsy of the skin lesion revealed perivascular IgA deposition, reflecting underlying IgA-related pathology. Therefore, HSP rather than RA was thought to be responsible for MM in this patient.

The relatively poor prognosis of HSP in adults makes its

prompt recognition and adequate management important. Furthermore, organized and systematic data regarding MM would facilitate the management of vasculitic neuropathies including HSP in the future.

Supplementary Materials

The online-only Data Supplement is available with this article at https://doi.org/10.3988/jcn.2018.14.1.112.

Conflicts of Interest

The authors have no financial conflicts of interest.

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