

Polythelia in a 13-year old girl

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SUMMARY: **Polythelia in a 13-year old girl.**

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We present a rare case of a 13-year old girl with a bilateral polythelia. We would like to draw attention to this particular mammary malformation.

KEY WORDS: Polythelia - Supernumerary nipples.

Introduction

The mammary gland, considered a highly modified sweat gland, is developed from epidermal thickening, normally along the path of the mammary line, milk line, extending from the axilla to the inguinal region. The milk line degenerates shortly after the development except for a small portion that persists in the pectoral region, where a single pair of glands usually develop and which later become the breasts (1-3).

The presences of supernumerary nipples, *polythelia*, or supernumerary breast, *polymastia*, are commonly developed along the milk lines, and can be either uni- or bilateral. Whereas polythelia is evident at birth, polymastia is evident after hormonal stimulation that occurs at puberty or during pregnancy. The incidence of polythelia varies between 0,2% and 5,6% in the general population, depending on factors such as sex, inheritance, ethnicity and geographical area. Polythelia is usually seen more often in males than females and more commonly occur in black than whites (3-6, 8). Polythelia may be

associated with other congenital disease, most notably urologic malformations or urogenital malignancies (1-3, 5, 7).

Case report

The patient, a 13-year old African female, presented herself with cosmetic complaints due to supernumerary nipples. Our patient informed that she was born with the extra nipples and had not before been bothered by these, no symptoms of pain or swelling. Three to four months before, she had started getting her menstruation and her breast had evolved. As the breast evolved, two of the extra nipples grew until she felt uncomfortable showing to others. There was no information on other family members with the same condition. No information on family history of physical anomalies, malignancy, cardiovascular or renal disease.

During the examination we found two, one on each side, approximately 8-10 cm above the right and the left nipple, 15x20 mm accessory nipples. Two other accessory nipples were found near the armpit on both sides, with the size of 1-2 mm. The one accessory nipple in the right armpit was a bit more visible than the one in the left armpit. All of the accessory nipples were found along the milk line. There was no sign of underlying neither swelling nor palpation pain (Figure 1 a, b, c).

There were no biochemical abnormalities. The patient was a healthy child, and the ultrasound showed no further anomalies.

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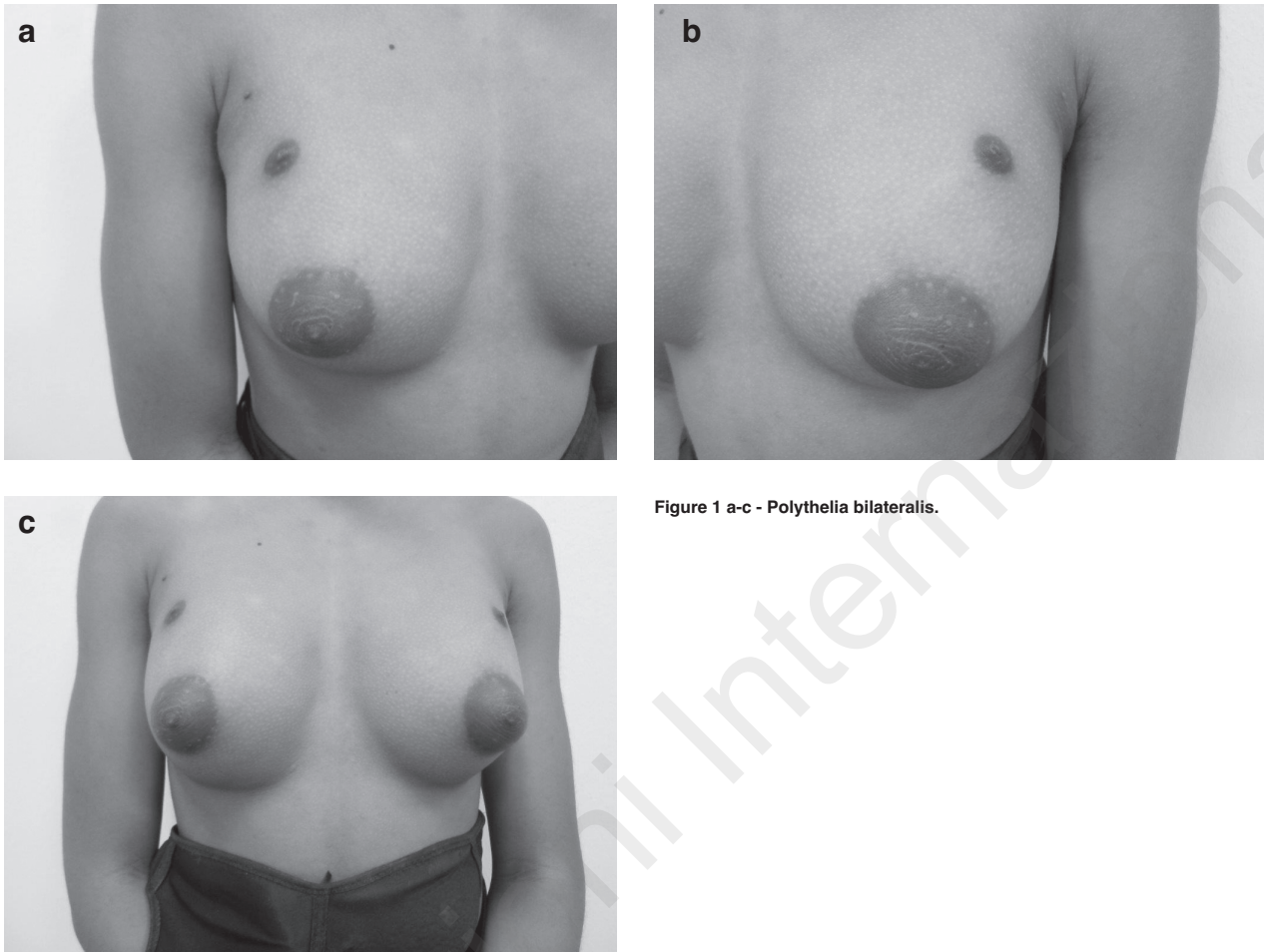


Figure 1 a-c - Polythelia bilateralis.

Surgical treatment was performed, removing the bigger accessory nipple on the chest and the smaller accessory nipple in the armpit, one side at the time, starting with those on the right side. We decided to operate in two steps with the intention to evaluate the cosmetic result because the patient due to her ethnicity was with a high risk of developing keloids. The specimen was sent to the pathologist for further investigation (Figure 2 a, b).

Gross pathology

Skin specimen with the diameter of 21 and 7 mm. Both with a small papilla in the middle (Figure 3).

Discussion

Polythelia are one of the most common forms of mammary tissue malformation. Most of the cases of polythelia appear to be sporadic, but some hereditary

forms are known (1, 2). The specific mutations responsible for the sporadic or the hereditary forms of polythelia are generally not known. Some suggest the genetic transmission to be a heterogeneous, autosomal dominant with incomplete penetrance and a dominant X-linked chromosome (7). One rare genetic disorder associated with accessory nipples is the Simpson-Golabi-Behmel syndrome, and this syndrome is caused by and an X-linked gene (2, 7).

The first time this malformation was described in a medical report was in 1878 by Leichtenstern (9). But the most known classification was presented by Kajava in 1915 (10):

- Complete supernumerary nipple - Nipple and areola and glandular breast tissue
- Supernumerary nipple - Nipple and glandular tissue (no areola)
- Supernumerary nipple - Areola and glandular tissue (no nipple)
- Aberrant glandular tissue only
- Supernumerary nipple - Nipple and areola and



Figure 2 a - Anterior photo: 3-month postoperative with an acceptable non-keloid scar-formation, as a concentric resulting line, no deformation or dysconfiguration of the breast shape.



Figure 2 b - Oblique photo: 3-month postoperative with an acceptable non-keloid scar-formation, as a concentric resulting line, no deformation nor dysconfiguration of the breast shape.

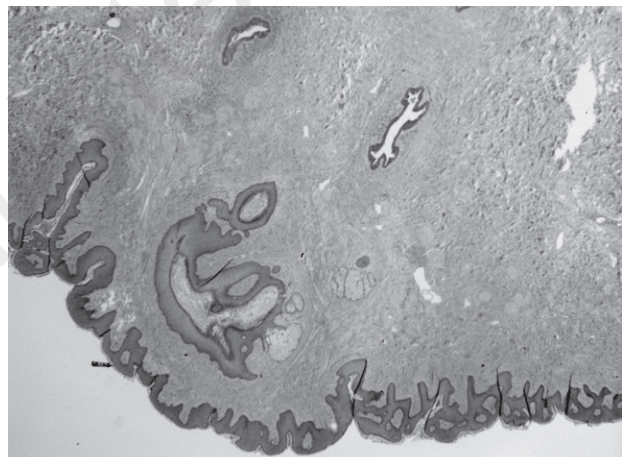
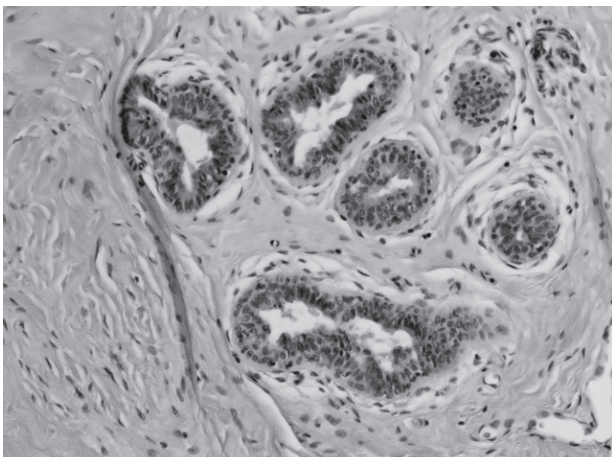


Figure 3 - Histopathology showed ducts within stroma in dermis with normal overlying epidermis, confirming the diagnosis of accessory nipples.

pseudomamma (fat tissue that replaces the glandular tissue)

- Supernumerary nipple - Nipple only (the most common supernumerary nipple)
- Supernumerary nipple - Areola only (polythelia areolaris)
- Patch of hair only (polythelia pilosa).

Several studies reported that the incidence of Polythelia is quite similar in males and females.

The association between polythelia and other anomalies has been investigated. Most frequently the authors indicate an association between polythelia and urologic/urogenital malformation or malignancies, that occurs in 23-27% of the supernumerary nipples cases (11, 12).

Other congenital disease such as cardiac malformation, vertebral malformation, central nervous system and dental anomalies has also been reported (1, 3, 5). It is

not well reported if the sporadic form of polythelia is more associated with malformation than the hereditary one. The numbers or the site of the accessory nipples may also be a predictor for the possibility of associated malformations.

It has earlier been noted that the frequency of supernumerary nipples in black Americans is almost 7,4-fold greater than in white Europeans. However, the association of the wide range of anomalies reported in whites with polythelia has so far not been reported experienced in black Americans (1, 6, 8).

The lack of reports of associated malformation in black people may be due to the difficulty of spotting small accessory nipples on dark skin, or from some cultural point of views, extra nipple may be a sign of better fertility and therefore the patient does not see doctors. Our patient was not examined for anomalies, this maybe because of the rarity and lack of knowledge of a possible asso-

ciation of polythelia and other congenital disease. In the routine physical examination of every newborn, it may be a suggestion to search for accessory nipples due to the possibility of association with malformation that may require other interpretations or treatment. It might be an idea to prepare guidelines/algorithms on how to best handle newborn with supernumerary nipples.

Conflicts of interest statement
None.

Ethical approval statement

We hereby state that written consent has been obtained from the patient.

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