

Breast Augmentation in Swyer Syndrome Patient

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Background: The number of breast augmentation in Indonesia has been raised, especially in young women. Women with disorders of sex development (DSD) can be one of our patients that come for breast augmentation. These patients may also have other problems that can lead to serious disease in her later life, such as malignancies which has 30% probability.

Patients and Methods: We report one case of female with 46 XY karyotype and normal female phenotype. She appeared to be normal female but did not develop secondary sexual characteristics at puberty with Tanner classification M1P1, did not menstruate, and had streak gonads in ovarian localization. This gonadal dysgenesis syndrome is also called Swyer syndrome.

Result: A clinical team consists of plastic surgeons, gynecologist, psychiatrist, geneticist was build to manage our patient comprehensively. We performed breast augmentation, laparoscopic gonadectomy, and psychological support.

Summary: Patient with disorder of sex development (DSD) can be one of our patients who come for breast augmentation. One must pay attention to subtle sign leading to DSD patients such as, history of amenorrhea, wide chest and lack of women body curve. Complete evaluation of sexual development is needed before performing breast augmentation.

Keywords: DSD, Swyer Syndrome, Breast Augmentation, Laparoscopic Gonadectomy

Lat ar belakang: Jumlah augmentasi payudara di Indonesia semakin bertambah, terutama pada wanita muda. Wanita dengan gangguan perkembangan sex mungkin saja menjadi salah satu dari pasien kita yang datang untuk dilakukan pembesaran payudara. Pasien dengan kelainan DSD ini bila tidak ditemukan, ke depannya dapat menjebak kita ke dalam masalah lain yang lebih serius misalnya keganasan gonad yang memiliki kemungkinan hingga 30%.

Pasien dan Metode: Kami melaporkan sebuah kasus, seorang wanita dengan kariotipe 46XY. Pasien ini secara fenotip adalah wanita normal, namun tidak memiliki perkembangan ciri seks sekunder yang sesuai dengan tingkat klasifikasi Tanner M1P1. Pasien tidak mengalami menstruasi, dan hanya memiliki streak gonad. Kelainan disgenesis gonad ini disebut juga sebagai sindroma Swyer.

Hasil: Sebuah team klinis yang terdiri dari ahli bedah plastik, ginekologis, psikiatris dan genetisis dibangun untuk mentatalaksana pasien ini secara komprehensif. Pasien ini kami lakukan augmentasi payudara, gonadektomi laparoskopis dan pemberian dukungan kejiwaan.

Ringkasan: Pasien dengan gangguan perkembangan seks (DSD) mungkin menjadi salah satu pasien kita yang meminta untuk dilakukan augmentasi payudara. Kita harus memeperhatikan beberapa tanda yang mengarah kepada gangguan perkembangan seks seperti, dada yang bidang, riwayat amenorrhea dan lekukan tubuh yang kurang mirip pada wanita pada umumnya. Evaluasi pasien secara menyeluruh mengenai perkembangan ciri seks, penting untuk dilakukan sebelum dilakukannya augmentasi payudara.

Kata Kunci: DSD, Swyer Syndrome, Breast Augmentation, Laparoscopic Gonadectomy

More than 300,000 women in the United States and many more around the world undergo surgery with breast implants every year, either to replace breasts lost to cancer or to increase the size of healthy breasts.¹ The number of breast augmentation also increasing in Indonesia, especially in young women. In this high technology era, women all over Indonesia can search for information to get the body figure that she

wants not only from plastic surgeons but also from beautician.

When facing women with breast hypoplasia, several things should be considered and discussed with the patient before plastic surgeons decide to perform breast augmentation. Clinical evaluation of patient history and physical examination play an important role before the decision of surgery was made. Knowing the expectation of patient

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and her psychological status can help us to determine whether the patient is a good candidate of breast augmentation and will not develop other problems later after surgery.

As a plastic surgeon, sometimes we forgot that women with breast hypoplasia might also have other problems that can lead to serious disease in her later life. Women with disorders of sex development (DSD) can be one of our patients that seek for breast augmentation. When women with these problems seek for our help, it will be our consideration to make the diagnosis of DSD and consult this patient for a multidisciplinary approach to get a holistic management.

Swyer syndrome, or 46 XY gonadal dysgenesis, is one of DSD patient that can come to plastic surgeons for breast augmentation. At birth, patients with the XY female type of gonadal dysgenesis (Swyer syndrome) appear to be normal females; however, they do not develop secondary sexual characteristics at puberty, does not menstruate, and have streak gonads.^{2,3} Many of these patients did not realize that they have bigger problems that can lead to malignancies instead of only small breast size. Early diagnosis of Swyer syndrome is important to decrease the risk of gonadal malignancy. The risk of malignant transformation by the age of 40 is 30%.³

PATIENT AND METHODS

We report a 27 years old "female" with breast hypoplasia. She came to our hospital asking for breast augmentation because she was ashamed of her body figure and planned to get married in several months after her first visit to our hospital. She was a tall young lady, with feminine face figure but had a wide chest and lack of women body curve. By the time of her first visit, we didn't realize that she is an XY gonadal dysgenesis, until she explained that beside plastic surgeon, she also follows a treatment program with gynecologist due to her primary amenorrhea.

Work up of the etiology of primary amenorrhea, was done by the gynecologist.

Knowing that this patient is a XY female, we decide to manage this patient simultaneously and also with psychiatrist. With the help of psychiatrist, the patient can get a psychological help that she need because of her disease. Patient acceptance of what she is, was an important factor whether to proceed with breast augmentation or not. After several discussions with the psychiatrist, she still wants to undergo breast augmentation.

Preoperative design was done bedside (Figure 1), patient was involved to decide how much volume can be implanted into her breast. By calculating her medial pinch, lateral pinch, and also simulating the implant onto her breast, we decided to implant around 195-210 cc of silicon. The exact number will be determined using sterilized silicon implant seizer by the time the operation was performed.

Breast augmentation (Figure 1) was performed under general anesthesia simultaneously after gynecologist performed laparoscopic gonadectomy (Figure 2). Two hundred cc of round high profile silicon implants were inserted under subglandular plane. Pressure garment was used after the surgery and patient was discharge after 1-day observation. Pathological analysis was performed for the gonads that have been removed. No signs of malignancies at the time the gonads were removed.

RESULTS

At one month follow up (Figure 3), there is neither sign of infection nor complication of breast augmentation in this patient. Overall satisfaction of her breast after the augmentation is good. The scar is minimal and the shape is acceptable. One thing that is less aesthetic is that her nipple is too small.

DISCUSSION

Disorders of Sex Development (DSD) are a congenital condition in which development of chromosomal, gonadal and anatomical sex is atypical. It is estimated that DSD affects 1 in 4500 to 5000 live births in the general population although with variability



Figure 1. (Left) Preoperative view & measurement. (Right) Breast augmentation with 200 cc round high profile silicon implants.



Figure 2. Bilateral laparoscopic gonadectomy was performed by



Figure 3. One month follow up

regarding the various DSD subtypes. DSD patients are subdivided into different entities: 46 XY DSD, 46 XX DSD, and sex chromosomal DSD.⁴

Swyer syndrome (46 XY DSD) was a complete gonadal dysgenesis. The exact incidence of the condition is unknown but can be estimated at 1:80.000 births.⁵ Other reference shows that the incidence is about 1:100.000. Woman with Swyer syndrome diagnosed in early adolescence with delayed pubertal development. Clinically they have tall stature, female external genitalia but they do not develop secondary sexual characteristics at puberty, do not menstruate and have streak gonads. Minimal breast enlargement reflects peripheral aromatization of androgens. Modern studies show patients with 46, XY pure gonadal dysgenesis are at a higher risk of developing gonadoblastoma and dysgerminoma, and may occur even in young ages. A bilateral gonadectomy should be done especially by laparoscopy when a Swyer syndrome is discovered in order to avoid the risk of malignant transformation.⁶ Ovarian cancer in patients with Swyer syndrome is found frequently when the patient is under 25 years of age (one third), and this incidence increases with age, overall affecting 30% to 70% by the third decade of life and reaching 80% by the fourth decade.⁷

Due to the rarity of DSDs, it is important that their management is undertaken in tertiary referral centers where a multidisciplinary team can provide the necessary medical and surgical support.⁸ In this condition, team work with gynecologist, psychiatrist together with plastic surgeons will provide a better result physically and psychologically for the patient. Gynecologist will take part assessing patient hormonal and internal organ status, while plastic surgeons planning for the technique that will be performed for breast augmentation. With prophylactic laparoscopic gonadectomy, the risk of malignancies can be reduced.

Congenital breast deformities may be hyperplastic, hypoplastic or deformational. Breast augmentation is the treatment choice for bilateral mammary hypoplasia.⁹ The key of satisfactory breast augmentation by traditional

planning are deciding the plane, incision and implant type, form and size. Patient with thick soft tissue cover (>1.5 cm pinch thickness at the upper pole) and no more than mild ptosis is a good candidate for subglandular plane dissection. As in our patient, with such thickness on the upper pole (2.2cm right breast, 1.8cm left breast) it is indicated for subglandular plane for inserting the silicon. Infra mammary incisions were choose because it gives easier approach to inserted the silicon implant, and the scar will be hide under the neo-infra mammary fold. 200 cc of silicon implant was choose after calculation of medial and lateral fold of each breast and converting to the diagram of silicon implant product. Before the actual insertion of the implant, silicon seizer was inserted to assure that the pocket that has been made was not to tight.

SUMMARY

Patient with disorder of sex development (DSD) can be one of our patients come for breast augmentation. Complete evaluation of sexual development is needed before performing breast augmentation. It is important is how to diagnoses this syndrome and performed multidisciplinary management for a batter quality life for the patient and preventing the risk of malignancies that can be developed later.

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