An Aged Man With One Big Chest Lesion and a Scattering of Smaller Ones

What's Your Diagnosis? | August 03, 2009
Unilateral polythelia is suggested by a sequence of supernumerary nipples that follow the embryonic milk line. No areolae are associated with these accessory nipples.

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HISTORY

A 78-year-old man is seen upon admission to a behavioral hospital unit. Agitation and visual hallucinations led to this admission. A similar prior episode, complicating his established Lewy body dementia, cleared with hospitalization and clozapine therapy.

PHYSICAL EXAMINATION

Mildly obese man who is conversationally capable and friendly, though dementia is also evident. Vital signs normal. Has a large central sternal seborrheic keratosis. Skin about right breast as shown. Murmur consistent with aortic stenosis. Ventral hernia.

What's Your Diagnosis?
(answer on next page)
WHAT'S YOUR DIAGNOSIS?
ANSWER: POLYTHELIA (accessory nipples)
The right breast is slightly pendulous, consistent with his overweight. It bears a normal areola and nipple and an ordinary amount of hair. Some 12 cm above the nipple lies a round papule with a granular or stippled surface and the same pigmentation as the nipple; another such structure is found immediately inferior to the normal nipple. The locale of all 3 structures in the embryonic milk line confirms our impression that these are supernumerary nipples constituting unilateral polythelia. There are no areolae about the accessory nipples. They display no elevation that could suggest any element of associated accessory breast tissue (polymastia).

The striking resemblance between the “offspring” and the “parent” structure is emphasized on closer examination (Figure 1), which also discloses tiny papules, keratoses, and nevi on nearby skin, none of which resemble the nipples. The red dot on the areola at 4 o’clock is a minute capillary hemangioma.

Medial to the areola (Figure 2), a pigmented lesion appears very dark brown and might be melanocytic, or might represent a comedo or even a bit of clotted blood atop a traumatized lesion of any type. Concern that this might be a dysplastic nevus prompted our consulting a dermatologist. The left side of the trunk showed no polythelia (Figure 3), but this survey view reveals the large icosahedral, minimally pigmented lesion that both distracted us and struck us as a typical seborrheic keratosis. The face shows heavy wrinkling from solar injury that one would expect in a person who had farmed apples for decades (Figure 4).
WHAT ELSE MIGHT IT BE?
Nothing else perfectly mimics an accessory nipple. Given the grainy surface, one could think about a conventional verruca vulgaris (wart), but these often have small filiform protuberances and less
regular papillae. A seborrheic keratosis often shows more pigmentation and has the familiar characteristics of looking stuck on to the skin surface, feeling greasy, and giving the impression that one could pick it off the skin—an impression furthered by a tendency to partial stalk formation or at least a mushrooming on the outer surface, features distinctly lacking here. Verrucous carcinoma would not be diagnosable at this tiny size unless it ulcerated deeply or produced some other atypical pattern of growth. Having said this, we often rightly dismiss many skin lesions of this size that bear no features suggesting possible malignancy. So the significance of the polythelia lies not with potential for harm per se, but with possible implications of internal disease.

**WHY IS THIS MORE THAN A CURIOSITY?**

Accessory breasts and polythelia have commanded extraordinary attention since antiquity, sometimes being interpreted as a sign of augmented fertility. For instance, the Phoenician goddess Astarte and her Greek descendants, including Artemis, are often depicted as having polymastia.
Polymastia and polythelia were regarded at other times and places as marks of the Devil. In one extraordinary rumor, Anne Boleyn was said to have a third breast over midsternum. Of course, this could have been an element of false evidence adduced by King Henry's minions to justify the royally sanctioned murder. The broad historical review of accessory breast tissue by Grossli\textsuperscript{2} offers further instances.

Cosmetic considerations sometimes lead to surgical interventions,\textsuperscript{4} but this patient expresses no such concerns nor does his wife, who is highly articulate. One paper in the literature entitled “Polythelia Is Not a Mere Aesthetic Issue”\textsuperscript{5} fails to prove the title assertion. Other authors give more compelling evidence.
Supernumerary breasts can be the site of any of the diseases of ordinary breasts, including carcinoma. Since pure (non-Paget) cancer of the nipple occurs with such rarity, the cancer concern with pure polythelia is not local but distant.

**INTERNAL DISORDERS**

Numerous reports detail an association with renal and ureteral abnormalities, including renal cell carcinoma. The numbers of associated nephrourinary abnormalities vary widely, and non-neoplastic lesions predominate: unilateral renal agenesis, reduplication of collecting systems, and unilateral or bilateral stenosis of the ureteropelvic junction. Noninvasive assessment for these problems has become considerably safer since the ultrasound scan of the abdomen and pelvis replaced most uses of the excretory urogram (often called the intravenous pyelogram, or IVP). For children and younger adults in whom polythelia is detected, this follow-up is recommended. In the total context of our patient’s care and preferences, we exhibited diagnostic restraint and did not order the study. We discussed the association with the primary care physician so that our decision could readily be revisited and reversed if the parties so chose. It is not yet clear to what degree polythelia is a marker for internal disease in adults and even in pediatric patients; nor what difference it makes if the polythelia is familial versus sporadic. At least one study includes nocturnal enuresis and urinary incontinence without structural anomaly among the “renal-urinary abnormalities” group, thereby clouding the conclusion that polythelia is more common in children with urinary tract disorders than in those without. We don’t know if the proposed association with fetal alcohol syndrome is established. However, we note inclusion of polythelia among one respected listing of genodermatoses with malignant potential.

**SECONDARY ISSUES**

A great many other associations with polythelia have been proposed, but many fail to withstand scrutiny: eg, hypertension, with its very high background rate. One could postulate that the associated renal abnormalities themselves cause hypertension; however, many of the associated urinary tract disorders do not (eg, renal cancers, solitary kidney, reduplicated collecting systems). The pigmented lesion medial to the lower of the two accessory nipples was seen by the dermatologist, but his index of suspicion was low enough that he recommended only serial clinical observation.

Finally, we present this case because it yet again exemplifies the principle that the largest or most striking lesion need not be the most important: the tiny pigmented lesion mattered more than the giant seborrheic keratosis, and if we had chosen to pursue the associations of polythelia, so would these small aberrations in the skin.


**References:**

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