Polythelia: Anatomic and clinical implications.

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Abstract

Background: Polythelia or supernumerary nipple (also called third or accessory nipple) is an additional nipple occurring in mammals including humans. These additional nipples develop during embryonic life as part of abnormal development of mammary glands.

Objectives: To describe the polythelia and it's most frequent locations also to give a perspective of polythelia in a series of observations. And lastly to asses if there is any genetic inheritance present.

Methods: Forty three cases of polythelia were collected from attendants of general practice clinic in Baghdad. The polythelia was observed during routine physical examination, which included examination of the chest and abdomen**.**

Results: From 43 cases of polythelia, males constituted 23 (53.5%) of cases. Regarding the anatomical location of polythelia, 2(4.65%) were on the anterior axillary fold, 28(65.1%) on the anterior thoracic wall, 12(27.9%) on the anterior abdominal wall and one (2.3%) was in the inguinal region. Only five cases (11.6%) had family history of previous similar conditions.

Conclusion: Polythelia is a fairly common abnormality. Men and women may have extra nipple, but no significant difference was detected that can be related to gender difference. Nevertheless presence of extra nipples was sometimes linked to heart disease, no such relationship was noticed. All cases in this study had their polythelia along the milk line. Nevertheless, there had been reports on polythelia presenting as far away as the foot.

Keywords: polythelia, mammary gland, Supernumerary nipple.

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Introduction

Polythelia or supernumerary nipple (also called third or accessory nipple) is an additional nipple occurring in mammals including humans. These additional nipples develop during embryonic life as part of abnormal development of mammary glands [(1)].

The first indication of mammary glands is found in the form of a band like thickening of epidermis along the mammary line or mammary ridge. In a 7-week embryo this line extends on each side of the body from the base of the forelimb to the region of the hind limb. Although the major part of the mammary line disappears shortly after it forms, a small portion in the thoracic region persists and penetrates the underlying mesenchym. Here it forms 16 to 24 sprouts, which in turn give rise to small, solid buds. By the end of the prenatal life, the epithelial sprouts are canalized and form the lactiferous ducts, and the buds form small ducts and alveoli of the gland. Initially the lactiferous ducts open into a small epithelial pit. Shortly after birth, this pit is transformed into the nipple by proliferation of underlying mesenchyme [(2)]**.**

Polythelia refers to the presence of an additional nipple alone while polymastia denotes the presence of additional mammary gland. Polythelia often looks like moles or freckles and do not always have a connection with breast tissue or milk ducts [(3)].

Polythelia is classified into eight levels of completeness from a simple patch of hair to a milk bearing breast. This study will investigate the condition of polythelia as regards the anatomical location of the additional nipples, the presence of family history

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of such condition in the first and second degree relatives, gender differences and the possible relation to other diseases[4] .

Material and Methods

This study was performed on 43 cases with polythelia. They were collected from attendants of general practice clinic at Baghdad. The polythelia was observed during routine physical examination, which included examination of the chest and abdomen. Once observed, detailed information were recorded about the condition in a study form which includes:

- Personal data of name and residence
- Age and gender
- Physical examination of the condition, included inspection, palpation and fluctuation test for presence of fat. Data regarding presence of glandular tissue, nipple, pigmented areola, fatty tissue and presence of hair.
- The anatomical position was recorded as being on anterior axillary fold, thoracic wall, anterior abdominal wall and the inguinal region (groin).
- Presence of similar condition in family, including first and second degree relatives.
- Presence of other notable systemic diseases.

Results

The total number of cases enrolled in this study was 43. The age was very variable; however most of observed cases were in 3rd or 4th decade of life. Males constituted 23(53.5%) cases and females were 20(46.5%).

Regarding the anatomical location of polythelia, 2(4.65%) were on the anterior axillary fold, 28(65.1%) on the anterior thoracic wall, 12(27.9%) on the anterior abdominal wall and one (2.3%) was in the inguinal region (Table 1).

Regarding the family history of such condition in first degree relatives (parents and offspring) and second degree relatives (brothers and sisters) only five (11.6%) had such family history.

No notable systemic diseases were recognized in these cases who were seeking medical consultation for no major systemic disease.

Careful inspection was done for the condition and palpation was performed to detect underlying glandular tissue. Two fingers fluctuation test was done to identify fatty material if a lumpy structure was present.

Table 1: Sites of polythelia (supernumerary nipples) in mammals including humans

Total number (43)		**Sites of polythelia**			
Gender		**AAW**	**AAF**	**ACW**	**IR**
male	23 (53.5%)				
female	20 (46.5%)	12 (27.9%)	2 (4.65%)	28 (65.1%)	1 (2.3%)

The abbreviations: AAW: Anterior abdominal wall, **AAF**: Anterior axillary fold, **ACW**: Anterior chest wall and **IR**: Inguinal region.

Discussion

The aim of this study was to give a perspective of the condition of additional nipple or polythelia in a series of observation. According to our observations, polythelia is a fairly common abnormality. It is proposed that this type of congenital abnormality occurs at rate of 1 in 18 human [5].

Larger study is required to investigate the prevalence of this

condition since many mistake it as mole, navus, freckle or pigmented skin condition. Men and women may have extra nipple. In this work no significant difference was detected that can be related to gender difference.

All the cases included in this study had no glandular tissue, so they were not polymastia. The observed additional nipples can be designated as pseudo mamma when they possess a nipple, areola and fat tissue.

They are considered classical polythelia when they are just a nipple. Polythelia areolaris, when only an areola is present and polythelia pilosa when it consists of a conglomerated patch of hair [(6)].

From the anatomical point of view, the most frequent location of polythelia is the thorax. Commonly an additional nipple was found to be present inferior and medial to the location of the genuine nipple.

The second most common site was on the anterior abdominal wall. Other sites observed were along the anterior axillary fold and above medial inguinal region.

All cases in this study had their polythelia along the milk line. Nevertheless, there had been reports on polythelia presenting as far away as the foot [(7)].

The development of breast is under genetic control, and the gene coding for this process is called Scaramanga gene. This gene is responsible for the expression of a protein called Neuregulin-3 (NRG3) which provides a signal to embryonic cells to differentiate into mammary cells [(8)]. Although polythelia tends to occur sometimes in families [(9)], it is more likely to develop at random. Only five cases with familial background in the form of the presence of polythelia was found in first and second degree relatives, in the whole study group. This finding does not support a genetic inheritance or predisposition to this condition.

In this study no concomitant pathological conditions with polythelia were found and unable to link it to other disease entities. Nevertheless presence of extra nipples was sometimes linked to heart disease, and a possible relationship with mitral valve prolapse had been proposed [(10)], no such relationship was noticed in the study group.

However in general any breast tissue, whether it appears in the standard location or elsewhere is vulnerable to the same diseases that can affect typical breast tissue. Extra mammary Paget's disease of the nipple can affect these additional nipples [(6)].

In most people, extra nipples are benign and may never be noticed. But if they change, develope a lump, rash or discharge, they should be taken seriously, otherwise polythelia may be surgically removed, just like a mole.

In western folklore, an extra nipple was held to be indicative that the women concerned was a witch, the nipple used to suckle the devil!

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