Ectopic mammary tissue in vulva

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Abstract

Background. Ectopic mammary gland tissue is a residual tissue that persists during the embryologic development along ectodermal primitive milk streaks. Incomplete involuc- tion anywhere along the primitive milk streak can result in accessory or ectopic mammary tissue. Case report. A woman, 27-year old, admitted to Obstetrics and Gynecology Clinic Kragujevac for surgery, of goose-egg size, vulva tu- mor, of elastic consistency. Menarche started in 12 years of age, with the regular menstrual cycle, without previous gyn- ecological diseases. The woman had one pregnancy termi- nated by cesarean section because of the multiple (twin) pregnancy. Excision of the tumor was completely done in the total endotracheal anesthesia. Pathohistologic (PH) findings was: Dysplasia fibrosa cystica simplex mammae, with fo- cuses of sclerosing adenosis. Expression of estrogen (ER) and progesterone receptors (PR) were positive. Conclusion. Ectopic mammary tissue in vulva in adult period is very rarely seen, and can be changed pathologically as well as normally positioned breast tissue into benign cystic changes, benign tumors, adenomas and fibroadenomas and tumors. Cells with low ER/PR receptor level grow independently of estrogen stimulation and they could be resis- tant to hormonal therapy effects.

Key words: breast; vulvar neoplasms; choristoma; histology; gynecologic surgical procedures.

Introduction

Mammary gland is the biggest skin gland. It is a modi- fied sweat gland, which is developed in the functional organ in women, while in men it remains undeveloped.

It has been thought for centuries, that polymastia is a synonym for greater fertility and feminity. In the nine- teenth century medical literature noted greater fertility in- cluding more multiple (twin) pregnancies with those women ¹.

Ectopic mammary gland tissue is a residual tissue that persists during embryologic development, found in 1–2% of women in general population, with the greatest rate de- scribed as 6 % ².

This is more often the case with women than with men. During the fifth week of the embryologic development along ectodermal primitive milk streaks the given forms are created on ventral side from axilla to inguinal region. Along the line, from both sides, equally remoted from the middle line, two ridges are developed on thorax, at the places of future


Ključne reči: dojka; vulva, neoplazme; choristoma; histologija; hirurgija, ginekološka, procedure.
breasts, at the same time with the regression of the given lines. Further differentiation into complete breast parenchyma happens along gestation. Incomplete involution anywhere along primitive milk streak could result in accessory or ectopic mammary tissue. Most often ectopic place of the gland tissue is axilla, probably as the center of big lymph nodes, and then vulva. Ectopic tissue can be found outside milk streaks, at the back, thighs, face, hips, upper part of the arms, shoulders and feet."

The tissue can be stretched from little focus, parenchyma to complete structure that includes the areola and breast nipple. In children, accessory tissue usually cannot be identified because the tissue mass is small. During menarche the tissue can be enlarged and become symptomatic and requires the doctor's attention. Symptoms generally depend on normal hormon stimulation. They are most often described as palpable mass along milk streak with pre-puberty and puberty girls.

Fibrocystic mammary disease or aberration in normal development and involution (ANDI), chronic cystic mastitis, cystic hyperplasia, cystic mastopathy, fibroadenosis, fibrocystic changes, hyperplastic cystic disease, breast dysplasia, mammary dystrophy, are the synonyms for this disease. The disease could appear in normal and in aberrant positioned breast tissue.

Case report

A woman, 27-year old, admitted to Obstetrics and Gynecology Clinic Kragujevac, Clinical Centre in Kragujevac, in December 2007, for surgery, because of goose-egg size, vulva tumor, at the position of the left lip, of elastic consistency.

The mass, first noted about three years ago, used to grow, especially during the pregnancy and lactation. The only local finding was itching.

Pubic distribution of hairiness and secondary sexual characteristics were normal. In gynecological anamnesis, menarche occurred in 12 years of age with the regular menstrual cycle which lasted 29 days, and the menstrual duration was 5 days. The patient denied previous gynecological diseases. Pregnancy anamnesis showed one preg-

nancy, terminated a year ago by cesarean section because of multiple (twin) pregnancy. The pregnancy had usual course, without complications. She gave birth to two mature children, the first, a female of the weight 2500 g, Apgar score 8, and the second, a male of the weight 2800 g, Apgar score 8.

In the family anamnesis, there were neither similar changes nor other significant diseases.

The basic laboratory analyses of urine and blood were carried out, and their values were in normal limits. The urinary tract ultrasonography findings were regular. Excision of the tumor was completely done in the total endotracheal anesthesia. The incision healed per primam.

Macroscopic findings of a tumor: grayish, kidney knot, dimensions $58 \times 30 \times 17$ mm, at the intersection with white, chop-shape.

Pathohistological findings: at the serial intersections of the analyzed samples the following was present: ectopic mammary tissue (embryonic development of milk streak) with multiplied stromal bind and gland parenchyma. Ductuses were randomly dilated, and their epithelium only focal showed a mild level of hyperplasia. The acini were also of regular histomorphology, rarely multiplied, compacted and “captured” into a hypocellular bind. The cells showed weak expression of estrogen receptors (ER) and a high progestrone receptors (PR). The final diagnosis was: **Dysplasia fibrosa cystica simplex mammae** with the focuses of sclerosal adenosis (Figure 1).

For detection of ER and PR receptors, monoclonal human antibodies were applied (DAKO Denmark) and a highly sensitive, specific streptavidin-biotin immunohistochemical method (LSAB+/HRP, DAKO, DENMARK), in which streptavidin was marked by peroxidase while 3,3 diamine-benzidine was used as a chromogene. During the coloring, known negative and positive tissue samples were tested, at the same time and one tissue intersection from the examined paraffin mould, was tested by a reagent which replaced primary antiestrogenic and antiprogesterone antibodies. Before the very procedure, suitable proceedings for unamasking of antigenes were applied, which were recommended for routinely prepared tissue samples, fixated in formalin and firmly moulded into paraffin.

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Discussion

Ectopic mammary tissue in adult period is usually presented in single cases. The appearance of the ectopic gland mammary tissue in vulva in England from 1900 to 1970 was found in 17 cases. By the insight into the data base in the USA, on the appearance of the abberant mammary tissue, 16 cases were noted in the period of 32 years, from 1971–2003.

While polythelia is originally congenital disease, expression of the ectopic mammary tissue depends on the sexual hormones in the specific life periods, such as puberty, pregnancy and puerperium and breastfeeding.

The existence of the additional nipple–polythelia, or breast–polymastia is not unknown fact. This could be found along mammary line, but also it could be found at other ectopic places. No matter what their location is, ectopic mammary tissue could be pathologically changed, as well as normally positioned mammary tissue into benign cystic changes, benign tumors adenomas, fibroadenomas, carcinomas, and these could present a challenge for the surgeons and for the pathologists to make correct diagnosis.

Fortunately, ectopic mammary tissue is most often a cosmetic problem. In that sense, usually tissue excision is done, and in the case of malignant alteration along with radical surgery, additional therapy is needed. Symptoms could vary as mild or local unpleasantness, sometimes joined with milk secretion.

Polythelia is often connected with abnormalities of the urinary tract. Each extra nipple in child should invoke suspicion to possible kidney or urinary tract anomalies. Connection with multiple kidneys is described, mistakes in a the kidney form, as well as with renal carcinomas, then vertebral anomalies, pylorus stenoses, testicular carcinoma. Polymastia and polythelia are found sporadically, they are described as familiar over the different generations.

The most often benign changes in breast tissue are fibrocystic diseases and they could be found up to 60%. Cysts (microscopic and macroscopic) are only components of fibrocystic diseases. Other histological components are stromal fibrosis, typical and atypical metaplasia. The estrogen influence on tissue proliferation and obstruction of ductus in breast is blamed for the appearance of cystic formation. Epithel secretion from these obstructions results in appearance of cystic formations. They are usually microcystic, thought macrocystic changes could be found, and they are up to 20%. The changes are usually found in reproductive period, 35–50 years of age, and rarely with those below 30 years and postmenopausal women, regardless of the possible substitutional therapy. Benign breast diseases present important factor for breast carcinoma appearance, less as nonproliferative and more as proliferative product, with or without athiopia.

Cells with low ER/PR receptor level grow independently of estrogen stimulation and could be resistant to the effects of hormone therapy.

Conclusion

Ectopic mammary gland tissue in vulva in adult period is very rarely found and can be pathologically changed as well as normally positioned breast tissue into benign cystic changes, benign tumors, adenomas and fibroadenomas and carcinomas.

 REFERENCES


The paper was received on February 05, 2008.