Paraneoplastic polyneuropathy associated with inflammatory myofibroblastic tumor in a pediatric patient

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OBJECTIVES

Inflammatory myofibroblastic tumor (IMT) is an uncommon, usually benign (non-cancerous) tumor made up of cells called myofibroblastic spindle cells. It usually develops in children or young adults but can affect people of any age. An IMT can occur in almost any part of the body but is most commonly found in the lung, orbit (eye socket), peritoneum (lining of the abdominal cavity and internal organs), and mesentery. The term ''paraneoplastic'' means that the disorder is not caused by the tumor itself, but by the immunological reactions that the tumor produces. The effects of paraneoplastic polyneuropthy can remit entirely, although there can also be permanent effects. We present the first case of a 14-year-old girl who developed paraneoplastic polyneuropathy associated with uterine inflammatory myofibroblastic tumor.

METHODS

Diagnosis, treatment and clinical course of the case with polyneuropathy due to uterine inflammatory myofibroblastic tumor are discussed.

CASE

A 14-year-old female patient with 4 months of abnormal uterine bleeding was diagnosed with an inflammatory myofibroblastic tumor of uterus in August 2021. The patient was given two cycles of chemotherapy protocol including ifosfamide, vincristine, actinomycin-D and doxorubicin. Then she was admitted to the intensive care unit due to febrile neutropenia and septic shock after chemotherapy. Symptoms suggestive of neuropathy such as pain in the legs and paresthesia started 1-2 days before admission to the intensive care unit. She remained intubated for 18 days. During the intensive care follow-up, the patient developed hypertension and had focal seizures. Her electroencephalogram revealed slow background rhythm for her age but no epileptiform anomaly. Brain magnetic resonance imaging (MRI) was performed due to left hemiparesis. MRI was compatible with posterior reversible encephalopathy syndrome (PRES). Antihypertensive treatment was started. PRES improved and control brain MRI was normal. One week after PRES improves, she developed severe leg pain, paresthesia, and inability to walk. On physical examination, she was conscious and emotionally depressed. Upper and lower extremity muscles were atrophied. Deep tendon reflexes could not be obtained. There was a significant distal-proximal difference in the superficial sensory examination, especially in the lower extremities. Superficial sensory loss was more prominent in the distal regions than in the proximal. Vibration sense durations were reduced in all extremities. With these findings, polyneuropathy was considered clinically. Because more than 3 months have passed since the use of vincristine, polyneuropathy due to vincristine was not considered. (She was started on gabapentin treatment for severe pain. Differential diagnosis was made in terms of paraneoplastic neuropathy, critical illness neuropathy and CIDP. Despite being on a mechanical ventilator for more than a week, the diagnosis of critical illness neuropathy was dismissed due to the onset of neuropathy findings before the intensive care unit admission. The protein level in the cerebrospinal fluid (CSF) was 19 mg/dl and no cells were seen in CSF. Sensory and motor responses could not be obtained in nerve conduction studies so it was compatible with severe sensorimotor polyneuropathy. Needle EMG could not be evaluated because she had severe paresis. She developed respiratory distress and then she was intubated. Due to recurrent, unsuccessful extubation attempts, she underwent tracheotomy. On suspicion of chronic inflammatory demyelinating polyneuropathy (CIDP), plasmapheresis together with intravenous immunoglobulin (IVIG) was applied but she did not benefit from treatment. The diagnosis of CIDP was ruled out due to the inability to show specific EMG findings, failure to respond to IVIG and plasmapheresis, and normal CSF protein level. She underwent total abdominal hysterectomy. There was no residual intra-abdominal tumor. The paraneoplastic antibody panel that studied from her serum was negative.

In the control nerve conduction studies, motor conduction velocities could not be measured, sensory conduction velocities in median and ulnar nerves were found at the lower limit.

There was no change in polyneuropathy findings after tumor resection. The patient died 1 year after starting treatment.

CONCLUSIONS

In our case, it has been shown that paraneoplastic polyneuropathy may develop in inflammatory myofibroblastic tumor cases and tumor resection may not cause a significant improvement in clinical findings.