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SYMPTOMATIC AND RECURRENT PERICARDIAL EFFUSION IN SYSTEMIC SCLEROSIS, THE TREATMENT AS A CHALLENGE

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BACKGROUND

Pericardial involvement in systemic sclerosis (SSc) is generally mild and asymptomatic, in symptomatic patients it varies from 7 to 20% of cases. Pericardial effusion represents 15–43% of echocardiographic findings and 78% in autopsy studies of SSc patients.

CASE REPORT

Female, 72 years old, diagnosed in October 2019 with limited cutaneous SSc (lcSSc) with pulmonary (nonspecific interstitial pneumonia – NSIP) and cardiac (pericardial effusion) involvement. She started treatment with cyclophosphamide (CP) 500 mg/m² in December 2019. The patient was regular follow-up, with clinical and laboratory improvement, but in March 2020 she opted to suspend treatment for fear of contamination by coronavirus disease 2019 (COVID-19). In May, she complained of progressive dyspnea, chest pain and fatigue. At clinical examination, hypophonic sounds, jugular turgency, slight edema of the lower limbs were noted. Doctors decided for repetition of the transthoracic echocardiogram (TTE), that revealed a worsen of pericardial effusion, with imminent risk of cardiac tamponade, associated with an increase in CRP 140 mg/L and ESR 80 mm/h. Doctors opted in conjunction with cardiologists by pericardial drainage, that removed 700 mL of liquid. At first, the patient showed clinical improvement, but 4 days after, her symptoms increased. After carrying out a new TTE, recurrence of significant pericardial effusion was found, and a new pericardial drainage was performed with removal of 300 mL. The patient was negative infectious screening, pericardial fluid inflammatory (597 cells/mm³, 95% lymphocytes, proteins 4.9 g/dL, glucose 90 mg/dL, LDH 377 UL), negative tuberculosis test and negative culture. Even after drainage, the patient had cardiac activity with recurrent pericardial effusion, then doctors opted for introduction of prednisone 20 mg/day, colchicine 1 mg/day and pulse therapy with CP dose 600 mg/m². The patient keeps on outpatient follow-up basis, receiving monthly pulse therapy with CP 733 mg/m² and weaning from prednisone. Follow-up in conjunction with cardiology to the last TTE, maintaining moderate to significant pericardial effusion, but without clinical symptoms.

CONCLUSION

We reported a case of massive pericardial effusion with signs of cardiac tamponade, that evolved with recurrence after pericardial drainage, but it was stabilized after reintroduction of immunosuppressive therapy. In our clinical practice, the challenge in drug versus surgical management in face of massive and symptomatic pericardial effusions associated with SSc remains. In this case, we highlight the importance of immunosuppressive therapy to stabilize the patient, as well as the presence of pericardial effusion in SSc is well tolerated by patients with this diagnosis, unlike other etiologies of pericardial effusion.