Juvenile papillomatosis of the breast
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Section: Breast imaging
Area of Interest: Oncology Breast Abdomen Abdominal wall
Procedure: Diagnostic procedure
Procedure: Biopsy
Imaging Technique: Ultrasound
Imaging Technique: MR
Imaging Technique: CAD
Imaging Technique: PET
Special Focus: Neoplasia Abscess Acute Trauma
Case Type: Clinical Cases
Authors: Santos-Magadán S, López-Casañas AM, Martínez-González J, Taborda Ramírez LF, Moreno-Torres A, Carreira-Gómez C
Patient: 15 years, female

Clinical History:

A 15 year-old girl presented with palpable mass in her right breast for the last 2 months. She had positive background for breast carcinoma (grandmother at the age of 50 years).

Imaging Findings:

Ultrasound revealed a multicystic and not well circumscribed mass in the upper quadrant of the right breast (Fig. 1). We performed an ultrasound-guided core-needle biopsy (14 gauge, 22mm) obtaining nonspecific results. Contrast-enhanced MR showed a segmental distribution of the lesion (Fig. 2) with multiple microcysts, slow and progressive enhancement (Type 1 uptake curve) and high signal on DWI with high ADC values (1, 7x10-3) mm/sg (Fig. 3).

Due to the lack of radio-pathologic correlation, an additional sample was obtained performing vacuum assisted biopsy (10-gauge) which revealed breast tissue without fatty components, cysts with apocrine metaplasia in some of them with focal changes of hyperplasia and a micropapillary pattern. The cysts contained a dense material with a proteinaceous appearance and focally showed a chronic periductal inflammation (Fig. 4). These results were consistent with focal juvenile papillomatosis (JP).

The patient subsequently underwent wide local excision of the lesion, which confirmed the diagnosis of JP and is currently on follow-up examination.

Discussion:

JP is an infrequent disorder among young women (under 30 years) with a mean age at diagnosis of 19 years [1]. It consists of a localised benign proliferative lesion of the breast also known as “Swiss Cheese” disease due to its fibrocystic appearance [2]. The most frequent clinical presentation is the palpation of a nodule, similar to a fibroadenoma, usually unilateral.
Approximately 26-58% of the patients have a family history of breast cancer [3]. Ultrasound appearance is a well or ill-defined mass with multiple small cysts, especially at the periphery [1].

The macroscopic aspect of the lesion is a multicystic mass without capsule, and its size ranges from 1 to 8 cm [1, 4]. Microscopically, the disease is characterised by a heterogeneous mixture of proliferative changes. Papillary hyperplasia of the ductal epithelium (papilloma or papillomatosis) and cysts of different sizes (<2 cm), with or without apocrine metaplasia are constant findings [4].

To diagnose JP a breast biopsy should be performed; however, imaging methods may play an important role in the preoperative orientation and follow-up of these patients (mainly US and MRI due to its early onset) [5]. On MRI, the most specific finding is the presence of numerous small cysts on T2WI. Contrast enhanced T1WI with fat suppression permits a proper visualisation of contour and internal matrix. Contrast media uptake pattern is that of a benign disease, with type 1-2 curves [6, 7, 8] and high diffusion.

Despite its benign nature, JP has been associated with an increase in the incidence of breast cancer. In approximately 4-15% JP and breast carcinoma coexist in the diagnosis (usually intraductal carcinoma, but also ductal invasive carcinoma, lobular carcinoma and especially secretory carcinoma of the breast) [5]. Around 10% of the patients will develop carcinoma in the future [4].

Wide surgical excision to prevent recurrence with clinical follow-up is considered to be the treatment of choice and incomplete excision leads to recurrence. Patients with bilateral, multifocal or recurrent disease and family history of breast cancer (in particular those with pathologic atypia) are at increased risk of developing breast cancer and should be closely monitored [4, 9].

There is no evidence so far that patients with non-recurrent unilateral JP and complete excision will develop subsequent ipsilateral carcinoma, despite family history or pathologic risk factors [4, 9].

In our case, MRI was crucial to characterise the lesion. It also allowed us to establish the precise size and existence of associated pathological enhancements, and rule out the presence of additional lesions in the other breast.

When finding a multi-cystic lesion in a young patient, it is necessary to consider JP, perform a core needle biopsy (10-gauge if possible), MRI, and propose for full excision with close follow-up.

**Differential Diagnosis List:**
- Juvenile papillomatosis of the breast.
- Fibrocystic breast disease.
- Fibroadenoma.
- Phyllodes tumour.
- Intracystic papillary carcinoma

**Final Diagnosis:** Juvenile papillomatosis of the breast.

**References:**


Wang T, et al. (2014) Bifocal juvenile papillomatosis as a marker of breast cancer: A case report and review of the
Figure 1

(a) Description: Ultrasound image. Panoramic view. Palpation area in the upper inner quadrant of the right breast showing a multicystic mass. Origin: Santos-Magadán S, Department of Radiology, Hospital Universitario de Fuenlabrada, Madrid, Spain.

(b) Description: Ultrasound image. Palpation area with multiple small cysts. The mass is non-well circumscribed. Origin: Santos-Magadán S, Department of Radiology, Hospital Universitario de Fuenlabrada, Madrid, Spain.
**Figure 2**

**a**

Description: Axial fat-suppressed T2WI (fat suppressed). Multiple small clustered cysts in the upper inner quadrant of the right breast. **Origin:** Santos Magadán, Sara, Department of Radiology, Hospital Universitario de Fuenlabrada, Madrid, Spain

**b**

Description: Axial T1WI. The mass is hypointense. **Origin:** Santos Magadán, Sara, Department of Radiology, Hospital Universitario de Fuenlabrada, Madrid, Spain
**Description:** Axial post gadolinium T1WI (fat suppressed) showing a pattern of slow and progressive enhancement (type I curve).

**Origin:** Santos Magadán, Sara, Department of Radiology, Hospital Universitario de Fuenlabrada, Madrid, Spain

**Description:** Axial post gadolinium kinetic curve showing progressive uptake curve (type 1).

**Origin:** Santos Magadán, Sara, Department of Radiology, Hospital Universitario de Fuenlabrada, Madrid, Spain
Description: Axial DWI. High signal with high values on ADC. **Origin**: Santos Magadán, Sara, Department of Radiology, Hospital Universitario de Fuenlabrada, Madrid, Spain

Description: ADC. High signal with high values (1.7x10^-3 mm/s). **Origin**: Santos Magadán, Sara, Department of Radiology, Hospital Universitario de Fuenlabrada, Madrid, Spain
Figure 3

Description: Sagittal view. Segmental distribution of the mass (larger diameter 6 cm). Origin: Santos Magadán S. Radiology Department. Hospital Universitario de Fuenlabrada, Madrid, Spain
Description: Cysts with apocrine metaplasia in some of them, hyperplasia and micropapillary pattern. The cysts contained a dense material with proteinaceous appearance and focally showed chronic periductal inflammation. Origin: Moreno-Torres A, Pathology Department, Hospital Universitario de Fuenlabrada, Madrid, Spain
Description: Intraductal hyperplasia with micropapillary pattern. Origin: Moreno-Torres A, Pathology Department, Hospital Univeristario de Fuenlabrada, Madrid, Spain