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History of Neurology

The locked-in syndrome: The early French descriptions

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ABSTRACT

The classic 1966 description of locked-in syndrome was performed by Plum and Posner. Here, we revisit the world's first case report of this condition, which was presented in 1875 by Camille Darolles, an intern supervised by François Damaschino, at a monthly meeting of the Société Anatomique de Paris chaired by Jean-Martin Charcot. We also review the fascination of classic writers with this syndrome, including Alexandre Dumas, a genius of literature and known admirer of the medical sciences who, in the book "The Count of Monte Cristo" published in 1846, described a character with this condition.

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1. Introduction

The term locked-in syndrome (LIS) was coined in 1966 by Plum and Posner in their classic text "Diagnosis of Stupor and Coma" [1]. The syndrome is characterized by the presence of quadriplegia associated with paralysis of the lower cranial

nerves, with preservation of vertical eye movements, blinking, and consciousness [1,2]. Most commonly, LIS results from bilateral infarction in the anterior pontine region due to occlusions of the paramedian arteries or the basilar artery [2]. After the classic form was described, other authors published descriptions of cases they encountered in practice with

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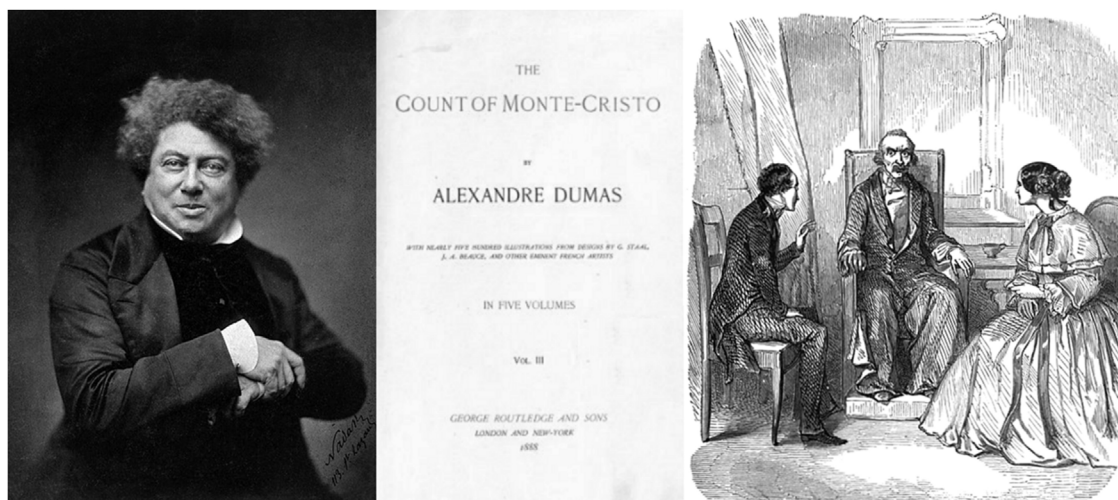


Fig. 1 – Alexandre Dumas (1802–1870) and illustration of the character M. Noirtier de Villefort, from the book “The Count of Monte Cristo”, with locked-in syndrome. (Composition: Nadar, public domain, via Wikimedia Commons and The Count of Monte-Cristo 1887 Volume 3 [PD US expired], via Wikimedia Commons).

atypical clinical presentations [2–4]. Historical aspects related to LIS have been widely discussed in the literature since the twentieth century [5].

This text reviews a case report of LIS presented in 1875 Paris at the *Société Anatomique de Paris* by Camille Darolles, an intern under François Damaschino, at the monthly meeting of the *Société Anatomique de Paris* that was chaired by Jean-Martin Charcot. We also describe how this syndrome has fascinated writers even prior to this probable first classical description.

2. Locked-in syndrome: an 1875 case report

In one of the sessions of the *Société Anatomique* in Paris held in June 1875 under the presidency of Jean-Martin Charcot (1825–1893), a clinical-pathological case was presented by a hospital intern named Camille Darolles (1847–1915) who later practiced as a clinician in Provins (Seine-et-Marne), 90 km east of Paris [6]. The presentation was entitled “*Ramolissement de la protubérance : thrombose du tronc basilaire*” [“Softening of the pons: thrombosis of the basilar trunk”]. It described a 36-year-old woman admitted to the service run by François Damaschino (1840–1889). She was previously healthy and began experiencing an excruciating occipital headache eight days prior to admission. Six days after the headache began, she experienced a brief loss of consciousness that lasted roughly 15 minutes; when she regained consciousness, she presented with right-sided hemiplegia and dysarthria [6]. The next day she was anarthric and tetraplegic, only able to communicate by moving her eyes: “*Ce n’est que par les mouvements qu’elle imprime aux globes oculaires qu’elle tâche de répondre aux questions qu’on lui adresse, et d’accuser les sensations qu’elle oyenn.*” [6] Through this communication, she was able to express to the physicians that her senses were preserved and that she still suffered from the headache. On the third day of symptoms, she had a fine, regular, and constant arterial pulse and

developed a fever (38.2 °C). She was treated with blood extraction through successive application of thirty leeches over her mastoid regions. She later experienced recurrent tonic-clonic seizures and left-side mydriasis; she became irresponsive and her breathing became stertorous the next day, and death was confirmed in the afternoon [6]. An autopsy was subsequently performed and found a subarachnoid hemorrhage over the left hemisphere secondary to the rupture of a pial vein and, more importantly, an obstruction (“*...Une incision longitudinale de l’artère met à jour une concretion gisâtre de forme ovoïde de la grosseur d’un pépin de raisin. Ce corps obturant, de consistance assez ferme, est situé à la partie moyenne de l’artère certainement per la périphérie à la tunique interne du vaisseau*”) in the mid-portion of the basilar artery, along with a lesion in the right portion of the pons marked by tissue softening [6].

3. Locked-in syndrome in literature

Three notable depictions of LIS are found in the non-medical literature [5,7–10]. The first, considered the first non-medical description of LIS in the world, is found in the classic book *Le Comte de Monte Cristo* (“The Count of Monte Cristo”) published by the French writer Alexandre Dumas (1802–1870) in 1846 [7,8]. One character in this book, M. Noirtier de Villefort, suffers a stroke and the description of his condition is characteristic of LIS: “...he was a corpse with living eyes.” “...He was able to communicate through blinking, closing his eyes for “yes” and winking several times for “no” (Fig. 1) [7,8].” There is evidence that Dumas was greatly interested in medicine and made several visits to hospitals at that time, guided by his physician and friend Dr. Thibaut [7]. Likewise, his interest in the subject of stroke (which was incidentally very common among the characters in his books) stemmed from the fact that this disease frequently affected members of his family, including his mother; Dumas himself died of a

stroke in 1870 [7]. Another publication, this time by Emile Zola (1840–1902) in his 1868 novel *Thérèse Raquin*, featured a character (the protagonist's mother) with a stroke that was clinically very similar to LIS [9]. Finally, more recently in 1997 a book was published under the title *Le scaphandre et le papillon* [The Diving Bell and the Butterfly] based on a true story of the journalist and writer Jean-Dominique Bauby (1952–1987), who suffered a brainstem stroke at age 43 in 1995 and developed LIS [10]. This book was made into a movie that was released in 2007, garnering worldwide attention as well as many awards [10].

4. Conclusion

The case presented by Camille Darolles in 1875, ninety-one years before the description by Plum and Posner, can be considered the first description of LIS. This disease has fascinated French writers, and Alexandre Dumas, a literary genius and admirer of the medical sciences, had already described a character with LIS in 1846.

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Statement of ethics

We confirm that approval of an institutional review board was not required for this work. We confirm that we have read the

Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

Data availability statement

This article is a historical report. There is no data to be deposited.

Disclosure of interest

The authors declare that they have no competing interest.

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