INTRODUCTION

Gigantomastia is a rare pathologic condition characterized by excessive breast development. There is no universally accepted definition of this disease. Many authors define gigantomastia as excessive breast tissue contributing to more than 3% of body weight, or that requires reduction of more than 1800 g of breast tissue. Such definitions are limited because the diagnosis of gigantomastia can be confirmed only after surgery, despite estimating the magnitude of the resection preoperatively. Netscher et al believe this pathology is better defined by the symptoms than the amount of tissue removed.

The pathogenesis of this condition is also not agreed upon. The two most widely accepted theories are hypersensitivity of breast receptors to normal levels of circulating hormones versus excessive hormone production. Various forms of gigantomastia have been described and include: gestational gigantomastia, in which breast enlargement occurs soon after pregnancy; juvenile gigantomastia, which occurs at puberty; idiopathic gigantomastia; and gigantomastia induced by drugs or associated with autoimmune diseases such as lupus erythematosus and rheumatoid arthritis.

The responsible drugs commonly described are penicillamine, bucillamine, neothetazone, and cyclosporine.

Another rare form of gigantomastia is the one caused by pseudoangiomatous stromal hyperplasia (PASH). PASH is a benign pathology of the breast due to collagen proliferation. Often diagnosed incidentally in breast biopsies, it rarely manifests clinically and no follow-up is necessary.

The treatment of gigantomastia is also controversial. Surgical resection is the usual treatment and options include reduction mammoplasty or mastectomy with reconstruction. The purpose of this article is to present a case of extreme and disabling gigantomastia caused by PASH treated at Hopital Universitaire de Mirebalais (HUM) in March 2021. This case is unique because of its association with severe morbidity considerably impacting the patient’s life.

CASE REPORT

We present a 29-year-old G0P0 woman without known medical history and a body mass index of 34.25 kg/m² (Fig. 1). Her story dates back 6 years when she noticed a marked growth in her breasts which had accelerated in the previous 2 years, associated with shoulder and back pain. As her breasts increased in size, her daily activities became more and more strenuous. Subsequently, she experienced constant pain in the breasts and nipple hyperesthesia and developed multiple skin ulcerations. Her breasts became so heavy that she finally became bedridden. Workup included a core needle biopsy which revealed an adenofibroma with a focus of fibroadenosis, a mammogram suggesting mammary dysplasia with cyst and fibroadenoma formation, and normal hormonal profile except for a high progesterone level (progesterone: 11.53 ng/ml).

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The patient was physically and emotionally distressed by the disease. Her thorax was asymmetrical with grade 3 breast ptosis. Her very enlarged and flattened nipple areola complex was located at the lower pole of the breasts, which reached below the anterosuperior iliac spine. Two ulcerations measuring 6 and 4 cm in diameter were present on the right breast. Palpation revealed two firm, 3-cm mobile and regular shaped masses in the right upper quadrant. After discussion and counseling on the possible failure of the free nipple areola grafts and loss of nipple sensation, bilateral breast reduction was planned.

Informed consent was obtained and she underwent bilateral reduction mammaplasty with free nipple graft under general anesthesia. The entire inferior pole of the breast was excised without pedicle and the breast mound was reconstructed with upper medial and lateral skin flaps. A JP suction drain was placed in each breast. The removed specimens weighed 8.6 and 7.3 kg, respectively, for the right and left breasts. The patient was not transfused during the operation, which lasted 3 hours.

Upon awakening, the patient described a complete resolution of neck, back, and breast pain. She was discharged the next day for follow-up in an outpatient clinic. The drains were removed on postoperative day 4. No complications occurred. The patient recorded a very significant improvement in her quality of life and remains very pleased with the aesthetic result (Fig. 2).

**DISCUSSION**

The first case of gigantomastia was described in 1648 by Palmuth. Although there is no universal definition of gigantomastia, our patient, with a massive 15.9 kg resection weighing 21.1% of her body weight, far exceeds all minimal criteria for gigantomastia. For comparison, the average weight of specimens after breast reduction is 1277 g in the United States. Our patient experienced the most severe symptoms associated with this pathology, including necrosis and ulceration of the breasts, and exceptionally for this pathology, inability to participate in daily activities.

The differential diagnosis of gigantomastia includes phyllodes tumor, giant fibroadenoma, fibrocystic disease, non-Hodgkin lymphoma, and lymphoblastic lymphoma. In our case, preoperative imaging and biopsy were unremarkable; thus, the preoperative diagnosis of idiopathic gigantomastia was made. The histopathological study of the resected specimen, however, revealed slit-like, anastomosing spaces in a dense collagenous stroma consistent with PASH (Fig. 3).

Surgery is the treatment of choice for gigantomastia. The two surgical options are subcutaneous mastectomy and breast reduction. The aim of surgical intervention is to improve the quality of life of patients from both symptomatic and aesthetic points of view. Several studies have demonstrated the effectiveness of breast reduction in improving the physical and psychological well-being of patients with gigantomastia. This procedure is associated with a very high rate of patient satisfaction. However, compared with mastectomy, the risk ratio of a recurrence after breast reduction is 7.0. For this reason, some
authors recommend mastectomy as a treatment for gigantomastia. But this treatment is too radical and the psychological sequelae of a mastectomy are too considerable for a benign pathology without completely eliminating the risk of recurrence.\textsuperscript{7,11} This is why we opted for breast reduction. The technical challenge of breast reduction is to result in adequate projection and fullness with adequate blood supply given the extreme degree of ptosis.\textsuperscript{12} Despite inclusion in the literature, we believe medical treatment for gigantomastia is not particularly effective in most forms of gigantomastia. However, prophylactic hormone therapy with tamoxifen may be useful to prevent recurrence.

CONCLUSIONS

Gigantomastia caused by PASH is rare. The treatment of choice for most forms of gigantomastia is free nipple graft breast reduction. Nevertheless, adequate counseling regarding the risk of recurrence is essential. Prophylactic hormone therapy should be considered.

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Fig. 3. Postoperative histopathological slides of resected specimen showing pseudoangiomatous stromal hyperplasia. A, Slit-like anastomosing spaces in a dense collagenous stroma. B, Myofibroblast that simulates endothelial cells.