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The association between a primitive Sjogren's syndrome and infiltrating ductal carcinoma: a case report

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Abstract: Sjogren's syndrome (SS) is an autoimmune disease characterized by lymphocytic infiltration of exocrine glands and non-glandular structures with a known risk of developing malignant lympho proliferative syndrome. In addition to this lymphoproliferation, patients with an SS can also develop other types and other cancer sites. We report a case of association between SS and infiltrating ductal carcinoma (IDC). The particularities of this association will be discussed through a literature review.
Keywords: Sjogren's syndrome- infiltrating ductal carcinoma - breast.

INTRODUCTION:

Sjogren's syndrome (SS) is a benign disease of slow growth with low morbidity and mortality, mainly characterized by specific autoimmune manifestations of organ and systemic manifestations which include cancers. We report a case of association between primary SS and infiltrating ductal carcinoma (IDC).

CASE REPORT:

A 63 years old woman, without specific medical history, who consults for mechanical back pain associated with nerve root pain in the right leg and a symptomatic dry eye and mouth. The general examination showed a patient in good general condition. Physical examination objectivated a lumbar spinal syndrome without radicular syndrome. The Laboratory tests showed an erythrocyte sedimentation rate at 11 mm, serum calcium to 2.13 mmol / l and a negative proteinuria. For his symptomatic dry eye and mouth, a biopsy of the salivary glands was performed with an objectivated Sialadenitis stage 3, a Schirmer test were also asked which returned positive. The patient has also benefitted from an electromyogram of the lower limbs in favor of sensorimotor axonal polyneuropathy. The patient was put under antimalarials with symptomatic treatment of dry eye and mouth with a good evolution.

Furthermore, during its follow a breast examination objectified the presence of a left breast nodule, a mammogram was requested showing a breast lesion of the left upper outer quadrant ACR5 complemented by an excisional biopsy with histological examination for an infiltrating ductal carcinoma measuring 3/2 cm SBR grade III with absence of vascular emboli. A tumor marker assays (CEA, CA15, 3) was also performed and returned negative. The association between primary SS and IDC of the left breast was established. The patiente was sent to Oncology department for medical care.

DISCUSSION:

SS is an autoimmune disease characterized by lymphocytic infiltration of glandular and extra-glandular structures. Patients with SS have a greater risk of developing a malignant lymphocyte proliferation; it is essentially non-Hodgkin lymphoma (NHL) low grade. This risk is significantly greater in the primary SS. Many histological types of NHL in patients with SS have been reported in the literature (centro folliculaire, plasmacytoid, diffuse large B cell). However, lymphoma marginal zone developed at the expense of mucosa of lymphoid tissues remain the histological type most commonly found, followed by lymphoma diffuse large B cells. Exocrine salivary glands are usually affected; other tissues may also be affected such as stomach, nasopharynx, skin, liver, kidney, lung and breast. Predictive factors of this degeneration are multiple: the presence of poly-lymphadenopathy, splenomegaly, neutropenia, and mixed cryoglobulinemia, a decrease of C3 fraction of complement, radiation or chemotherapy history [1, 2].

Besides lympho proliferative risk, other cancers are also possible in patients with SS, such as gynecological, gastrointestinal, renal, breast and skin cancer with an estimated global incidence to 1.42-2.6 times that found in the general population and RR = 0.67 [3-5]. For all breast cancers, the incidence in patients with SS is estimated at 0.46 [6]. Considering the incidence and RR of developing breast cancer. It remains very under-diagnosed or diagnosed at an advanced stage as was the case of carcinoma in situ diagnosed at the stage of lymph node metastases in a

patient with long history of SS [7] .For our patient, the diagnosis of breast cancer was made at an early stage, contrary to what the literature reports. According to a literature review, Lazarus and al reported 3 cases of breast cancer in a retrospective study of 112 patients with SS and including 25 cases of malignant disease [4]. A recent study by Zhang and al on 1320 patients with SS, 29 cases of malignancy were recorded including 4 cases of breast cancer [3]. These two studies did not put the point on the histological type or grade of these cancers.

It should also be recalled that the IDC is the histological type most frequently encountered in women with a frequency of 50 to 70%. It affects women whose average age is 56 years. The circumstances of discovery of this type of cancer are represented primarily by the discovery during the physical examination of a palpable breast mass or during a screening mammography. 30% of IDC is classified grade III SBR like our patient and is generally good prognosis [8,9].

CONCLUSION:

The association between SS and breast cancer is possible, but it still uncommon. This association should be seriously considered inciting to take stock in search of neoplasia. This also requires vigilance of clinician in monitoring of patients with SS.

CONFLICT OF INTEREST STATEMENT

The authors have no conflicts of interest

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