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EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS COMPLICATED WITH CHOLECYSTITIS AND HEPATITIS: A CASE REPORT.

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BACKGROUND

Eosinophilic granulomatosis with polyangiitis (EGPA), formerly known as Churg-Strauss syndrome, is a systemic vasculitis with peripheral eosinophilic infiltration of undetermined etiology. The American College of Rheumatology (ACR) proposed six diagnostic criteria, where the presence of four criteria are necessary to define EGPA: allergic asthma, transient pulmonary infiltrates, hypereosinophilia, neuropathy, paranasal sinus abnormalities and extravascular eosinophils in biopsy. However, some cases may manifest with atypical symptoms or uncommon complications, which make it difficult to establish the diagnosis of EGPA

CASE REPORT

We report the case of a 53-year-old man with a history of asthma and allergic rhinitis at the beginning of the study 2 years ago, initially admitted with abdominal pain clinically suggestive of acute cholecystitis. He presented with peripheral neuropathy and associated alveolar hemorrhage during hospitalization. Laboratory tests showed leukocytosis with sustained eosinophilia (highest count: 25% of leukocytes or in absolute count: 6154 eosinophils/mm3). Although CT scan of the abdomen suggested only acute calculous cholecystitis, histopathology of gallbladder evidenced calculous lithiasis cholecystitis associated with cholecystitis due to eosinophilic granulomatous vasculitis (Figure 1), as well as chronic liver disease with fibrosis and septal formation associated with eosinophilic granulomatous hepatitis (Figure 2). Antineutrophil cytoplasmic antibodies (ANCA) positive with mixed pattern (non MPO/non PR3). Electroneuromyography was compatible with multiple mononeuropathy of upper and lower limbs. Induction of remission was performed with pulse therapy with methylprednisolone 1 g per day for 3 days, followed by prednisone 1 mg/kg/day plus cyclophosphamide (CP) 750 mg/m2 of body surface, two first doses with a 15 day interval followed by 5 monthly pulses of cyclophosphamide. Azathioprine 2 mg/kg/day was used to maintain remission. After one year of illness, the corticosteroid was completely weaned off. Currently, the patient has no disease activity only in use of azathioprine, with lower limbs dysesthesias as sequelae. Patients with EGPA who present with cardiac, gastrointestinal or neurological impairment usually have a worse prognosis. In the absence of adequate therapy, the syndrome can lead to death. However, an effective treatment can lead to remission of symptoms in large number of patients.

CONCLUSION

EGPA is a rare vasculitis, difficult to diagnose, especially in the case of patients presenting with cholecystitis. Alerting to the possibility of this entity in patients with chronic asthma or chronic sinusitis can provide early treatment, lower morbidity and increase the chances of survival.