Case Report

Multiple Sclerosis Revealed by Intrapontine Axial Lesion of Peripheral Nerves

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Abstract

Multiple sclerosis (MS) is most often revealed by motor, sensitive symptoms with paresthesia, ocular symptoms, and more seldom by symptoms with rapidly installed hearing loss and vertigo. We report an observation with initial symptoms of a peripheral pathology mimicking a meningo-neuritis. A 22 years old young woman was addressed as an emergency for a sudden peripheral symptomatology associating a sudden right hearing loss with ear fullness, vertigo with vomiting and a right peripheral facial palsy. The patient had a spontaneous nystagmus beating toward the left side suppressed by fixation and sensory neural hearing loss on low frequencies. The Fukuda tests initially deviated toward the right side. The caloric test showed a right hypofunction at 75% and a correlated consistent left preponderance. The head shaking test (HST) and skull vibration induced nystagmus test (SVINT) revealed a left nystagmus. Blood samples results and cervical vestibular evoked myogenic potentials (cVEMP) were normal. Brainstem evoked response audiometry (BERA) demonstrated a retro-cochlear disease with a Wave V prolonged latency on the right side. A CSF lumbar puncture revealed a nodular image on the VII and VIIIth nerve pathway in the immediate vicinity of the right vestibular nucleus (hyper signal T2 and T1 Gadolinium contrast fixation). The evolution surveyed in neurology was totally regressive. A further disease relapse confirmed dissemination in time and location of the pathology.

Conclusion: Initial vestibular symptoms are seldom in MS. The ensnaring initial pseudo-peripheral presentation of this case is explained by the plaque location in close vicinity on the pons intra-axial cranial nerves pathway route. BERA are essential to reveal a neuronal conduction impairment on auditory nerve related to a demyelination process.

INTRODUCTION

Multiple sclerosis (MS) described for the first time in 1868 by Charcot [1] is a rather frequent pathology in young adults (45 cases for 100,000 inhabitants), twice as frequent in women as in men, and constitutes 3 to 10% of the central nervous system diseases [2-4]. This chronic inflammatory pathology of the central nervous system (CNS) is more often initialized by motor or sensitive symptoms (45% of cases), visual symptoms (optic neuritis, double vision, inter nuclear ophthalmoplegia are observed in 20% of cases) than by properly isolated cochleo-vestibular symptoms [4, 5]. We report in this work a seldom presentation of a first attack of MS disease associated with isolated audiological, vestibular and then facial symptoms mimicking a peripheral EN patholgy.

CASE PRESENTATION

A young woman of 22 years old, student in nursery, having no peculiar medical history was addressed at the ENT consultation by the Emergency Unit for a right unilateral hearing loss with ear fullness and a vertigo associated with vomiting (one throw up). The examination revealed a horizontal left beating spontaneous nystagmus observed with the eye in median straight sighting position and in the left sided sight (gaze) (degree 2 of Alexander classification). This nystagmus was minimized with fixation. The examination revealed a delayed right peripheral facial palsy (House-Brackmann stage 2). The anamnesis revealed that 4 days ahead the patient complained of paresthesia in the Vth nerve territory. The Fukuda-Unterberger test was initially deviated rightward. The audiogram showed a sensory neural hearing loss (SNHL).

Fifteen days later the facial palsy disappeared and bedside examination tests revealed at the head shaking test (HST) a horizontal left beating nystagmus. The skull vibration induced nystagmus test (SVINT) showed a left beating horizontal nystagmus (Figure 2).

Videonystagmographic (VNG) recordings (Figure 3) demonstrated at the caloric test a right hypofunction at 75% associated with a left nystagmic preponderance at 21°/sec. The cVEMP were normal with symmetric amplitudes on both sides and normal thresholds at 90 dB HL.

The BERA (recorded 3 weeks later) were demonstrative of a right retro-cochlear pathology. Wave V had a prolonged latency (6.7 ms measured at 80 dB) and the interaural I-V difference was 1.2 ms. (Figure 4).

The MRI demonstrated a spontaneous hyper signal in T2 weighted sequence and a post gadolinium T1 weighted hyper signal (1 cm diameter) on the right and anterior part of the pons (Figure 5).

The Biological blood samples were normal including blood cells count, CRP, glycemia, creatinine, ionogram, neurotrope viral serologies (VZV, HSV, HIV, EBV, CMV), Lyme, toxoplasmosis, Syphilis tests, HLA and blood proteins electrophoresis.

The lumbar puncture (LP) revealed an oligoclonal band on electrophoresis. Normal cytology (6 white cells/ml). Other biological Tests in the CSF (Lyme, Syphilis, toxoplasmosis) and PCR were negative and there was no cytologic sign of carcinomatous meningitis.

The initial symptoms were suggestive of a cerebello-pontine angle syndrome mimicking initially a peripheral disease. The patient was initially treated in neurology with 1 gr IV steroid treatment per day during 3 consecutive days. The evolution surveyed in neurology was totally resolute regressive 6 weeks later with recovery of hearing, facial palsy, disappearance of vertigo. The electrophysiological explorations were normalized in the same time. A MRI control 5 months after onset showed a partial regression of the lesion (Figure 6). The long term evolution was favorable.

**DISCUSSION**

Multiple sclerosis refers to the numerous scars or sclerae, better known as “en plaque sclerosis” [1] that develop the white matter of the brain and spinal cord [6]. It is the most common autoimmune disorder affecting the CNS [7]. A genetic component of the disease has been demonstrated with a higher probability in developing the disease with relatives of an affected person [5]. In identical twins both are affected in about 30% of cases. The genes are linked with the HLA on chromosome 6 and alleles of the major histocompatibility complex defined as DR15 and DQ6 [8].
Figure 3: Videonystagmography (VNG).
The caloric test shows a right hypofunction at 75% correlated with a left directional predominance.
B. The Sinusoidal rotatory test confirms a correlated left directional predominance and the fixation index confirms the nystagmus suppression as in a peripheral disease. VVOR= Visuo-vestibulo-oculare Reflex. VOR= Vestibulo-oculare Reflex. OF= ocular fixation (or gaze fixation) during rotation (the subject fixes his thumb at 50 cm in front of him).

Figure 4: Brainstem evoked response audiometry
On the right side, the different waves appear desynchronized. Wave V is recognizable still to 70 dB and its latency is increased (6.7 ms). On the left side all waves are identified with normal latencies. The interaural I-V delay is 1.2 ms, suspecting a right retrocochlear pathology.

Figure 5: MRI
a. There is a lesion in the right Pons with post Gd enhancement showing a demyelinated lesion (indicated by a circle L1).
b. There is a hyper signal of the white matter on the right part of the pons visible on coronal and axial view.

Figure 6: MRI 5 months after onset
The reduction of the size of the lesion is significant.

The criteria for MS have been described by McDonald 2005 [9], modified in 2010 [10] and are based altogether on clinical and radiological criteria. The diagnosis of MS disease is based on progression from onset:

1) One year of disease progression (retrospectively or prospectively determined).

2) Plus two of the following: a) Positive brain MRI(nine T2
lesions or four more T2 lesions with positive visual evoked potentials (VEP). b) positive spinal cord MRI (two focal T2 lesions). c) positive CSF (isoelectric focusing evidence of oligoclonal IgG bands or increased IgG index or both).

The inaugural symptoms may be isolated and monosymptomatic in 85% of cases: most often sensitive or motor symptoms (45%), visual (20%), autonomic (urinary retention, constipation) or ataxia (cerebellar) [5]. In 15% of cases the inaugural attack associates more than one neurological location [11].

Inaugural Brainstem signs are observed in 12 to 22% of cases [11] and are most often represented by VIth nerve. Intrapontine sensitive root of the Vth nerve is implicated in 12 to 22% of cases [11]. A viral or bacterial infection of the brainstem may be the cause. The involvement of the Facial and cochlea vestibular cranial nerves contributes to a cerebello-pontine angle syndrome suggesting several etiologies such as infectious, inflammatory or tumoral. Meningo neuritis involving the same cranial nerves and responsible for painful shingle rash with facial palsy and possible hearing loss are observed after Herpes Zoster oticus (Ramsay Hunt syndrome (RHS)). But other viral infections can reveal similar neural symptoms (Herpes simplex, Coxackie, measles, mumps, rubella, EBV, varicella) [14]. Viral infections are more frequent than after bacterial infections (Lyme disease with borrelia bacteria) or Syphilis. The Ramsay Hunt syndrome (RHS) is also called Sicard syndrome in European literature. Differential diagnosis depends on clinical symptoms at presentation [15,16]. Devic’s disease (recently named neuromyelitis optica spectrum disorder), neurosarcoïdosis (MRI images are different), neurological manifestations of Lupus or Sjögren syndrome, Whipple disease, Favre disease.

In MS, the lumbar puncture is positive in 75 to 85% of cases with an oligoclonal band of IgG on electrophoresis which is significant of an inflammatory marker [9].

Figure (7) depicts why a pseudo-peripheral symptomatology of this lesion is observed since in our observation the pons plaque involved only the intra-axial course of the nerves (V, VII, VIII) and not their nuclei observed.

A vertigo and hearing impairment as inaugural symptom is unusual as in our observation. Pula and coll. (2013) report 4% of MS related with acute vestibular disorders as inaugural among a population of 170 patients examined for acute vertigo. Their 7 cases of MS involved often the intrapontine 8th nerve fascicle [17].

Caloric test is modified in 63% of patients with MS [18]. Unsteadiness and cerebellar symptoms with coordination difficulties and balance disorders (ataxia) are more often reported than acute pure rotatory vertigo [5].

The modifications are more often described as an irritative type or pseudo-peripheral syndrome in 60% of these cases [18]. SVINT is modified in less than 30% of MS with brainstem symptoms but is not specific to a peripheral or central disease though its modifications are more often observed in peripheral pathologies [19-21].

Rare cases of sudden hearing loss have been related to MS and are signaled in 1% to 10% of cases [22]. Recurrent attacks of bilateral SSNHL can be a warning sign of MS [23]. Our case with unilateral apparently peripheral symptoms is particularly ensnaring since at the very beginning the patient reported a low frequency hearing loss with a feeling of ear fullness as observed in Hydrops like Menière’s disease.

BERA are essential to demonstrate a neuronal conduction alteration and are modified in 60% of MS cases associated with brainstem symptoms [24,25]. BERA were the sole objective examination to discriminate between central and peripheral pathology in our observation and were decisive to indicate the need of a LP and MRI.

The nervous system in MS may respond less actively to stimulation sensory nerves such as the auditory nerve due to demyelination on their pathways [25]. Visual or somesthetic evoked potentials may also be altered [26].
Gadolinium injection in MRI shows active plaques and also the existence of historic lesions not associated with the symptoms at the moment of the evaluation (time and space dissemination in the MRI follow up) [27, 28].

Clinically the evolution is marked by dissemination in time and space. Either it processes in a single and progressive evolution without remission or more frequently with successive attacks interrupted by partial or total remissions. Secondary MS attacks occurs in about 65% of cases. They eventually have progressive neurologic decline between acute attacks without definite periods of remission [5, 12].

Inaugural cochleo-vestibular symptoms are seldom in MS. The ensnaring initial pseudo-peripheral presentation of this case is explained by the plaque intra-axial location on cranial nerves pathway or course in the pons. BERA are essential to reveal a neuronal conduction impairment on auditory nerve related pathway or course in the pons. BERA are essential to reveal a neuronal conduction impairment on auditory nerve related pathway or course in the pons.

REFERENCES

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