Medicina de Familia
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CLINICAL LETTER

COVID-19 and myotonic dystrophy type 1: Case report

COVID-19 y distrofia miotónica tipo 1: caso clínico

Myotonic muscular dystrophy type 1 or Steinert disease (SD) [ORPHA 273] is an autosomal dominant myopathy of low prevalence (<5/10,000) with almost complete penetrance and multi-organ involvement (neurological, cardiac, respiratory, endocrine and digestive). It is the most common form of muscular dystrophy in adults.[1]

A 55-year-old woman presenting, since March 24, 2020, with malaise, anorexia, increased habitual hypersomnia and thermometric evening fever with labored and hurried nocturnal breathing. She is referred to the hospital emergency department on March 27, 2020 by her primary care physician.

Her past medical history includes the diagnosis of SD at 33 years of age. Hypothyroidism diagnosed in 1996 with progressive thyroid failure. Hyperprolactinemia of unknown etiology without morphological image on MRI (1997) treated for several years with bromocriptine and later cabergoline. She has been wearing an anti-equin orthosis due to repeated falls since 2004. Phakectomy of both eyes in 2002 and 2005. Grade 1 AVB with implantation of bicameral pacemaker (PM) with response sensor (DDDR) in 2018. Moderate obstructive sleep apnoea (AHI 28.2) diagnosed in 2012, CPAP indicated with zero tolerance. Oxygen therapy with nasal prongs (NP) at 2 l/min at night. On chronic treatment with levothyroxine 100 mcg/24 h five days a week and levothyroxine 75 mcg/24 h two days a week; calcifediol 0.266 mg one dose/month and omeprazol 20 mg/24 h.

On arrival at the emergency room, the patient's vital signs were BP 93/68 mm Hg, HR 71 bpm and O_2 Sat 94%. She was afebrile, normal color and eupneic. Pulmonary auscultation showed preserved vesicular murmur and good bilateral ventilation without added pathological noises. There were no other relevant findings in the physical examination. Radiologically, peripheral ground glass opacities were identified in the right upper and lower right and lower left fields, with diffuse interstitial pattern. Laboratory parameters and chest X-rays imaging on admission to the emergency department are shown in Table 1 and Fig. 1. Given the findings suggestive of COVID-19 infection, it was decided to admit the patient to internal medicine under isolation conditions.

Table 1 Analytical parameters at admission.

Full blood count	
White blood cells (3.5–11 × 10^3 μl)	6.02
Hemoglobin (12–15 g/dl)	13.7
Lymphocites (20–45%)	17.8
Platelets (150–450 × 10^3/μl)	122
Chemistry	
CRP (<0.5 mg/dl)	11.79
Ferritin (13–150 ng/ml)	529
LDH (<250 U/l)	452
D-dimers (68–494 μg/l)	724
Fibrinogen (200–400 mg/dl)	602
Creatinine (0.51–0.95 mg/dl)	0.75
AST (<32 UI/l)	38
ALT (<35 UI/l)	32

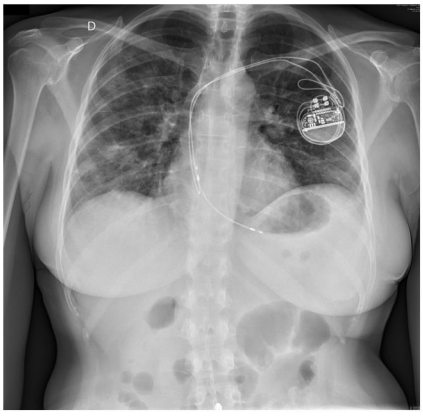

Figure 1 Chest X-rays imaging at admission.

The patient was not admitted to the intensive care unit (ICU). The patient was not considered candidate for ICU and invasive ventilation, as by the judgment of the critical care team.

After ruling out other aetiologies (negative total HIV antibodies; negative pneumococcal and legionella antigenuria), bilateral SARS-CoV-2 pneumonia was suspected. Since no diagnostic test for SARS-CoV-2 was available due to the limitations of the time, clinical diagnosis was assumed. Considering the findings of the complementary tests, after obtaining the patient's verbal consent and taking into account the risk/benefit ratio, treatment was started for

https://doi.org/10.1016/j.semerg.2023.102127

5 days with hydroxychloroquine (HCQ), azithromycin and systemic glucocorticoids in a descending regimen (250 mg/24 h in rapid IV injection on day 1; 80 mg/24 h on days 2, 3 and 4 and 60 mg/24 h on day 5 according to current local protocol by the early moment of the pandemic management. He remained glucocorticoid-free for 48 h prior to discharge. In addition, cyclosporine 50–100 mg/day was also administered for 5 days.[2]

On April 1, 2020 the patient was discharged home after being afebrile for 48 h, well saturated and eupneic. The discharge diagnosis was bilateral pneumonia due to probable SARS-CoV-2.

Drug treatment was completed at home with HCQ 200 mg/24 h for 2 more days after discharge. The patient was given written recommendations to maintain home isolation for 14 days after discharge. Treatment was completed with 14 more days of oxygen therapy with NP at 2–3 l/min for as long as possible. On May 29, 2020, during radiological follow-up after discharge, positive IgG and negative IgM antibodies to SARS-CoV-2 were detected and the diagnosis of bilateral COVID-19 pneumonia was serologically confirmed.

Since then the patient has received four doses of SARS-CoV-2 mRNA vaccine on May 13, 2021 (BioNTech/Pfizer), November 17, 2021 (BioNTech/Pfizer), June 14, 2022 (BioNTech/Pfizer/Comirnaty), and November 9, 2022 (Pfizer/BioNTech). No adverse side effects attributable to this immunization have been identified.

Given that the main cause of mortality in patients with SD is respiratory infections[3] it has been suggested that SARS-CoV-2 infection during the COVID-19 pandemic may have been an additional risk factor for these patients. All three cases in a series of SD patients hospitalized for COVID-19 between April 1, 2020, and April 30, 2020, died of SARS-CoV-2 infection.[4] Two other cases of SD hospitalized with COVID-19 overcame respiratory infection.[5]

Several factors could favor an unfavorable evolution of COVID-19 infection in patients with SD, such as the myotoxicity of the association of drugs such as chloroquine (CQ) and azithromycin, used empirically at the beginning of the pandemic, the immune alteration inherent to SD or the concomitance of this disease with other entities.[6]

On the other hand, SARS-CoV-2 can produce not only pneumonia with respiratory and flu-like symptoms, but can also affect other systems, with cardiological, dermatological, autoimmune, psychiatric, olfactory and taste alterations, as well as neurological manifestations.[7] To date, no sequelae attributable to COVID-19 have been identified in the reference patient.

The episode described above should be interpreted within the chronology of its healthcare context. As shown by the dates indicated, it occurred at the beginning of the first pandemic wave of COVID-19 in Spain. At that time there was a lack of clinical experience to support the efficacy of certain pharmacological treatments, so molecules such as CQ, HCQ or azithromycin were used empirically, the limited usefulness of which has subsequently been demonstrated. However, as long as no studies were available to justify the efficacy of HCQ, its use – as in the case described – was adjusted to the knowledge available at the time.[8] For this reason, the patient's consent was requested as it was an indication not contemplated in the technical data sheet. At that time, there was also no evidence of the usefulness of thromboprophylaxis.[9]

Due to the small number of described cases of COVID-19 infection in patients with SD, it seems necessary to analyze larger sample size in order to establish that this neuromuscular disease is a specific risk factor in the prognosis of SARS-CoV-2 infection.

The protocols of the workplace on the publication of patient data have been followed and the privacy of the subject has been respected.

Informed consent

The patient's consent has been obtained.

Funding

This work has not received funding.

Conflict of interest

The author declares no conflict of interest.

Acknowledgments

To Dr. Gloria Pindao Quesada, Internal Medicine, Hospital Universitario General de Villalba, Madrid. To Dr. Javier Sayas Catalán, Mechanical Ventilation Unit, ALS Multidisciplinary Unit, Hospital Universitario 12 de Octubre, Madrid.

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A. Rosado-Bartolomé

Medicina Familiar y Comunitaria, Grupo de Trabajo de Neurología de SEMERGEN, Sociedad Española de Médicos de Atención Primaria SEMERGEN, C/ Jorge Juan 66, 28009 Madrid, Spain
E-mail address: alfredorosado76@gmail.com