Case Report

A Case of Juvenile Papillomatosis- "Swiss Cheese Disease" of Breast

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Abstract :
Juvenile papillomatosis (JP) is a breast tumor of young women featuring atypical papillary duct hyperplasia and numerous cysts, first described as a clinicopathologic entity in 1980. The disease is of interest because of the youth of these patients and the fact that the pathologic elements resemble those considered to be precancerous in older women. Recently a case of JP in a 16 years old female was treated and followed in this hospital. The patient noticed a large tumor, 6cm in diameter, on the medial side of the right breast. She underwent an excisional biopsy and pathology demonstrated JP. According to literature, wide local excision is adequate to control the lesion in most cases. Careful clinical surveillance is indicated for any woman who has juvenile papillomatosis and for her female relatives.

Introduction

Papillary lesions of breast have varied morphological, radiological, and pathological features. Such lesions are characterized by formation of epithelial growth that have both the luminal epithelial and the outer myoepithelial cell layers, supported by a fibrovascular stroma[1]. Papillomas of the breast can be divided into solitary papillomas, juvenile papillomatosis, and multiple papillomatosis[2]. Their malignant potentials vary and may have an impact on patients' decision making process.

Case Report

A 16-year-old woman presented with a lump in right breast. Her past medical history unremarkable and she had no significant breast cancer-related family history. Previous breast cytology revealed fibroadenomatoid and fibrocystic changes. Ultrasound (USG) of the lesion revealed an irregular but circumscribed mass containing solid and cystic regions thought to possibly represent thought to possibly represent an atypical (cystic) fibroadenoma (Fig. 1). The patient was elected to undergo excisional biopsy of the mass. Gross evaluation of the specimen revealed a 5.8X3.8X2.8 cm well-circumscribed nodule with diffusely cystic cut surfaces. Some of the cysts contained cloudy fluid and there were scattered foci of chalky yellow-white fat necrosis. Microscopic evaluation revealed a well-demarcated lesion with an admixture of fibrocystic changes including multiple apocrine cysts, adenosis, ducts with prominent fibrovascular cores suggestive of benign intraductal papilloma and prominent duct expansion by usual ductal hyperplasia. Chronic inflammation and foamy macrophages were present in areas of previous cyst rupture.

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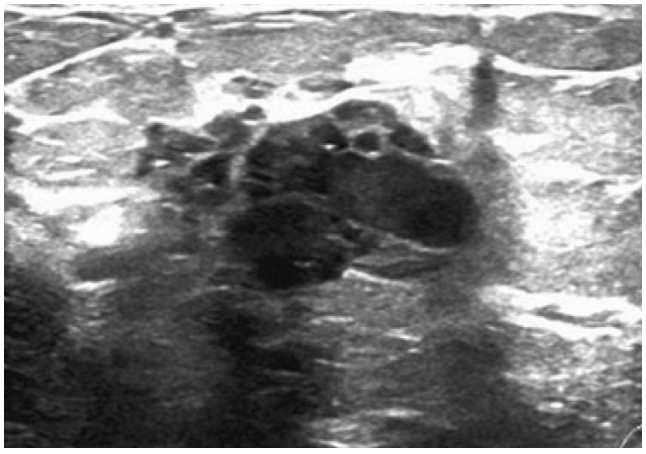

Fig-1: Ultrasound image demonstrating an irregular mass with circumscribed margins. Presence of multiple areas of hypo-echogenicity indicating cystic areas.

No significant architectural or cytologicatypia was present. These histopathologic features were characteristic of juvenile papillomatosis (JP) or "Swiss cheese disease" as it is sometimes referred due to its multicystic appearance (Fig. 2).

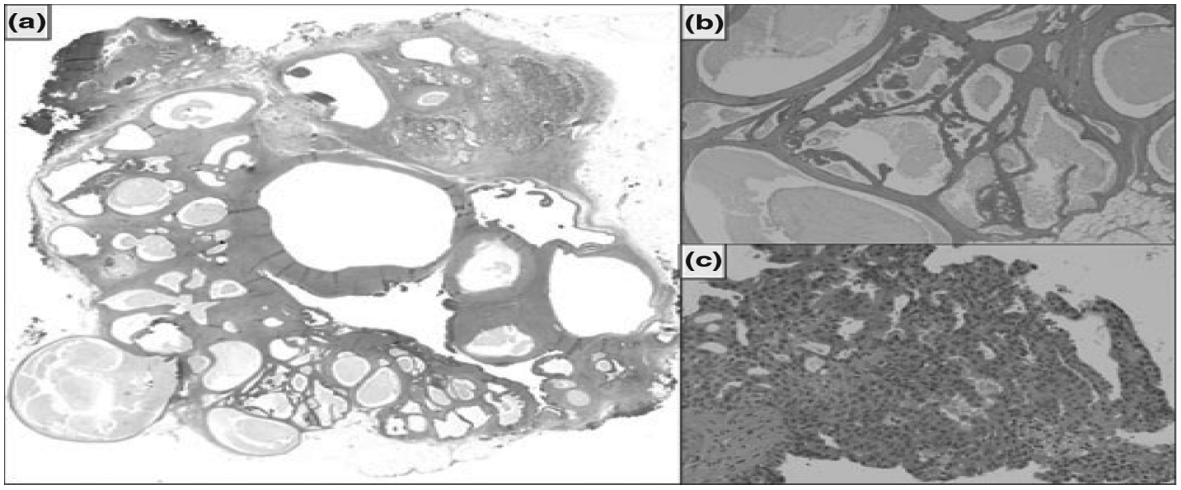

Fig-2 : Whole-mount section from the excisional biopsy specimen showing multiple cysts interspersed with more solid regions, imparting a "Swiss cheese" appearance (a). Higher power images of the lesion including a cluster of simple and apocrine cysts filled with inspisated secretions (b) and ducts expanded by florid usual ductal hyperplasia with intermixed apocrine metaplasia (c).

Discussion

This case is a diagnostic and therapeutic challenge, taking into account the patient's age and the controversial treatment recommendations.

Juvenile or cellular fibroadenomas are an uncommon variant of fibroadenoma. JF are well-circumscribed lesions and, generally, surgical excision is advised for any rapidly growing mass in the adolescent breast, even if it has been previously characterized as benign by core biopsy[3]. The surgical management of JP is a complete excision with histological confirmation. This process is usually effective, and incomplete excision invariably leads to recurrence. The concern should be greatest for women with a positive family history of breast cancer and recurrent bilateral JP[4].

In 1980, Rosen et al[5] reported 37 cases of papillomatosis in young females with a mean age of 19 years (range, 10–44 years), and defined this novel disease as JP due to its clinical and microscopic features. Thus far, ~400 cases of JP have been reported, the majority in Caucasian females aged <30 years at the time of diagnosis. Cases of JP in Asian individuals were rare and thus, fewer reports exist. Whether the difference in incidence between Caucasian and Asian populations is due to genetic or environmental factors remains unclear.

The typical manifestation of JP is a unifocal tumor, commonly located in the upper outer quadrant or outer half of the breast, and is firm, well-circumscribed, mobile, painless and generally measures <3 cm in diameter. Reports of bloody nipple discharge were unusual[6]. When the clinical diagnosis was determined prior to surgery, it was typically fibroadenoma. Mammography is not routinely recommended for diagnosis or follow-up in females <35 years; however, the few reported mammographic findings regarding JP revealed a well-circumscribed homogeneous opacity, which is similar to that observed in fibroadenomas and cysts[7]. Ultrasonography is the preferred imaging technique for JP patients as it facilitates with the differentiation between JP and similar cystic lesions, fibroadenomas, phyllodes tumors, intracystic papillomas and breast cancer[8].

Sonographically, the JP lesion presented as a poorly-defined heterogeneous mass with various small, round, echo-free areas, predominantly observed close to the border of the lesion[9]. Microscopically, the typical histopathological features are duct papillomatosis with or without epithelial atypia, apocrine and non-apocrine cysts, duct stasis and sclerosingadenosis[5]. Papillomatosis and cysts are the dominant diagnostic criteria of JP. A case report describing the fine-needle aspiration cytology of JP revealed the tumor to be comprised of sheets of hyperplastic breast epithelium with areas resembling fibroadenoma, and containing macrophages and apocrine cells. Although it is difficult to diagnose JP solely by its cytology, a combination of clinical and cytological findings may facilitate with the diagnosis of JP. There is no evidence to associate hormonal agent use or reproductive history with the occurrence of JP in young individuals, nor to associate JP with the maternal use of teratogenic agents during pregnancy[5].

Various breast disorders in children and young adults must be distinguished from JP. Rosen[5] described rare types of papillary duct hyperplasia, which are observed in adolescence, including papilloma, papillomatosis and sclerosing papillomatosis.

The most common symptom of papillary duct hyperplasia is the presence of a mass, although certain cases also exhibited nipple discharge, or presented with nipple discharge alone; however, all of these lesions lacked the cystic component that is characteristic of JP. Breast cancer is rare in children, however, when it does occur it most commonly takes the form of a secretory carcinoma and presents as a long-standing breast mass, which is occasionally painful. Nipple discharge is rarely identified. Secretory breast cancer is characterized by the presence of abundant intracellular and extracellular secretions, and intracytoplasmic vacuoles. Furthermore, immunoperoxidase staining for α-lactalbumin is typically positive in secretory carcinoma, but negative in JP[10]. Rosen et al6 reviewed 84 cases of JP in 1982 and identified that 26% of the patients had a family history of breast cancer in at least one female relative. The majority of breast cancer cases were observed in older, secondary relatives (for example grandmothers or great aunts), although instances of maternal breast carcinoma were also reported. This may have been due to the young age of the JP patients and, therefore, the patients' mothers or young female relatives may not have reached the peak age of breast cancer incidence. Bazzocchi et al[11] observed that 33% of JP patients had a family history of breast cancer. These findings indicate that JP may be a marker of breast cancer in the family of the JP patients, thus, a thorough medical follow-up is recommended for JP patients and their families. Furthermore, microscopic evaluation revealed that breast carcinoma coexisted with JP in certain cases. Bazzocchi et al[11] identified that 15% of the JP patients presented with a coexisting carcinoma and Rosen et al[6] described three cases of other types of cancer coexisting with JP (n=84). Two of the patients exhibited secretory carcinoma (one arising from JP and another with contralateral secretory cancer) and the two patients had a maternal history of breast cancer. In addition, although the follow-up data was insufficient, the JP patients appeared to be at an increased risk of developing breast cancer. A previous study of 41 patients with a median follow-up period of 14 years demonstrated a 10% incidence of subsequent breast carcinoma in patients with JP[10]. Although the risk of breast cancer should not be exaggerated, patients exhibiting any one of the following characteristics should be closely monitored for the subsequent development of breast cancer:

i) positive family history of breast cancer
ii) atypical proliferative lesions
iii) bilateral lesions
iv) multifocal lesions
v) recurrence of JP.

Patients with a positive family history of breast cancer and recurrent bilateral JP are considered to be at the greatest risk. Further studies with a longer follow-up period are required to determine the incidence of breast cancer in patients with JP.

In conclusion, the recommended treatment strategy for JP is complete excision of the cancerous lesion to reduce local recurrence. On consideration of the current literature, it is prudent to advise an annual clinical follow-up, including a physical examination and/or ultrasonography of the breasts for JP patients, and for the patients' female relatives, particularly those with a family history of breast cancer and with recurrent or bilateral JP.

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