Sporadic Unilateral Intra-areolar Polytelia
Report of an Additional Case and Review of the Literature

Sir,
Supernumerary nipple, or polytelia, is a minor cutaneous developmental anomaly, usually located along the embryonic mammary mammary lines, which extends longitudinally from axillae to inguinal folds; infrequently it also occurs in ectopic sites. Although the incidence of supernumerary nipples is fairly common, being recorded as 1–2%, their occurrence within the areola is an extremely rare condition. To the best of our knowledge only 9 cases, all having a bilateral pattern, of intra-areolar polytelia (IAP), or “paired nipples” or “dysplastic divided nipples”, have been published in the English literature (1). Family history has been observed in only one report (1). We describe an additional patient affected with unilateral IAP, pointing out its embryogenetic mechanisms.

CASE REPORT

A 32-year-old Caucasian man was routinely examined for clinical evaluation of a congenital "nipple-like" protuberance located within the left areola (Fig. 1). The breasts and their nipple-areolar complexes were morphologically normal, without any evidence of mammary hypoplasia. No lateral displacement of the nipples was noted. Both major pectoral muscles were well developed. His general health was good and there was no other personal or family history of relevance.

Occurrence of supernumerary nipple or any other forms of accessory mammary tissue was ruled out both in the patient and in his relatives (parents and a daughter). The lesion, soft in consistency and asymptomatic, was surgically treated because of aesthetic discomfort and a mild psychologic embarrassment claimed by the patient. He had no family history of such a disease.

The histopathological picture confirmed the clinical suspicion of polytelia. It was characterized by mild hyperkeratosis and acanthosis, vacuolar degeneration throughout the epidermis and slight basal pigmentation. Smooth muscle bundles were randomly interfaced in the reticular dermis, and a number of mammary ducts, embedded in a hyalinized fibrous connective tissue stroma, were seated in mid-dermis.

Complete ultrasound examination of the abdomen, routinely performed in patients affected with accessory mammary tissue (2), excluded the occurrence of both malformations and malignancies of the kidney and urinary collecting system.

COMMENT

The embryological background of IAP is different from that of polytelia. Embryogenetically, we may define polytelia as a "quantitative" anomaly and IAP as a "qualitative" defect. In fact, polytelia in its "usual" form is due to the persistence of the redundant accumulations - or anlagen - within the ectodermal longitudinal thickenings - the mammary lines - which normally regress during the third month of embryonic development. IAP, on the contrary, is due to an intrauterine dichotomy of the developing nipple (3) or to an incomplete development, or disruption, of the epithelial pit, or mammillary anlage (1).

REFERENCES


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Fig. 1. Clinical picture of intra-areolar polytelia. A well-structured nipple without any dysmorphic changes or structural anomalies is located within a morphologically normal left areola. No breast or major pectoral muscle hypoplasia is present.