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PHOTO QUIZ

A large lump in the left breast

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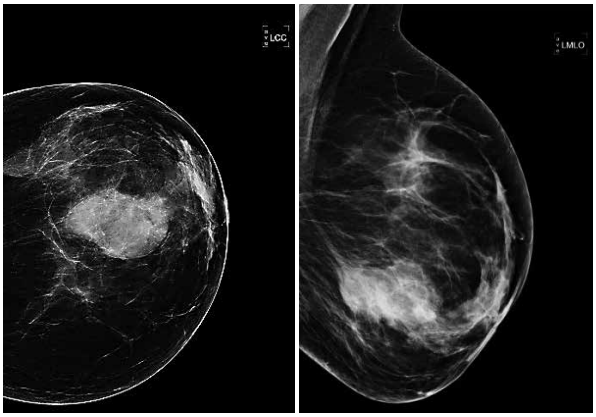
CASE REPORT

A 30-year-old woman with a history of traumatic tetraplegia was referred to our hospital with a palpable swelling of the left breast. On examination a painless swelling of the left breast and enlarged lymph nodes in the left axilla were observed. An ultrasound showed a solid inhomogenous lesion with a maximum diameter of 5.5 cm, highly suspicious of malignancy. A mammography showed similar findings (*figure 1*). A biopsy of the lesion showed no signs of malignancy, but a chronic granulomatous inflammation. Revision of biopsy material showed chronic fibrosing inflammation characterised by the presence of large amounts of histiocytes and plasma cells (*figure 2*).

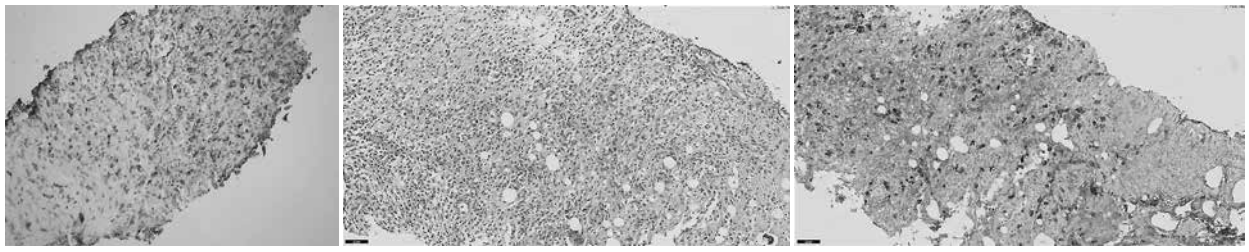
Figure 1. *Mammography shows a solid inhomogenous lesion suspicious of malignancy*



WHAT IS YOUR DIAGNOSIS?

See page 503 for the answer to this photo quiz.

Figure 2. *Biopsy of the lesion shows chronic fibrosing inflammation containing large amounts of histiocytes and plasma cells. Left: CD68 immunostaining. Middle: Haematoxylin and eosin (HE) staining; Right: IgG4 staining. 30% of the plasma cells stained positive for IgG4*



ANSWER TO PHOTO QUIZ (PAGE 500)

A LARGE LUMP IN THE LEFT BREAST

DIAGNOSIS/DISCUSSION

The patient was diagnosed with IgG4 related disease. Our differential diagnosis included a malignancy of the breast or Langerhans cell histiocytosis. There were no other signs of IgG4-related disease elsewhere.

At biopsy 30% of the plasma cells stained positive for IgG4, suggesting IgG4-related disease (IgG4-RD). Furthermore the CD68 immunostaining showed a reactive, partly histiocitic inflammatory infiltrate. Staining was negative for CD1a and S100; this excluded a Langerhans cell histiocytosis. In conclusion histology showed an infiltrate with lymphocytes, plasma cells and histiocytes. The diagnosis IgG4-related disease (IgG4-RD) was made. This was done in spite of the normal IgG4 serum levels.

IgG4-RD is a newly recognised condition of unknown aetiology, which is comprised of a collection of disorders that share specific pathological, serological and clinical features.[1] The incidence is about 2.63-10.1 patients per million people per year.[2] In 2001, autoimmune pancreatitis was related to infiltration with IgG4-positive plasma cells for the first time.[3] Furthermore, since 2003, IgG4-RD has been identified in multiple organ systems making this a systemic autoimmune condition.[1] IgG4-RD is characterised by dense lymphoplasmacytic infiltrates rich in IgG4-positive plasma cells, and tumour-like lesions with storiform fibrosis and destruction of the original architecture. Moreover, in more than 60% of the cases, serum IgG4 concentrations are elevated.[4] By now, IgG4-RD has been described in almost every organ system, most commonly in the pancreas and biliary tree. Regardless of the organ affected, the histopathological findings are similar.[1] The current therapy is treatment with high-dose prednisolone, based on data from several case reports.

Our patient refused treatment and a wait-and-see policy was followed. Follow-up for 18 months showed spontaneous regression of the palpable swelling in the left breast. This was confirmed by mammography.

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