Intraareolar polythelia: a rare anomaly

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Abstract

Polythelia, the presence of supernumerary nipples or nipple-areola complexes, is the most common anomaly of the pediatric breast. Although polythelia is common, intraareolar polythelia (nipple dichotomy) is an extremely rare congenital malformation. Surgical treatment is performed as a prophylaxis against breast cancer which has a higher prevalence in polythelia or polymastia. We describe a case of a young woman with polythelia in the right breast. The patient was a 25-year-old young woman who had two nipples by birth on the areola of the right breast. An abdominal ultrasound examination showed no urorenal malformations. Surgery was performed with elliptical excision under local anesthesia. A management of such cases with polythelia or polymastia was discussed in the view of recent literature.

Keywords: Intraareolar polythelia, Supernumerary nipples, Nipple malformation.

Introduction

Polythelia, the presence of supernumerary nipples or nipple-areola complexes, is the most common anomaly of the pediatric breast. The frequency of polythelia ranges from 0.22% to 5.6% in the general population.[1] Although polythelia is common, intraareolar polythelia is an extremely rare congenital malformation.[2,3] Rarely, cases with polythelia have been described in the medical literature. Polythelia is generally asymptomatic, but may sometimes be associated with urological malformations. Excision is indicated for diagnostic, treatment or cosmetic reasons.[4] We report a case of a young woman who had two nipples by birth on the areola of the right breast.

Case Presentation

A 25-year-old young woman who had two nipples by birth on the areola of the right breast. She desired surgical correction of the nipples for aesthetic and psychological reasons. Both the nipples were similar in size and appeared normal (Figure-1). The size of the right breast and its areola was normal and similar to the left breast. The pectoralis major muscles were bilaterally present and they were normally developed. She had no family history of similar anomalies. No history of breast cancer in her first degree relatives or personal history of breast pathology was present. No regional adenopathy, in the axillary and supraclavicular basins, was detected. Her menarche was at age 14. No hormonal treatment was conducted in the past. Her general health was good and she had normal secondary sexual characteristics. She did not have any other supernumerary nipples along the 'milk line' or elsewhere. An abdominal ultrasound examination showed no urorenal malformations. Surgery was performed with elliptical excision under local anaesthesia after the baseline haematologic test. Specimen for histopathologic study was sent which revealed only normal nipple tissue and diagnosis of an accessory nipple was made. Follow-up visits at 4 and 12 months after the procedure showed a satisfactory result.

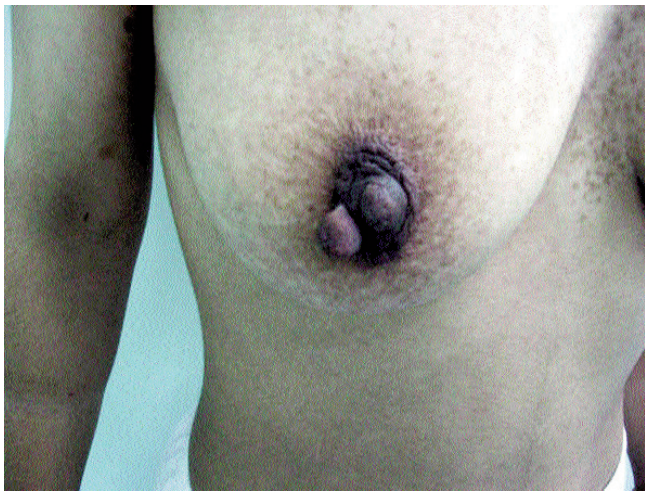

Figure: View of right breast with two nipple.

Discussion

Intraareolar polythelia is a congenital malformation characterized with one or more supernumerary nipples within the areola. It must be classified as different from supernumerary nipples or polythelia, in which the nipples

are along the milk line from the axillae to the inguinal fold.[4] During the development of embryo, the mammary glands develop from the mammary line or ridge extending from the axillary region on either side of the body down to the region of the medial extent of the inguinal ligament. Normally in homo-sapiens, most of the ridge disappears shortly after its formation and leaves a small portion in the thoracic area that develops ultimately into normal human breasts. Yet, other small areas of this ridge may sometimes remain, leading to an increase in polythelia or accessory nipples. The remnant may also evolve into a complete mammary gland resulting in the condition which is known as polymastia. These glands can develop everywhere along the mammary line.[5]

It is believed that polythelia results from the persistence of ectodermal ridges along the milk lines which normally regress during the third month of intrauterine development. In intra-areolar polythelia the embryologic origin is a dichotomy of the nipple that develops during foetal life.[6,7]

Intra-areolar polythelia is usually bilateral in the sporadic cases explained in the literature, and these cases are often associated with hypoplastic breast.[8] A case of intraareolar polythelia has also been reported in a male.[9] In our case, the patient has not reported any familial or personal history related to other congenital malformations. Polythelia is generally recognized at birth, although it may initially be confused with nevi or other skin lesion.[10] Polymastia may not be evident until the influence of sex hormones during puberty. The presence of supernumerary nipples has been associated with the occurrence of genitourinary anomalies.[11] Surgery is currently suggested in cases of suspected malignancy, in symptomatic cases and for cosmetic problems. No detailed surgical technique has been reported for correction of this rare malformation. For cosmosis, supernumerary nipples may be removed by excision along the lines of Langer.[12] Surgical correction may be performed by transposition flaps sutured to one another in the center of the areola.[2] Supernumerary nipples should be excised before puberty because after the onset of puberty in girls, resection may require wider tissue excision as a result of glandular growth.[10]

Although we did not demonstrate any renal anomalies in this report on polythelia, we feel that a thorough physical examination, urine analysis, and renal ultrasound should be performed in any patient with polythelia. Surgical treatment is performed as a prophylaxis against breast cancer, which has higher prevalence in polythelia or polymastia.

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