

Guillain-Barré syndrome: celebrating a century

Síndrome de Guillain-Barré: celebração de um século

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ABSTRACT

A hundred years ago, Guillain, Barré and Strohl described a syndrome with a predominant motor acute or subacute polyneuritis, albumin-cytologic dissociation in the cerebrospinal fluid, and a benign course. Before them, many other authors, such as Landry, Duménil, Osler, and Grainger Stewart had described similar cases although they had not performed lumbar punctures. In this work, we outline certain features of the beginning of this famous syndrome.

Keywords: Guillain-Barre syndrome; neuritis

RESUMO

Há 100 anos três médicos franceses, Guillain, Barré e Strohl descreveram um tipo de polineuropatia aguda ou subaguda de predomínio motor com dissociação albumino-citológica no líquido cefalorraquiano e de evolução benigna. Antes destes, outros autores, como Landry, Duménil, Chomel, Osler e Grainger Stewart, estudaram casos similares, porém não realizaram punção lombar. Neste trabalho procuramos sintetizar alguns fatos importantes no início da descrição desta síndrome.

Palavras-chave: síndrome de Guillain-Barré, neurite.

In 2016, a century had passed since the famous description of a syndrome of an acute, ascendant and symmetrical paralysis involving the peripheral nerves with an albumin-cytologic dissociation in the cerebrospinal fluid and a good recovery, by three French neurologists: Guillain, Barré and Strohl. This syndrome, well known as “Guillain-Barré syndrome”, is the most common cause of acute flaccid paralysis in the world. In June 2016, a symposium in Glasgow celebrated a century of Guillain-Barré syndrome, patronized by the Peripheral Nerve Society¹.

Guillain-Barré history

In 1916, Georges Charles Guillain, Jean Alexandre Barré and André Strohl (Figure 1) described in *Bulletins et Mémoires de la Société Médicale des Hôpitaux de Paris*², a new syndrome in two young French soldiers characterized by motor disturbance, abolition of the tendon reflex, paresthesia with mild disturbance of sensation, slight changes in electric reaction of nerves and muscles and hyperalbuminosis of the cerebrospinal fluid with absence of cytological reaction (albumin-cytologic dissociation). The cases were presented at the Medical Society of the Hospitals in Paris on October 16th 1916, under the title “*Sur un syndrome de radiculonévrite avec hyperalbuminose du*

liquide céphalorachidien sans reaction cellulaire. Remarques sur le caractères cliniques et graphiques des réflexes tendineux” (Figure 2). They thought that this syndrome was due to involvement of the spinal roots and nerves, probably infectious or toxic in nature. The treatment consisted of rest, massage and injections of strychnine. Both patients recovered after a few months.

Guillain became professor of Neurology at the Salpêtrière Hospital in Paris. Barré was professor of Neurology at Strasbourg. They became friends at the “Centre Neurologique” of the 6th army near Amiens during the first world war, which is where they saw these cases³. Strohl was forgotten, probably because of his young age and his participation was limited to electrophysiology.

Early description

In 1850, Jean Baptiste Octave Landry de Théizillat (1826-1865) (Figure 3) wrote “Note sur la paralysie ascendante aiguë” where he reported on 10 patients, five of whom he had personally examined⁴. He described three types of presentation: ascending paralysis without sensory signs or symptoms; ascending paralysis with ascending anesthesia, and a progressive disorder characterized by paralysis and slight sensory loss. His first patient, a 43-year-old

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Figure 1. Georges Charles Guillain (1876-1971), Jean Alexandre Barré (1880-1967) and André Strohl (1887-1977).

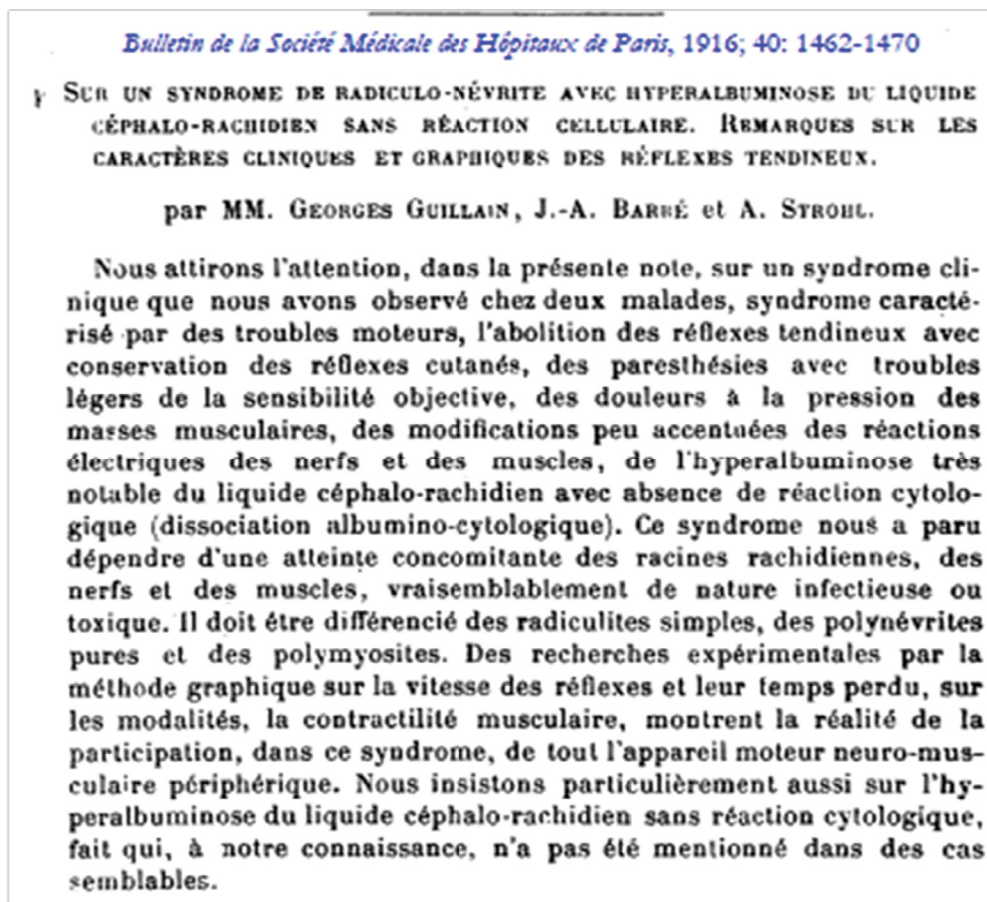


Figure 2. Original article by Guillain-Barré-Strohl.

man, after a premonitory fever, complained of “weakness, tingling his fingers and toes”. Touch sensation was lost in the fingers and feet but was little impaired in the upper two thirds of the lower limbs. The paralysis ascended, with sensory loss. By the third week, his limbs were paralyzed, he developed difficulty in breathing and swallowing, with fever and coughing. Touch was lost in the feet and in the finger tips, but without pain and temperature loss. He died

in the third week. Necropsy included histological examination only of the cord and soleus muscle. The main difference between that report and the syndrome described by Guillain, Barré and Strohl was that Landry did not perform a lumbar puncture, as this was introduced by Quincke only in 1891.

Landry was a gentle and modest man. At the age of 40, while attending the victims of a cholera epidemic in Paris,



II.

TRAVAUX ORIGINAUX.

NOTE SUR LA PARALYSIE ASCENDANTE AIGUE, par le docteur O. LANDRY.

L'objet de cette note est de signaler un état morbide assez rare et généralement inconnu, mais qui mérite de figurer parmi les affections les plus remarquables des cadres pathologiques.

Dans un assez grand nombre de paralysies, auxquelles convient la qualification générique d'*extenso-progressives*, les troubles fonctionnels, d'abord restreints à une partie limitée du corps, s'irradient graduellement plus ou moins loin de leur point de départ. Cette propagation s'effectue tantôt de proche en proche, et d'après un ordre bien déterminé; tantôt, au contraire, sans régularité et comme au hasard. On peut appeler les paralysies de ce dernier groupe : *extenso-progressives irrégulières*, et à celles du premier, bien plus importantes à connaître, donner le nom d'*extenso-progressives ascendantes*, ou, plus simplement, de *paralysies ascendantes* ou *centripètes*. Dans ces affections, en effet, les symptômes

Figure 3. Jean Baptiste Octave Landry de Thézillat (1826-1865).

Landry contracted the illness and died a few days later⁵. Haymaker called this syndrome the "Landry-Guillain-Barré syndrome" and included the name of Landry as one of the fathers of Neurology⁶. Landry's description probably included all types of acute or subacute ascending paralysis, such as some cases of poliomyelitis or ascending myelitis. In 1864, Louis Stanislas Duménil, in Rouen, described one case of an acute and symmetrical ascending paralysis. He thought it was due to nerve atrophy. Three years later he described three other cases¹.

Sir William Osler (1849-1919) in "The Principles and Practice of Medicine" (Figure 4) wrote about an ascending paralysis he referred to as an acute infectious polyneuritis³. Guillain and Barré did not accept that description, as Osler's patient had fever in the beginning of the disease and had no lumbar puncture⁷.

In Paris in 1828, Auguste François Chomel (1788-1858) described a patient with disturbed sensations in their hands and feet, ascendant weakness with acute evolution, becoming paralytic within a few days¹. In 1881, Thomas Stewart (1837-1900), in Edinburgh, reported on three cases of that same illness, calling attention to the loss of tendon reflexes¹.

Soon after 1916

Draganesco and Claudian were the first to use the term "Guillain-Barré syndrome" in 1927¹.

In 1936, Guillain published ten nonfatal cases and established the main conditions of this syndrome: 1) albumin-cytologic dissociation in the cerebral spinal fluid; 2) onset with paralysis and paresthesia or pain, with or without premonitory symptoms; 3) flaccid paralysis of the lower limbs and later in the upper limbs; 4) occasional fibrillatory twitching and slight atrophy of the distal muscles; 5) abolition

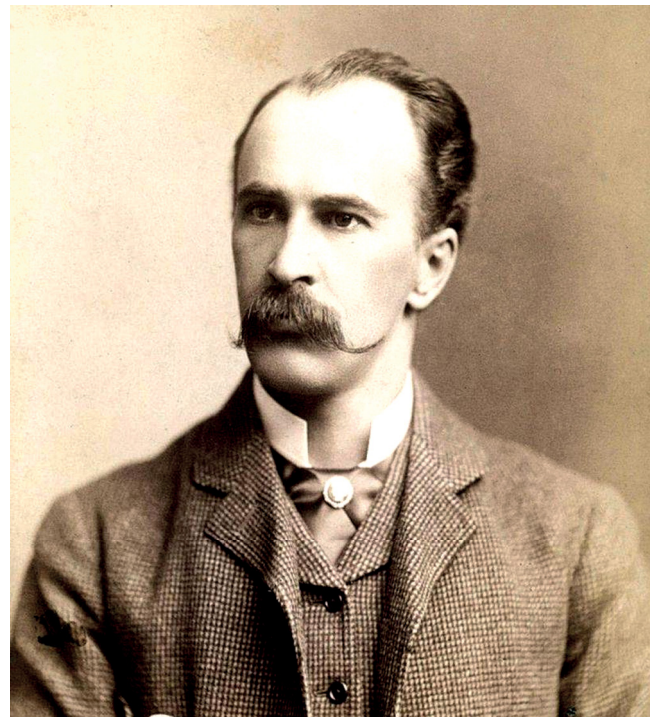


Figure 4. Willian Osler (1849-1919).

of tendon reflexes; 6) presence of subjective sensory disturbance; 7) objective minor or transitory changes in sensibility; 8) transitory paralysis of the cranial nerves; 9) transitory sphincter disturbance⁸.

In the same year, Alajouanine published a similar case in which death had occurred⁹.

Guillain and Barré⁷ refused to accept the cases described without albumin-cytologic dissociation in the cerebral spinal fluid, and in the cases with fatal outcome, Landry and Alajouanine were the objects of attack by Guillain¹³. Firstly

because they did not mention the albumin-cytologic dissociation in the cerebral spinal fluid, and secondly, because the patients died from the disease. Two years later, Guillain accepted that this syndrome could be fatal³.

The first case in Brazil was described by Antonio Rodrigues de Mello (Figure 5) in a soldier who, without prodromal symptoms, presented with a subacute ascending paralysis, slight sensory disturbances in the limbs with paralysis of the trigeminal and facial nerves. The cerebral spinal fluid showed an albumin-cytologic dissociation. He was treated with intravenous and intrathecal vitamin B and recovered in a few weeks. Mello believed that the disease was due to lack of vitamins or due to a neurotrophic virus¹⁰.

Conclusion

The history of Guillain-Barré syndrome continues today. Many advances have been made in the epidemiology, clinical variants, atypical cases, electrophysiology findings, pathogenesis, preceding infections and vaccinations, nerve images, treatment and rehabilitation. These are another story.



Figure 5. Antonio Rodrigues de Mello (1911-1988).

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