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MODESTUM

Case Report **OPEN ACCESS**

A rare case of gigantomastia caused by pseudoangiomatous stromal hyperplasia

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ABSTRACT

Gigantomastia is a rare pathologic condition characterized by an excessive and abnormal breast hypertrophy. To date, there is no exact definition or classification of this disease. Pathogenesis still unclear. The purpose of this article is to present a case of extreme and disabling gigantomastia caused by pseudoangiomatous stromal hyperplasia, one of the rarest etiology of gigantomastia.

Keywords: breast hypertrophy, gigantomastia, pseudoangiomatous stromal hyperplasia, reduction mammoplasty

INTRODUCTION

Gigantomastia is a rare and disabling condition characterized by excessive breast growth. Massive breast hypertrophy leads to physical and psychological discomfort, such as neck, back and breast pain, muscle strain, skin maceration and ulceration, emotional and psychological disturbances with significant impact incapacitating social life [1]. Gigantomastia may be associated with my benign breast condition, however association with pseudoadenotous stromal hyperplasia (PASH) is rarely reported [1, 2].

We present a case of extreme and disabling gigantomastia caused by PASH aiming to document the unusual presentation and delineate the challenges encountered in the management.

CASE PRESENTATION

A 31-year-old mother of two (ages five and nine years old) not known to have any comorbidities presented with bilateral extensive breast enlargement over the past two years. Patient gave history of regular menstrual cycle. Menarche at 13 years. She never breast fed her children. No similar family history of breast hypertrophy. The progressive enlargement of the breast has resulted in neck and back pain, inframammary excoriation, breast edema and skin ulceration. The progressive growth of both breasts caused debilitating hindrance for all basic social activities. No associated other symptoms. There was no history smoking or alcohol consumption.

On Examination

Body weight was 70 kg and height was 157 cm (BMI 28 kg/m^2). Breast patient walking with positional kyphosis. General examination was within normal limits. Local examination revealed extremely large pendulous breast, grade III ptosis (**Figure 1**). Sternal notch to nipple areola (SN:NAC) distance 44 cm for the right breast, 42 cm for left breast. Skin edema, distended veins, skin stria, and marked maceration beneath both breasts (**Figure 2**).

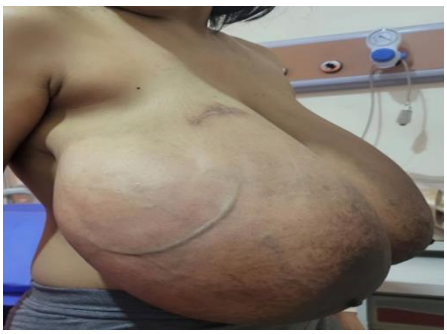

Figure 1. Preoperative appearance of bilateral macromastia with marked skin changes & dilated veins caused by PASH (reprinted with permission of the patient)

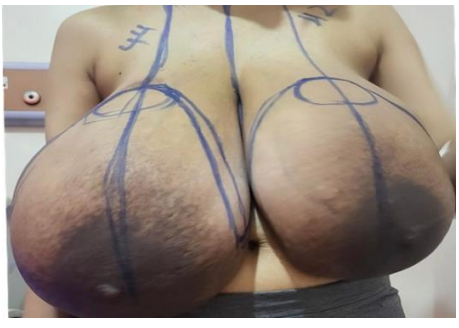

Figure 2. Preoperative marking showed asymmetry NAC:SN distance 44 cm, 42 cm for right & left breast, respectively (reprinted with permission of the patient)

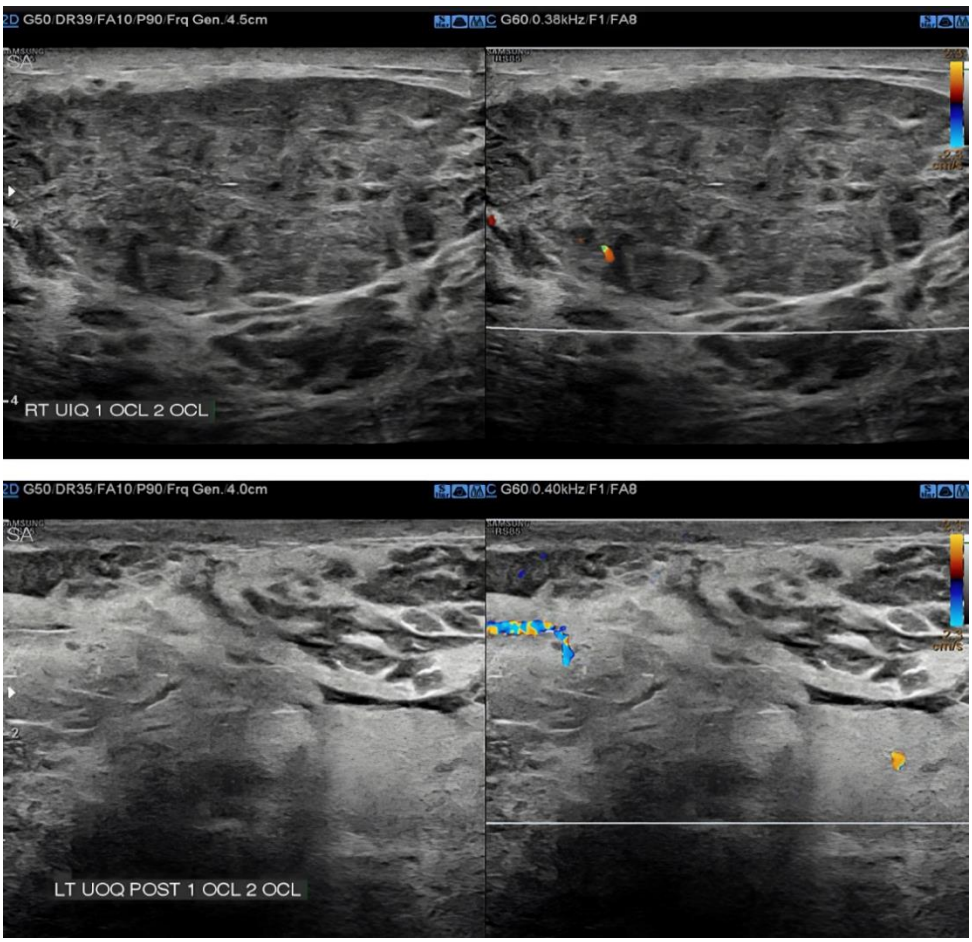

Figure 3. Selected grey scale and color doppler images both breasts (reprinted with permission of the patient)

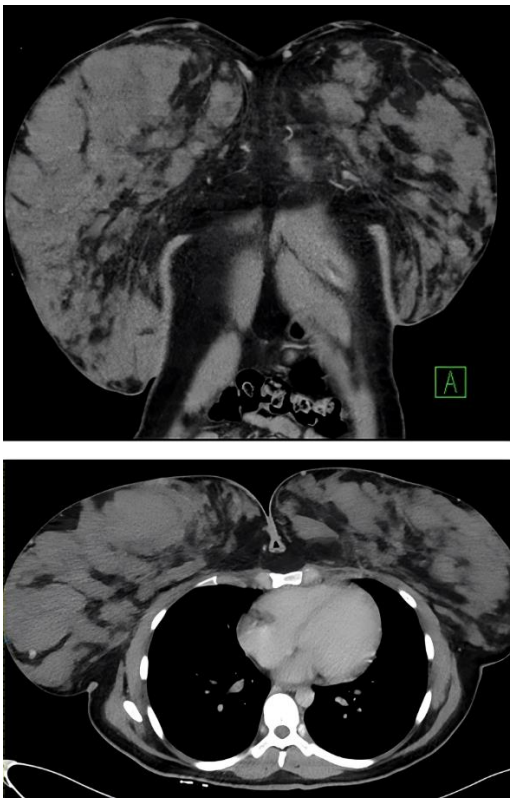

Figure 4. Axial and coronal images of contrast enhanced chest CT scan show bilateral breasts hypertrophy (reprinted with permission of the patient)

On palpation

Bilateral palpable non tender masses of variable sizes with multiple palpable axillary lymph nodes. Inframammary areas of both new and healed maceration.

Investigations

All biochemical parameters were within normal limits.

Ultrasonography

Right breast

Large well-defined heterogenous mass with peripheral internal vascularity.

Left breast

Area of echogenic parenchyma with linear serpiginous hypoechoic areas (**Figure 3**).

CT Scan With Contrast

CT scan showed bilateral breast hypertrophy with multiple innumerable soft tissue masses scattered in both breasts (**Figure 4**).

Axial gadolinium enhanced MR images of both breasts, show bilateral breasts hypertrophy with multiple innumerable enhancing round masses of variable sizes scattered in both breasts (**Figure 5**).

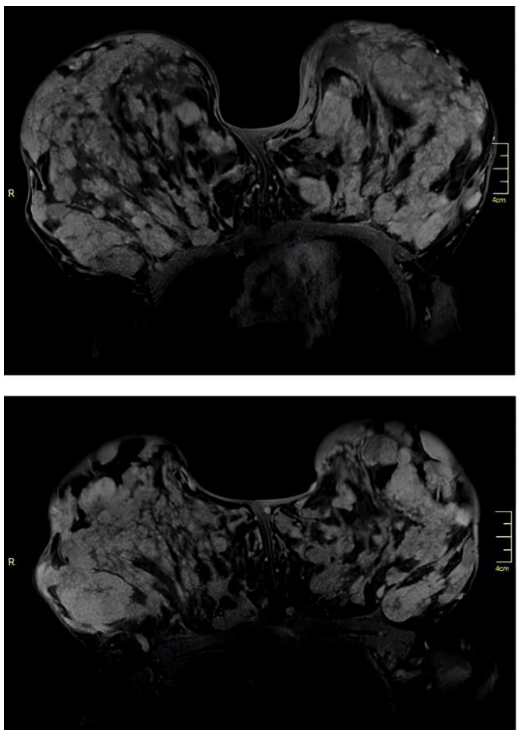

Figure 5. Axial gadolinium enhanced MR images of both breasts show bilateral breasts hypertrophy (reprinted with permission of the patient)

Core Needle Biopsy

Core needle biopsy was taken from multiple masses and was reported as PASH, fibro adenomatous changes, focal lymphocytic mastitis, focal fibrocystic changes.

Patient's case was discussed at the multidisciplinary meeting and the consensus was bilateral mastectomy versus reduction mammoplasty based on the intraoperative findings. The excised tissue weigh was 5 kg in total. 3.2 kg from the right breast and 2.8 kg from the left breast (**Figure 6**).

Reduction mammoplasty with free nipple graft was the most suitable for this case.

The patient had a smooth postoperative recovery and on regular follow up in outpatient clinic.

Two months post-surgery she showed acceptable healing with great patient satisfaction (**Figure 7**).

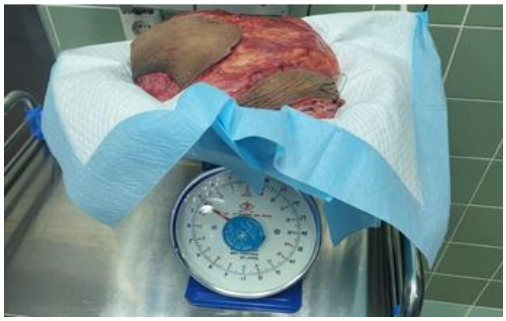

Figure 6. Right breast on the scale estimated weight 3.2 kg (reprinted with permission of the patient)

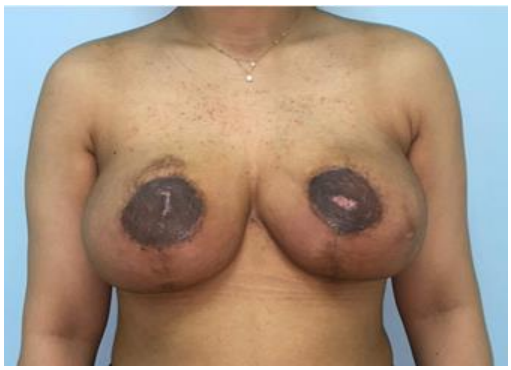

Figure 7. Post operative appearance two months back after surgical excision and reduction mammoplasty with healing partial nipple necrosis (reprinted with permission of the patient)

DISCUSSION

The term gigantomastia was first introduced by Palmuth in the German literature in 1648. To date there is no specific definition or classification of gigantomastia is reported, yet it is agreed upon that gigantomastia is considered as massive breast hypertrophy that requires a reduction of over 1,500 g per breast [1, 2] or excessive breast tissue contributing to more than 3% of body weight [3].

Attempted to classify gigantomastia as gestational, juvenile (virginal), idiopathic and drug induced gigantomastia have been employed [1]. There are numerous hypotheses for the pathophysiology of gigantomastia, such as hormonal hypersensitivity, excessive release of endocrinologic hormone (estrogen or prolactin), autoimmune diseases, such as myasthenia gravis, rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), or the use of pharmacologic agents, such as penicillamine or cyclosporine [2]. ERα and PR oversensitivity in women with gigantomastia is questionable as ERα and PR expressions in the glands of these women did not differ from the receptor expressions in control women [2].

The differential diagnosis of gigantomastia includes phyllodes tumor, giant fibroadenoma, fibrocystic disease, sarcoma and PASH disease [3, 4].

PASH is uncommon disease with few reported cases in the medical literature. It is a benign proliferative mesenchymal lesion of the breast found as an incidental of breast biopsies as a palpable or detectable mass in mammography. As demonstrated by Ibrahim et al. incidental microscopic PASH can be found in up to 23% of consecutive breast specimens [5]. The pathogenesis is unclear yet but most probably due to hormonal hypersensitivity and treatment modality are still controversial [2, 5]. Rarely PASH may present with extreme gigantomastia.

This reported case presented with unique clinical manifestations of bilateral multiple nodular disease on top of massive bilateral breast hypertrophy (gigantomastia). The clinical presentation influences PASH treatment. If PASH is an unexpected histologic discovery in a patient's specimens with other lesions, no additional special treatment is necessary. The recommended treatment for tumorous PASH is excision with a sufficient yet close margin. When diffuse PASH causes chronic suffering and pain or requires extensive removal of breast tissue, a mastectomy may be necessary. After excision, the recurrence rate ranges from 0% to 22% [5-7].

Gigantomastia treatments options are still controversial. Ranging from medical conservative hormonal therapy to

surgical reduction mammaplasty, and simple/subcutaneous mastectomy.

It was preferred surgery, especially in patients with large tumors and higher risks for breast malignancies while Raza et al. treated their patient conservatively [6].

But most authors conclude that gigantomastia can only be treated surgically. The aim of surgical intervention is to improve the quality of life of patients, improving the psychological well-being of patients and enhance quick alleviation of their symptoms. Reduction mammoplasty or mastectomy followed by breast reconstruction are viable options for management.

Inverted T scar pattern and superior-medical pedicle were chosen among the preferred surgical options [7].

Breast reduction is associated with high rate of patient satisfaction however, some surgeons prefer t mastectomy as a treatment option for gigantomastia. This radical treatment may be accompanied by psychological sequelae of a mastectomy as most patients within the reproductive age group. Gigantomastia is unlikely to recur after reduction surgery [8, 9].

The risk of recurrence higher in breast reduction surgery compared to prophylactic mastectomy, risk ratio 7.0 [3].

It was reported a case of recurrent PASH-caused gigantomastia in a 33-year-old female who had undergone bilateral breast reduction surgery four years back for gigantomastia [8]. Mastectomy was done and histopathology was suggestive of PASH [8].

Long-term follow-up observations are required to monitor for recurrences after surgery [10].

CONCLUSION

Gigantomastia caused by PASH is rare. Diagnosis of the underlying etiology my aid in success of the management. Surgical intervention is mandatory in the improvement of the affected quality of life.

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Ethical statement: The authors stated that written informed consent was obtained from the patient for publication of this case report and accompanying images.

Declaration of interest: No conflict of interest is declared by authors.

Data sharing statement: Data supporting the findings and conclusions are available upon request from the corresponding author.

REFERENCES

1. Cho MJ, Yang J-H, Choi H-G, Kim WH, Yu Y-B, Park KS. An idiopathic gigantomastia. Ann Surg Treat Res. 2015;88(3):166-9. https://doi.org/10.4174/astr.2015.88.3.166 PMid:25741497 PMCid:PMC4347046
2. Kasielska-Trojan A, Danilewicz M, Strużyna J, Bugaj M, Antoszewski B. The role of oestrogen and progesterone receptors in gigantomastia. Arch Med Sci. 2019;18(4):1016-20. https://doi.org/10.5114/aoms.2019.88280 PMid:35832720 PMCid:PMC9266871
3. Jean-Louis WF, Bowder A, Dupont CR, et al. Extreme gigantomastia caused by pseudoangiomatous stromal hyperplasia at Hopital Universitaire de Mirebalais: A case report. Plast Reconstr Surg Glob Open. 2021;9(12):e3960. https://doi.org/10.1097/gox.0000000000003960 PMid:35070603 PMCid:PMC8769100
4. Morone I, de Andrade GP, Cardoso PS, et al. Bilateral pseudoangiomatous stromal hyperplasia in childhood gigantomastia: A challenge in reconstruction and management. JPRAS Open. 2018;19:106-10. https://doi.org/10.1016/j.jpra.2018.08.003 PMid:32158862 PMCid:PMC7061678
5. Krawczyk N, Fehm T, Ruckhäberle E, et al. Bilateral diffuse pseudoangiomatous stromal hyperplasia (PASH) causing gigantomastia in a 33-year-old pregnant woman: Case report. Breast Care (Basel). 2016;11(5):356-8. https://doi.org/10.1159/000450867 PMid:27920630 PMCid:PMC5123024
6. Tahmasebi S, Zadeh AKR, Zangouri V, Akrami M, Johari MG, Talei A. Pseudoangiomatous stromal hyperplasia of the breast: A case report. Clin Case Rep. 2022;10(8):e6221. https://doi.org/10.1002/ccr3.6221 PMid:35957783 PMCid:PMC9361800
7. Oppenheimer AJ, Oppenheimer DC, Fiala TGS, Noori S. Pseudoangiomatous stromal hyperplasia: A rare cause of idiopathic gigantomastia. Plast Reconstr Surg Glob Open. 2016;4(1):e593. https://doi.org/10.1097/gox.0000000000000572 PMid:27104092 PMCid:PMC4801101
8. Vashistha A, Rundla M, Khan F, Om P. Idiopathic gigantomastia with pseudoangiomatous stromal hyperplasia: A case report. Int J Surg Case Rep. 2020;77:915-9. https://doi.org/10.1016/j.ijscr.2020.09.151 PMid:33162384 PMCid:PMC7775963
9. Johnson R, Haywood R. A case report of prepubescent idiopathic gigantomastia with pseudoangiomatous stromal hyperplasia. Int J Surg Case Rep. 2022;101:107793. https://doi.org/10.1016/j.ijscr.2022.107793 PMid:36434871 PMCid:PMC9685276
10. Lee JW, Jung GS, Kim JB, et al. Pseudoangiomatous stromal hyperplasia presenting as rapidly growing bilateral breast enlargement refractory to surgical excision. Arch Plast Surg. 2016;43(2):218-21. https://doi.org/10.5999/aps.2016.43.2.218 PMid:27019818 PMCid:PMC4807181