IGG4-RELATED DISEASE: A REPORT OF PERITONEAL FIBROSIS

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BACKGROUND

IgG4-related disease (RD) may be characterized by lesions with inflammatory reaction associated with fibrosis and lymphoplasmacytic infiltration with predominance of positive IgG4 tissue plasmocytes. The phenotypes of the disease were recently identified and divided into four groups, according to the involvement: “pancreato-hepatobiliary”, “retroperitoneal-aortic”, “limited to the head and neck” and “Mikulicz-systemic”. The patient in question can be classified as the “retroperitoneal-aortic” subtype. The findings of pathological anatomy characterize a dense lymphoplasmacytic infiltrate and the presence of fibrotic cells immersed in lymphoplasmacytic infiltrate. There may also be the presence of obliterating phlebitis. The manifestation of retroperitoneal fibrosis requires an urgent approach, as its non-control and progression can lead to irreversible damage to ureter, nerves, and renal damage. The first line of treatment is corticosteroid and the dose varies according to severity. The combination of immunosuppressants is recommended when it is not possible to reduce corticosteroids by still active disease. Recently, new approaches with the use of biological agents have emerged, and rituximab has proved interesting. In the patient in question, the clinical treatment with corticosteroids associated with methotrexate had a great response, with reduction of mass, clearance of urinary flow, leading to improvement of the renal condition and possibility of removal of double J catheter.

CASE REPORT

Male patient, 48 years old, hypertensive, evolved in 2018 with insufficiency acute renal function requiring “TSR” and implantation of a double J catheter. During investigation, ultrasound was performed that showed bilateral pyelocaliceal dilation and, in computerized tomographic of the abdomen, bilateral nephrolithiasis was visualized and the presence of a retroperitoneal mass was performed. In mass biopsy, dense fibroconjunctive tissue with a predominance of plasmocytes was evidenced. In immunohistochemistry, myofibroblastic proliferation without atypia and absence of malignancy criteria was identified. Due to retroperitoneal fibrosis and anatomopathological report, the hypothesis of IgG4 was suggested. Corticosteroid therapy was initiated associated with methotrexate, folic acid and vitamin D. With treatment, the patient’s creatinine improved significantly and there was regression of periureteral mass. Currently it is with controlled creatinine levels, preserved renal function and without the use of double J.

CONCLUSION

The IgG4-RD is a complex and difficult-to-manage entity. Among its various manifestations, retroperitoneal fibrosis can lead to severe and irreversible sequelae. Several approaches have been proposed. The patient in question obtained a significant improvement in the condition with drug therapy.

KEYWORDS

Inflammatory disease, Hyper IgG4, Retroperitoneal mass.