Background

Polymyositis (PM) is a connective tissue disease characterized by dysimmune inflammatory involvement of striated muscle with bilateral, symmetrical motor deficits, typically proximal to the limbs, which may generalize to all muscles of the body. Guillain–Barré syndrome (GBS) is an acute polyradiculoneuropathy of autoimmune origin. In its typical form, it comes out as an acute ascending and symmetrical flaccid paralysis associated with hypo- or areflexia occurring in a post-infectious context. In its pure motor form, GBS may mimic PM making the diagnosis difficult.

We report the unusual association of GBS and polymyositis in a young patient.

To our knowledge, this association has not been described in the literature.

Clinical Case

A 19-year-old woman presented 3 weeks after the onset of an influenza syndrome with an acute onset paraparesis which is rapidly progressive.

Her clinical examination revealed a symmetrical and ascending flaccid motor deficit with muscle strength rated at 1 in the quadriceps, hamstrings, leg muscles, biceps, triceps, and forearm muscles leading to tetraplegia. Paresthesias of the extremities in the form of tingling were reported by the patient. Thermo-algesic sensitivity
The osteo-tendinous reflexes and the cutaneous-plantar reflexes were abolished. Bulbar damage with deglutition disorders and acute respiratory failure complicated the case, indicating the need for mechanical ventilation. The electromyogram showed a slowing of nerve conduction velocities on the peripheral sensory and motor nerves in favor of a demyelinating sensory-motor polyradiculoneuritis (PRN). The CSF revealed an albuminous-cytological dissociation with a normal cellularity of 4 leukocytes/mm³ and a hyper-albuminorrachia of 0.8 g/L.

In front of this picture of (PRN), toxic causes (alcoholism) were eliminated at the interrogatory. Serologies for hepatitis B, C, HIV, and Lyme were negative. The dosage of vitamins B6 and B12 was normal. Metabolic disorders such as hypoglycemia, hypokalemia, and hypomagnesemia were also eliminated.

The diagnosis of Guillain–Barré syndrome in its severe form was retained, and the patient was treated with plasmapheresis and vitamin therapy in addition to mechanical ventilation. However, the evolution was marked by the persistence of bilateral and symmetrical proximal muscular deficit and swallowing disorders. A biological assessment noted elevated CPK at 4915 IU/L (33-fold normal), LDH at 1200 (fivefold normal), and hepatic cytolysis at sixfold normal. Antinuclear antibodies (ANA) were positive at 1/1280 with positive PM/SCL70 antibodies. Muscle MRI showed T2 hypersignals in the muscle compartments of the thighs, arms, trunk, and neck (Figure 1). The electromyogram showed the previous abnormalities in addition to myogenic tracing. Histological study of a muscle biopsy of the deltoid muscle showed an aspect compatible with polymyositis (Figure 2). We ruled out toxic, traumatic, medicinal, and infectious causes that could explain this muscular disorder.
The diagnosis of PM was made based on the proximal, bilateral and symmetrical muscle deficit, including that of the respiratory muscles, biological myolysis, radiological, electrophysiological, histological data, and immunological findings. The patient was treated with general corticosteroids at a dose of 1 mg/kg/day initiated by 3 pulses of Methylprednisolone maintained for 2 months followed by a slow taper, methotrexate at a dose of 15 mg/week and rituximab at a rate of 2 courses of 1 gram spaced 2 weeks apart in view of the severity of the initial life-threatening presentation. The evolution was marked by a spectacular improvement with the disappearance of the muscular deficit, the regression of swallowing disorders and respiratory distress, the normalization of muscle enzymes.

3 | DISCUSSION

GBS is an acute autoimmune polyradiculoneuritis reacting to a mostly infectious event resulting from a cross-immune response. It was first reported in 1916 by Georges Guillain, Jean-Alexandre Barré, and André Strohl who published the case of two soldiers who presented with rapidly progressive paralysis associated with tendon areflexia and paresthesias.2,3

Epidemiologically, the incidence of GBS is estimated at 1.8–2.8/100,000 persons/year and can affect people of any age with a male predominance. It occurs in 75% of cases within 3 weeks of an infectious syndrome.4

The pathophysiological mechanism of GBS is based on a molecular mimicry involving an homology between the antigenic epitopes of certain bacteria or viruses and certain constituents of the peripheral nerves responsible for a cross-activation of self-reactive T lymphocytes against membrane surface antigens of the Schwann cell, leading to the influx and activation of macrophages, major actors in demyelination.2,5,6

Typically, the diagnosis of GBS is based on the presence of acute flaccid paralysis with rapidly progressive ascending motor deficit of proximal and distal muscles associated with areflexia. It is supported by lumbar puncture data revealing hyperproteinorachia without hypocytosis and EMG data consistent with demyelinating peripheral neurogenic involvement showing decreased nerve conduction velocity, prolonged or absent F-wave, and increased distal latencies.7

Respiratory distress, an element of clinical severity, may occur in 10% of cases at the time of discovery of the disease and in 30% of cases during its course.8

In our patient, the diagnosis of GBS was certain in view of the context of occurrence, the typical clinical presentation, the electrophysiological data, and the albumino-cytological dissociation in the CSF. However, the absence of clinical improvement under appropriate treatment (plasma exchange, cardiorespiratory monitoring, vitamin therapy) led us to consider the differential diagnoses of this entity or a syndromic association.

The discovery of biological myolysis, radiological stigmata of inflammation, a myogenic syndrome on EMG, an histologically confirmed inflammatory myositis, and positive NAAs of the anti-PM/SCL70 type pleaded in favor of a PM associated with GBS.

This association does not appear to be coincidental. Indeed, PM and GBS are two autoimmune diseases that may have interrelated pathophysiological mechanisms involving a virus-induced immune response through molecular mimicry. An inflammatory infiltrate and T-cell hyperreactivity has been demonstrated at the endomysial and Schwann cell level in PM and GBS, respectively.9

The coexistence of these two diseases is a challenging diagnosis as the distinction between these two entities, which are clinically superimposable due to the presence of symmetrical muscle deficits affecting proximal and distal muscles in the same patient, is quite delicate.

GBS and PM are both severe due to the risk of life-threatening complications that they can cause. Severe forms are managed in an intensive care unit with cardiorespiratory support. Intravenous immunoglobulin (IVIg) and plasma exchange have similar efficacy in the treatment of GBS as demonstrated in several studies, while the combination of the two treatments has not shown superiority.10

The recommended total dose of IVIg is 2 g/kg, divided over 5 days at a rate of 0.4 g/kg/day. Five plasma exchange sessions are recommended every other day. Trials of corticosteroid administration have not shown efficacy.4

Corticosteroids remain the cornerstone of treatment for PM. Conventional immunosuppressants are combined with steroids in severe forms.11

Rituximab is an effective treatment alternative for severe and life-threatening or refractory forms of both diseases11,12

The simultaneous occurrence of GBS and acute myositis has been rarely reported in the literature in the context of acute viral myositis caused by Mycoplasma pneumoniae, Cytomegalovirus, and dengue virus.13,14,15

However, the association of GBS and PM has been described only once in the literature. It leads to a confusing and intricate clinical presentation in which myogenic and predominantly motor neurogenic damage are superimposed. A meticulous clinical examination from the onset and paraclinical explorations guided by the clinical data are necessary to achieve a precise diagnosis and to initiate an adequate therapy at an early stage, given the diagnostic
and therapeutic emergency that each entity presents and the specificity of the management of each one.

4 | CONCLUSION

The clinical presentation of GBS is very variable, which makes its diagnosis delicate. The originality of this case lies in the exceptional association of two rare autoimmune diseases, GBS and polymyositis. The synchronous occurrence of the two diseases and the similarities between their clinical features make the diagnosis difficult and encourage a rigorous search for associations in any patient with a dysimmune disease, especially in the absence of improvement under adequate treatment.

The good clinico-biological evolution under plasmapheresis, corticosteroid therapy, methotrexate, and Rituximab has consolidated the diagnosis.

AUTHOR CONTRIBUTIONS

Imen Chabchoub: Conceptualization; investigation; writing – original draft. Mouna Snoussi: Methodology; project administration. Rania Ammar: Funding acquisition; investigation. Raida Ben Salah: Methodology. Chifa Dammak: Resources; supervision. Faten Frikha: Supervision; validation. Zouhir Bahloul: Project administration; supervision; validation.

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CONFLICT OF INTEREST

The authors declare that there are no conflicts of interest.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal’s patient consent policy.

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