

## Case Report



## OPEN ACCESS

**Received:** Feb 9, 2023

**Revised:** Apr 7, 2023

**Accepted:** Apr 16, 2023

**Published online:** May 8, 2023

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### Conflict of Interest

The authors declare that they have no competing interests.

### Author Contributions

Conceptualization: Kim EK; Data curation: Woo SH; Investigation: Kang H, Choi WJ; Methodology: Woo SH, Kim EK; Project

# Pseudoangiomatous Stromal Hyperplasia of the Breast in a Female Adolescent Presenting as Bilateral Gigantomastia

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## ABSTRACT

Pseudoangiomatous stromal hyperplasia (PASH) is a rare idiopathic proliferative mesenchymal breast disease related to hormonal imbalance, and thus extremely rare in children and adolescents. In addition, PASH manifests as a bilateral gigantomastia in some cases with no established cause or treatment. Here, we report a case of a rapidly developed PASH presenting with bilateral gigantomastia in a 14-year-old premenarchial female patient. Considering the patient's age and emotions and the need for nipple-areolar complex repositioning, we performed reduction mammoplasty rather than total mastectomy despite the possibility of recurrence. Although some masses could not be completely removed, no complications, such as infection, wound dehiscence, or hematoma occurred postoperatively. The patient was stable during the 18-month follow-up period, although an evidence of recurrent and residual disease was noted upon ultrasonography.

**Keywords:** Breast Diseases; Gigantomastia; Mammoplasty; Pseudoangiomatous Stromal Hyperplasia

## INTRODUCTION

Pseudoangiomatous stromal hyperplasia (PASH) is a rare benign proliferative mesenchymal breast condition of an unknown etiology and was first described in 1986 [1,2]. Its clinical manifestations vary widely from incidental microscopic findings to palpable masses, gigantomastia, and asymmetry [3]. Gigantomastia, reported unilaterally as well as bilaterally [4-7], was identified incidentally on mammography screening in approximately 53% of the patients, and as a palpable mass in approximately 44% of the patients [1].

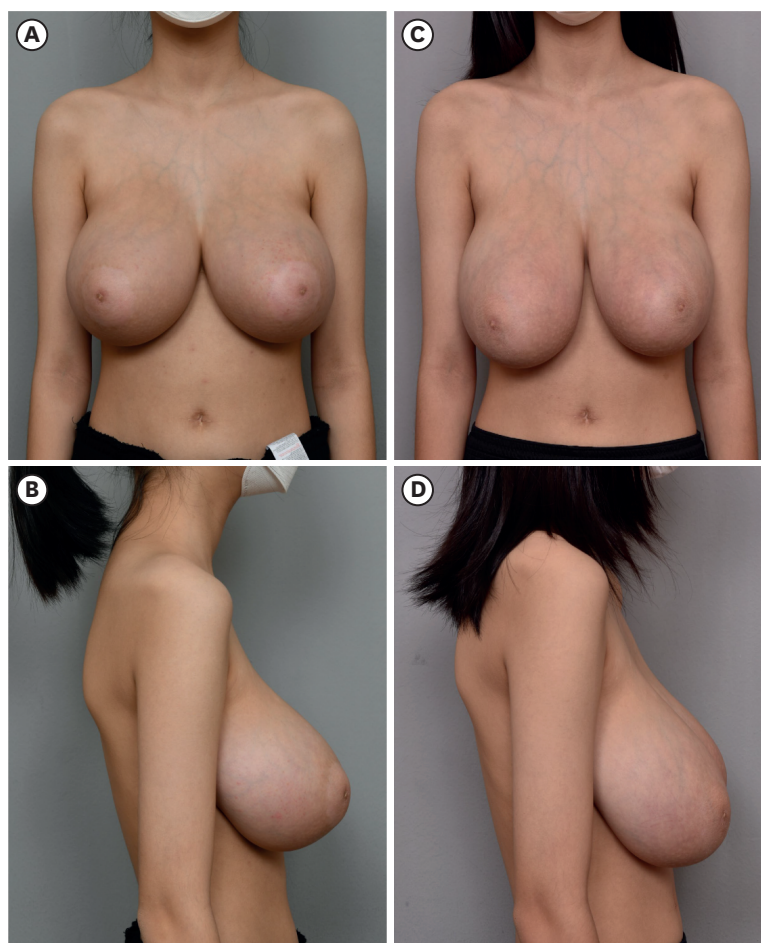
It is difficult to accurately diagnose and determine the incidence of PASH, as its presentation is diverse, non-specific, rare, and benign. However, since PASH occurs mostly in premenopausal women aged 30–40 years, it is considered to be related to hormones and is extremely rare in children and adolescents [2,4]. However, treatment methods for PASH have not yet been established. Here, we report a case of bilateral breast reduction performed in a patient with a diffuse PASH, who showed rapid bilateral breast enlargement during adolescence before reaching menarche.

administration: Kim EK; Resources: Choi WJ,  
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Kang H, Choi WJ; Writing - original draft: Woo  
SH; Writing - review & editing: Woo SH, Kim  
EK.

## CASE REPORT

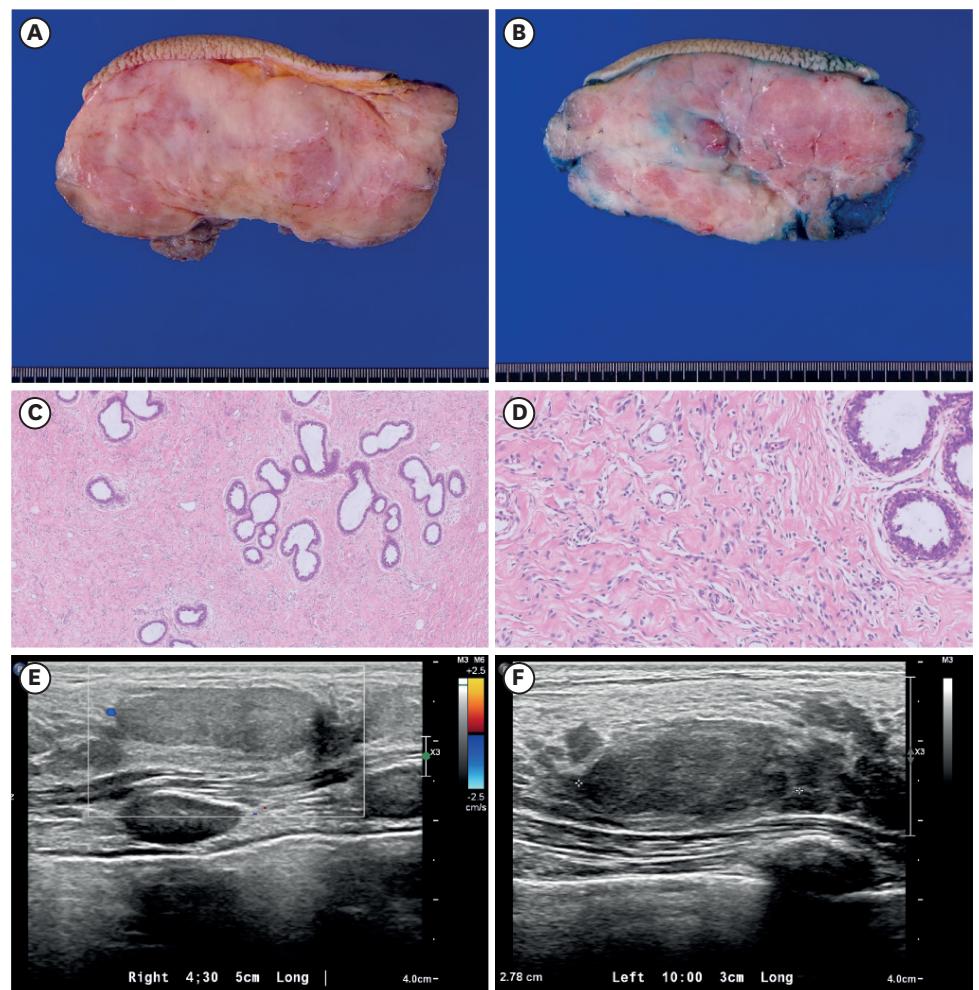
A 14-year-old girl presented to our center with a symmetrical bilateral breast enlargement. No breast development occurred until one year ago; however, the breasts started to increase in size rapidly six months ago (**Figure 1A and B**). Physical examination and ultrasonography performed at another hospital revealed no structural abnormalities related to breast. Her height and weight were 161 cm and 45 kg (body mass index [BMI] = 17 kg/m<sup>2</sup>), respectively. She had no history of drug use or any remarkable past or family history and did not experience menarche. The blood tests showed no hormonal imbalance.

PASH was diagnosed based on an ultrasound-guided core needle biopsy of both breasts (five pieces from each side). Breast enlargement continued for approximately three months after her first visit to our clinic, showing rapid changes in size (**Figure 1C and D**). We considered total mastectomy or breast reduction for surgical treatment. However, considering the patient's age and emotional well-being, and because the nipple-areolar complex (NAC) was extremely ptotic for implant-based reconstruction after nipple-sparing mastectomy, we performed bilateral breast reduction to simultaneously reduce the breast volume and raise the NAC.

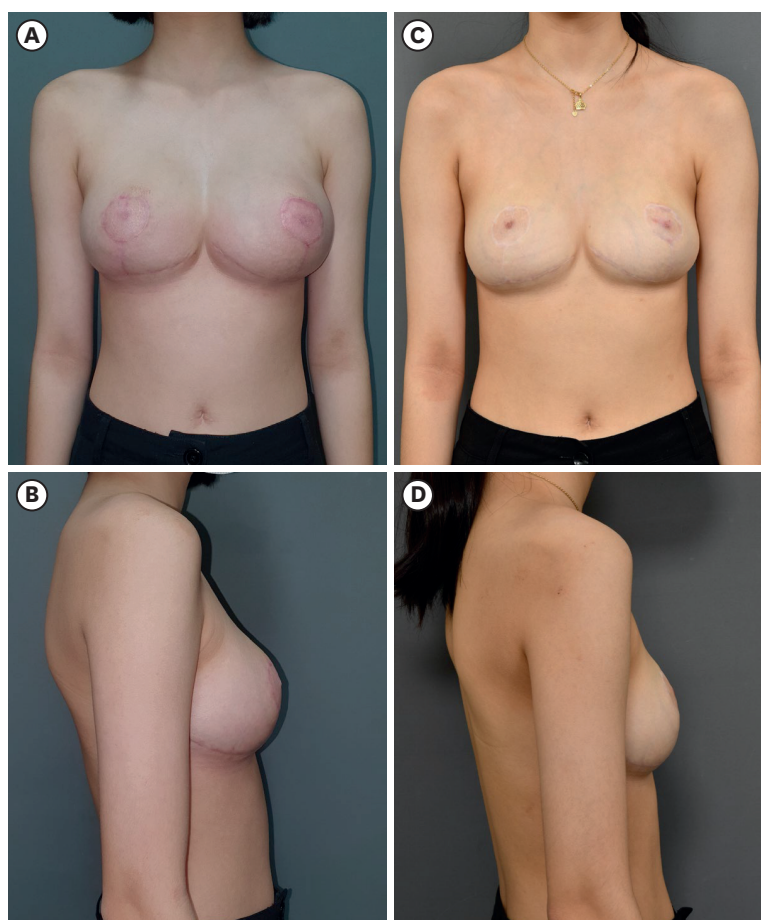


**Figure 1.** Preoperative clinical photos. (A, B) Photographs at the first visit. (C, D) Photographs at three months after the first visit, showing a rapid size increase.

Breast reduction was performed using the Wise pattern incision, and superomedial pedicles were used. Total 840 g of the right breast and 740 g of the left breast was resected (**Figure 2A and B**). No immediate complications such as infection, hematoma, skin necrosis, or wound dehiscence occurred postoperatively. Hematoxylin and eosin staining revealed pseudoangiomatous spaces that invaded the perilobular and intralobular stroma, and a myofibroblast proliferation was observed between the normal lobules in both breast specimens. The final diagnosis of PASH was made (**Figures 2C and D**). Ultrasonography revealed new and re-grown benign-looking masses in the right breast 18 months postoperatively (**Figure 2E and F**). However, no rapid growth of the breasts on gross examination was observed, and the patient was found stable on follow-up (**Figure 3**).



**Figure 2.** Specimen, pathology, and imaging test results. (A) Specimens obtained after reduction mammoplasty in a wise pattern. 840 g removed from the right side, and (B) 740 g removed from the left side. Multiple masses of various sizes were observed. (C) H&E stain,  $\times 50$  magnification (right breast). Histological findings showing pseudo-angiomatous spaces that involve perilobular and intralobular stroma. (D) H&E stain,  $\times 200$  magnification (right breast), showing an anastomosing space penetrating a dense collagenous stroma. (E) Ultrasound with color Doppler findings at 18 months postoperatively. Compared with the ultrasound findings at five months postoperatively, few new or re-grown benign-looking masses in the right breast were observed. (F) No interval changes of a benign looking mass in the left breast were observed. H&E = hematoxylin-eosin.



**Figure 3.** Surgical results and follow-up clinical photographs. (A, B) Clinical photographs at five months postoperatively. (C, D) Clinical photographs at 18 months postoperatively.

## DISCUSSION

PASH is rare and has diverse clinical manifestations; therefore, physicians treating breast lesions are often unfamiliar with it. PASH may not be accompanied by breast enlargement, which may manifest unilaterally or bilaterally [3-6]. Additionally, PASH sometimes occurs in men [3]. Although PASH occurs mainly in the 30- and 40-year-old age group, studies have reported an age range of 13–67 years; PASH occurs mostly in pre-menopausal women and is often misdiagnosed as fibroadenoma in its early stages [2]. Most studies are case reports related to diagnosis, especially imaging tests and pathology; however, treatment methods have not yet been established. Surgical resection is not required in all cases, and the excision of the masses may be therapeutic. However, surgery involving the breast parenchyma is required if the breasts are too large or rapidly growing or if there is an asymmetry due to a unilateral involvement. In some cases, total mastectomy is performed [6]. Moreover, some patients who underwent breast reduction did not develop complications [7], while others required total mastectomy because a recurrence occurred after reduction [8]. It is difficult to reach an accurate consensus on the recurrence rate and prognosis according to treatment methods. In the present case, the overall breast size remained stable for 18 months, even with some new and re-grown benign-looking masses on ultrasonography that increased in a slow pattern, unlike the preoperative rapid growth pattern.

Kurt et al. [9] reported the distribution of the incidence and tumor size according to age and reported that < 7% of patients were under the age of 20 years, and a mass of  $\leq 2$  cm was most commonly found. In contrast, we reported a very rare case that showed multiple large masses in both breasts at a very young age. Thus, the actual incidence in children and adolescents may be higher than that previously reported, as PASH is sometimes diagnosed incidentally, and screening ultrasonography or mammography is rarely performed in children and adolescents. Therefore, when our patient underwent ultrasonography for the first time, no structural abnormalities other than a breast parenchymal hypertrophy were identified. Both breasts appeared isoechoic without any lesions and with clear boundaries, and PASH could not be diagnosed without active suspicion.

Moreover, the patient did not want any further breast development when the breast enlargement became excessive; therefore, she desperately tried to lose weight. Nevertheless, the breast enlargement progressed rapidly. After breast reduction, she was relieved of the psychological stress, and at 18 months postoperatively, her height and weight reached 167 cm and 51 kg (BMI = 18.29 kg/m<sup>2</sup>), respectively. This indirectly suggests that even in children and adolescents, the breasts occupy a very important place in the body image, self-esteem, and quality of life. No consensus regarding treatments for PASH in children and adolescents has been established yet; therefore, treatment principles should not be different from those in adults. In cases of bilateral PASH in children and adolescents, both breast reduction and total mastectomy are performed as in adults [5]. Both approaches have their advantages and disadvantages. However, total mastectomy can cause a great emotional shock to children and adolescents and their parents even if a reconstruction is performed, particularly because PASH is a benign disease with an unknown recurrence rate.

When deciding a surgical method other than mass excision, sufficient consultation with the patients and their parents is necessary preoperatively. In cases of recurrence after breast reduction surgery, re-reduction may be performed, depending on the patient's age, tumor growth rate, and tumor size. In addition, reconstruction after total mastectomy may be performed, in which it is safer to reposition the NAC to an ideal position and tailor the skin before definitive surgery than to move it simultaneously during a skin-sparing mastectomy, especially when the breast is large and the NAC is in a very ptotic position, as in the present case. Hammond et al. [10] reported that premastectomy mastopexy or reduction should be performed in large or ptotic breasts to prevent mastectomy flap formation or NAC necrosis during subsequent nipple-sparing mastectomy with immediate breast reconstruction. The theory that premastectomy reduces stress on the remaining skin flap after mastectomy has been introduced in several studies [11-13]. Hammond et al. [10] delayed mastectomy by 39–96 days (average, 55 days) after mastopexy or reduction, and achieved safe results. However, waiting was not very dangerous because PASH is a benign disease. Therefore, we suggest that reduction mammoplasty, or at least the repositioning of the NAC, should be considered as the first treatment option for large ptotic breasts with PASH, especially in children or young women, although mastectomy and reconstruction might be indicated afterwards due to the rapid recurrence of PASH.

In our case, although ultrasonography showed a recurrence, the patient was stable on follow-up and showed no rapid growth of the breasts on gross appearance for 18 months. A longer follow-up period and additional studies are required, as little is known about the long-term prognosis of PASH.

## REFERENCES

1. Raj SD, Sahani VG, Adrada BE, Scoggins ME, Albarracin CT, Woodtichartpreecha P, et al. Pseudoangiomatous stromal hyperplasia of the breast: multimodality review with pathologic correlation. *Curr Probl Diagn Radiol* 2017;46:130-5.  
[PUBMED](#) | [CROSSREF](#)
2. Jaunoo SS, Thrush S, Dunn P. Pseudoangiomatous stromal hyperplasia (PASH): a brief review. *Int J Surg* 2011;9:20-2.  
[PUBMED](#) | [CROSSREF](#)
3. Drinka EK, Bargaje A, Erşahin CH, Patel P, Salhadar A, Sinacore J, et al. Pseudoangiomatous stromal hyperplasia (PASH) of the breast: a clinicopathological study of 79 cases. *Int J Surg Pathol* 2012;20:54-8.  
[PUBMED](#) | [CROSSREF](#)
4. Shehata BM, Fishman I, Collings MH, Wang J, Poulik JM, Ricketts RR, et al. Pseudoangiomatous stromal hyperplasia of the breast in pediatric patients: an underrecognized entity. *Pediatr Dev Pathol* 2009;12:450-4.  
[PUBMED](#) | [CROSSREF](#)
5. Koksall H, Kara B, Eren Karanis MI, Sen S, Koksall Y. Pseudoangiomatous stromal hyperplasia of breast in an adolescent: a case report and review of the literature. *J Pediatr Adolesc Gynecol* 2021;34:209-12.  
[PUBMED](#) | [CROSSREF](#)
6. Dai H, Connor C, Cui W, Gatewood J, Fan F. Bilateral diffuse tumorous pseudoangiomatous stromal hyperplasia: a case of bilateral mastectomy in a 29-year-old woman. *Case Rep Pathol* 2014;2014:250608.  
[PUBMED](#) | [CROSSREF](#)
7. 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.  
[PUBMED](#) | [CROSSREF](#)
8. Xu X, Persing SM, Allam O, Park KE, Mozaffari MA, Lannin DR, et al. Management of recurrent bilateral multifocal pseudoangiomatous stromal hyperplasia (PASH). *Breast J* 2020;26:1814-7.  
[PUBMED](#) | [CROSSREF](#)
9. Kurt E, Turanlı S, Markoç F, Berberoğlu U. How to manage pseudoangiomatous stromal hyperplasia: our clinical experience. *Turk J Med Sci* 2017;47:1410-5.  
[PUBMED](#) | [CROSSREF](#)
10. Hammond DC, Little AK. The role of premastectomy mastopexy and breast reduction in the reconstruction of the enlarged or ptotic breast. *Plast Reconstr Surg* 2022;150:270-80.  
[PUBMED](#) | [CROSSREF](#)
11. Spear SL, Hannan CM, Willey SC, Cocilovo C. Nipple-sparing mastectomy. *Plast Reconstr Surg* 2009;123:1665-73.  
[PUBMED](#) | [CROSSREF](#)
12. Salibian AA, Frey JD, Karp NS, Choi M. Does staged breast reduction before nipple-sparing mastectomy decrease complications? A matched cohort study between staged and nonstaged techniques. *Plast Reconstr Surg* 2019;144:1023-32.  
[PUBMED](#) | [CROSSREF](#)
13. Gunnarsson GL, Bille C, Reitsma LC, Wamberg P, Thomsen JB. Prophylactic nipple-sparing mastectomy and direct-to-implant reconstruction of the large and ptotic breast: is preshaping of the challenging breast a key to success? *Plast Reconstr Surg* 2017;140:449-54.  
[PUBMED](#) | [CROSSREF](#)