

Manifestation of Lermoyez’s syndrome in Meniere’s disease patient

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Although Lermoyez’s syndrome (LS) has been described already at the beginning of the 20th century there are not many cases found in the literature and not much is written about its aetiology, diagnosis and treatment. Is there really such a low incidence or is the syndrome simply unrecognized? Some authors consider it as a variant of Meniere’s disease (MD) and some as a sole entity.

Case report

A 34-year old male with a history of a sudden hearing loss, tinnitus and feeling of pressure in the left ear was admitted to our hospital. On his first examination he had no vertigo and no spontaneous nystagmus was seen. His hearing on the right ear has been damaged for years, but on the left ear it had been normal. Anamnestically right-sided hearing loss happened gradually over years with episodic attacks of hearing fluctuations and mild nausea. Audiogram revealed both-sided sensorineural hearing loss. According to an old audiogram hearing on the right side was stable, but deterioration of hearing threshold in the low frequencies of the left ear was noticed. Tympanometry showed curve A on both sides. He was treated with oral corticosteroid (methylprednisolone 48mg/d for 1 week, followed by a taper every 2 days) and betahistine (24mg/12h).

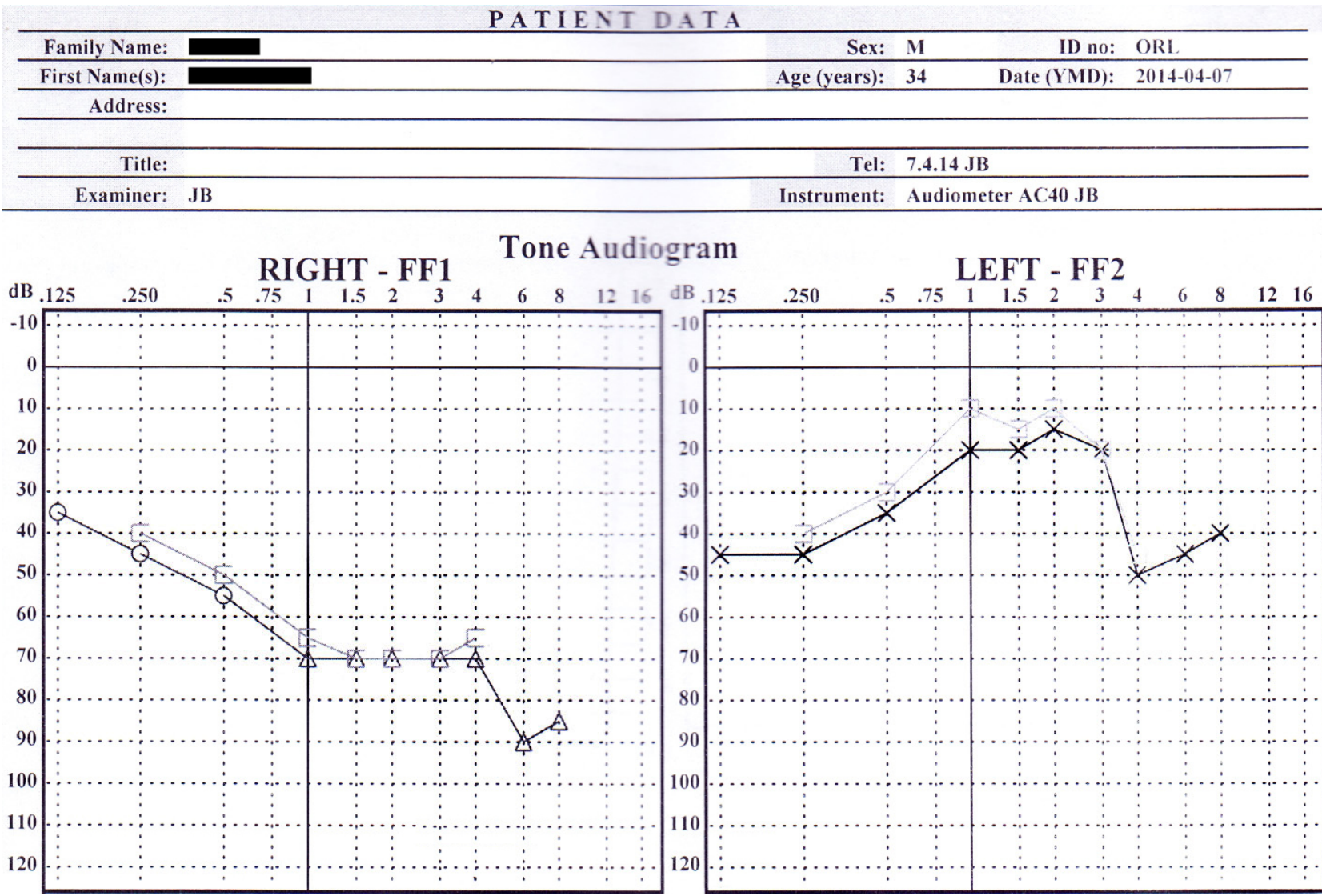


Figure 1: Audiogram on admission: previously known sensorineural hearing loss on right ear and recent on the left ear.

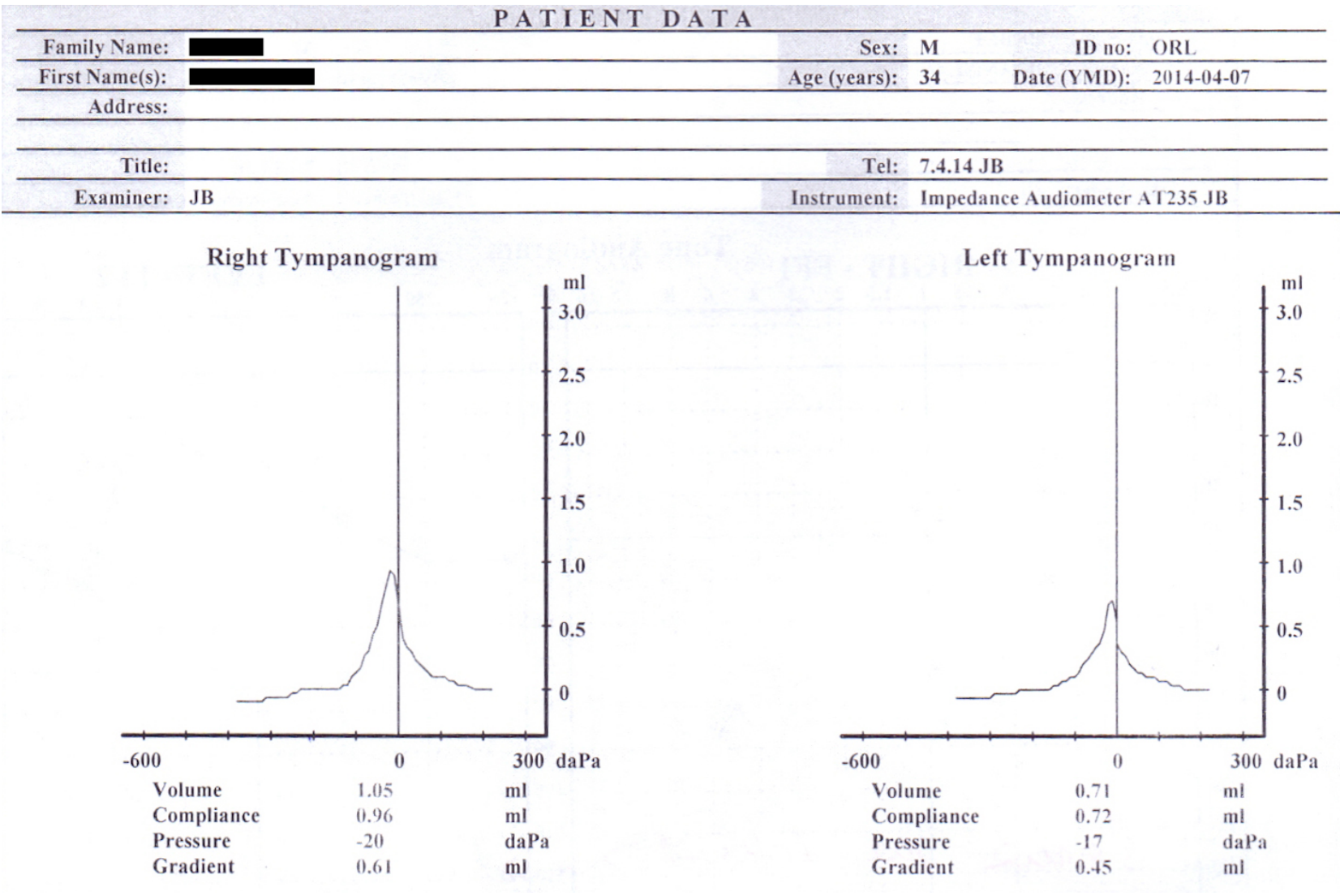


Figure 2: Tympanogram on admission: tympanogram A on both sides.

During hospitalization the patient noticed improvement of hearing in his left ear but at the same time vertigo with spontaneous nystagmus toward the affected ear had appeared. On examination he had no signs of ear infection. Check-up pure tone audiometry confirmed improvement of the hearing in the low frequencies from 125Hz-500Hz. Caloric test revealed symmetric response,

with preponderance of left-sided nystagmus.

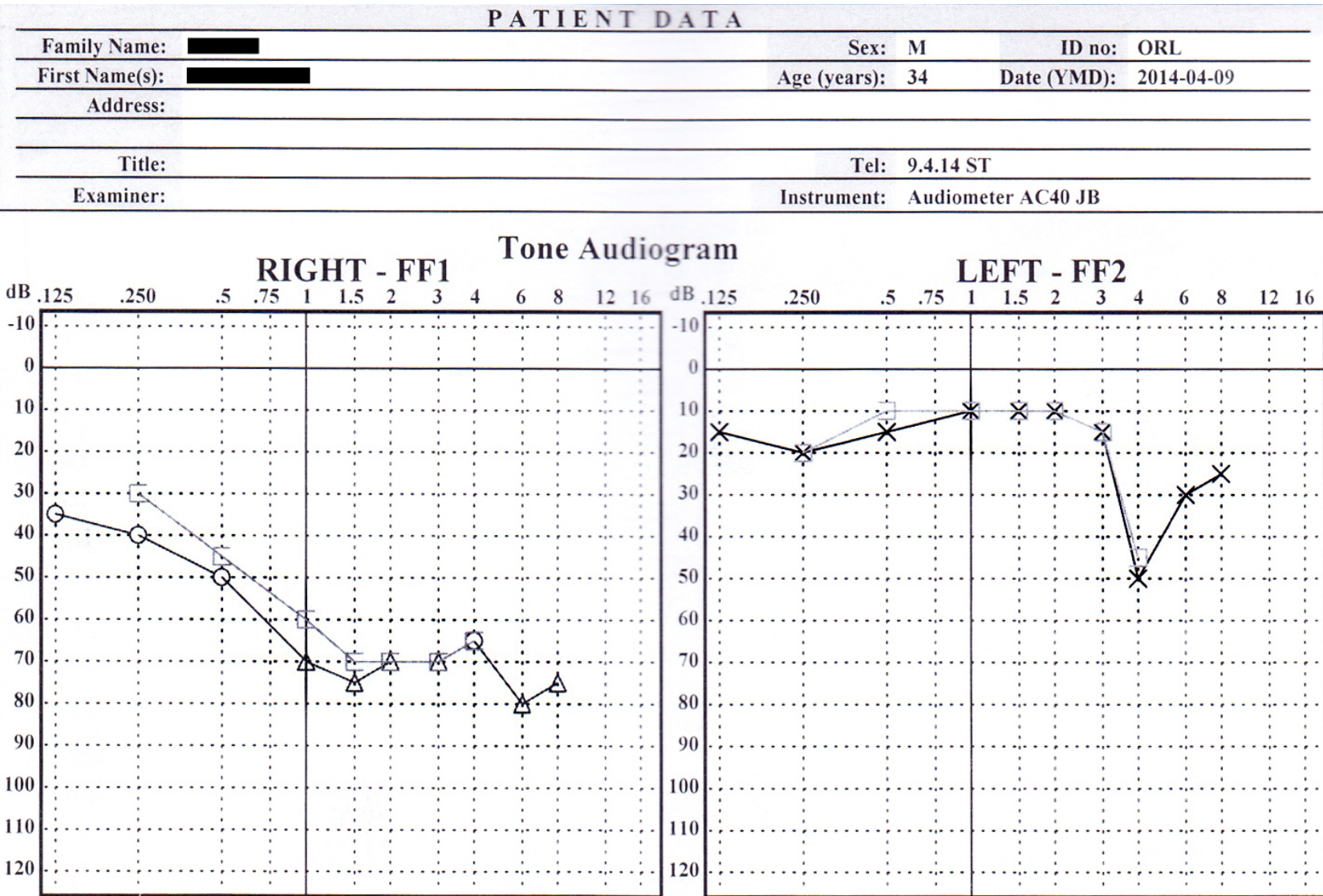


Figure 3: Audiogram 2 days after admission: recovery of hearing threshold in low frequencies on the left ear.

On follow up he complained of frequent day to day fluctuations of hearing in the left ear. Hearing fluctuations were sometimes accompanied with vertigo and vomiting, sometimes not. Usually as the vertigo attack appeared, spontaneous nystagmus towards the affected side was detected and his hearing improved. Even higher doses of betahistine (24mg/8h) and intratympanic injection of dexamethasone (24mg in a single dose) did not relieve his attacks. He was prescribed a hearing aid for his right ear.

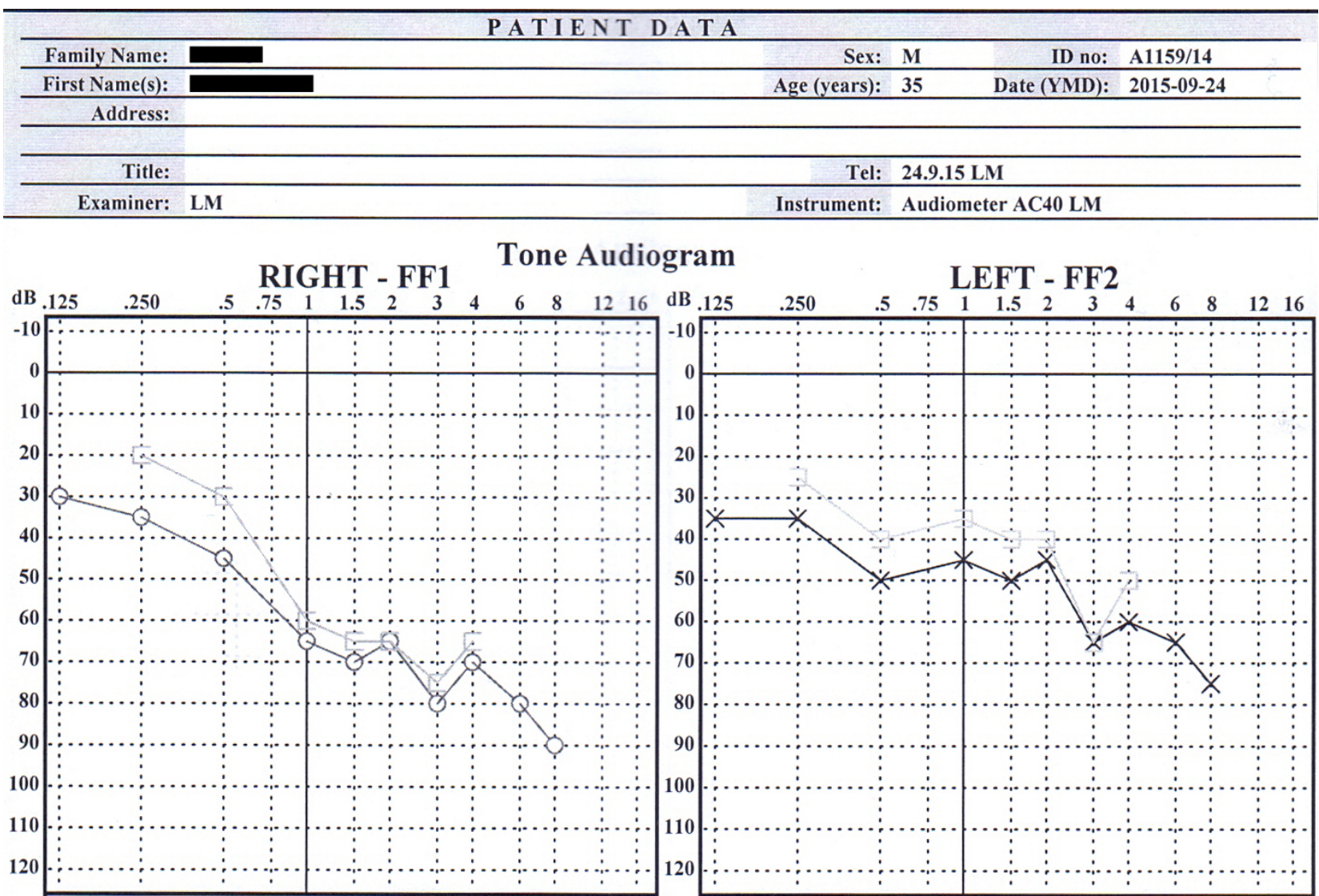


Figure 4: The patient had frequent fluctuations of hearing. Audiogram on last deterioration of hearing.

Magnetic resonance imaging excluded pathologic changes in the brain, pontocerebellar angle and in the course of the vestibulocochlear nerve. Serology for Borrelia Burgdorferi and rheumatologic tests were negative. The cervical vestibular evoked myogenic potential (cVEMP) showed symmetric response. The ocular vestibular evoked myogenic potential (oVEMP) unfortunately is not at our disposal.



Figure 5: Head MRI was normal.

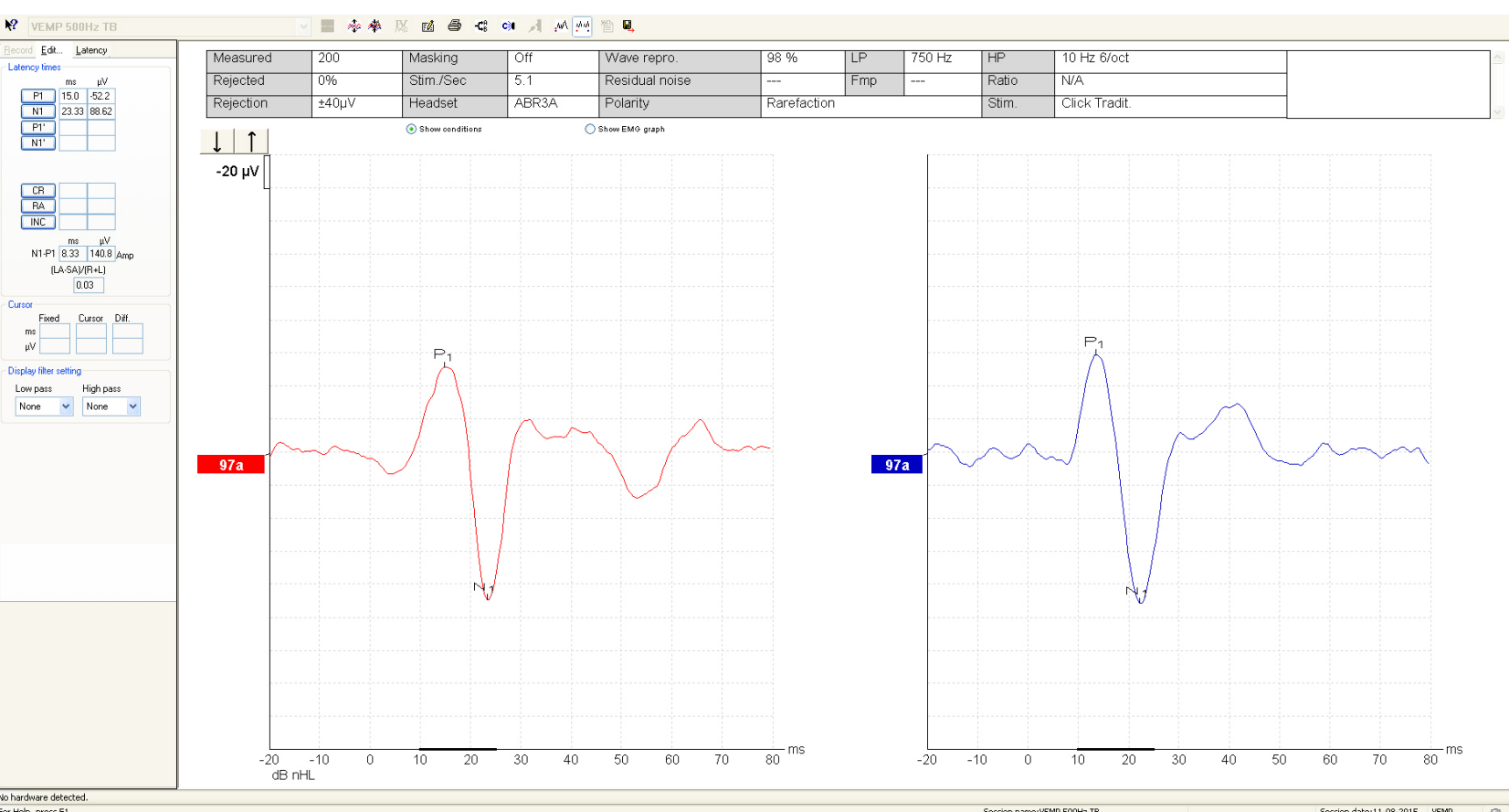


Figure 6: Symmetric response on cVEMP.

In 1919 French otolaryngologist Lermoyez described a syndrome of progressive hearing loss and buzzing in the ear followed by a sudden attack of vertigo and fast recovery of hearing. As opposed to MD at the time of the vertigo attack hearing in the low frequencies improves. Lermoyez also described it as “The vertigo that restores hearing” (“Le vertige qui fait entendre”). It can affect one ear or both ears.

Our patient presented with aural fullness, tinnitus, fluctuating hearing loss and vertigo attacks which are otherwise characteristics for MD. The direction of spontaneous nystagmus (SN) in MD patients at the attack is usually away from affected ear. In our case the nystagmus was towards the affected ear. There have been reports that in LS patients the direction of SN is towards the affected ear because of an increase of volume of endolymph in the horizontal semicircular canal after the opening of the ductus reunions. This fluid shift could cause displacement of the horizontal canal cupula towards the utricle and consequently a SN with fast phase beating towards the affected ear.

Recently objective measures of dynamic otolith and dynamic semicircular canal function in a LS patient have been reported that could help differentiate LS from MD. The amplitude of the n10 potential of the oVEMP to bone conducted vibration in MD patients at the time of attack was found to be increased since the utricular function in the affected ear is enhanced. On contrary in LS patient it has been shown that utricular function is decreased and that the n10 potential is reduced.

The presence of ipsilateral nystagmus and hearing improvement at the time of vertigo (with exclusion of some other underlying causes) made us suspect of a variant of Meniere’s disease – Lermoyez’s syndrome. The patient’s hearing on the right ear was most probably affected by the same reason.

References:

- Manzari L, Burgess AM, Curthoys IS. Vestibular function in Lermoyez syndrome at attack. *Eur Arch Otorhinolaryngol* 2012; 269: 685-691.
- Manzari L, Tedesco AR, Burgess AM, Curthoys IS. Ocular and cervical vestibular-evoked myogenic potentials to bone conducted vibration in Meniere’s disease during quiescence vs during acute attacks. *Clinical Neurophysiology* 2010; 121: 1092-1101.
- Young YH, Wu CH. Electronystagmographic findings in a case of Lermoyez’s syndrome. *Auris, Nasus, Larynx* 1994; 21: 118-121.
- Zhang Q, Xu M, Zhang X, et. al. A case of Meniere’s disease in the left ear and Lermoyez syndrome in the right ear - a 32-month longitudinal observation and literature review. *Acta Oto-Laryngologica* 2010; 130: 1084-1088.