

History of the Emergence and Recognition of Syringomyelia in the 19th Century

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Abstract

Charles-Prosper Ollivier d'Angers coined the term "syringomyelia" in 1827 to describe the presence of a cavity in the spinal cord, which he considered pathological in all cases. In 1882 in Germany, Otto von Kahler and Friedrich Schultze defined the clinical syndrome, which associated Duchenne-Aran muscular dystrophy of a limb with sensory dissociation. They explained the syndrome by the presence of an abnormal cavity, distinct from that found in the spinal cord of healthy adults. Although Guillaume Duchenne de Boulogne and Jean-Martin Charcot had observed cases in France, both failed to identify syringomyelia, whereas a family physician in Brittany, Augustin Morvan, described the clinical symptomatology using the term "analgesic whitlow" in 1883. Based on several dozen observations that they collated in their remarkable theses, Anna Bäumlér in Zurich in 1887 and Isidore Bruhl in Paris in 1890 established the complete clinical picture of syringomyelia, covering anatomic functional and pathological aspects. Whereas Charcot isolated pathologies by correlating clinical signs with anatomic-pathological lesions, the isolation of syringomyelia initially involved an anatomic-pathological concept before the semiology was defined. In addition, this work would later enhance physiological understanding of sensory spinal pathways.

Résumé

Le mot syringomyélie a été proposé en 1837 par Charles-Prosper Ollivier d'Angers (1796-1845) pour décrire la présence d'une cavité au sein de la moelle épinière. Il faut attendre 1882 pour qu'en Allemagne, Otto von Kahler et Friedrich Schultze individualisent le syndrome clinique associant l'amyotrophie d'un membre de type Duchenne-Aran et la dissociation sensitive en l'expliquant par la présence d'une cavité anormale, distincte du canal de l'épendyme. En France, c'est Georges Debove qui publie, en 1889, la première observation d'une malade, précédemment hospitalisée dans le service de Jean Martin Charcot, où le diagnostic n'avait pas été porté. Pourtant, un médecin de famille breton, Augustin Morvan avait, lui, décrit la symptomatologie clinique sous le nom de panaris analgésiques dès 1883 mais n'acceptait pas l'unicité des deux pathologies décrites. Le concept de syringomyélie fut d'abord anatomopathologique, bien avant que la sémiologie s'y rapportant ne soit précisée. La syringomyélie permet également de préciser les connaissances anatomo-physiologiques des voies médullaires de la sensibilité, peu connues auparavant.

Jean Martin Charcot (1825-1893) Tuesday Lesson on 28 June 1889 at La Salpêtrière Hospital

"It is a major achievement for a physician to bring a morbid entity, previously ignored and unknown, out of chaos, to show for the first time that it possesses a symptomatic attribute that will make it recognisable for all, and to give clinical and nosographical life to a group of phenomena that, until then, were not taken into account".

The syndrome of syringomyelia has been perfectly defined and involves cervical-occipital pain, atrophy and anaesthesia most frequently affecting one or both upper limbs, with loss of the ability to feel temperature or pain. Tactile sensation is maintained, there is loss of tendon reflexes in the affected limb, often associated with muscular atrophy in the hand and fasciculations and finally scoliosis and trophic disturbances in some cases. Damage to the medulla oblongata is referred to as syringobulbia and involves rotational nystagmus, dissociated facial anaesthesia in the territory of the

trigeminal nerve, paralysis of the velum, palate, pharynx and larynx by damage to the *nucleus ambiguus*, and unilateral tongue atrophy. Neurological damage to the lower parts of the body is inconsistent, in the worst cases presenting as spastic paraplegia. Progressing insidiously over several dozen years, this syndrome has a variety of causes: post-traumatic haematomyelia, ischaemia in the territory of the anterior spinal artery, traumatic lesions, multiple sclerosis, spinal cord tumours and, though currently rare, tuberculous meningitis. The syndrome is acquired and thus non-genetic, although disorders related to neural tube closure favour its development (Arnold-Chiari malformation, spina-bifida); hydrodynamic abnormalities in the circulation of cerebro-spinal fluid may also be pre-disposing factors.

Origin of the Term "Syringomyelia"

The term "syringomyélie" appeared for the first time in 1827 in the second edition of *Traité des maladies de la moelle épinière* (Treatise on Spinal Cord Diseases) by Charles-Prosper Ollivier d'Angers (1796-1845).



Picture 1. Charles-Prosper Ollivier d'Angers 1796-1845 by Guillaume Bodinier 1821 Université d'Angers, public collection. Photo P. Leroy.

He derived the new word from the Greek terms, “syringo” meaning tube-shaped and “myelia” meaning spinal cord. On 12th June 1823 in Paris, Ollivier defended his thesis, *Essai sur l'anatomie et les vices de conformation de la moelle épinière chez l'homme* (Essay on the Anatomy and Conformational Defects of the Spinal Cord in Humans). He used this first work as the basis for his *Traité des maladies de la moelle épinière contenant l'histoire anatomique, physiologique et pathologique de ce centre nerveux chez l'homme* (Treatise on Spinal Cord Diseases, including the anatomical, physiological and pathological history of this nervous centre in humans), published in three, progressively expanded editions from 1824 to 1837. Ollivier reviewed spinal cord anatomy and macroscopic anatomical pathology and described malformations such as spina bifida as well as inflammatory and infectious conditions of the meninges (Ollivier d'Angers, 1823). His thesis and book are milestones, given that Ollivier wrote the first treatise exclusively dedicated to spinal cord pathology (Ollivier Charles-Prosper, 1837). As early as 1823, he argued that “examination of the inner structure of the spinal cord

demonstrates that it does not contain a central canal, as some anatomists have maintained”. He would never change his position. This error is probably explained by the technical difficulties involved in sampling and dissection that prevented all but a macroscopic examination of the spinal cord several days after death. In his chapter on pathology, he added: “several anatomists have argued that a canal exists in the centre of the spinal cord [...]. This canal is presumed to be a prolongation of the fourth ventricle and similar to the canal observed by Mr. Portal”. In 1804, Portal wrote: “Based on examination of the spinal cord in various subjects, a narrow canal has been observed at its centre that descends to varying levels, and whose upper end opens into the fourth ventricle. It seems reasonable to believe that this canal exists naturally, but it only becomes clearly visible in disease states, as in subjects who have died of spina-bifida” (Portal 1804). In 1828, Louis Florentin Calmeil (1798-1895) noted that “several vertebrate animals (birds, reptiles, fish) maintain throughout their lives a canal in the centre of the spinal cord. Certain anatomists have made a similar observation in adult humans, which has been contested. I have performed a number of dissections which help to clarify this point in human anatomy. It is clear that even at an advanced age, a central canal is often found in the spinal cord” (Calmeil, 1828). Despite these observations, Ollivier maintained the position of his thesis in his book, published in 1827 and 1837. With regard to Calmeil's observations, he merely remarked: “Several examples show the variety of primitive spinal cord organization, whereas others show that the canal observed in the spinal cord was clearly the product of pathological damage occurring after birth”. For Ollivier, the observation of a cavity in the spinal cord was always a pathological feature. Eventually he would be proved right, but since he did not recognize any clinical features associated with the tube shape in the spinal cord, Ollivier can only be credited with the term “syringomyelia”, a purely anatomopathological description.

The Spinal Cord Canal and its Anatomical Anomalies before 1880

Until the middle of the 19th century, observations were made from time to time of abnormal dilation in the spinal cord canal, referred to as hydromyelia. For example, in 1838, Auguste Nonat (1804-1887) published one such observation in *Archives de Médecine*.

“A 34-year old man had presented the symptoms of a chronic myelitis for a year: he was paraplegic, suffering from cramps and spasmodic movement in his lower limbs, with loss of sensation and edema; his rectum and bladder were paralyzed; and bedsores were present in the sacral region. The patient died in October 1836. Upon autopsy, [...] the centre of the spinal cord was found to be hollowed out from the eighth dorsal vertebra to above the calamus with a canal that upon incision released a serous liquid. The walls of this cavity consisted of cellular bands and were lined with a fibrous membrane”.

In his discussion, Nonat suggested that hemorrhaging had occurred in the abnormal cavity and had been resorbed (Nonat, 1838). The existence of a canal in the healthy spinal cord had been definitively accepted, by 1860, after the work of the German anatomist and surgeon Benedict Stilling (1810-1879) as well as Jean Cruveilhier (1791-1874) in France (Stilling, 1859; Cruveilhier, 1862-1867). In England in 1859, Lockhart Clarke (1817-1880) made an important contribution to the description of the various structures of the spinal cord and brainstem. His name is associated with the description of the posterior thoracic nucleus, or Clarke's column. Incidentally, he also described a second spinal cord canal, thereby observing syringomyelia without putting a name to it:

“In the human spinal cord, the canal is often completely filled up by what would appear to be the debris of the epithelium; [...] sometimes in the midst of this heap there remains a small opening or canal, which, strange to say, is still lined or surrounded at its margin by the usual regular layer of columnar cells; and what is still more curious, I occasionally find, particularly in the cervical region, two such secondary canals, each lined in the ordinary way” (Clarke, 1859).

The writings of Guillaume Duchenne de Boulogne (1806-1875) suggest that he had observed patients with syringomyelia as early as 1860. In the 1861 edition of his book *De l'électrisation localisée* concerning progressive muscular atrophy, he focused on how sensation is affected in certain patients. In one passage (p. 448-449), he described syringomyelia without realizing it:

“All authors who have written on progressive fatty muscular atrophy have maintained that sensation is always normal [...]. In a good third of the cases I have observed, I have noted that electro-muscular sensation was more or less lost as was cutaneous sensation. This anesthesia is sometimes so significant that the patients are insensitive both to the strongest faradic currents and to fire. I have observed some patients sustain serious burns in the anesthetized areas, because they did not perceive the action of the incandescent objects; they were not visually warned that these objects were in contact with their bodies. This anesthesia is generally found in the upper limbs, decreasing from the hand to the shoulder” (Duchenne, 1861).

It is interesting to read the chronology written by Gilles de la Tourette (1857-1904) in 1889 of a patient examined successively by Duchenne, who diagnosed her with progressive muscular atrophy, then by Charcot who in 1879, first diagnosed her with amyotrophic lateral sclerosis, but was unsure because of the slow development and also suspected multiple sclerosis. Finally, in 1889, Charcot diagnosed her with syringomyelia after reading the semiology described by the German authors (Gilles de la Tourette, 1889) (see drawing by P. Richer).

Henri Hallopeau (1842-1919), at that time an *interne* (house officer) under Alfred Vulpian (1826-1887), published in 1870 in *Gazette Médicale de Paris* an observation of a 62-year old woman with asymmetrical atrophy of the muscles in both forearms and paretic, claw-like hands, as well as facial droop, nystagmus and pupillary inequality. Upon autopsy, he observed:

“The medulla oblongata shows remarkable damage; it is hollowed out on the right, toward its centre part, with a cavity whose anterior wall initially corresponds to the olive, then continues obliquely back and into the floor which is reached close to the median line”.

He concluded with a microscopic examination of the spinal cord:

Specifically, inflammation occurred in the conjunctive substance around the spinal cord canal, with a considerable mass of morbid tissue

forming in the centre of the spinal cord. Later this tissue underwent regressive metamorphoses leading to partial destruction [...]. As a result of sclerosis, the spinal cord atrophied. Serious damage to the anterior horns resulted in secondary atrophy of part of the nerve roots and corresponding muscles” (Hallepeau, 1870).

Here Hallepeau is the first to describe a form of syringobulbia affecting the medulla and pons, as he proved in his autopsy notes, but without identifying a specific clinical semiology.

Charcot’s initial confusion between Amyotrophic Lateral Sclerosis and Syringomyelia

The theory of “ependymal sclerosis secondarily leading to the formation of an internal cavity” can be found in an 1869 article by Charcot and Alix Joffroy (1844-1908), one of the publications that would lead to the description of amyotrophic lateral sclerosis (Charcot, Joffroy, 1869; Guinon, 1894). They showed that damage to the nerve cells of the anterior horn causes peripheral muscular atrophy and paresis. In his thesis defended in 1873 and entitled *De la pachyméningite cervicale hypertrophique d’origine spontanée*, Joffroy described cases of chronic myelitis in which he observed the following: “In areas where fibrinoid transformation has taken place, mainly in the grey matter, one frequently finds cavities with generally irregular contours and variable diameter”. Joffroy provided a detailed account of cervical pain irradiating into the arms of patients with “pachyméningite” and noted that

“sensation in these patients has not been sufficiently well studied, but one can argue that it does not present consistent modifications. In general anaesthesia is initially observed, but later either anaesthesia or hyperesthesia is observed”.

Joffroy and Charcot provided a remarkable description of amyotrophy causing “preacher’s hand”, but seemed not to have examined in depth the associated sensory disturbances which distinguish two different clinical pictures: amyotrophic lateral sclerosis and syringomyelia (Joffroy, 1873). It is therefore likely that in 1873, Charcot examined patients suffering from syringomyelia without recognising the disease. Later in 1891, Charcot provided

the following explanation:

“The confusion between syringomyelia and muscular atrophy is certainly not the only diagnostic error committed. Given the multiplicity of its locations, forms and dimensions, the lacunar lesion is capable of producing the most disparate phenomena. It is clear that gliomatosis shows a marked predilection for the central grey matter and the adjacent white elements. As a result, a clinical form can be recognised that is to some degree typical and characterised by symptoms consistent enough to be grouped together under the term ‘syringomyelic syndrome’” (Charcot, Brissaud, 1891).

In the middle of the 19th century, anatomo-pathological interest in spinal cord cavities existed in all European countries. In 1875, Theodore Simon in Hamburg was studying the cavities existing independently from the central canal and formed by the disintegration of tumors which he referred to as telangiectasic gliomas. He proposed the term hydromyelia be reserved for the simple dilation of the spinal cord canal, by comparison with hydrocephalus, and that syringomyelia be used for pathological cavities (Simon, 1875). The same year, his colleague Carl Westphal (1833-1890) confirmed the concept of a cavity independent from the spinal cord canal and potentially resulting from necrotic resorption of a gliomatous tumour in the spinal cord. In his clinical description of a 37-year old man, he described atrophy in an upper limb, predominantly at the extremities, with loss of sensitivity to hot and cold (Westphal, 1875; Westphal, 1883), but he did not suggest any links between his clinical and anatomopathological observations.

Clinical picture of Syringomyelia in Germany in 1882

In his Tuesday lesson on 28 June 1889, Charcot said:

“Today I propose that we study a few examples of an organic spinal disease newly introduced in the neuropathological clinic, where from now on it will occupy a distinguished place, given that it is not much rarer than amyotrophic lateral sclerosis, for example. I have named it syringomyelia. As I noted above, the disease’s introduction in the clinic is very recent. While for some time certain cavities have been known,

with varying degrees of anatomical precision, to form in the central parts of the spinal cord, it was not known until recently which symptoms could point to these cavities during a patient's lifetime. In summary, until now, syringomyelia has been seen as a simple anatomopathological curiosity; it has not been considered in practice. This has changed with the work of two German authors – Professor Schultz in Dorpat and Professor Kahler in Prague – who contributed a series of important studies initiated in 1882 and linking the syringomyelic lesion with certain functional or organic problems which, when patients present them in the clinic, signal the existence of the damage and even allow determining the main details of location and



extent” (Charcot, 1889) (Picture 2).

Picture 2. Patient depicted by Jean-Martin Charcot during the Tuesday's lesson 1889, 28 juni by Paul Richer (1849-1933) « Cas de syringomyélie gliomateuse » O. Walusinski's library.

In 1882 in Prague, Otto Kahler (1849-1893) described a patient with paresis of the left arm involving contractures; he noted the loss of temperature sensation located initially in the paralysed side, then

extending in a cape-like fashion to the opposite side and to a thigh. He intuitively attributed this pathology, which linked dissociated sensory and motor problems and evolved progressively to other territories, to a high spinal cord lesion (von Kahler, 1882). He may have read the observation of Friedrich Schultz (1848-1934), published a few months earlier. In a 37-year old woman, Schultz watched the development of the following symptoms over four years:

“Weakness accompanied by muscular atrophy in the upper limbs, partial loss of temperature sensation, analgesia in the upper limbs and thorax, then in the lower limbs, and analgesia in the tongue, in addition to intermittent medullary symptoms and trophic disturbances in the skin (bullae in the right hand)”.

Schultz explained the motor and sensory deficits and the trophic disturbances by the presence of an abnormal vertical cavity in the spinal cord:

“Hyperplastic neuroglia are found along the edge of the cavity, which exclusively invades the grey matter of the posterior horns; the white matter was normal in appearance.” (Schultz, 1882).

Kahler and Schultz are thus indeed to be credited with the description in 1882 of the clinical symptoms of syringomyelia and their causal links with an abnormal spinal cord cavity. In the years thereafter, several observations evoking syringomyelia were published. In 1883, W.E. Fürstner and H. Zacher garnered attention by advancing general paralysis as an aetiology (Fürstner, Zacher, 1883). In 1884, Ernst Remak (1849-1911) described a “central gliomatosis” for which the clinical picture involved a loss of pain and temperature perception in the left arm and the upper left part of the body as well as a muscular atrophy, trophic disturbances of the skin and shoulder disarticulation. Remak clearly noted that tactile sensation was preserved in territories with loss of pain sensation (Remak, 1884; 1889).

In 1885, Hermann Oppenheim (1858-1919) postulated syringomyelia in a 32-year old male patient in whom anaesthesia developed over two years with loss of temperature sensation in the upper limbs, thorax, neck and nape of the neck. The patient also had muscular atrophy in his left hand, bullae on his fingers and tenacious paraesthesia in his upper extremities (Oppenheim, 1885). Also in 1885, shortly before leaving

for Paris to attend Charcot's lessons, Sigmund Freud (1856-1939) described "a case of muscular atrophy with extensive sensation disturbances", which were in fact located in the upper thorax and upper limbs. In these territories, he described loss of pain and temperature sensation, whereas the sense of touch was preserved, except around the left shoulder. Freud compared his observation with those of earlier authors, and based on their cases followed by autopsy, he proposed the diagnosis of central gliomatosis with syringomyelia for his patient (Freud, 1885). In 1887, Martin Bernhardt (1844-1915) published observations of two patients aged 35 and 54 years respectively, with muscular atrophy of the arm and a claw-like hand, totally insensitive to pain during injections or electrical stimulation. His older patient had had anaesthesia for 22 years when he examined her and had progressively developed scoliosis and various trophic disturbances. He diagnosed a syringomyelia clinically (Bernhardt, 1887).

Wladimir Roth (1848-1916) published his paper, which

excavé de la moelle" Paris 1890

O.Walusinski's library

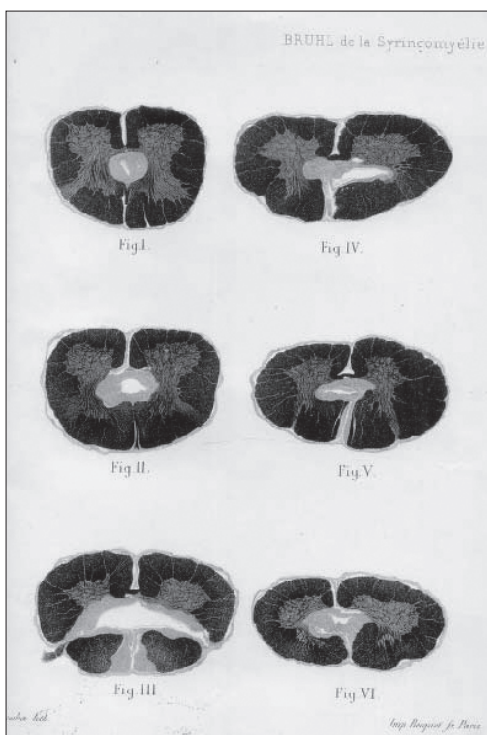
he proposed a series of ten very detailed observations. Based on these several cases, he presented the complete clinical symptomatology and discussed the semiological value of each symptom (Roth, 1887; 1888). One of his students, Azriel Raichline, would defend a thesis in Paris in 1892 on the clinical description of syringobulbia (Raichline, 1892).

All of the work published at this time, around 100 observations, are collated in the remarkable thesis of Anna Bäumlér (1852-1934), defended in Zurich in 1887. This thesis would serve as the framework for the thesis defended in 1889 in Paris by Isaac Bruhl (1863-?), *interne* under Georges Debove (1845-1920), as well as the thesis of Daniel Critzman (1863-1928) defended in 1892 and directed by Charcot, while Critzman was an *externe* (non-resident student) under Maurice Letulle (1853-1929) (Bäumler, 1887; Bruhl, 1890; Critzman, 1892) (Picture 3).

French Neurology adopts the Diagnosis of Syringomyelia

As noted by Charcot, most of these descriptions came from Germany. Paul Berbez (1859-?), at the time an *interne* under Charcot, presented a paper to the Société Médicale des Hôpitaux on 9 July 1885, entitled *Essai de diagnostic d'une affection de la moelle indépendante du tabès avec arthropathie du coude* (Diagnosis of a spinal cord disease independent of tabes with arthropathy of the elbow). The autopsy was carried out by his successor Paul Blocq (1860-1896) and presented to the Société anatomique on 18 February 1887. The autopsy report points to syringomyelia but does not specifically name it (Berbez, 1885; 1889; Blocq, 1887). This case was covered in the Tuesday lesson of 28 June 1889 and published again in 1891, when Charcot wrote the preface to the magnificent spinal cord atlas of Blocq and Albert Londe (1858-1917). Plate XLII shows

"gliomatous syringomyelia in the dorsal region. There is a central, cordiform cavity delimited by a wall with budding and surrounded by the glioma that pushed back the grey matter, partially destroying it. Despite the extent of the lesion, there is no clear degeneration of the white tracts" (Blocq, 1891) (Picture 4).



was presented at the 2nd congress of Russian physicians held in Moscow in 1887, in *Revue Neurologique*. Therein

Picture 3. Isaac Bruhl's thesis: « Contribution à l'étude de la syringomyélie ». "Gliome central



Picture 4. Planche XLII, from « L'anatomie pathologique de la moëlle épinière » Paul Blocq et Albert Londe. 1891 "Syringomyélie gliomateuse - région dorsale" O. Walusinski's library.

It was not until 1889 that Debove published the first observation in France, describing a 38-year old man suffering from Duchenne-Aran type muscular atrophy of both hands over 5 years, associated with loss of pain and heat sensation. The patient was briefly hospitalized in Charcot's department in 1887 and underwent electrotherapy and sulphur baths, but the etiological diagnosis was not established. This is the first observation in Bruhl's thesis (Debove, 1889). At the same session of the Société Médicale des Hôpitaux, on 22 February 1889, Dejerine presented another similar case (Dejerine, 1889). Bruhl's thesis includes observations of 36 cases. The first 10 collated cases were novel and involved French patients, many of whom were observed by Charcot's *internes*: Paul Blocq (1860-1896), Gilles de la Tourette (1857-1904) and Adolphe Dutil (1862-1899). The other observations were copied

from Bäumlér's thesis. Thus, in 1889, syringomyelia was an established and accepted clinical entity in France.

Analgesic panaris as observed by Morvan: a French description

In 1883, Augustin Morvan (1819-1897), a country physician in Lannilis, Brittany published the following:

"The disease which we will study involves paresis with analgesia in the upper extremities, initially limited to one side, then moving in most cases to the other side and always resulting in the production of one or more whitlow inflammations".

Morvan details a novel clinical picture characterised by the successive appearance of multiple whitlow inflammations, resulting in necrosis and definitive deformation of the fingers, accompanied by muscular atrophy in the hands and upper limbs, as well as disturbances in tactile and temperature sensation. Morvan noted that there was complete lack of pain sensation for these whitlow inflammations, which could be operated on without causing the patient any pain. He named the clinical condition "analgesic paresis with whitlow". In five successive papers from 1883 to 1889 presented to the Académie de Médecine, he completed his description with new observations published in *La Gazette Hebdomadaire* (Morvan, 1883), adding trophic disturbances to the initial clinical picture: bone fragility, hyperhidrosis, hemorrhage and bone/joint deformations (without noting any scoliosis). Probably unaware of the articles of Kahler and Schultze, he suggested a pathophysiological explanation based on the first publication of Augusta Klumpke (1859-1927, the future Madame Dejerine), which described radicular paralysis in the brachial plexus (Klumpke, 1885). Morvan wrote:

"The disease started in the posterior tract and, probably at a later time, moved to the anterior tract. I believe the natural progression of the disease to be the following: 1) analgesia; an initial analgesia, because it is incomplete, can be found alone, never paresis; 2) when the two orders of nerves are involved, the sensory paralysis is always one degree more advanced than the motor paralysis, the former being complete whereas the latter is not yet complete and may never become so" (Morvan, 1886).

Morvan indicated the location of the lesions based on

the perspicacity of his judgment alone, unaware of the anatomical pathology of the disease he was describing. In 1888, Georges Charles d'Oger de Spéville dedicated his thesis to Morvan's disease, after writing an observation of a case when he was an *externe* under Victor Hanot (1844-1896). He reviewed the 29 cases previously published. Due to the slow progression and gradual extension, he posited a cause in the central spinal cord and refuted Morvan's explanation of peripheral neuritis (d'Oger de Spéville, 1888). Louis Hallion (1862-1940), an *interne* under Charcot, confirmed in his 1892 thesis the frequent presence of scoliosis during syringomyelia (Hallion, 1892). The first anatomical study was published in 1889. Mathieu Prouff (1849-1931), with the help of Albert Gombault (1844-1904), noted the existence of scoliosis:

“Considerable deviation of the vertebral column [...] The question as to whether the central grey matter contains abnormal cavities resembling those that characterise syringomyelia is more difficult to judge definitively because the spinal cord was contused during extraction. [...] It is, however, possible to rule out syringomyelia” (Prouff, 1889).

With regard to this observation, Charcot remained evasive in his Tuesday lesson of 28 June 1889:

“There is still much discussion as to whether the disease of Morvan of Lannilis, otherwise known as analgesic whitlow, should fall entirely within the definition of syringomyelia, or should be classified apart” (Charcot, 1889).

Dejerine was more peremptory and stated the following at the 5 July 1890 meeting of the Société de Biologie:

“There is no doubt that syringomyelia may sometimes involve a symptomatic complex similar to that of analgesic whitlow, and certain authors, Roth in Moscow in particular, have come to this very conclusion. Morvan has taken a strongly opposed position by showing that sensory dissociation does not exist in the disease to which he has given his name. Furthermore, loss of tactile sensation is very rare in syringomyelia and only occurs at a very advanced stage in the disease. The frequency of analgesic whitlow in certain areas is yet another factor arguing against the syringomyelic nature

of the process and tends to prove that Morvan's disease belongs to a family of neuritis caused by infection or toxicity. In this way, analgesic whitlow resembles anaesthetic leprosy, which also is found in certain areas. We know that nothing similar exists for gliomatosis in the spinal cord, which is relatively rare in large urban centres such as Paris. In summary, Morvan's disease increasingly appears to be linked not to syringomyelia, but rather to a peripheral neuritis whose nature and causes have yet to be determined” (Dejerine, 1890).

Two years later, Charcot summed up how this debate was decided:

“It became clear that only the pathological anatomy could provide a definitive solution to the problem. It has spoken, and in my opinion, has done so peremptorily in favour of the single doctrine” (Charcot, 1891).

Joffroy and his *interne* Charles Achard (1860-1944) published an observation of Morvan's disease with autopsy in July 1890, including microscopic preparations of the spinal cord and upper limb nerves. Therein they note, contrary to Dejerine, that anaesthetic dissociation is the rule rather than the exception in Morvan's disease, which shares with syringomyelia paresis and muscular atrophy, scoliosis and various trophic disturbances of the skin and joints (Joffroy, Achard, 1890; Achard, 1890). In 1890, Georges Guinon (1859-1932), *chef de clinique* (specialist registrar) under Charcot, and Adolphe Dutil (1862-1899), his *interne*, described two cases of Morvan's disease examined by Charcot. They maintained that the disease was accompanied by hysteria due to visual, olfactory and gustatory disturbances, but they did not know how to interpret the nystagmus, which indicated syringobulbia (Guinon, Dutil, 1890). Shortly thereafter, Raichline translated from Russian another paper by Roth for Guinon. Based on eight new cases, Roth's paper formally concluded that syringomyelia and Morvan's disease were a single condition (Guinon, Raichline, 1891). Thus in 1891 Charcot drew the following conclusion, accepting that the two diseases were in fact one, which he had doubted in 1889.

“The symptomatology associated with Morvan's disease can result entirely from the spinal lesions of syringomyelia. The dispute now appears to me

to be empty. There are not two distinct diseases: there is only one, and however novel it may appear clinically, Morvan's disease merely represents an atypical form of syringomyelia. [...] The major characteristic of the lesion, and its originality, lies primarily in its location. It initially limits itself to the grey matter, generally starting behind the central canal (often unaffected) then preferentially invading the posterior horns, either on one or both sides, followed by the anterior horns, either by histological propagation or by compression".

Diverse Etiological Hypotheses and Differential Diagnoses

Demetrius Zambaco-Pacha (1832-1913) in Constantinople argued that Morvan's disease was a variety of the leprosy that causes nerve damage, a hypothesis he believed was confirmed by a case of Albert Pitres (1848-1928) with leprosy bacteria in a neuritic nodule, and also by the endemic nature of Morvan's disease in certain areas of Brittany (Zambaco, 1891; Pitres, Sabrazès, 1893). The conclusion of Joseph Babinski (1857-1932) on this question is clear:

"There are two forms of syringomyelic dissociation: perfect and imperfect. In the first form, pain and temperature sensation is totally lost, whereas tactile sensation remains normal. The second form has several varieties, one of which is characterised by loss of pain and temperature sensation, while tactile sensation is incompletely maintained. [...] Pending new information, we can formulate two propositions: first of all, in peripheral neuritis (cf. leprosy), the diverse modes of sensation may undergo imperfect syringomyelic dissociation; secondly, it is not rigorously demonstrated that perfect syringomyelic dissociation can result from peripheral nerve damage, and it is entirely exceptional if it can" (Babinski, 1892).

In his lesson of 1 April 1898, Fulgence Raymond (1844-1910), Charcot's successor, taught that "syringomyelia may simulate the symptomatology of tabes with striking similarity; this is when syringomyelic gliomatosis has the same topography as the spinal cord damage in tabes". Raymond based this lesson on a publication of Max Nonne (1861-1959), Wilhelm Erb's assistant in Heidelberg, whose patient had a comparable clinical

picture to that of the 52-year old woman Raymond presented to his students. Raymond's patient had scoliosis with amyotrophy and loss of pain and temperature sensation in her hands but also shooting pains in her limbs as well as Argyll Robertson's pupil and Romberg's sign, which indicate tabes (Raymond, 1900; Nonne, 1892).

Georges Guillain (1876-1961) defended his thesis in 1902 to a jury that included Raymond. He had studied five patients presenting parietic amyotrophy with pyramidal hypertonia in the limbs (exaggerated reflexes and Babinski's reflex), together with dissociation of tactile and temperature/pain sensation. This description stood in contrast to the clinical picture of paresis previously reported, with amyotrophy and absent reflexes, leading Guillain to conclude:

"If one considers the motor functions in many cases of syringomyelia or hydromyelia, the resulting conclusions are paradoxical. There are syringomyelia patients with enormous cavity-causing lesions that destroy much or all of their grey matter, with considerable lesions in the pyramidal tracts, who do not present motor problems in keeping with the intensity of their lesions. In light of these facts, one wonders whether it is reasonable to accept that nervous pathways strictly correspond to either sensory or motor conduction, and whether the theory of spinal cord localisations as it is taught should be regarded as absolutely accurate. It would appear that, in the spinal cord and the nerve centres, sensation can use different paths; we also believe that motor functions can use paths other than the pyramidal tract" (Guillain, 1902).

Description of Syringomyelia increases Understanding of the Physiology of Sensation

Guillain's ideas had already been highlighted in the 1891 thesis of Frédéric Caillet, who was an *interne* under Raymond: "Clinical medicine has come to the aide of physiology" (Caillet, 1891). While it was established that the dorsal roots carried sensation based on the experiments of François Magendie (1783-1855) and that the different types of perception could be separated into sensitivity to pressure, touch and pain, there was no knowledge of the physiological perceptive mechanisms and the existence of different spinal tracts within the grey matter. Roth thus wrote in *Revue Neurologique* in

1888:

“It is regrettable that our knowledge of how the sensory pathways in the spinal cord function is of little help in correcting the insufficiency of facts with theoretical suppositions. We don't even know with certainty if there are in fact isolated anatomical conductors for tactile, temperature and pain sensation, not to mention the direction these different paths take after the roots enter the spinal cord and their later function in the white and grey matter. But cases of gliomatosis give us hope of solving these physiological problems”.

After a speaker stated before the *Société de Biologie* on 8 February 1890 that “in physiology, we do not know where the different types of sensation pass; the problem is not resolved”, Dejerine responded:

“Clinically, the problem is absolutely resolved as established by Kahler and Schultze; touch is absolutely independent from temperature sensation on the whole, and temperature sensation is by contrast linked to pain sensation”.

Whereas the clinical and anatomopathological examinations for tabes and Pott's disease did not improve physiological understanding, the isolation of syringomyelia made it possible to recognize the dissociation of temperature and pain pathways from tactile and proprioceptive pathways (Roth, 1888; Dejerine, 1890).

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