Juvenile Papillomatosis of the Breast in a Pre-Pubertal Girl: An Uncommon Diagnosis

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ABSTRACT

Juvenile papillomatosis of the breast represents a rare benign proliferative disorder that affects women younger than thirty years of age. Although it is a localized lesion, it does not have well-demarcated margins. These patients tend to have a strong family history for cancer. As it has similar clinical presentation with that of a fibroadenoma, it usually receives the diagnosis of the latter in the preoperative period. Nonetheless, it has distinct microscopic features such as ductal papillomatosis and cysts that are helpful in the diagnosis. In this article, a case of juvenile papillomatosis diagnosed in a young girl who presented due to a mass of the breast was presented. For the fairly rare case, a total mass excision was performed with preserved breast tissue. The exact diagnosis was made by postoperative histopathological examination.

Keywords: Breast, juvenile, papillomatosis

Introduction

Juvenile papillomatosis (JP) of the breast was diagnosed as Swiss cheese disease due to multiple cystic formations on the fraction based on the macroscopic examination. Some of the epithelial ducts demonstrated apocrine metaplasia and papillary formations expanding toward the lumen, which were dilated in cystic formation as typical characteristics upon macroscopic examination. This lesion usually causes a mass of the breast. They are similar to fibroadenoma in clinical and macroscopic aspects. Carcinoma of the same of the opposite breast has also been observed besides JP. In addition, it has been observed that breast carcinoma developed during follow-up of patient diagnosed with JP in the past (1). In this article, a 11-year-old female patient who was operated on due to preliminary diagnosis of fibroadenoma of the right breast was presented.

Case Presentation

A 11-year-old female patient presented to the General Surgery outpatient clinic due to rapidly growing mass of the right breast without pain for about two months. During the physical examination, a hard mass with irregular contour and with a dimension of approximately 7 cm, which infiltrated the right breast almost completely, was palpated (Figure 1a). No pathology was noted for the left breast and bilateral axillary regions. No specific characteristics were noted in the patient's familial medical history and laboratory test results. She had no nipple or skin changes associated with the mass, and there were no symptoms suggestive of a history of an abscess. A solid lesion in dimensions of 7x6 cm that infiltrated the right breast almost completely was seen on the breast ultrasonography (US). A Tru-cut biopsy of the right breast was performed because of suspected fibroadenoma at the first stage. The result of the pathological examination was reported as fibroglandular proliferation and an operation was decided upon. Under general anesthesia, the mass was totally excised preserving the breast tissue (Figure 1b). The resulting deformity was filled using the patient's own breast tissue. The patient was discharged on the post-operative first day without complications and no further treatments were provided. The patient was seen to have recovered...
Relationship was found between JP and endocrinological variations (3).

Juvenile papillomatosis is seen at the ages of 10 to 48 (average 23). It is commonly seen above 30 years of age. It has three types including intraductal papilloma, intraductal papillomatosis and sclerosing papillomatosis (7). All characteristics such as the patient’s age being below 30, identification of a mass with regular contours in the breast (although it is considered fibroadenoma in clinical setting), presence of ductal formations with some of them being cystic by dilation in the connective tissue areas on macroscopic examination, and the lumens of ducts containing partial papillary formations lined with epithelium that demonstrates single or several apocrine metaplasia are consistent with the diagnosis of juvenile papillomatosis as it was in our patient. In a 18 years of follow-up of patients with papillary ductal hyperplasia, carcinoma was found in none of them. However, when all the papillary lesions of the breast are considered, it is estimated that papillary carcinoma will emerge later on in 14% of the patients with papillomatosis in average (8).

Our patient had not yet menstruated and the patient’s mother did not take any medication during pregnancy.

When family members of patients with JP are questioned for breast diseases, breast carcinoma has been found to have a ratio of 58% in their first and second degree relatives. However, the presence of carcinoma in the relatives of patients with JP can be considered as a tumor marker of JP, allowing the possibility of following up their close relatives without waiting until they are over 50 years of age where the cancer risk is most (2). No specific characteristics in terms of breast carcinoma were found in our patient’s familial medical history. Bazzochi et al. (3) have found a familial history of breast carcinoma in 33% of patients with JP.

Patients present usually with the complaint of a mass in the breast regarding JP. Hemorrhagic discharge and pain of the nipple are rarely seen (4). The reason of presentation of our patient was the presence of a single, unilateral and growing mass of the breast. In situ or invasive carcinoma at a ratio of 4% was diagnosed in the same or opposite breast regarding JP (5). During 59- to 274-month follow-up of patients with JP, breast carcinoma was diagnosed in 5 patients within about 14 years. In all of these patients, the lesion was bilateral with recurrence or multifocal and all patients had positive familial history (6). Juvenile papillomatosis is a clinical entity different from childhood papillary ductus hyperplasia of the breast. Papillary ductus hyperplasia is commonly seen above 30 years of age. It has three types including intraductal papilloma, intraductal papillomatosis and sclerosing papillomatosis (7). All characteristics such as the patient’s age being below 30, identification of a mass with regular contours in the breast (although it is considered fibroadenoma in clinical setting), presence of ductal formations with some of them being cystic by dilation in the connective tissue areas on macroscopic examination, and the lumens of ducts containing partial papillary formations lined with epithelium that demonstrates single or several apocrine metaplasia are consistent with the diagnosis of juvenile papillomatosis as it was in our patient. In a 18 years of follow-up of patients with papillary ductal hyperplasia, carcinoma was found in none of them. However, when all the papillary lesions of the breast are considered, it is estimated that papillary carcinoma will emerge later on in 14% of the patients with papillomatosis in average (8).

As patients affected by these lesions are usually young, US appears to be the optimal tool for their diagnosis and follow-up. Ultrasoundography is useful for differentiating it from other cystic masses, fibroadenomas, intracystic papillomas, phyllodes tumors, and breast cancer. Although mammography is usually not routinely used for women younger than 35 years of age, its findings in rare cases that present include a homogenous opacity with sharp borders, which is also common in fibroadenomas and cysts. Magnetic resonance imaging also aids in the diagnosis by showing multiple small internal cysts on T2-weighted sequences (9).

Treatment of juvenile papillomatosis is usually accomplished by complete excision followed by histologic confirmation. Prognosis is uncertain. No additional treatments are necessary if no carcinogenic lesions exist simultaneously. Recurrence is inevitable following incomplete excision. Annual monitoring for possible breast cancer development should be done until studies on this subject would determine the value of this practice. Breast carcinoma most commonly develops in those who had a family history for breast cancer and recurrent bilateral JP lesions (10).
In conclusion, juvenile papillomatosis of the breast is rarely encountered in clinical practice. Although it is a benign lesion, monitoring of the patient and her family is an appropriate practice given that there is an association between family history and the risk of future development of breast cancer. In order to decrease the rate of recurrence, JP lesions should be excised completely.

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References


