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PULMONARY HYPERTENSION AS FIRST MANIFESTATION OF AUTOIMMUNE DISEASE: 2 CASE REPORTS

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BACKGROUND

Pulmonary arterial hypertension (PAH) is defined as a resting mean pulmonary arterial pressure ≥ 25 mmHg or 30 mmHg with exercise. It is complication of autoimmune diseases and is becoming a major cause of morbidity and mortality. To confirm the diagnosis it is recommended to perform a right heart catheterization. The annual incidence ranges from 5 to 15 cases per million, when associated with systemic autoimmune diseases. Concerning the therapeutic approach, it is important to treat the underlying disease and focus on a better stratification of the PAH.

CASE REPORT

CASE 1: A 22-year-old woman presented with exercise-induced shortness of breath, syncope, dry cough, sudden-onset tachycardia, constrictive chest pain and peripheral swelling. The cardiac investigation presented with a negative stress test for myocardial ischemia and also a negative CAT scan for PE, although the transthoracic echocardiogram demonstrated a severe PAH and pericardial effusion. Laboratory tests showed negative inflammatory markers and autoantibodies, however a positive ANA with 1: 320 homogenous nuclear pattern with reagents nucleus and metaphase plate. Fluid restriction was initiated (up to 1000 ml per day), anticoagulation (1 mg/kg twice-daily Enoxaparin), methylprednisolone pulse therapy followed by oral corticosteroids, Azathioprine 2 mg/kg/day, Hydroxychloroquine 400 mg/day and Sildenafil 60 mg/day. Initially the PASP estimated by transthoracic echocardiogram (TTE) was 120 mmHg (severe PAH), which was confirmed by heart catheterization as 113 mmHg with a transpulmonary gradient of 45 mmHg. After 3 months of medication it settles down to 65 mmHg.

CASE 2: A 37-year-old man presented activity-related dyspnea and unilateral painful swollen right leg. Doppler ultrasound showed extensive deep vein thrombosis in the femoropopliteal segment with a PASP of 35 mmHg in the TTE. Laboratory tests revealed ESR 58, positive ANA with 1: 160 thin dotted nuclear pattern with reagents nucleus and metaphase plate, positive anticardiolipin IgG positive (150) and reactive lupus anticoagulant. Treatment included anticoagulation (1 mg/kg twice-daily Enoxaparin), followed by Warfarin and Hydroxychloroquine 400 mg/day.

CONCLUSION

The association of PAH with systemic autoimmune diseases worsens the prognosis of the disease, increasing the mortality rate. It is the third most common cause of death in systemic lupus erythematosus. Multiple etiological factors are probably involved, such as chronic recurrent thromboembolic phenomena and pulmonary thrombosis due to the presence of antiphospholipid antibodies and possible endothelial dysfunction with abnormal vascular response.