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Case Report

Neuromuscular diseases in intensive care

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Summary

Guillain-Barre syndrome and myasthenia gravis are common in the Intensive Care Unit (ICU) and are not always easy to diagnose in the emergency department. The short-term risk is the occurrence of respiratory muscle paralysis, which must be monitored by repeated measurement of vital capacity, and the existence of respiratory impairment requires transfer to intensive care. Apart from specific therapeutic options (plasma exchange, intravenous immunoglobulins and anticholinesterase agents), management is mainly symptomatic. Our work is a retrospective study, concerning 23 cases of neuromuscular diseases in the ICU from a period from 2018 to 2022. The various epidemiological, clinical, paraclinical, therapeutic, and evolutionary data were collected from the files of the Intensive Care Department A1 of the Hassan II University Hospital of Fez for the study. Osteomas of the paranasal sinuses are benign, often asymptomatic, tumors that progress very slowly. Endocranial development of an osteoma can breach the dura mata, allowing air to enter the cranium producing pneumocephalia which leads to severe neurological deficiencies. Pneumocephalia is an exceptional complication of osteoma.

Introduction

Guillain-Barré Syndrome (GBS) has become, since the eradication of acute anterior poliomyelitis, the main cause of extensive acute paralysis in developed countries. Given the risk of short-term severity, it is a neurological emergency. Diagnosis is generally clinical, with additional tests used mainly to rule out a differential diagnosis. Symptomatic treatment is paramount, aimed at preventing complications of motor deficit [1,2].

Myasthenia is a chronic autoimmune disease linked to a defect in the transmission of nerve impulses between the motor nerve and striated muscle. A disease of the neuromuscular junction due to blockage of motor plate receptors by anti-acetylcholine receptor antibodies. It manifests as muscular weakness that worsens with exertion and improves with rest. Diagnosis is straightforward, but treatment is more difficult due to the unpredictable course of the disease. Its severity lies in the risk of respiratory complications that can be life-threatening [3,4].

Materials and methods

The aim of our study is to describe the clinical, diagnostic, therapeutic, and evolutionary aspects of neuromuscular diseases in the Intensive Care Department A1 of the Hassan II University Hospital of Fez between 2018 and 2022. Our work is a retrospective study, containing 23 cases from a period from 2018 to 2022. The various epidemiological, clinical, paraclinical, therapeutic, and evolutionary data were collected from the files of the Intensive Care Department A1 of the Hassan II University Hospital of Fez.

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Results

For our study, 23 cases were included (Table 1). The mean age was found to be 49.72, with extremes ranging from 17 to 81 years (Figure 1) and males predominated. An infectious syndrome was present in 45% of the cases (Figure 2). The main symptom (80% of cases) was heaviness of the limbs. Respiratory distress was reported in 8 patients, swallowing disorders in 6, and sphincter disorders in 3. Only one patient was admitted in a state of cardiocirculatory shock of hypoxic origin (Sd de guillan barré diagnosed by EMG) and another for a myasthenic crisis. The electroneuromyogram showed 3 types of electromyography (demyelinating in 52% of cases, axonal and post-synaptic neuromuscular block in 4%). In addition to symptomatic treatment, which was the norm in all our patients depending on the type of disease detected, specific treatment was instituted depending on the type of pathology: plasmapheresis in 17.39% of cases, immunoglobulins in 79%, anticholinesterase agents and corticosteroids in 13.04% of cases (Table 2). The evolution was marked by complications, especially of infectious order made nosocomial pneumonia in 50% of the cases, the mortality in our series was 21.73% of the cases. The majority of patients died in respiratory distress.

Discussion

Guillain-barré syndrome and myasthenia gravis are neuromuscular diseases of variable incidence, estimated at between 0.81 and 1.89 per 100,000 inhabitants per year in Western countries [1,2]. Myasthenia classically affects women between the ages of 20 and 30, which is consistent with the results of our series [3,4]. Diagnosis is predominantly clinical, with additional tests performed mainly to rule out a differential diagnosis. GBS evolves in three phases: an extension phase, a plateau phase, and a recovery phase; generally progressive weakness of more than one limb associated with areflexia that evolves in less than 4 weeks [5]. Heaviness of the limbs was found in 18 patients with major respiratory distress in 30% of cases. The technique most commonly used to assess neuromuscular transmission is repeated nerve stimulation. Stimulation at 2 Hz to 5 Hz results in a progressive decrement of the muscle component of the action potential in the myasthenic patient (from at least 10% between 1st and 5th stimulation). This test is abnormal in 75% of patients with generalized myasthenia but in less than 50% of patients with a purely ocular form, and post-synaptic neuromuscular block was found in only one patient in our series. Polyvalent immunoglobulins are as effective as plasma exchange in terms of mortality and clinical recovery (weaning from mechanical ventilation, recovery of walking) in the short (four weeks) and long (one-year) term [6-8]. Immunoglobulins were administered to 18 patients (78.26%). We had 4 deaths out of a total of 20 patients hospitalized for Guillain-Barré syndrome, i.e. 25% of cases, a high percentage given the limited number of cases in our series. The reported mortality for Guillain-Barré syndrome is 6% in the article by Doets, et al. 2018 [9]. Tolerance is reported to be better, with fewer discontinuations than with plasma exchange. IV immunoglobulins were administered in 18 of our patients. Risk factors for mortality were: age, nadir disease severity, need for mechanical ventilation, and hyponatremia.

Table 1: Incidence of neuromuscular diseases in our series.

Number of patients hospitalized	2819
Number of patients with Guillain-Barré syndrome	20
Number of patients with Myasthenia Gravis	3

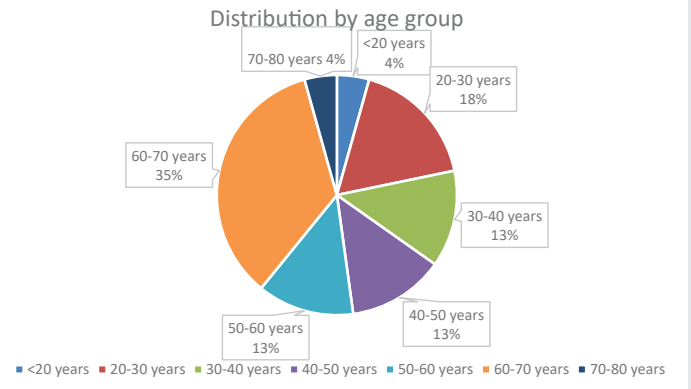

Figure 1: Age distribution.

Table 2: Specific treatment in our series.

Type	Workforce	Percentage
Plasmapheresis	4	17.39%
Immunoglobulin	18	78.26%
Anticholinesterase	3	13.04%
Corticosteroid therapy	3	13.04%

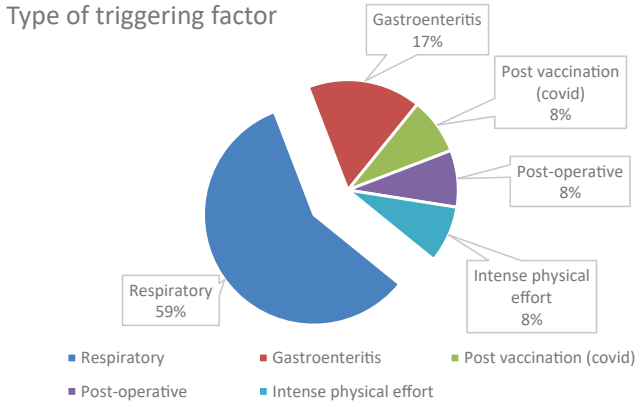

Figure 2: Type of trigger.

Conclusion

Patients with neuromuscular diseases are at greater risk of ICU admission, especially those with chronic respiratory failure, cardiac impairment, reduced mobility, or swallowing disorders. Early detection and prompt initiation of appropriate treatment can improve morbidity and mortality in these patients.

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