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RESEARCH ARTICLE

THE ASSOCIATION OF MYASTHENIA AND GOUGEROT-SJOGREN SYNDROME

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ABSTRACT

The association of Grave disease of with the syndrome of Gougerot remains rare. The purpose of the presentation of this observation is to make it a meta-analysis through the literature and to work towards an early treatment. We are reporting a case of Myasthenia associated with a syndrome of Gougerot-Sjögren in a 32-year-old patient that were lately diagnosed. The diagnosis was retained in front of the myogenic syndrome with muscular deficit, a left (awkward) unilateral ptosis on the left side, a poly arthralgia and a Sicca oculo-buccal syndrome. These signs were associated with a positivity of antibodies. This association seems exceptional. A Good therapeutic result was first obtained with the triple prostigmine-prednisolone-azatioprin association.

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INTRODUCTION

Myasthenia is an auto immune neuromuscular disease caused by anti-bodies directed against receptors to the acetylcholine at neuromuscular junction (Eymard, 2014). Myasthenia can be associated with other auto-immune affections. Its association with Gougerot Syndrome is not exceptional but has been very rarely described in the literature (Dourov *et al*., 1968; Sellami *et al*., 2008; Tsai *et al*., 2013). We are reporting a new case of auto-immune myasthenia associated with Gougerot syndrome.

Observation

A 32-year old patient has presented since April 2013 muscular weakness associated with an abnormal fatigability to effort leading to several consultations during crises that can last for one to three months. In March 2015, the signs have exacerbated. She has been admitted in the internal medicine Unit at the University Hospital Centre of Le Dantec (Dakar). The questioning disclosed a fatigability to effort preventing some activities like walking, household chores, climbing stairs, by relieved when resting. This symptomatology was often associated with mastication and deglutition disorders.

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The exam emphasized the following:

- A myogenic syndrome with a muscular deficit valued at 3/5 right lower limb and 4/5 at the left lower limb
- A left unilateral ptosis
- A high cervical dysphagia with mastication and deglutition disorders associated with pulmonary aspirations.
- An inflammatory looking polyarthralgia directed towards the knees, shoulders, and the chronic non-deforming bilateral symmetric ankles
- A dry oculo-buccal syndrome

The exam did not find any skin or muscular signs. Any sign of dysthyroid or goiter was noticed. No history of autoimmune disease was found in the family. The anti-receiver Ac of the acetylcholine were higher than 24.8 nmol (Normal <0.40 nmol/L), no signs of dysthyroid, or goiter. The questioning did not reveal any auto immune history disease in the family. The anti-receiver Ac of the acetylcholine were positive at 24.8 nmol (Normal <0.40 nmol/L). The electromyogram showed an electrical table compatible with a myasthenia. The thoracic scanner came back to normal and did not show any thymoma. The autoimmune myasthenia diagnosis was retained over the myogenic syndrome association, enophtalmia and dysphagia associated with antibody positivity and the evocative aspect of the electromyogram. Additionally, she was presenting a poly arthralgia and a Sicca syndrome in favor of a Gougerot-Sjögren syndrome confirmed by the Ac and positive anti SSa and SSb

and the biopsy of the salivary glands showing a Stage III of Chisholm. A research of other autoimmune diseases associated was conducted. The TSHus was normal after researching a dysthyroid as well as the anti-nuclear Ac, native anti DNA and anti ECT researching connectivity concluded negative. The rhymatoid factors were also negative. The hemogram was normal, there were no sign of microcytic anemia that could allow thinking about a Biermer disease, as well as the muscular enzymes while researching an inflammatory myopathy. The patient was put under prostigmine with a 180 mg/day associated with a corticotherapy: Prednisolone at 1mg/kg for 2 weeks and azatioprine 100 mg/day for 6 months. The corticotherapy was stopped gradually after 3 months. The prostigmine associated with azathioprine has been extended. The evolution was favourable during the first quarter with an improvement of clinic signs, but followed by a recurrence. Finally, the patient is still under prostigmine and is being regularly monitored every three months.

DISCUSSION

The myasthenia is an affection of the autoimmune neuromuscular junction characterized by the presence of antiantibodies anti-receivers of acetylcholine (El Midaoui *et al*., 2010). We are reporting a case of myasthenia associated with a Gougerot syndrome. The diagnosis of these two illnesses was exposed before the clinical criteria (myogenic syndrome, ptosis, dysphagia, chronic polyarthritis and Sicca syndrome), immunology (positivity of antibodies anti receivers of choline acetyl and antibodies anti Ssa and SSb) electrical (electromyogram) and anatomo-pathological (stage III of Chisholm). The association myasthenia/other autoimmune disease is not exceptional. The illnesses are varied (Eymard, 2014). On a same patient, many autoimmune illnesses can be associated with myasthenia, each evolving independently. The associations that are the most described are those with dysthyroids (Basedow disease, thyroid) on 5 to 10% of patients. Other pathologies can be encountered during the myasthenia: rheumatoid arthritis, systemic lupus erythematosus, Biermer anemia, Gougerot-Sjogren syndrome, inflammatory myopathy, neutropenia and thrombopenia (Eymard, 2014). The Gougerot myasthenia syndrome association is a clinical entity rarely described in the literature. To our knowledge only three cases of myasthenia gravis associated with Sjogren's syndrome have been described. Indeed, Durov *et al*. (1968) in France had described myasthenia gravis associated with Sjogren and malignant thymoma in 1968, in 2008 Sellami *et al.* (2008) described another case associated with spondylitis and Sjogren and a third case was described by Tsai (2013) associating thymoma and myasthenia Gougerot Sjogren. We report a new rare case of association of myasthenia and Sjogren's syndrome. The explanation for the simultaneous presence of both diseases in the same individual is not yet clear. A genetic predisposition influences most likely the occurrence of the disease (Berrih-Aknin and Le-Panse, 2014). The analysis of the literature could allow to consider a common pathogenetic link. There is a genetic predisposing to the single or multiple occurrence of immune dysfunction diseases. (Dieudé, 2007) This is probably not the case of our patient; no history of autoimmune disease was found in the patient's immediate family. The most frequently described association is the one with thyroid dysfunction. Some cases with thyroid dysfunction have been reported in the literature in Senegal by Pouye *et al.* (2014), in the Maghreb countries by Trabelsi *et al.* (2006) in Côte d'Ivoire by Kouame (2009) and by Fantin (2010). This frequent association led to the research of clinical signs of thyroid dysfunction on the patient that did not exist and the physical examination was normal, and the TSHus done was normal. Myasthenia being an autoimmune disease, other autoimmune diseases that could be associated although rare, such as lupus, rheumatoid arthritis, Biermer disease, scleroderma and inflammatory myopathies were sought. In our patient none of these conditions were found. The clinical examination did not find any signs for these diseases. Nuclear antibodies, anti-native DNA, anti ECT and rheumatoid factors were negative. The blood count as well as muscle enzymes were normal. Myasthenia illustrates the involvement of the thymus in autoimmune diseases because of its frequent association with thymic abnormalities and the important role of the thymus in its pathogenesis (Puissant, 2004). Indeed, association with thymoma - Gougerot - myasthenia has been described in the literature by Durov (1968) and Tsai (2013). A thymic abnormality was investigated in our patient; the scan did not find any thymoma although the simultaneous presence is often described.

Diagnosis is often difficult in the early stages of the disease; everything will depend on what was attained in the foreground (2014). In fact there has been a misdiagnosis for two years in our patient; a diagnosis was put together when signs became richer. The evolution is capricious, interspersed with outbreaks that can be life-threatening when reaching respiratory muscles (El Midaoui *et al*., 2010). The anticholinesterase constitutes the basis for the symptomatic treatment of the disease in our patient treated with prostigmine, which allowed early improvements of the signs. The azathioprine (AZA) improves 70-90 % of cases of patients affected by myasthenia (El Midaoui *et al*., 2010). In addition to the anticholinesterase treatment, our patient was put under azathioprine at an oral dose of 100mg / day for 8 months. Combining this with immunosuppressive anticholinesterases has allowed for a clear regression of the signs for up to six months. The evolution was marked by the resumption of signs, which is consistent with the literature.

Conclusion

The diagnosis of myasthenia is not easy especially if the signs are not complete. Its diagnosis requires systematic research with other autoimmune diseases. Its association with the Gougerot syndrome is rare. The treatment is not always easy due to the evolution of the myasthenia. Monitoring should be regular to improve the quality of life of patients.

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