Intermittent bilateral superior palpebra ptosis in a 20-month-old infant

Sirs,

In June 2009, a previously healthy Caucasian fully immunised 20-month-old girl was referred to our Department with a 10-day history of intermittent bilateral ptosis of the superior palpebra. Each episode lasted 3–6 hours and recurred at different intervals during the day; sleeping was regular and the patient was otherwise well (Fig. 1A, 1B). Three weeks before, she had been admitted to the regional hospital with persistent high fever lasting seven days, refractory to antibiotics and antipyretics, macular rash on the trunk, and eyes redness. Laboratory tests showed: erythrocyte sedimentation rate (ESR) of 80 mm/h, C-reactive protein (CRP) 10.6 mg/dl, and white blood cells (WBC) 22.4×10^9/L with 82% neutrophils. As the fever dropped and the other symptoms resolved, on day eleven from hospitalisation she was discharged with the diagnosis of viral infection. On admission at our department she was in a good general condition, afebrile and vital. A sheet-like desquamation of both hands and feet was the only remarkable sign. The ocular fundus of both eyes was normal, pupils bilaterally symmetrical, round and reactive to light and accommodation. Extra ocular movements were intact, ruling out ocular muscle palsy. Electroencephalogram and neurological assessment were normal. Full laboratory work-up revealed a mild increased ESR (30 mm/h), reduced haemoglobin for the patient’s age (10.1 g/dl), and high platelet count (1000×10^9 per mm^3). Serological tests for the most common infantile viral infections were unremarkable. The photographs show the child with intermittent palpebra ptosis at different times on the same day.

Neurological manifestations including, irritability, seizures, cranial nerve palsies, transient haemiplegia, and ataxia are not uncommon in KD (1-6) but palpebra ptosis has been described only in a case report in the Chinese literature (7). The pathophysiology of neurological manifestations of KD is unknown and mechanisms such as aseptic choriomeningitis, leptomeningitis, ganglionitis, ischaemic vasculitis of cranial arteries and neuritis of both central and peripheral nerves, have been suggested (5). Palpebra ptosis might be related to ischaemic vasculitis of arteries supplying the elevator muscles of the palpebra and to immunological mechanisms that may induce palpebra nerve dysfunction. Neurological manifestations of KD are usually transient and resolve spontaneously, while definitive alterations are relatively rare (1-6). At the last follow-up, our patient is growing well with no residual neurological symptoms, and no coronary alterations even though the appropriate therapy had been omitted. This is not in contrast with the diagnosis, as coronary aneurysms occur in about 15%–25% of KD patients who do not receive IVIG therapy (8). Our patient presented an incomplete clinical picture of KD as she had three typical clinical manifestations (conjunctivitis, rash, and digit peeling) along with high fever of seven days duration (8-10). Intermittent bilateral palpebra ptosis resulted to be critical in the diagnosis. These neurological signs should be kept in mind in children with doubtful KD as a helpful clue to guess the diagnosis.

Fig. 1A, 1B. The photographs show the child with intermittent palpebra ptosis at different times on the same day.

All the authors were involved in this report. FF wrote the manuscript, GCa made the diagnosis, FLa and GCa cured the patient, MMF and VA made the neurological evaluation.

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References