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Case Report

Bilateral pseudoangiomatous stromal hyperplasia in childhood gigantomastia: A challenge in reconstruction and management

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ABSTRACT

The presence of mammary nodules during childhood and adolescence is somewhat unusual, generally consisting of benign lesions. Despite the range of possible diagnoses, they are generally similar in clinical terms. In the rare cases where these lesions constitute a case of gigantomastia, the differential diagnosis must be between juvenile fibroadenomatosis, phyllodes tumours and other even less common diseases of the mammary gland, such as Pseudoangiomatous Stromal Hyperplasia (PASH). This is caused by the exacerbated proliferation of mature fibroblasts and myofibroblasts, with the trigger factor still unknown. This study reports on a rare case of bilateral PASH in a young patient, describing its diagnosis and the surgical technique used.

An eleven-year-old patient with no previous comorbidities presented exacerbated breast growth during the six months immediately after the menarche, associated with mastalgia. She was referred to the Mastology Department at the HUCFF-UFRJ in November 2016 with enormous breasts, larger on the right and with multiple mobile masses of assorted sizes with a fibrous elastic texture throughout

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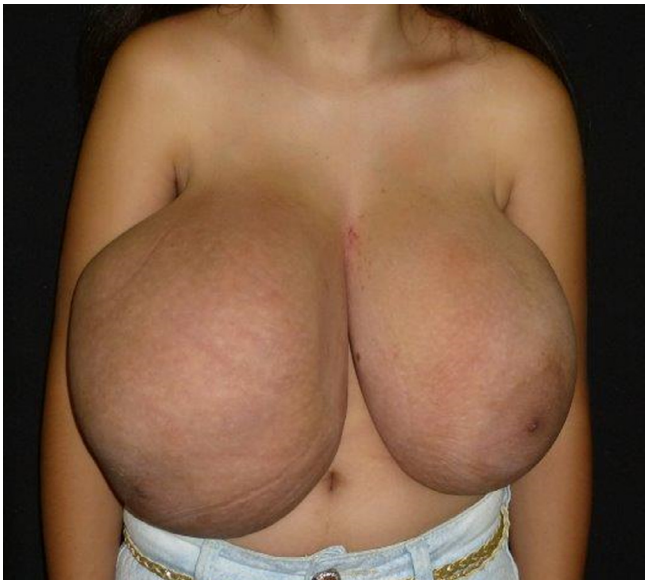

Figure 1. Pre-operative.

the entire mammary parenchyma. There were no signs of papillary discharge or lymphadenopathy (Figure 1). Unaware of any family history of neoplasia, she stated she was not taking any regular medications. The hormone profile was compatible with the normal levels for this age, and a breast ultrasound scan (USS) and Magnetic Resonance Imaging (MRI) presented findings compatible with fibroadenomatosis.

An ultrasound guided core biopsy was taken of the tumour with the largest diameter (12.3 cm in the right breast), with the histopathological report indicating fibroadenoma. On a joint assessment, an approach was proposed through which the Mastology Department would excise the lesions, followed by immediate reconstruction by the Plastic Surgery Department. This assessment foresaw difficulties in quantifying the amount of healthy breast tissue and lifting the Nipple Areolar Complex (NAC), due to severe ptosis. The pre-surgical planning was steered by the applicability of gigantomastia treatment techniques, deciding on an incision preserving the lower pedicles for access as well as for dissection and resection of the lesions. There was a concern to maintain the maximum possible dermal and glandular tissue in the inferior pedicle of the breast without separating it from the thoracic wall, in order to preserve perfusion and innervation since the dissection of the upper pole was more aggressive. In planning reconstruction, an inverted T marker was used, together with the incision marker for subsequent surplus skin excision (Figure 2). In the right breast, twelve tumorectomies were performed (2340 g) and thirteen in the left breast (860 g) (Figure 3). The patient required a three-unit blood transfusion during surgery. The breasts were reconstituted by lifting the lower pedicle to the upper pole, positioning it on the chest muscles, repositioning the NAC and adjusting the inverted T marker for excising surplus skin. Suction drains were placed bilaterally, followed by dressing with sterile pads and bandages.

During the first day post-surgery, there was no measurable bleeding and the drains were removed. Breast oedema was within the expected limits and the NAC showed no signs of ischemia. She was discharged from hospital on Day 3 after surgery, subsequent to **electrolyte** and volume replacement. During out-patient follow-up visits, she presented pallor of the right areole, but with satisfactory post-surgery progress, with relief of the pre-surgery mastalgia. At eight months post-operative there was no further increase in breast size and she did not want further surgery (Figure 4).

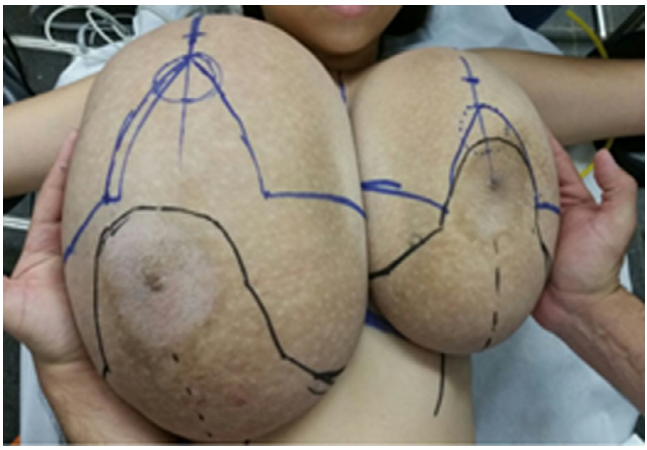

Figure 2. Planning surgery. In black, lower pedicle marker. In blue, forecast skin flaps for breast reconstruction (Inverted T). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

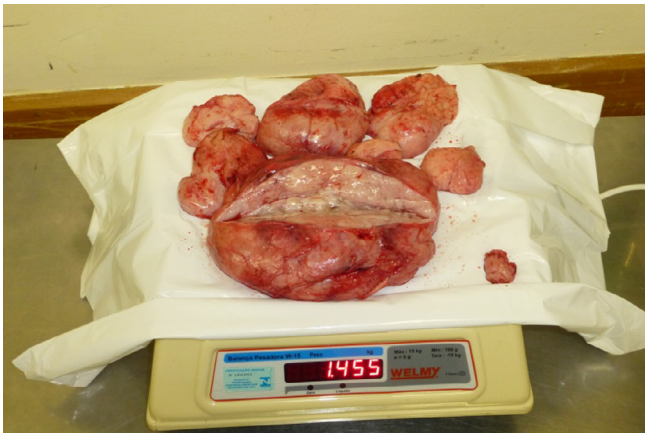

Figure 3. Nine out of twelve lesions excised from the right breast. Largest tumour with cross-section.

Discussion

The definitive histopathological diagnosis of the lesions was PASH with only one of the lesions identified as juvenile fibroadenoma after several pathology opinions and immunohistochemical study. The surgical treatment of pseudoangiomatous hyperplasia in its tumoral form and juvenile fibroadenomatosis is the same, based on complete tumour excision.[1–3] In this specific case, large breast volumes were a matter of concern, together with tumour size and diffuse involvement, together with preserving the viability of the nipple-areola complex. There is a shortage of data supporting the observation and non-surgical treatment of diagnosed cases, in terms of psychological aspects as well as the need to distinguish between other breast tumours with uncertain behaviour.[4,6,7] The patient is being followed up closely, in order to observe the biological behaviour of the lesion and to plan a larger intervention if needed.[8]

PASH is a finding that is generally incidental during histopathological evaluations of breast nodules; although benign, it may resemble vascular tumours, such as angiosarcomas.[3,5,9,10] This is due to its histological aspects which simulate vascular channels, as well as rapid lesion growth.[3] Despite the rapid progress of its tumoral form, there is no association between this disease and breast cancer[3].

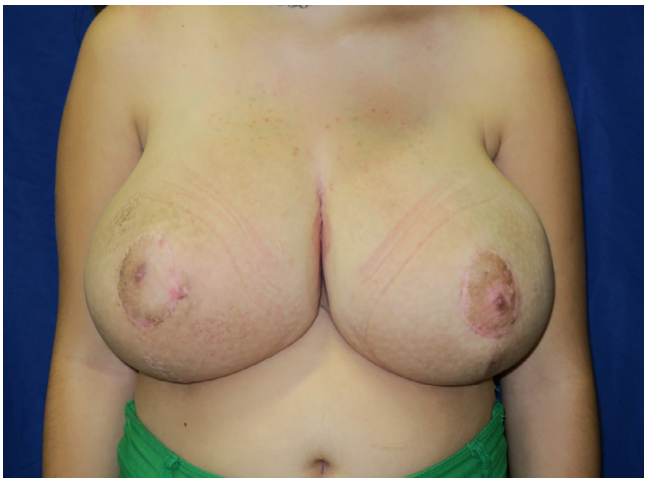

Figure 4. Six months post-operative.

Use of the lower pedicle flap proved an interesting treatment option in this case, as it was possible to lift the NAC and reconstruct the breasts. In this case, direct amputation associated with NAC grafts would be the fastest and simplest option,[11] but would result in definitive loss of sensation and inability to lactate. This case underscores the challenge of treating an uncommon mammary disease in a pubertal patient with the concern of minimising damage to developing breast tissue,[4,5] and the applicability of the versatile surgical technique that is normally used for idiopathic mammary hypertrophy. This consequently underscores the importance of mastering well-established mammoplasty techniques for handling rare and complex cases requiring breast reconstruction.

Conflict of interest

None.

Funding

None.

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