Pseudomamma of the inguinal region in a female patient: A case report

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ABSTRACT

INTRODUCTION: Supernumerary breasts are relative common benign congenital anomalies. General population occurrence rates vary up to 6% according to ethnicity and gender. Higher incidence is recorded in Asian individuals, especially Japanese. Embryonic breast development of the mammary ridge (milk line) is explained and supernumerary breast tissue resulting from involution failure of any portion of the embryonic mammary folds is described.

PRESENTATION OF CASE: We report a case of supernumerary breast (pseudomamma) in a female occupying her right inguinal region that was treated in the breast unit of our hospital. Differential diagnosis, imaging methods, operative approach, surgical treatment and histological verification are specified.

DISCUSSION: Classification system for supernumerary breast tissue is presented, high risk population is identified and congenital malformations linked to it are outlined. Evaluation of diagnostic workup and limitations are stated. Cancerous degeneration and justification for surgical removal of the accessory gland is discussed.

CONCLUSION: Differential diagnosis of lesions along the milk line should always be inclusive of developmental abnormalities such as any type of supernumerary breast, often overlooked due to small size, although carrying a malignant potential equal to normally positioned breasts. Surgical correction is a sensible approach, often encouraged by the patients. Additional evaluation is recommended due to the frequent accompanying urinary tract and cardiac anomalies.

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1. Introduction

Supernumerary nipples or breasts are relative common benign congenital anomalies. First descriptions are dated in ancient Greece, where supernumerary breast was regarded as sign of fertility. However, during medieval age, individuals with supernumerary breasts were considered as bad omen. In modern literature, general population occurrence rates vary up to 6% according to ethnicity and gender. Higher incidence is recorded in Asian individuals, especially Japanese, this being twice as common in females than in males [1–3]. Embryonic breast development begins during the fourth week of gestation, when ectodermal tissue forms a ridge across the ventral surface, extending from the axilla towards the midline of the groin. This ridge is called mammary ridge or milk line. In normal breast development, the mammary ridge recedes, leaving only bilateral mammary tissue at the level of the fourth intercostal space. Supernumerary breast tissue results from involution failure of any portion of the embryonic mammary folds [4]. Approximately two-thirds of affected patients have solitary accessory breast tissue and 30% of women with polymastia have two or more locations of ectopic breast tissue [5]. In most of the cases the thoracic or abdominal portions of the milk line are involved, often just below the inframammary crease, another 20% occur in the axilla, while other infrequent sites documented include the face, posterior thigh, foot and vulva [6–12].

2. Presentation of case

A 56 year old Caucasian postmenopausal female, gravida 2, parity 2, presented to the outpatient department of our hospital for the first time, in order to perform her annual gynecological screening. During physical examination, a palpable raised lesion was easily distinguished, occupying her right inguinal region and upper anterior aspect of her thigh. (Picture 1) The exact position of the mass was inferior of the inguinal ligament and lateral to the adjacent inguinal vessels. In palpation the lesion was painless, smooth, soft and mobile. In closer observation, a skin-colored region rep-
Pseudomamma of the right inguinal region in a female. Representing a hypoplastic nipple along with surrounding areola was identified. (Picture 2) No other similar lesions could be identified in her body and in clinical examination breasts and axillary regions were found to be normal.

After a brief history, the patient revealed that she initially noticed this lesion during puberty, gradually increasing in size after her first delivery. She was then referred to a general surgeon, reassured that this was a case of lipoma and no further treatment was necessary. There were no complains of cyclic pain, enlargement or fluid discharge from the mass. The patient reported that there was light discomfort and oversensitivity all over the lesion when pressure was applied. Past medical history of the patient included appendectomy in younger age and hyperlipidemia under atorvastatin treatment. Family medical history for either supernumerary nipple or breast was negative.

Medical imaging techniques were implemented for further investigation. The patient underwent ultrasound scan and diagnostic mammography of the lesion that were unable to differentiate between lipoma and breast tissue. (Picture 3) An additional bilateral mammography of the breasts was performed to exclude breast lesions. Urinanalysis was negative and routine hematological and biochemical parameters were within normal range.

After a multidisciplinary discussion and extensive counseling, the patient consented for surgical removal of the lesion. Under general anesthesia, a tear-drop-like incision over the mass followed by wide local excision of the excessive tissue and overlying skin was performed. (Picture 4) Inguinal vessels and underlying fascia were left intact, while reapproximation of the skin was achieved. Due to the rich vascular and lymphatic supply of the area, wound drainage was not performed.
was subcutaneously positioned for 24 h. Histopathological examination of the specimen confirmed the diagnosis of ectopic breast tissue class V (pseudomamma), consisting of nipple and areola but without any glandular tissue, being replaced by fat. Postoperatively the patient was discharged in perfect health, two days later. (Picture 5) After three months follow-up, no complications were reported.  

3. Discussion

Initial classification system for supernumerary breast tissue was first published by Kajava in 1915 and still remains in use today in current practice. According to Kajava’s system, supernumerary breast tissue can be categorized to: Class I (polymastia) consists of a complete breast with nipple, areola, and glandular tissue. Class II consists of nipple and glandular tissue but no areola. Class III consists of areola and glandular tissue but no nipple. Class IV consists of glandular tissue only. Class V (pseudomamma) consists of nipple and areola but no glandular tissue (with fat replacing glandular tissue). Class VI (polythelia) consists of a nipple only. Class VII (polythelia areolaris) consists of an areola only. Class VIII (polythelia pilosa) consists of a patch of hair only [13]. More recently ectopic breast tissue has been described by differentiating between polythelia and aberrant glandular tissue. Polythelia includes cases of accessory mammary gland remnants of mammary ridge regression, while by the term aberrant glandular tissue are represented cases of ectopic breast tissue without ductal system or relationship to the overlying skin [2]. Our patient resembles a typical case of pseudomamma.

Commonly, polymastia and polythelia occur sporadically, but familial cases have been reported [2]. Such cases are inherited in an autosomal dominant fashion with variable penetrance, although X-linked dominant transmission has also been described [9]. Supernumerary nipples can exist as a single finding or as a part of syndrome like Simpson–Galabi–Behmel syndrome, Char syndrome, Turner’s syndrome and fetal alcohol syndrome [14]. Several studies support the association between polythelia and genitourinary tract malformations. Supernumerary kidneys, renal aplasia, hydronephrosis, polycystic kidney disease, duplicated renal arteries and ureter stenosis are some of the congenital malformations linked to polythelia. A possible explanation lies on the fact that both regression of the milk line and urogenital system development happens around the third month of gestation [15]. Other anomalies such as cardiac conduction abnormalities or congenital heart disease have been rarely reported. It has been suggested that individuals with supernumerary breast tissue should undergo further investigation in order to rule out possible urinary or cardiac abnormalities [9,14]. In the present case a detailed abdominal and vaginal ultrasound scan along with preoperative electrocardiography did not reveal such an association.

Accessory breast tissue is characterized with a high incidence of being misdiagnosed and most women being unaware, until clinically examined by a breast expert. In the absence of the areola-nipple complex, a clinical diagnosis is extremely difficult. Common presumptive diagnoses include lipoma, lymphadenopathy, hydadenitis, sebaceous cyst, vascular malformations and malignancy [16,17]. If any level of uncertainty exists, ultrasonography, mammography or biopsy is proved to be helpful in differential diagnosis [4,12]. Tissue diagnosis is obviously the gold standard for accessory breast tissue. Pathologists may look for typical stroma, lobules and ducts; however these may be poorly organized in aberrant breast tissue. 

Most cases of polythelia are diagnosed since birth; however ectopic breast tissue may remain latent until enhanced by sex hormones during menarche, pregnancy or even lactation [18]. Cyclical enlargement, tenderness, discomfort, pain, milk secretion, and local skin irritation are the chief complaints, while psychological embarrassment seems to be the main cause leading to surgical removal [19]. Accessory breast tissue is prone to the same pathology as normal breast tissue and a number of both benign and malignant diseases have been described [1]. Cancerous degeneration has been reported and provides additional justification for surgical removal of the accessory gland [11,12]. The most frequently reported malignant subtype is infiltrating ductal carcinoma,
followed by medullary and lobular carcinoma [14,17]. Postoperatively further follow-up is inevitable because of the possibility of developing cancer in any retained breast tissue. Ectopic cancer usually presents as advanced stage disease due to late diagnosis, however there is no evidence to support increased incidence of neoplasia in accessory breast tissue [2,20].

4. Conclusion

Managing lesions along the milk line should keep practitioners in alert and differential diagnosis should always be inclusive of developmental abnormalities such as any type of supernumerary breast. General therapy guidelines are not definitive. Supernumerary breast tissue may be overlooked due to small size although carrying a malignant potential equal to normally positioned breasts. Surgical correction is a sensible approach, often encouraged or even demanded by the patients themselves. Ectopic breast tissue whenever encountered must be documented and additional evaluation is necessary, due to the frequent accompanying urinary tract and cardiac anomalies.

Conflicts of interest

All authors disclose no conflicts of interest.

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Ethical approval

This is not a research study, no ethical approval is required.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Spyridon Marinopoulos: first author, breast surgeon.
Ioannis Arampatzis: studying and presenting references and bibliography.
Flora Zagouri: writing, team oncologist.
Constantine Dimitrakakis: head breast surgeon, correcting manuscript.

Guarantor

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