Immunoglobulin G4 related mastitis: A case report

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ABSTRACT

Immunoglobulin (Ig)G4-related sclerosing disease is a recently recognized condition characterized by mass forming lesions associated with storiform fibrosis, obliterate phlebitis, lymphoplasmacytic infiltrate rich in IgG4 positive plasma cells and elevated serum IgG4 levels. IgG4-related mastitis (IgG4-RM) is exceedingly rare with only thirteen cases reported in the literature to date. Immunoglobulin G4-RM is diagnosed exclusively on histological analysis. It is a benign chronic inflammatory process that can be treated sufficiently with excision or steroid. However, conservative treatment should be preferred and unnecessary surgery should be avoided as IgG4-RM respond to simple and effective steroid treatment. Herein, we presented a 28-year-old patient with IgG4-RM. She was the youngest patient in the literature at the time of her diagnosis.

Keywords: Mastitis, IgG4, Steroid therapy, Inflammatory pseudotumour

INTRODUCTION

Immunoglobulin (Ig)G4-related sclerosing disease, first described in the pancreas is now recognized as a systemic entity that can involve the liver, salivary gland, lymph nodes, mesentery, breast, and others [1-3]. The milestone of this disease is increased serum concentration of IgG4 levels and tumor-like swelling of organs, infiltration of lymphocytes that are enriched in IgG4-positive plasma cells, and storiform-patterned fibrosis. Differentiating this process from malignancy clinically can be a challenge as the lesions often present with suspicious radiographic features. It is important to recognize this entity as patients often respond well to steroid therapy and can avoid unnecessary surgery. To the best of our knowledge, the IgG4-related sclerosing disease of the breast is exceedingly rare, with 13 cases reported in the literature [4]. The well-known treatment of IgG4-related mastitis (RM) is glucocorticoid therapy. However, most cases undergo excision. Herein, we presented a 28-year-old patient with IgG4-RM. She was the youngest patient in the literature.

CASE REPORT

A 28-year-old female applied to our clinic. She suffered from a mass in upper inner quadrant of the right breast with no pain. She has two kids and no additional family history. Her first pregnancy was when she was 21 years old. She never used oral contraceptives. The patient noticed the mass one month ago. Red fluffy painful lesions increased around the abdomen and face, simultaneously. The mass size was 3 cm and palpable with an irregular shape. Preoperative laboratory values were C-reactive protein 1.2 mg / dL, white blood cell 9.3 K / uL and neutrophil 6.0 K / uL.

Malignancy could not be ruled out by examination and radiological findings in our patient, therefore, excisional biopsy was performed. The specimen revealed that the lesion infiltrated the ducts around the areas of fibrosis in the breast tissue by invading the normal breast tissue in most areas. In addition, multinuclear giant cells, which eliminated lobular structures, intense lymphoplasmaciyd inflammation with histiocytes in situ with neutrophils and lymphoid follicle structures were observed. Immunohistochemical examination revealed multiple vascular structures in the area of inflammation with CD34. CK 5/6 (+) in natural-looking breast duct epithelial cells. CD68 showed histiocytes (+) but granuloma structures were not observed. However, CD20 and CD8 focal (+) and fewer lymphocytes; CD3, CD4 with common (+) lymphoid follicles around the ductus were detected. CD138 was detected (+) in plasma cells.
Dense lymphoplasmacytic infiltrate with storiform fibrosis
Case Report

Histopathological findings were consistent with IgG4-RM. Findings showed no malignancy.

When the IgG4-related mastitis was diagnosed the patient was consulted to the Department of Rheumatology in our hospital. Rheumatologists started methotrexate and steroid therapy. IgG4 value was detected as 44 mg/dL. two months after the surgery. 7.5 mg methotrexate was used weekly and folic acid was added to the therapy. Prednisolone loading dose 40 mg/day was initiated and gradually tapered to 5 mg/day maintenance dose. The control imaging after treatment was completely normal. Extensive whole-body imaging was not recommended unless the patient is symptomatic. The patient had no recurrence 2 years after diagnosis.

DISCUSSION

IgG4-related sclerosing disease of the breast is rare. However, diagnosing this disease is a key point. Clinical and radiological findings of this disease resemble those of a malignant tumor. Therefore, timely diagnosis and appropriate therapy can be effective in majority of patients. We would like to underline that unneeded surgical biopsy should be avoided and conservative therapy should be preferred.

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Conflict of interest

The authors declare that they have no competing interests.
REFERENCES


