Painful Mononeuritis Multiplex in Idiopathic Thrombocytopenic Purpura

An 11-year-old boy presented with severe paresthesia involving the left leg and the right hand of three weeks duration. He had severe pain and mild numbness in the lateral aspect of left leg and right forearm and hand. General examination showed petechial and purpuric skin lesions over the trunk and extremities. There was no hematoma in the vicinity of affected nerves. Neurological examination revealed severe dysesthesia and diminished sensations in the left peroneal and right ulnar nerve distribution. Peripheral nerves were not enlarged. He had no history of fever, joint pain, oral ulcers, diabetes mellitus or contact with leprosy. Though the child had a history of intermittent nasal bleeding and petechial lesions for seven months, he was diagnosed to have idiopathic thrombocytopenic purpura (ITP) only four weeks ago. As he did not adequately respond to corticosteroids and immunoglobulins, he was admitted for a splenectomy considering a diagnosis of chronic ITP. There was no history of intake of any other drugs.

Nerve conduction studies showed evidence of asymmetric axonal sensorimotor neuropathy (mononeuritis multiplex). Needle electromyography and sural nerve biopsy were deferred because of thrombocytopenia. Hemoglobin was 12.4 g%, platelet count was 3000/cu.mm and ESR was 25 mm/hour. Fasting blood sugar was 90 mg%. Antinuclear antibody, rheumatoid factor and LE cells were negative. Serum complement was 90%. A final diagnosis of mononeuritis multiplex related to chronic ITP was made. The child was treated with carbamazepine and amitryptiline for symptomatic relief and advised follow up. Meanwhile, he underwent splenectomy uneventfully. His platelet counts improved subsequently. At a follow-up after three months, his neuropathic symptoms were markedly better and at six months, he was asymptomatic. He has since been followed up for three years, without recurrence of neuropathy.

Neuropathy occurring in the setting of bleeding and coagulation disorders is uncommon. Neuropathy occurs in 15% of patients with hemophilia, which is related to hematoma occurring in the vicinity of peripheral nerves(1). However, our patient did not have any evidence of hematoma. There are occasional reports of Guillain-Barre syndrome (GBS) occurring in patients with ITP(2). It has been proposed that the association between GBS and ITP is plausible as both can occur following viral infections, are auto-immune diseases, and respond well to immunoglobulins. However, the association of mononeuritis multiplex and ITP, as seen in our case, is intriguing. There was no evidence of hematoma in previous cases too presenting with mononeuritis multiplex and ITP(3). However, histo-pathological examination showed evidence of intraneuronal hemorrhage located beneath epineurium between nerve fascicles(3). This finding suggests that neuropathy in ITP is a direct effect of thrombocytopenia causing hemorrhage in the affected nerves. However, one should exclude conditions such as vasculitis(4) and systemic lupus erythematosus(5), which can result in both thrombocytopenia and mononeuritis multiplex. All previously reported cases of ITP-associated mononeuritis multiplex occurred in adults, and our case, to the best of our knowledge, is the first such instance in a child.
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REFERENCES


