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Case Report

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The outcome of a patient with locked in syndrome in countries with a limited technical platform: about an observation

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Abstract

The locked in syndrome is a diagnosis that puts the clinician to the knee because of his prognosis for most cases, especially when its origin is vascular by occlusion of the basilar artery or a pontine haemorrhage. The victims often remain in a coma for some time, then little by little, wake up, but remain paralyzed and dumb, reminiscent of a vegetative state. Scarcity and prognosis make it a special entity. We report in this article the case of a patient admitted at the neurological clinic I.P NdiayeUniversity Hospital of Fann, Senegal, Dakar, for a locked in syndrome of vascular ischemic origin and whose evolution was unfavorable marked by a death on day 17 of his admission in a context of vomito negro and cardiac arrest.

Keywords: Locked in syndrome, Outcome, Limited technical platform.

INTRODUCTION

The Locked-In Syndrome (LIS), introduced by Plum and Posner [1], refers to patients who are locked from inside. This condition is due to an interruption of the corticospinal and corticobulbar pathways, thus inducing bilateral supranuclear palsy, which then precludes voluntary motor control of the four limbs and communication by speech or gesture by paralysis of the mixed nerves, leaving the patient free to communicate his state of consciousness only by the movements of verticality or the blinking of the eyes, by preservation of the motor nuclei of III (oculomotor nerve). This state can then be confused with coma, vegetative state and akinetic mutism, which can then explain the not insignificant course consisting of stays in different institutions, sometimes interrupted by brief returns to the home of the patient who is victim following. The diagnostic wondering. Even if medicine manages to increase the life expectancy of patients up to 27 years [2]. This is still not the case in countries with limited resources and technical platform. We report in this article, the case of locked in syndrome of a patient who had been admitted to the neurological clinic Ibrahim Pierre Ndiaye of Dakar, Senegal, and whose prognosis was unfavourable.

MEDICAL OBSERVATION

This was the patient AW, 55 years old, right-handed of who had been admitted to the neurological clinic IP Ndiaye of the University Hospital Center of Fann dated 07.06.2018, for motor deficit of 4 members. The history of his illness started on the 30th May 2018 after he had collapsed in the bathroom in the morning, lost consciounsness for about 3 hours, and then regained. The same day, in the evening he was taken to the main hospital of the place for a treatment where he received ceftriaxone, Perfalgan, junior aspirin as well as normal saline 0.9% and dextrose 5%. Given the worsening of his status he was subsequently transferred to the neurology department for further management. He had no cardiovascular risk factors in his history. Upon his arrival, the examination found a patient awake, aphonic, with swallowing disorder, quadriplegic with muscular hypotonia, the vertical eye movements were preserved, a bilateral babinski. Tachycardia at 117 beats / min and body temperature at 38.9 °C were noted. With this, the diagnosis of locked in syndrome and an infectious syndrome was made, so a cerebral computed tomography was requested which showed an area of hypodensity in the ventral midbrain (Fig 1). The patient was admitted to the intensive care unit other test were ordered which revealed CRP of 173 mg / I, leukocytosis of 16,340 elements / mm³ and urine

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culture grew up isolate *E coli*. On day 17 the evolution will be unfavorable, marked by the occurrence of vomitonegro and cardiac arrest.



Figure 1: Cerebral CT- scan sagittal section, showed an area of hypodensity in the ventral midbrain (hypodensity in the basilar artery trunk).

DISCUSSION

The locked in syndrome is a neurological status often unusual, rare in the current practice of the neurologist doctor and therefore a difficulty to make a systematic study to identify specificities and epidemiological specificities. Already on his side, Leon-Carrion *et al.* (2002a)[3], in his study he reported that the doctor could only realize that the patient was able to communicate through the eyes only in 23%, while the family would do it in 55% of cases. The same author had pointed out that the delay between the diagnosis and the occurrence or the installation of the LIS would be 78 days on average [4]. This would then reflect one of the initial misdiagnosis.

This leads us to say, since the initial diagnosis is wrong, it could then impact the further treatment, and that it would be subject to medical chopping. This situation was present in our patient, diagnostic wondering and internment in more than two structures before admission to the neurological clinic Ibrahim P. Ndiaye, where he was admitted.

The cause of this LIS in our patient was vascular. It was on day 17 that his evolution was disastrous marked by a death in a challenge of respiratory distress and cardiac rhythm disorder.

This evolutionary modality has been reported by other authors in their studies, although ancient. For Ohry A, the long-term survival of these patients was rare [5]. Patterson and Grabois in their study had shown that [6], mortality in case of LIS was high in the initial phase in the order of 76% for cases of vascular origin and 41% for non-vascular cases and that in 87% of cases, occurred during the first four month.

Moreover, for its part, Doble *et al.* 2003 had reported in his study that after stabilization of the patient beyond one year, his life expectancy would vary from 10 to 20 years, in 83% and 40% of cases respectively[7]. The high mortality rate in the acute or initial phase would be multifactorial, including the widening of the infarct, an infection including pneumonia or decubitus ulcers, the bad localization of the infarct (vegetativecenter) which then imposes a catch in charge of a resuscitation unit with assisted ventilation, intubation and then tracheotomy if indicated. Repeated bronchial aspirations as well as regular chest physiotherapy to reduce congestion which is linked to an inability to voluntarily modify breathing times. To our knowledge, for many developing countries, there is a lack of infrastructure

(resuscitation unit well equipped), qualified resource personnel (specialists physiotherapist and resuscitator, neurologist).

In addition, in case of survival, the patient should be admitted to a functional rehabilitation center for an extended stay, but this structure is unfortunately often difficult to find in many countries and even, if available, these institutions are counted on fingers and are not able to accommodate many of these patients who weigh medically while their prognosis of recovery is reserved.

In our opinion, this would be the cause of the increase in the mortality rate for patients who suffer from it and therefore a question about the future of the patient once the diagnosis is made while living in regions with limited resources.

In addition, to our knowledge, there is no data to date in Dakar on the incidence of locked in syndrome due to lack of long series. A Dutch study estimated this prevalence at 0.7 per 10,000 beds. [8].

CONCLUSION

If the diagnosis of locked in syndrome is already a problem especially in the medical community unfamiliar with neuroscience, the patient in whom the diagnosis is made in countries with limited technical platform logically sees his life prognosis brought into play. A complication however avoidable or manageable occurring during evolution of this pathology, unsuitable or inadequate medical conditions compromise this prognosis. Should we then hope for a dangerous natural evolution for the survival of the patient? And even if this evolution is proved to be favorable, but its future remains at the level of the question which one poses in our circles.

Conflict of interest

None declared.

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