Study of the disorder formerly known as Barré's vestibulospinal tract syndrome or isolated imbalance syndrome

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ABSTRACT

On 14 March 1925, Jean Alexandre Barré gave a presentation titled 'Essay on a vestibulospinal tract syndrome' to the Société d'Oto-Neuro-Oculistique of Strasbourg. He described the case of a 54-year old man who experienced a sudden and disabling vertigo that led him to take complete bed rest for several days. Barré asserted that this was a new clinical entity, but also prudently mentioned that future studies were needed to confirm the syndrome. One of its most characteristic clinical manifestations was a gait with small steps, which was different from the gait abnormalities characteristic of pyramidal, extrapyramidal, and cerebellar signs.

In 1931, Antonio Subirana, one of Barré's students, presented his doctoral thesis, which included 12 cases of this syndrome while also detailing its symptoms and differential diagnosis.

The study of the vestibulospinal tract syndrome described by Barré once again shows us how important it is to identify symptoms, and that sometimes syndromes and signs which were described in the past have been forgotten or are underdiagnosed today.

KEYWORDS

Vestibulospinal tract syndrome of Barré, neurological syndromes, symptoms

On 14 March 1925, Jean Alexandre Barré (1880-1967) (Figure 1) gave a presentation titled 'Essay on a vestibulospinal tract syndrome'¹ to the Strasbourg Société d'Oto-Neuro-Oculistique.¹ He described the case of a 54year old man who experienced a sudden and disabling vertigo that led him to need complete bed rest for several days. Later, when he attempted to get out of bed, he was unable to stand, presenting astasia-abasia. After his symptoms remitted somewhat he was able to walk, but only slowly and with small steps.

Clinical examination ruled out signs of pyramidal, extrapyramidal, or cerebellar involvement. The patient demonstrated balance disturbances, pulsions, and Romberg sign with a tendency to fall to the left. In the extended arm test, past pointing to the left was also observed. No nystagmus was observed.

Motor symptoms were ruled out with Mingazzini and Barré tests; reflexes were normal and there was no spasticity, although motor deficit was observed in the psoas muscle and quadriceps. With the patient in the supine position, the heel-to-knee test showed normal results, but the patient was unable to change from the supine to the prone position and vice versa.

According to the author, clinical symptoms were due to vestibulospinal tract impairment. Among the symptoms shown, astasia-abasia, short strides with a characteristic pattern, and the difficulty in changing from a supine to a prone position were the most interesting.

Barré stated that it was a new clinical entity, but also mentioned prudently that future studies were needed to confirm the syndrome.

Antonio Subirana (1903-1992) was trained in the neurology department of the Hôpital Civil de Strasbourg, where he engaged in a close personal and professional relationship with Barré, who managed the department. In 1931, he presented a doctoral thesis titled "Barré's

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Figure 1. Prof. Jean Alexandre Barré (1880-1967). SEN historical archive.

vestibulospinal tract syndrome or isolated imbalance syndrome"² (Figure 2).

His thesis emphasized three points: 1) the originality of the topic although ignored at present, 2) its strictly symptomatological basis, and 3) the neurophysiological explanation, fundamentally based on a differential diagnosis.

The thesis mentioned that motor function had classically been associated with the pyramidal tract; however, the latest clinical and anatomical contributions of the time showed that the striospinal, reticulospinal, rubrospinal, and vestibulospinal tracts were all related to involuntary motor function.

Subirana pointed out that the vestibulospinal tract had been previously described by Marchi as part of the

extrapyramidal system, and several studies detailed the complete anatomy of this tract.

It was Barré who, in 1925, wrote that in patients with clinically detected cerebellar syndrome, a detailed examination revealed vestibular signs that were overlooked or were considered part of the cerebellar deficit. According to Subirana, Barré described the first case of vestibulospinal syndrome, or isolated imbalance syndrome.

Because some structures of the vestibular system are located in the cerebellum, the author justified attributing disorders due exclusively to a dysfunction of the vestibular system to a cerebellar lesion. The close anatomical link between the cerebellum and the vestibular system result in the comorbidity of clinical symptoms caused by deficits in both neurological structures in most pathological processes.

When he described the vestibular syndrome, he underlined that just as a cerebellar lesion causes symptoms only on one side, vestibular lesions always cause bilateral manifestations, although predominantly ipsilateral. Therefore, the author asserted a distinction between the two.

Its presentation is ictal or rapidly progressing, and intermittent claudication of the legs, or in Barré's words, *effondrement sur place*, stands out as the initial sign. Further symptoms included severe vertigo that lasted several days, and when the patient attempted to stand up, he presented astasia-abasia. Paradoxically, in the supine position he can control the movement of his legs and arms and perform the finger-to-nose and heel-toknee tests correctly. However, the patient cannot change from the supine to the prone position, and vice versa.

Study of pyramidal function with the Miganzzini and Barré's manoeuvres yielded normal results, ruling out a pyramidal deficit, though a mild proximal impairment was observed with Barré's psoas test. The patient, in a position similar to that of Mingazzini's test, presented proximal claudication of the thigh due to psoas involvement but with preservation of strength at the distal leg and foot level.

Symptoms slowly remitted, although the gait with small steps (*à petits pas*) persisted. Unlike in pseudobulbar palsy, the patient slightly raises the feet without dragging them. The base of support was barely widened and no lack of coordination was observed. The majority of the clinical symptoms significantly remitted, but gait impairment persisted.

Trabajo realizado en la Clínica Neurológica de Estrasburgo (Director Profesor J. A. Barrè), y terminado en Barcelona, en los servicios de Clínica Médica y Quirúrgica a cargo de los Profesores Ferrer Cagigal y Bartrina, respectivamente

El síndrome vestíbulo-espinal de Barré o síndrome de deseguilibrio puro

Tesis doctoral leída en Madrid, el 15 de diciembre 1931, ante el tribunal formado por los Profesores García del Real, Tello, Fernández Sanz y Torre Blanco, y calificada de sobresaliente,

por el doctor ANTONIO SUBIRANA Ayudante de clases prácticas, ex-interno por oposición de la Facultad de Medicina de Barcelona y exsalistente de la Cínica Neurológica de Estrasburgo

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Figure 2. Title page of the doctoral thesis "Barré's vestibulospinal tract syndrome or isolated imbalance syndrome", 1931. SEN historical archive.

The author mentioned the existence of associated entities, characterised by the presence of cerebellar and pyramidal signs. All this was due to the extension of lesions to adjacent nerve centres.

The author provided the study of 12 cases in his thesis. Medical records are very detailed and the examination was exhaustive, illustrating how the author attitude was in line with the Golden Age of symptomatology and clinical examination. The differential diagnosis is very extensive, and the author shows a profound knowledge of the symptoms (Figure 3).

According to Subirana, gait in patients with vestibulospinal syndrome shows unmistakable clinical signs, particularly characterised by the absence of pyramidal, parkinsonian, and cerebellar signs, which are in turn present in patients with pseudobulbar and lacunar syndromes.

Regarding the diagnosis of gait disorders caused by brain lesions, especially in Bruns apraxia, Subirana supported the opinion of Clovis Vincent. The French physician stated that frontal ataxia is secondary to a labyrinthine lesion due to intracranial hypertension secondary to a tumour or hydrocephalus, therefore ruling out ataxia due to a pure frontal lobe lesion.

Contributions by Schilder and Gerstmann were cited; they considered apraxia to be the result of gait disorders in the case of a frontal lesion. In these cases, the primary symptoms were astasia-abasia, hypokinesia, and psychiatric disorders. All these manifestations help distinguish these disorders from the syndrome described in the thesis.

Also in his thesis, the author mentioned that Vernière had defined astasia-abasia in 1909 as a syndrome characterised by the ability to perform different gait movements when the patient is in the supine position and the inability to remain standing (astasia) or walk (abasia). Furthermore, Dejerine classified these symptoms as functional, Bonnier reported the vestibular origin of astasia-abasia in 1903, and later Guillain and Barré showed the organic character of the syndrome due to labyrinthine lesion in cases of war head trauma.

Among the most important conclusions of the thesis are the following: 1) symptoms start as severe vertigo followed by astasia-abasia but the patient is able to perform voluntary movements with normal coordination and strength when in the supine position. However, the patient either cannot change position or can do so only with great difficulty. Results obtained with the Mingazzini and Barré manoeuvres were normal while impairment was observed with Barre's psoas test. No nystagmus was observed and effondrement sur place was the premonitory symptom. 2) During the active phase, a gait with small steps, a positive Romberg signs, and a Barré vertical axis test showed indications of vestibular origin; 3) Associated entities with cerebellar symptoms do not have as good a prognosis as isolated vestibular forms; 4) Regarding aetiology, the ages of the patients and the form of presentation lead directly to the consideration of a vascular origin.

To respond to doubts about the very existence of this syndrome, we literally translate the seventh and last conclusion:

One day, anatomical pathology might show that this hypothesis is not true and that the symptoms experienced by our patients are due to causes other than the involvement of the vestibulospinal tract. In that case, our master would still deserve the honour of having clinically identified a new syndrome, which even if it is no longer called 'vestibulospinal', should all the same bear the name of Barré's syndrome.^{2(p73)}



Figure 3. Picture taken by Dr Antonio Subirana (Barré's student) of a patient with vestibulospinal syndrome, March 1930. SEN historical archive.

Conclusions

The study of the vestibulospinal tract syndrome described by Barré and further detailed in Subirana's thesis once again shows us how important it is to identify symptoms, and that sometimes syndromes and signs which were described in the past have been forgotten or remain underdiagnosed today. On a side note, it is surprising that vestibulospinal or isolated imbalance syndrome was not included as one of the lacunar syndromes that C.M. Fisher³ studied so deeply. Probably an exhaustive clinical study would even today enable the diagnosis of a case of Barré's vestibulospinal syndrome.

Conflicts of interest

The author has no conflicts of interest to declare.

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