

SURGICAL CORRECTION OF POLYMASTIA AND POLYTELIA WITH AN UNCOMMON PRESENTATION

Correção cirúrgica de polimastia e politelia com apresentação incomum

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ABSTRACT

Polymastia is a rare condition that is present in 1 to 5% of the population. Clinically, it is characterized by the presence of two or more breasts, which can occur in males or females. The condition may appear with or without the presence of extra nipples, which is termed polythelia. Bilateral presentation is uncommon. Here we report a case of a patient who had more than one pair of breasts and extra nipples which were surgically corrected.

KEYWORDS: Breast; breast diseases; mammoplasty.

RESUMO

Polimastia é uma condição rara presente em 1 a 5% da população. Clinicamente se caracteriza pela presença de duas ou mais mamas e pode ocorrer em homens ou mulheres. Essa condição pode se apresentar com ou sem a presença de mamilos extras, ou seja, politelia. A apresentação bilateral é incomum. Aqui apresentamos um caso de uma paciente que tinha mais de um par de mamas e mamilos extras que foram removidos cirurgicamente.

PALAVRAS-CHAVE: Mama; doenças mamárias; mamoplastia.

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INTRODUCTION

Polymastia is a term used to describe the presence of more than two breasts in humans¹ and is synonymous with accessory or supernumerary breast tissue. This condition occurs when a failure occurs in the embryonic development of breast tissue. It is usually an asymptomatic condition but can cause anxiety, cosmetic problems, pain or restriction of movement².

Its incidence varies around 1-5% of the population, with the armpit being the most frequent site of onset³. It can be identified during or before puberty and during pregnancy. Usually, the accessory breast tissue appears sporadically, but it is also suspected that it may be a hereditary condition, once that the abnormality is often found among relatives⁴. Bilateral presentation is infrequent and no reports similar to the one described have been found in the literature, in which the patient presents more than one pair of extra breasts and nipples.

CASE REPORT

A 12-year-old female patient was referred to the mastology clinic for the presence of multiple nipples. Physical examination revealed the presence of extra breast in the bilateral axillary region of two pairs of areola papillary complexes (APC) on the breasts and two additional pairs of APCs (Figure 1), located bilaterally in the axilla and in the abdominal region. One of the paired mammary papillae does not have an areola.

It was decided to perform surgical correction, with excision of the accessory breast tissue and extra APCs (Figure 2). Removal of the breast tissue was performed through spindle

incisions, followed by subcutaneous approach and closure of the skin with intradermal suture with 4-0 monocryl wire. There were left two penrose drains, removed the next day. The histopathologic revealed to be a breast tissue with no signs of malignancy. The patient returned four months later for a follow-up and was asymptomatic and satisfied with the results (Figure 3).



Figure 2. Preoperative marking.



Figure 1. Extra breast in bilateral axillary region, pair of supernumerary mammary papillae in the thorax and pair of supernumerary mammary papillae in the abdomen.



Figure 3. Final result four months after surgery.

DISCUSSION

During the sixth week of embryonic development, the mammary line, which represents two ectodermal thickenings, develops along the sides of the embryo, extending from the axillary region to the groin. In normal development, most of the embryonic mammary peaks disappear, except for the two segments in the pectoral region, which later become breasts. Regression failure in any portion of the mammary ridge can lead to polymastia, with or without an APC. Therefore, the ectopic breast usually occurs along the “milk line” or breast line⁵.

Ectopic mammary tissue can also be located on the face⁶, the foot⁷, the back⁸, the perineum⁹, and the anus¹⁰. These locations outside the mammary line can be explained by the migration of primordial mammary cells during the development of the chest wall or through the modification of apocrine sweat glands⁴.

The accessory mammary tissue has the same capacity to undergo benign and malignant alterations as the normal mammary tissue¹¹; however, the incidence of carcinoma in the accessory breast tissue is rare, corresponding to only 0.3% of the breast carcinomas¹². There are multiple reports of benign alterations, such as fibroadenomas^{4,13-15} or hyperplasias^{16,17}, but the incidence of these alterations in supernumerary breasts is uncertain. Polymastia may present in an aberrant form that is not accompanied by a nipple or areola and is commonly mistaken for lipoma, lymphadenopathy, or hidradenitis¹¹.

Polytelia is the term used to designate the presence of accessory papillae, considered a benign congenital anomaly. They may bother the patient because of cosmetic concerns, pain and swelling during menstruation or milk secretion¹⁸. Polytelia has been associated with urinary abnormalities — such as supernumerary kidneys, renal insufficiency and renal carcinomas — which can be explained, in part, by the parallel embryological development of mammary structures and the genitourinary system⁵. In the present case, the patient had three pairs of extra nipples and no urinary malformations were identified.

Polytelia is more common in males than in females and more incident in the afrodescendant population than in the Caucasian¹⁹. Their incidence in the population varies between 0.22–5.60%, depending on factors such as genetic inheritance

and ethnicity²⁰. Because supernumerary papillae are often clinically obvious, occasionally, when only a papilla or areola and papillae are present, biopsy is needed to establish a diagnosis. Supernumerary papillae may also be clinically confused with fibroids, lipomas, warts and nevi²¹.

The oldest and most commonly used classification was described by Kajava in 1915, who classified supernumerary mammary tissue into eight categories:

1. complete supernumerary breast, with papilla, areola and glandular tissue;
2. supernumerary breast without a areola, but with papilla and glandular tissue;
3. supernumerary breast without papilla, but with areola and glandular tissue;
4. only aberrant glandular tissue, without papilla and areola;
5. only papilla and areola, with glandular tissue replaced by fat (pseudomama);
6. only papilla (polytelia);
7. only areola (areolar polytelia);
8. presence of only a tuft of hairs (polytelia pilosa)²².

Accessory breast tissue is present from birth, but is often only diagnosed after puberty, pregnancy or lactation, a period in which symptoms become more evident, such as swelling and pain during menstruation and elimination of secretions during lactation. Still, many cases are asymptomatic. In cases of doubt, definitive diagnosis should be made through fine needle aspiration or excisional biopsy, as it may be confused with abscess or another cause²³.

After being diagnosed, the recommended treatment is the surgical one, so that a simple excision of the tissue or a liposuction can be performed. Preoperative examinations should include ultrasonography to exclude the possibility of benign or malignant lesions. It is also recommended that patients with a family history of breast cancer undergo a radical resection instead of liposuction². In the present case, it was decided to perform the removal of the breast tissue through spindle incisions, followed by subcutaneous approach and closure of the skin with intradermal suture. The patient is satisfied with the results four months after the operation.

REFERENCES

1. Santos Junior LA, Eulálio Filho WMN, Medeiros Neto AM. Polimastia e politelia: imagem. *Rev Bras Mastologia*. 2016;26:24-5. <https://doi.org/10.5327/Z201600010006RBM>
2. Qian JG, Wang XJ, Yu AR, Zhou FH. Surgical correction of axillary accessory breast tissue: 12 cases with emphasis on treatment option. *J Plast Reconstr Aesthet Surg*. 2008;61(8):968-70. <https://doi.org/10.1016/j.bjps.2008.03.001>
3. Lourenço C, Brandão M. Polimastia Axilar: Um Diagnóstico no Puerpério. *Acta Med Port*. 2014;27(2):274.
4. Azoz MEH, Abdalla EA, Elhassan MM. Fibroadenoma in ectopic breast tissue: a case report. *Sudan Med J*. 2014;50(2):112-5. <http://dx.doi.org/10.12816/0015583>
5. Shin SJ, Sheikh FS, Allenby PA, Rosen PP. Invasive secretory (juvenile) carcinoma arising in ectopic breast tissue of the axilla. *Arch Pathol Lab Med*. 2001;125:1372-4. [https://doi.org/10.1043/0003-9985\(2001\)125%3C1372:ISJCAI%3E2.0.CO;2](https://doi.org/10.1043/0003-9985(2001)125%3C1372:ISJCAI%3E2.0.CO;2)
6. Koltuksuz U, Aydin E. Supernumerary breast tissue: a case of pseudomamma on the face. *J of Pediatr Surg*. 1997;32(9):1377-8.

7. Conde DM, Kashimoto E, Torresan RZ, Alvarenga M. Pseudomamma on the foot: an unusual presentation of supernumerary breast tissue. *Dermatology Online J*. 2006;12(4):7.
8. Shreshtha S. Supernumerary Breast on the Back: a Case Report: *Indian J Surg*. 2016;78(2):155-7. <https://dx.doi.org/10.1007%2Fs12262-016-1443-8>
9. Chung-Park M, Liu CZ, Giampoli EJ, Emery JD, Shalodi A. Mucinous adenocarcinoma of ectopic breast tissue of the vulva. *Arch Pathol Lab Med*. 2002;126(10):1216-8. [https://doi.org/10.1043/0003-9985\(2002\)126%3C1216:MAOEBT%3E2.0.CO;2](https://doi.org/10.1043/0003-9985(2002)126%3C1216:MAOEBT%3E2.0.CO;2)
10. Ho SP, Tseng HH, King TM, Chow PC. Anal phyllodes tumor in a male patient: a unique case presentation and literature review. *Diagn Pathol*. 2013;8:49. <https://doi.org/10.1186/1746-1596-8-49>
11. Burdick AE, Thomas KA, Welsh E, Powell J, Elgart GW. Axillary polymastia. *J Am Acad Dermatol*. 2003;49:1154-6. <https://doi.org/10.1016/S0190>
12. Rho JY, Juhng SK, Yoon KJ. Carcinoma originating from aberrant breast tissue of the right upper anterior chest wall: a case report. *J Korean Med Sci*. 2001;16(4):519-21. <https://doi.org/10.3346/jkms.2001.16.4.519>
13. Ayadi-Kaddour A, Khadhar A, Mlika M, Braham E, Ismail O, Zegal D, et al. Fibroadenoma in an ectopic vulvar breast gland: a common neoplasm in an uncommon site. *Pathologica*. 2014;106(1):32-4.
14. Mukhopadhyay M, Saha AK, Sarkar A. Fibroadenoma of the ectopic breast of the axilla. *Indian J Surg*. 2010;72:143-5. <https://doi.org/10.1007/s12262-010-0024-5>
15. Ortiz-Mendoza CM. Axillary ectopic breast tissue fibroadenoma: report of three cases and review of the literature. *Ginecol Obstet Mex*. 2012;80(2):99-103.
16. Shatzel J, Blum A, Khoury T, Milligan J, Skitzki JJ. Gynecomastia-like hyperplasia of axillary ectopic breast tissue in a young female. *Case Rep Pathol*. 2013;2013:634248. <http://dx.doi.org/10.1155/2013/634248>
17. Hayes MM, Konstantinova AM, Kacerovska D, Michal M, Kreuzberg B, Suvova B, et al. Bilateral Gigantomastia, Multiple Synchronous Nodular Pseudoangiomatous Stromal Hyperplasia Involving Breast and Bilateral Axillary Accessory Breast Tissue, and Perianal Mammary-Type Hamartoma of Anogenital Mammary-Like Glands: A Case Report. *Am J Dermatopathol*. 2016;38(5):374-83. <https://doi.org/10.1097/DAD.0000000000000498>
18. Grimshaw EC, Cohen PR. Supernumerary nipple and seminoma: case report and review of polythelia and genitourinary cancers. *Dermatol Online J*. 2013;19:4.
19. Kokavec R, Macúch J, Fedeles J, Ondriás F. Polythelia is not a mere aesthetic issue. *Acta Chirurgiae Plasticae*. 2002;44:3-6.
20. Galli-Tsinopoulou A, Krohn C, Schmidt H. Familial polythelia over three generations with polymastia in the youngest girl. *Eur J Pediatr*. 2001;160:375-7.
21. Cohen PR, Kurzrock R. Miscellaneous genodermatoses: Beckwith-Wiedemann syndrome, Birt-Hogg-Dube syndrome, familial atypical multiple mole melanoma syndrome, hereditary tylosis, incontinentia pigmenti, and supernumerary nipples. *Dermatol Clin*. 1995;13:211-29.
22. Leung AKC, Robson WLM. Polythelia. *Int J Dermatol*. 1989;28:429-33.
23. Farcy DA, Rabinowitz D, Frank M. Ectopic glandular breast tissue in a lactating young woman. *J Emerg Med*. 2011 Dec;41(6):627-9. <https://doi.org/10.1016/j.jemermed.2010.11.033>