A RARE CASE OF COLD AGGLUTININ SYNDROME AS AN EXTRAGLANDULAR MANIFESTATION OF SJÖGREN'S SYNDROME

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BACKGROUND

Autoimmune hemolytic anemia (AIHA) in Sjögren's syndrome (SS) is uncommon. Usually, it is associated with warm antibodies. Cold agglutinin syndrome (CAS) is a rare autoimmune hemolytic anemia mediated by antibodies that bind to erythrocyte surface at low temperatures, causing agglutination and complement-mediated hemolysis. It can be primary (cold agglutinin disease) or secondary mainly to *Mycoplasma pneumoniae* pneumonia, Epstein-Barr (EBV) virus infection, or aggressive lymphoma. It is rare occurring in association with autoimmune diseases. We reported here a case of CAS in primary SS.

CASE REPORT

A 56-year-old woman was admitted with 6-month history of asthenia, pallor, jaundice, dyspnea on moderate exertion and recent episodes of syncope. Laboratory evaluation revealed hemoglobin, 6 g/dL (normocytic and normochromic erythrocytes); white blood cell count, 6,960/mm³ (neutrophils 4,350, lymphocytes 2,262); platelet count, 120,000/mm³; erythrocyte sedimentation rate, 104 mm/h, C-reactive protein, 0.7 mg/dL; normal liver tests; creatinine, 0.79 mg/dL and normal urine routine. Further evaluation revealed high indirect hyperbilirubinemia (2.14 mg/dL), high serum lactate dehydrogenase (683 UI/L); reticulocytosis (14,5%), low serum haptoglobin (1:320), rheumatoid factor (> 450 IU/mL) and anti-SSA/Ro (45 IU/mL); anti-SSB/La, anti-dsDNA and anti-Sm were negative. Salivary scintigraphy showed severe hypofunction of glands and labial salivary gland biopsy evidenced focal lymphocytic sialadenitis (focus score 3.3). Thus, primary SS presenting with CAS was diagnosed. Prednisone 1 mg/kg/day, hydroxychloroquine 5 mg/kg/day and azathioprine 2 mg/kg/day were introduced. She also received lubricating eye drops and artificial saliva to control dry symptoms.

CONCLUSION

There were few cases of AIHA related to SS described in the literature. Here we reported a still more uncommon situation in SS, which is cold agglutinin-mediated AIHA. Cold agglutinin syndrome is a rare condition that, when occurring secondary to overlying disease, is commonly associated to infection or aggressive lymphoproliferative disorder.

KEYWORDS

Autoimmune hemolytic anemia, Sjögren's syndrome, Warm antibodies, Cold agglutinin-mediated.