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TAKAYASU ARTERITIS AND NON-AUTOIMMUNE HEMOLYTIC ANEMIA - CASE REPORT.

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BACKGROUND

Takayasu arteritis is an autoimmune granulomatous vasculitis that affects the aorta, its major branches, and pulmonary arteries. It may evolve with chronic disease anemia. Hemolytic anemia, which is characterized by increased reticulocytes, lactic dehydrogenase and indirect bilirubin, and reduced haptoglobin, may have an immune mechanism or not. Situations that lead to blood whirlwind within the vessels, with increased shear stress, may cause destruction of the red blood cells, due to mechanical trauma. The authors present a case of Takayasu arteritis associated with hemolytic anemia of probable mechanical cause.

CASE REPORT

A 38-year-old female patient, admitted on 08/20/2018 complaining of limb claudication, headache, difficult to control arterial hypertension and anemia, beginning 2 months early. On physical examination she was pale (2 + / 4 +); icteric (1 + / 4 +); with asymmetry of blood pressure in the upper limbs (BP in right arm = 220x60 mmHg, BP in left arm = 60x40 mmHg); systolic heart murmur irradiating to sternal furcula and carotids; trophic alterations in left arm; and diminished peripheral pulses. Chest angiotomography revealed diffuse atheromatosis of the thoracic aorta, specially in the descending aorta, with sequential luminal stenosis; hypoplasia of the left common carotid artery, with calcified plaques and mild to moderate stenosis; and hypoplastic left subclavian artery from the origin. Laboratory tests showed: hemolytic anemia, with negative direct COOMBS; ANA = fine dotted nuclear pattern (1:80); antiphospholipid antibody screening, anti-ENA, anti-DNA = non-reagent; hemoglobin electrophoresis = normal; G6PD deficiency = negative; karyotype = unchanged; immunophenotyping = no changes. Bone marrow biopsy revealed hypercellular bone marrow for age, with a relative increase of the erythrocyte series, without blasts. Search for nocturnal paroxysmal hemoglobinuria was negative. The patient was submitted to pulse therapy with methylprednisolone and maintenance therapy with prednisone, with no hematimetric levels improvement. Currently, she is in follow-up with cardiology and hematology.

CONCLUSION

Intravascular hemolysis is one of the main complications associated with cardiac valve prostheses. The turbulence of the blood flow creates a shear stress, destroying the red blood cells. Mechanism similar to that observed in valvular prostheses could explain the chronic hemolysis in this patient with Takayasu arteritis.