Short report

Pontocerebellitis—a rare manifestation of mononucleosis

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SUMMARY A patient with pontocerebellitis associated with infectious mononucleosis is described. The clinical picture with transitory limb and truncal ataxia was compatible with previous reports of cerebellitis. CT showed wide pontocerebellar cisterns and a hypodense area in the pons.

Central nervous system involvement in infectious mononucleosis is relatively rare, but various manifestations including meningoencephalitis, meningitis, transverse myelitis, and cerebellitis have been reported. We here report a case of pronounced cerebellar ataxia without other neurological abnormalities associated with infectious mononucleosis and describe its appearance in computed tomography (CT).

Case history

A 16-year-old schoolboy, previously in good health, fell ill with sore throat, mild headache and fever on 25 August 1981. Two days later he experienced unsteady gait, vertigo, nausea and vomiting. Because of increasing difficulties in walking he was admitted to the local hospital on 28 August. His speech was dysarthric. Incoordination was observed in both lower and upper extremities. The throat and tonsils were injected. A few small lymph nodes could be palpated in the neck. The ataxia increased and the patient could no longer sit. Laboratory studies showed a mild leukocytosis (12.1 × 10⁹/l) with 78% lymphocytes in the differential count, some of them being “atypical.” The liver transaminases S-GOT and S-GPT were mildly elevated. Lumbar puncture yielded clear cerebrospinal fluid (CSF) with a normal cell count and normal concentrations of glucose and protein. A commercial test for serum heterophil antibody (Monospot®) was positive. The patient was referred to the University Central Hospital, Helsinki. On admission clear-cut dysarthria was observed. There was marked ataxia of all limbs and of the trunk. He could neither sit nor write or hold a fork. His general muscle tone was diminished. No neck stiffness was noted. The patient was tired but alert. No other neurological abnormalities could be found. In particular no nystagmus was ever elicited. The ataxic symptoms reached their peak within ten days of the onset of the fever and pharyngeal symptoms. After another ten days the patient could walk, but he had to take sidesteps. His speech was normal. At discharge a week later he could write his name and walk with a mildly unsteady gait. At follow-up two months after the onset he was symptom-free and had resumed his normal activities including cycling.

The Paul-Bunnell reaction was positive twice. The titres were 112 and 224 before and 56 to 112 after absorption with guinea pig kidney. The titre was 0 after absorption with beef red cells. The serum IgM antibody titre to Epstein-Barr virus (EBV) was 1/640 nine days after the onset of the pharyngeal symptoms and gradually declined thereafter to zero within two months. Simultaneously with the highest serum anti-EBV-IgG titre (1/1280) a low titre of 1/1 of the same antibody could be detected in the cerebrospinal fluid. The total count of CSF showed 2 × 10⁹/l erythrocytes and 6 × 10⁹/l leukocytes. Ninety-five percent of the CSF cells were lymphoid cells. The proportion of large lymphoid cells was 60% of the total lymphoid cell population, implying a clear lymphoid reaction (the upper normal value in our laboratory is 31%). There were also some mature plasma cells in the CSF. The CSF-IgG index was 0-54 (normal). Oligoclonal IgG of the CSF was detected in polyacrylamide gel electrophoresis. A CT scan of the brain at the peak of the symptoms one week after the onset of the neurological symptoms (4 September) exhibited an area of lower density in the pons. In a further scan ten days later the hypodense area was still visible (fig). The last CT scan did not exhibit any hypodense areas, but the pontocerebellar cisterns were large as in the previous scans. The EEG was recorded on four occasions. Slight, transitory abnormalities without any focal signs were detected.
Discussion

In the case presented the classical features of EBV-mononucleosis were present, together with marked truncal and limb ataxia as the sole neurological finding. To our knowledge some twenty cases of cerebellar ataxia associated with EBV-mononucleosis have been reported so far. It is not known whether the encephalitic complications of infectious mononucleosis are due to direct invasion by the virus of the central nervous system, or to immunologic phenomena without viral invasion. The presence of EBV-antibodies has been demonstrated in the CSF of some cases of infectious mononucleosis with cerebellar ataxia, suggesting the presence of the virus in the central nervous system. We were not able to demonstrate any signs of local central nervous system antibody production to EBV, as only a very low titre of EBV-antibody could be detected in the CSF at the time when serum antibodies were at their peak.

Owing to the benign course of the cerebellar symptoms in infectious mononucleosis, few histopathological correlations are available concerning the affected structures. In the present case the CT scan of the brain exhibited wide pontocerebellar cisterns and a small hypodense area in the pons without any mass lesions. The type and localisation of CT findings were compatible with the neuropathological reports of two necropsy cases of mononucleosis with central nervous system involvement. These include Purkinje cell loss in the cerebellum and degenerative changes in the medulla, at the level of olivary nuclei. The essential neuropathological alterations of this condition are thus more precisely expressed by the term “pontocerebellitis” than by the term “cerebellitis.” It is interesting to note that similar neuropathological findings have been described in patients with classical olivo-ponto-cerebellar atrophy.

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References