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First presentation of multiple sclerosis: eight and a half plus syndrome

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Abstract

Isolated cranial nerve palsy as the first clinical relapse in multiple sclerosis is very rare. In multiple involvement, the abducens and facial nerve are most commonly affected together. Herein, we report a newly diagnosed case of multiple sclerosis presenting as eight and a half plus syndrome, which is very rare and involving the facial colliculus and its neighbourhood as the first clinical relapse.

Keywords: Multiple sclerosis, Internuclear ophthalmoplegia, facial colliculus, eight and a half plus syndrome

CASE REPORT

A 21-year-old woman presented four days ago with increasing dizziness and horizontal diplopia. Subsequently, unsteadiness in walking, postural tremors in the whole body and head, and weakness in the left facial half developed. There was no recent history of infection, trauma, malabsorption or toxication, or any known disease or medical treatment. Examination revealed a conjugate gaze palsy (left eye with complete restriction in horizontal gaze, right eye with restriction in inverted gaze) and rotatory nystagmus striking bilateral gaze direction. Light reflexes, pupils, fundus, and visual field were normal. There was peripheral mild fascial paralysis on the left side. While comfortable in the supine position, truncal ataxia and titubation were observed in sitting or standing.

3 T magnetic resonance imaging including thin slice T2-weighted, fluid-attenuated inversion recovery and contrast enhanced T1-weighted sequences revealed left occipital periventricular and temporal juxta-cortical hyperintense demyelinating lesions and a demyelinating lesion with mild diffusion restriction and mild contrast enhancement involving the left facial colliculus in the dorsal tegmentum of the caudal pons. (Figure 1)

Routine serum tests, infectious (bacteria culture tests, brucellosis, syphilis, borreliosis, human immunodeficiency virus (HIV), herpes simplex virus (HSV), varicella-zoster virus (VZV), cytomegalovirus (CMV), Epstein-Barr virus (EBV), toxoplasmosis, hepatitis, tuberculosis), and rheumatological (except antinuclear antibody (ANA) 1/100 endpoint positivity (ANA profile negative)) investigations and tests were normal. Rheumatology and cardiology (with transthoracic echocardiogram (TTE)) consultation was normal. In cerebrospinal fluid (CSF) examination, the viral panel was negative; there were no cells, the protein was 16.4 mg/dL, and glucose and chlorine ratios were within normal limits compared to serum. The oligoclonal band was positive (type 2), and the number of bands was 5. Anti aquaporin-4 antibody (AQP4-Ab) and anti myelin oligodendrocyte glycoprotein (MOG-Ab) (cell-based assay) were negative in serum. Thus, multiple sclerosis was diagnosed according to the revised McDonald 2017 criteria. Intravenous methylprednisolone (IVMP) was given one g per day for ten days, and then teriflunomide 14mg/day was started. In the second month, eye movements, ataxia, and titubation completely recovered, and the expanded disability status scale (EDSS) score became zero.

DISCUSSION

The one-and-a-half syndrome is a combination of horizontal conjugate gaze paralysis (involvement of the pontine reticular formation (PPRF) and abducens nucleus) and ipsilateral Internuclear Ophthalmoplegia (INO) (involvement of the medial longitudinal fasciculus (MLF)). All

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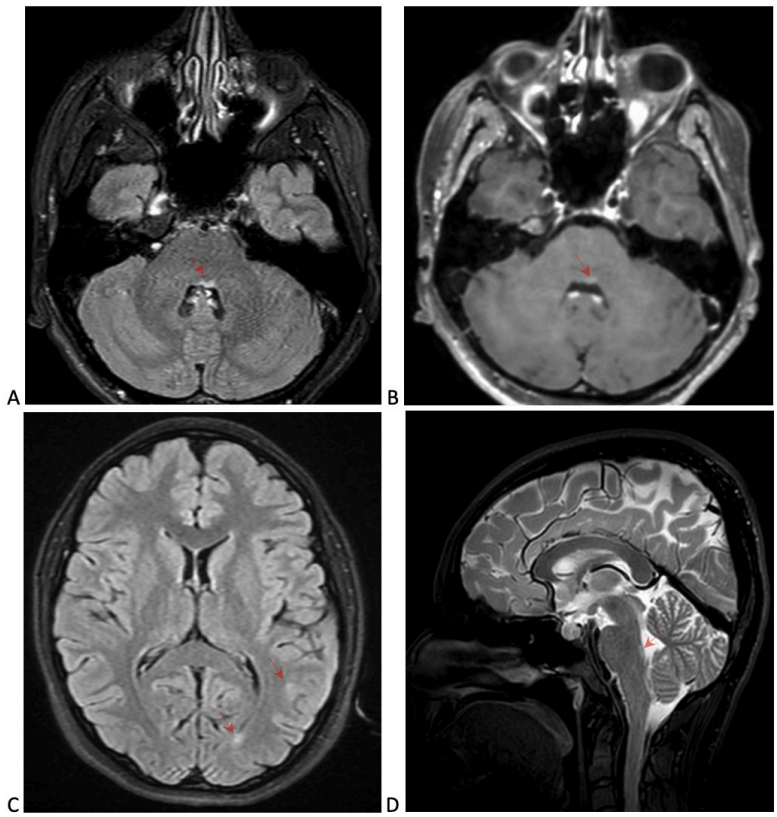

Figure 1. Red arrows; hyperintense demyelinating lesion in the facial colliculus (A and D), mild contrast enhancement (B), periventricular and juxtacortical demyelinating plaques (C). A,B and D; CISS, C; FLAIR MRI

horizontal movements are restricted except the abduction of one eye.[1] Eight-and-a-half syndrome occurs when peripheral-type facial paralysis is added to one-and-a-half syndrome due to ipsilateral involvement of the intraaxial fascicle of the facial nerve in the immediate neighborhood (ipsilateral dorsal tegmentum of the caudal pons).[2] It is infrequent; about thirty cases have been reported in the literature.[3,4] The most common causes of both are cerebrovascular diseases, demyelination, tuberculoma, tumors, or metastasis, respectively.[5,6] If the affected area is enlarged, other clinical findings may be added. Bilateral facial paralysis, complete loss of vertical saccades and pursuit, vertical gaze paralysis, contralateral hemiparesis have been reported and described as 'eight and a half plus' or 'nine syndrome'.[6-10]

The MLF receives projections from the vestibulocochlear nerve and provides connections to cranial nerves 3-4-6. In the reticular formation in the anterior neighborhood of the MLF in the dorsal tegmentum of the caudal pons, there are reticular nuclei mainly associated with the cerebellum and the median pontine reticulospinal tract. These projects to the reticulospinal tracts provide extensor-flexor coordination in the trunk and proximal limbs. They play a significant role in controlling our posture through their reflex relationship with the body's position in space.[11] Vertigo and ataxia may develop due to proximity to the reticulospinal tract and vestibulocochlear projections.[12]

Isolated cranial nerve palsies occur in one-tenth of MS patients. In order of frequency, the 5th, 7th, 6th, 3rd, and 8th cranial nerves are most commonly affected.[13] The combination of these is relatively rare. Sometimes, peripheral facial paralysis is accompanied by other cranial nerve palsies.[14] Although isolated cranial nerve palsies are rare symptoms in MS, it should be kept in mind that MS may be a cause.

In conclusion, the MS patient we described presented with eight-and-a-half syndrome, which developed as a result of the involvement of the facial colliculus, vertigo, truncal ataxia, and titubation, which probably developed as a result of the participation of these posture-regulating pathways or connections to the vestibular system. With these components, we define this

case as eight-and-a-half plus syndrome, which is unexpected and very rare as an initial clinical presentation of MS.

DISCLOSURE

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