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CASE REPORTS

Cystosarcoma phyllodes of the breast: a case report in a 12-year-old girl

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Abstract Breast tumors in adolescents are very rare and mostly benign. Fibroadenomas are the most frequent, but within the extensive differential diagnosis, the phyllodes tumor must be mentioned, which accounts for about 1% of breast tumors and the diagnosis of which is very rare in patients younger than 20 years. There are no specific symptoms or radiological images to distinguish phyllodes tumor from fibroadenoma; therefore, histological examination is mandatory for diagnosis. Histology also allows the classification of phyllodes tumor into benign, borderline, or malignant types for appropriate surgical treatment: free-margin excision in benign tumors and mastectomy in the other two types. Fortunately, the majority of these tumors are benign, and treatment maximizes breast conservation with free infiltration margins surgery, given that this fact is the most important factor to prevent local recurrence. In this article, we describe a rare case of borderline cystosarcoma phyllodes in a 12-year-old girl.

Keywords Cystosarcoma phyllodes · Borderline · Adolescence · Breast tumor

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Introduction

The differential diagnosis of a breast mass in adolescence includes physiological hypertrophy, inflammatory processes, benign proliferative lesions, malignant lesions and miscellaneous. There are no specific guidelines for evaluating a breast mass in adolescence. The diagnosis is usually based on clinical history and exploration [1]. Ultrasound is the imaging technique of choice at this age, and the definite diagnosis will be done based on the histological study through core needle biopsy (CNB) [2–4]. Cystosarcoma phyllodes is an unusual breast tumor in adult women, accounting for 0.3–1% of all breast cancer [5]. Most phyllodes tumors (PT) described in adolescence are benign [1]. Complete tumor excision is the treatment of choice for all grades of PT in adolescence [6, 7], during which time the goal of breast conservation should be maximized.

Case report

A 12-year-old girl came to our emergency department presenting disproportional right breast growth of 2 months evolution (Fig. 1) and recent pain. There was no remarkable family history. The patient's personal background included pre-menarche and 1 year of breast development. Physical examination was normal except for a voluminous right breast, with solid consistency, inflammatory aspect,

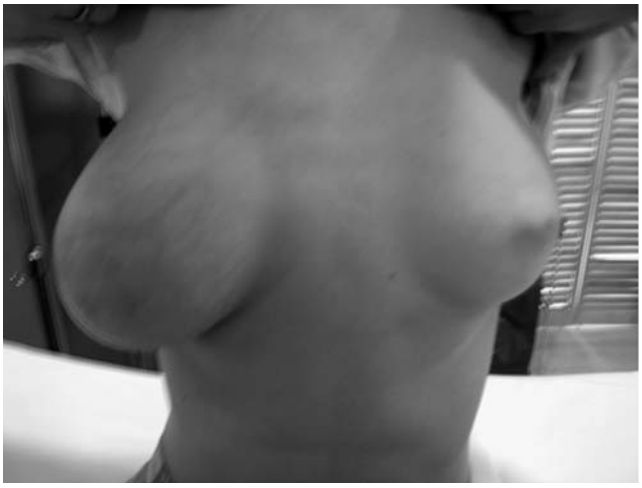

Fig. 1 A 12-year-old girl presenting disproportional right breast growth

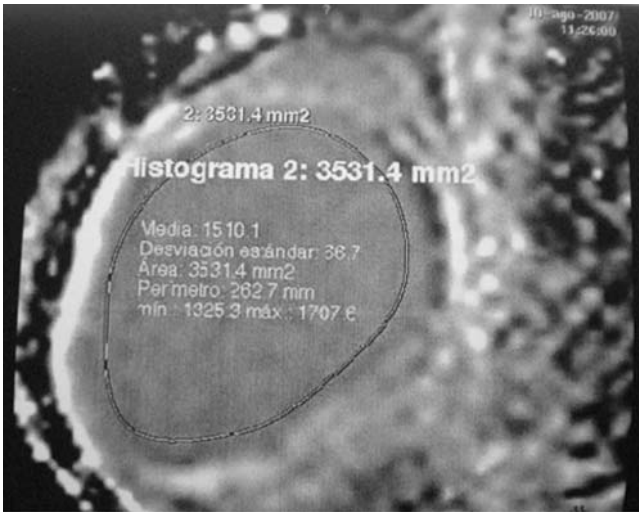

Fig. 3 Magnetic resonance imaging study

skin pressure, no clear tumor delineation, and increase palpable size. Lymph nodes were free.Ultrasound study confirmed a solid mass with low-level internal echoes, axis approximately 10 cm long (Fig. 2), with vascularized, apparently encapsulated, aspect in the Doppler study. A follow-up radiological study was conducted that revealed a mass that occupied nearly all of the right breast without associated microcalcifications. Biomarkers were carcinoembryonic antigen (CEA) 0.69 ng/ml and carbohydrate antigen-15.3 (CA-15) 4 U/ml. On magnetic resonance imaging (MRI), the right breast was virtually replaced by the lesion area, with a maximum diameter of 8.6 cm and showed small pockets within the lesion in the hyperintense T2 baseline study, which could correspond to small areas of necrosis or cystic degeneration (Fig. 3). The mass presented a relatively well-defined, smooth contour, with no data suggesting infiltration of adjacent tissues.

Under ultrasound (US) control, a CNB was performed, extracting three tissue samples from the solid mass that were sent to the pathologist for histological classification, which reported an epithelial and mesenchymal proliferation that revealed the existence of borderline PT. A simple mastectomy of the right breast was performed, and the pathology analysis of the surgical specimen revealed a final diagnosis of phyllodes tumor, with focal areas of slight atypia and borderline type with free tumor edge resection. Upon clinical follow-up at 12 months after surgery, the patient was disease free.

Discussion

PT represents 1% of all breast tumors and primarily occurs in patients around the time of perimenopause, appearing in only 8% of patients younger than 20 years [8]. Trauma, lactation, pregnancy, estrogen excess, radiation exposure, early menarche, as well as family history of breast cancer, gynecological or entities such as neurofibromatosis 1, and celiac disease, have been reported as risk factors for developing this entity, although none of these events were present in our patient.

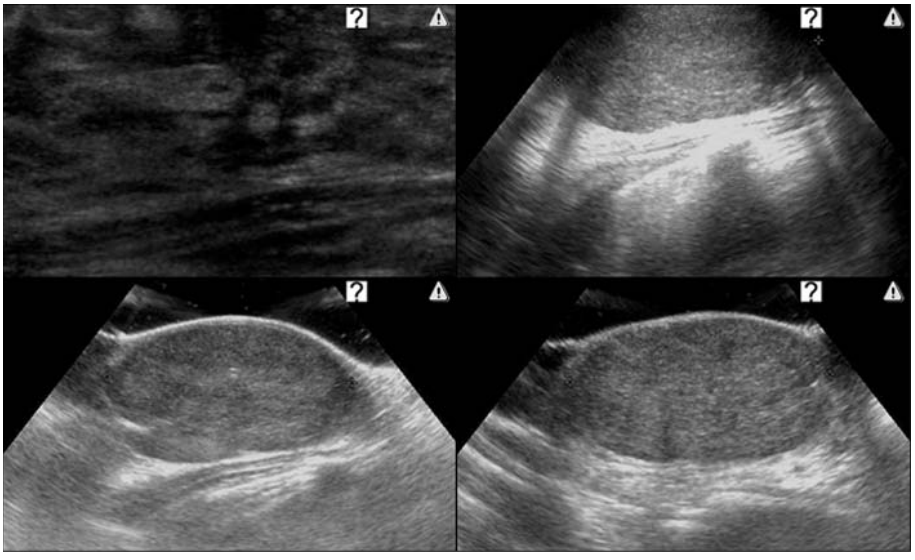

Fig. 2 Ultrasound study shows a solid mass with low-level internal echoes

Histological classification so far accepted is divided into benign, borderline, and subtypes of malignancy depending on tumor margins, stromal overgrowth, necrosis, atypia, and number of mitoses per high power field [3]. Our case was classified as a borderline PT due to the number of mitosis per high power field and the moderate pleomorphism nuclear increase of fibroblasts nucleus.

Diagnosis is often based on medical history and complete breast examination with chaperone signs and symptoms [3]. Other studies include mammography, US, MRI, and fine needle aspiration. Mammography is not recommended in adolescents because of the density of breast tissue at this age [1]. Ultrasound helps differentiate between solid and cystic masses does not differentiate between benign and malignant tumors. US usually shows a seemingly solid mass with low-level echoes and smooth edges, as in our case [1]. MRI is useful to study local extension and diagnose tumors >3 cm, which are more likely to be malignant. In our case, MRI showed that the mass did not infiltrate any adjacent tissue and defined its maximum diameter. Fine needle aspiration allows studies of the cytological characteristics of the tumor and can confirm the existence of a benign lesion and the need for a biopsy. Review of the literature concluded that thick needle biopsy is the research method of choice for diagnosis and is justified if there is a high degree of suspicion of PT [8].

Differential diagnosis, which must be performed on any teenager presenting a breast mass, includes physiological hypertrophy, inflammatory processes, benign proliferative lesions, malignant lesions, and miscellaneous. Fibroadenoma (FA) is the most frequent breast cancer found in adolescent women, which has a similar clinical presentation to PT [1, 3]. The existence of a PT should be suspected if there is a history of rapid breast growth [1, 4], but there are no defined clinical criteria to differentiate the two entities. In the case we present, there were no clinical or study characteristics sufficient to differentiate PT; therefore, thick needle biopsy was necessary for diagnosis. Adolescent cystosarcoma is usually differentiated from fibroadenoma by histological study due to mitotic activity, increased cellularity in the stroma, and the heterogeneous proliferation trend in PT, as shown in the pathology report of our case [4]. However, there is still controversy regarding differential diagnosis, and the rare cases that cannot be differentiated will be managed as if they were a PT to avoid underdiagnosis and to begin appropriate treatment [4].

The decision of how to treat young patients is a problem, and is even more difficult than reaching a correct diagnosis. The aim of treatment must be complete tumor removal with free margins (1 cm) for small lesions; breast lesion segment resection with free edges for lesions >3 cm, and mastectomy in the case of extensive disease and borderline or infiltrative findings [3]. Radiation and chemotherapy are not currently validated for clinical practice in this entity [6]. In our patient, mastectomy was performed, because it was an extensive, borderline tumor [1].

Recent research has focused interest on immunohistochemical markers and other techniques to elucidate clinical behavior and for development of new, additional prognostic indicators in PTs [9].

In conclusion, PT is a very rare entity in the adolescent, and an accurate histological classification and a correct differential diagnosis is essential, especially in juvenile fibroadenoma. The infiltrative tumor edges and positive surgical margins are the best predictors of local recurrence, requiring margins of 1–2 cm to prevent it [7]. It is important to maximize breast conservation in adolescent patients in cases where it is possible, thus avoiding the great psychological burden that accompanies a more aggressive management.

Conflict of interest The authors declare that they have no conflict of interest relating to the publication of this manuscript.

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