MONONEURITIS MULTIPLEX IN A CHILD WITH HENOCH-SCHÖNLEIN PURPURA

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SUMMARY : *A 13 year old boy presented with the complaints of left leg pain, mild muscle weakness, and a few, small, macular rashes on his right malleol associated with arthritis of right wrist and elbow. The diagnosis of Henoch-Schönlein Purpura (HSP) with kidney involvement was made. Shortly after he developed mononeuritis multiplex. The clinical spectrum of HSP and the involvement of peripheral nerves is discussed.*

Key Words : *Mononeuritis Multiplex, Henoch-Schönlein Purpura, Necrotizing Vasculitis.*

INTRODUCTION

Peripheral nervous system (PNS) lesions are the most common and often the earliest neurological manifestation of Connective Tissue Disorders (CTD) or Systemic Necrotizing Vasculitis (SNV) (9). However PNS complications occur very rarely in HSP (5, 8).

We report a patient in whom the peripheral nervous system lesion as mononeuritis multiplex occured early in the clinical picture of HSP.

CASE REPORT

A 13 year old boy was admitted to Gazi University Hospital with arthritis of both elbows and knees, lower extremity weakness, following an upper respiratory tract infection 20 days previously.

Physical examination revealed a well developed boy with high fever and blood pressure of 145/100 mm Hg. He had a few small maculer rashes on the right malleol and edema on the dorsum of both hands. All his joints and muscles were tender with palpation and movement. His deep tendon reflexes (DTR) were hypoactive and he has mild motor weakness of lower extremities.

Laboratory examinations : Urinalysis showed (++) proteinuria, leucocyturia, hematuria and silendiruria. Except leucocytosis, high CRP, ASO and high sedimantation rate levels, the rest of the laboratory findings were normal. Kidney needle biopsy was performed. The light microscopy findings showed focal segmental necrotizing glomerulonephritis. Immunofleurescent microscope revealed diffuse mesengial IgA deposition; characteristic of HSP.

Electrophysiological Findings : Nerve conduction Studies (NCS) performed on admission disclosed low amplitude Compound Sensory Nerve Action Potentials (CNAP) of the finger-wrist and wrist-elbow segments of the left median and sural nerves. Left Tibial Nerve Conduction Velocity (NCV) was slightly slow with a reduced Compound Muscle Action Potential (CMAP) amplitude. Right peroneal and left tibial F responses were unobtainable.

Needle EMG of the left deltoid and anterior tibial shaved fibrillation potentials and positive sharp waves, with small short, polyphasic Motor Unit Potentials (MUP). A repeat study seven months later, disclosed essentially the same underlying NCS abnormality with a complete resolution of the myopathic pattern. Fourteen months later, right ulnar nerve abnormalities were still persisting with an absent CNAP of the left sural nerve and a recovered right sural nerve function. Low CNAP amplitude of the left peroneal nerve was still observed (Table 1). Electromyography (EMG) was not remarkable except for the giant MUPs (> 3 mV) in the right dorsal interosseus and left anterior tibial and vastus lateralis muscles.

On follow up he was put on prednisolon therapy of 60 mg/m^2/day. One weak after, he developed hypoesthesia on his left foot and right hand. Three weeks later his pain and muscle tenderness vanished. When he was discharged eight weeks later from the onset of his sypmtoms, he only had hypoestesia of the right hand and left foot. His prednisolone therapy was tapered to 15 mg/m^2/day continued for another six months. Seven months later his neurological examination was normal except hypoesthesia in the distal right ulnar and left sural dermatomes. Fourteen months later he neither had any complaints nor any neurological abnormalities, but his EMG was still abnormal.

DISCUSSION

HSP is one of the most common type of CTD and SNV of childhood (2, 5). Unusal manifestations of HSP include ocular, oesophageal, pulmonary, scrotal, testicular and neurological involvoments (1). Clinically significant vasculitic neuropathy is most commonly observed with PAN, RA, or SLE as mononeuritis multiplex (3). Peripheral

	Amplitude (sensory μV, Motor mV)		Distal Latency (msec)		Nerve Conduction Velocity (m/s)		F-wave (msec)	
Nerve	Rt	Lt	Rt	Lt	Rt	Lt	Rt	Lt
Median sensory	> 10				> 39.4			
First exam		4.2				48.3		
Second exam	6.0				35.3			
Third exam	5.2				40.5			
Ulnar sensory	> 7				> 37.3			
First exam								
Second exam	NP							
Third exam	NP							
Sural	> 5				> 33.8			
First exam		4				35.1		
Second exam	NP	3.6				34.0		
Third exam	8	NP			38.9			
Median motor	> 4		< 3.8		> 49.7		< 24	
First exam		8.8		2.6		54.7		
Second exam	15		3.4		52.2		25.6	
Third exam	9		3.4		50.0		25.2	
Ulnar motor	> 7		< 3.3		> 49.9		< 24	
First exam								
Second exam	4.5		3.6		50.0		29.6	
Third exam	3.5		3.1		55.0		28.4	
Deep peroneal	> 3.5		< 5		> 40.9			
First exam	4.2		4.6		42.9		NP	
Second exam	5.6	0.7	3.8	6.4	44.1	37.4	44.4	52
Third exam	5.8	1.5	4.0	5.2	41.6	42.8	44.4	NP
Tibial	> 3.5		< 6.0		> 39.6			
First exam		0.9		5.5		38.9		NP
Second exam	13.8	7.7	6.6	6.2	43.9	46.7	47.6	
Third exam	7.0	5.0	5.2	6.8	48.1	42.4	48.4	50.4

NP : No potential

Table 1 : Nerve conduction sudies.

neuropathies may present as mononeuritis multiplex, extensive mononeuritis (overlapping), distal sensorymotor neuropathy or cuteneous neuropathy (4, 6). In most patients there is a rapid progression of the neuropathy resulting in complete nerve paralysis within hours or days. Generally complete recovery occurs, but permanent deformity although rare, remains (7).

EMG findings of our patient revealed the existence of a diffuse sensorimotor peripheral neuropathy with a predominant sensory involvement, together with myopathy. As myopathy resolved, markedly a symmetric involvement of several sensory and motor nerves in different limbs superimposed on the diffuse peripheral neuropathy. These findings were compatible with an overlapping mononeuropathy multiplex, as is commonly seen in peripheral nervous system vasculitis (4).

In children with HSP peripheral nervous system involvement is seldom seen. A few reports concerning femoral, siatic, peroneal, facial nerve involvement and polyneuropathy exist (2, 8). To our knowledge this is the first child descrebed with HSP and MNM. The petechial, purpuric rash and other common sypmtoms characteristic of HSP may be presenting signs of other serious vasculitic illness.

Differential diagnosis of large artery vasculitis or PAN or HSP-like illness is a great challenge and depends on a deep knowledge of the clinical and laboratory findings of CTD or SNV (5).

Patients with HSP or other types of SNV associated with peripheral neuropathy are treated with corticosteroids or cytostatics succesfully (4, 6). The diagnosis of HSP should be considered in children who develop MNM in association with or without cutaneous manifestations, arthralgia, muscle pain and weakness.

The neuritis may preceede, accompany or follow the clinical manifestations. A through evaluation for each patient and a carcful follow up should be implemented

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