Actas Dermosifiliogr. 2010;**101(5)**:453–467

ACTAS
Dermo-Sifiliográficas

Full English text available at
www.elsevier.es/ad

CASE AND RESEARCH LETTERS

Bilateral Familial Polythelia Without Associated Malformations

Politelia bilateral familiar sin malformaciones asociadas

To the Editor:

The presence of supernumerary nipples not associated with other anatomical breast structures (polythelia) is the most common malformation of accessory mammary tissue.[1] As well as the nipples that normally develop on the thorax, cases have also been reported of additional nipples, areolas, mammary glandular tissue (polymastia), or any combination of these.[2-4] The supernumerary elements are normally found along the mammary lines—imaginary lines running from the axillas to the groin.[5] Most cases of polythelia are sporadic, although several heredity patterns have been reported.[6,7] Furthermore, possible associations have been sought between polythelia and different congenital malformations, particularly malformations of the kidneys and the urinary tract.[4,8-10] However, reports of familial bilateral supernumerary nipples at sites other than the mammary lines, with no underlying mammary tissue, and not associated with congenital malformations, are very rare in the literature.[11]

We report the case of a 53-year-old woman who was allergic to sulfamides, had never been pregnant, and who had been diagnosed with well-differentiated intraductal carcinoma (ductal intraepithelial neoplasia stage 1c) of the left breast in 2002; the carcinoma was treated by lumpectomy, local radiation therapy, and oral tamoxifen. The woman was referred to the dermatology department due to radiodermatitis in the irradiated area. As well as the characteristic signs of radiodermatitis, the physical examination revealed 2 fully developed breasts with their respective nipples and areolae, accompanied by 2 clearly defined slightly hyperpigmented papular lesions with a firm consistency, corresponding to supernumerary nipples located near the axillas (Figures 1 and 2), and another 2 located below the breasts; none of these supernumerary nipples was accompanied by an areola and there was no palpable underlying glandular tissue. The patient stated that she had had these supernumerary elements since birth and that they had not developed during puberty. They were stable and asymptomatic. She did not remember having had episodes of secretion or experiencing changes during the menstrual cycle. Furthermore, the patient presented no facial, cranial, palatine, ocular, or limb malformations, and no systemic symptoms. When questioned regarding her family history, the patient stated that other members of her family had bilateral supernumerary nipples: mother (I:1), 6 sisters (II:1, II:2, II:5, II:7, II:8, and II:10), and a niece (III:1) (see family tree in Figure 3). She was unaware of supernumerary nipples in the grandparents, uncles and aunts, and other nieces and nephews.

Computed tomography of the cervical, thoracic, abdominal, and pelvic regions was performed, together with an ultrasound scan of the areas on which the nipples were located; no underlying mammary tissue or associated malformations were observed. A chest x-ray, skeletal x-rays including the long bones and lumbar-sacral spine, and abdominal and gynecological ultrasound scans were also performed with negative results.

The terms polythelia and polymastia are used to refer to the presence of multiple nipples and mammary glands, respectively, and are considered to be examples of atavism.[1] They form part of a broad clinical spectrum, depending on which of the different anatomical structures are present: breast, nipple, areola, glandular parenchyma, or a combination.[4] Nevertheless, polythelia is much more common than polymastia and is therefore considered to be the more frequent malformation of accessory mammary tissue; the incidence varies between 0.22% and 5.6% of the general population, depending on different factors (sex, ethnic group, and geographic region).[12] Most cases of polythelia are sporadic, although as many as 4 forms of genetic transmission have been described[6,7,13-15]: autosomal dominant with incomplete penetrance, X-linked dominant, autosomal recessive, and paradominant. The supernumerary nipples are most commonly found on the upper third of the mammary line, which is an imaginary line running from the axilla to the groin.[3] Bilateral polythelia has been reported but is less common that the unilateral presentation.[4,5,12]

Supernumerary nipples are mainly an esthetic problem of little medical significance. However, some cases of polythelia respond to hormonal changes in the same way as normal mammary tissue and are susceptible to diseases such as mastitis, cysts, abscesses, and cancer.[4] Also reported in the literature is a considerable number of associations between polythelia and an increased risk of systemic diseases including cardiac arrhythmias, pyloric stenosis, epilepsy,[2] Char syndrome,[16] Hailey-Hailey disease,[7] and different types of cancer (testicular, prostate, vesical,[9] and renal cancer[17]). However, most of the associations reported refer to the relationship between polythelia and

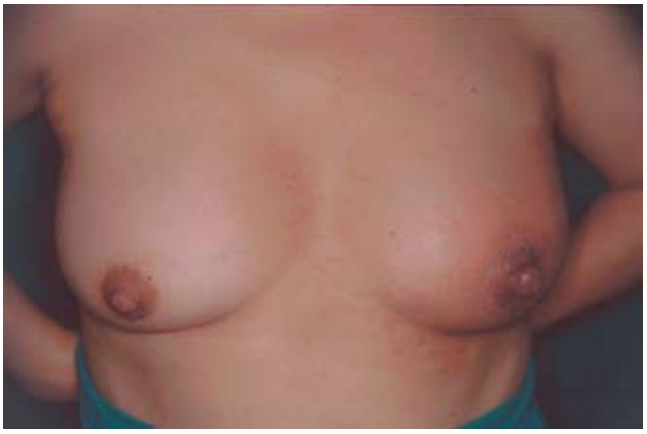

Figure 1 Bilateral polythelia near the axillas. Radiodermatitis in the left breast.

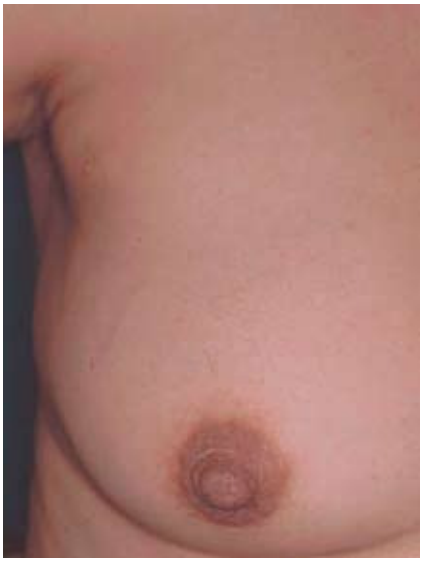

Figure 2 Detail of Figure 1.

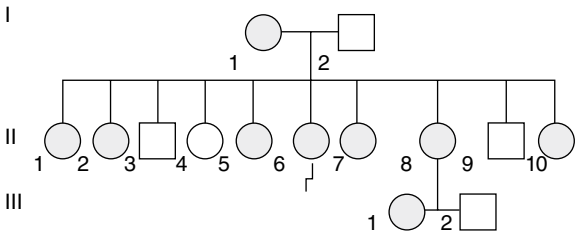

Figure 3 Family tree of the family being studied. The patient in the case description is II:6.

congenital renal and urinary tract malformations, such as adult autosomal dominant polycystic disease, unilateral renal agenesis, ectopic kidney, cystic renal dysplasia, and congenital dysplasia of the pelviureteric junction.[4,6,8-10]

The case of our patient is notable for the presence of bilateral familial supernumerary nipples at a distance from the typical location and apparently with no associated systemic disease or congenital malformation. Nevertheless, when polythelia is detected, a detailed family history should be taken, a careful physical examination should be performed, and a complete investigation should be undertaken using imaging studies to detect possible associated malformations and diseases.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

References

1. 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.
2. Iwai T. A statistical study on the polymastia of the Japanese. Lancet. 1907;2:753-9.
3. Cellini A., Offidani A. Familial supernumerary nipples and breasts. Dermatology. 1992;185:56-8.
4. Brown J, Schwartz RA. Supernumerary nipples and renal malformations: a family study. J Cutan Med Surg. 2004;8:170-2.
5. McLaughlin MR, O'Connor NR, Ham P. Newborn skin: Part II. Birthmarks. Am Fam Physician. 2008;77:56-60.
6. Toumbis-Ioannou E, Cohen PR. Familial polythelia. J Am Acad Dermatol. 1994; 30:667-8.
7. Benmously-Mlika R, Deghais S, Bchetnia M, Charfeddine C, Mokni M, Kassar S, et al. Supernumerary nipples in association with Hailey-Hailey disease in a Tunisian family. Dermatol Online J. 2008;14:15.
8. Leung AKC, Robson WLM. Polythelia. Int J Dermatol. 1989; 28:429-33.
9. Urbani CE. The significance of familial polythelia. J Am Acad Dermatol. 1995;32:687-8.
10. Urbani CE, Betti R. Accessory mammary tissue associated with congenital and hereditary nephrourinary malformations. Int J Dermatol. 1996;35:349-52.
11. Casey HD, Chasan PE, Chick LR. Familial polythelia without associated anomalies. Ann Plast Surg. 1996;36:101-4.
12. Schmidt H. Supernumerary nipples: prevalence, size, sex, and side predilection- a prospective clinical study. Eur J Pediatr. 1998;157:821-3.
13. Klinkerfuss G.H. Four generations of polymastia. JAMA. 1924;82:1247-8.
14. Leung AKC. Familial supranumerary nipples. Am J Med Genet. 1988;31:631-5.
15. Bortz J, Parker S, Ray TL. Lack of associated anomalies in familial polythelia. Am J Dis Chile. 1989;143:883.
16. Zannolli R, Mostardini R, Matera M, Pucci L, Gelb BD, Morgese G. Char syndrome: an additional family with polythelia, a new finding. Am J Med Genet. 2000;95:201-3.
17. Mehes K. Familial association of supernumerary nipple with renal cancer. Cancer Genet Cytogenet. 1996;86:129-30.

A.M. Castaño-León,* C. Eguren, and E. Daudén

Servicio de Dermatología, Hospital Universitario de la Princesa, Madrid, Spain

*Corresponding author.
E-mail address: alkimia86@hotmail.com (A.M. Castaño-León)