CASE REPORT

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Pseudoangiomatous stromal hyperplasia (PASH) in a 14-year-old girl: a case report

Hiperplasia estromal pseudoangiomatosa (PASH) en niña de 14 años: reporte de caso

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ABSTRACT
Pseudoangiomatous stromal hyperplasia (PASH) occurs between 12 and 75 years of age, being prevalent in the reproductive age. Its etiology is unknown. They are characterized by being large, deforming, unilateral and are diagnosed by ultrasound and histology. In adolescents, early surveillance, diagnosis and management is important in order to avoid radical surgeries that cause large scars, deformity and breast asymmetry, which have repercussions on the behavior and emotional state of the patients. Breast tumors are generally benign in adolescence, but their diagnosis in this age group is worrisome for the family and the patient. We present the case of a 14-year-old adolescent who underwent a submammary approach, preserving breast symmetry.
Key words: Breast neoplasms, Adolescent, Ultrasonography, mammary, Mastectomy, segmental.

RESUMEN
La hiperplasia estromal pseudoangiomatosa (PASH, por sus siglas en inglés) se presenta entre los 12 y 75 años de edad, siendo prevalente en la edad reproductiva. Se desconoce su etiología. Se caracterizan por ser grandes, deformantes, unilaterales y se les diagnostica por ecografía e histología. En las adolescentes es importante la vigilancia, diagnóstico y manejo tempranos, con la finalidad de evitar cirugías radicales que originen cicatrices grandes, deformidad y asimetría mamaria, que repercuten en el comportamiento y estado emocional de las pacientes. Los tumores de la mama generalmente son benignos en la adolescencia, pero su diagnóstico en este grupo etario es preocupante para la familia y la paciente. Presentamos el caso de una adolescente de 14 años a quien se le realizó abordaje submamario, conservando la simetría mamaria.
Palabras clave. Mama, neoplasias, Adolescente, Ultrasonografía mamaria, Mastectomía segmentaria, Mamoplastia.

Introducción

Pseudoangiomatous stromal hyperplasia, commonly known as PASH, is a rare benign disease and its etiology is currently unknown[(1)]. It was first described in 1986 by Vuitch et al[(2)], and appears frequently between the ages of 12 to 75 years[(3)]. It presents as a large, solitary, firm, painless, circumscribed, delimited and mobile nodular mass of variable size between 2 and 13 cm. Frequently unilateral, there are reports of bilaterality[(4)]. In some cases, it is diagnosed in the biopsy. PASH can also arise in the axillary tail of the breast and very rarely involves the nipple-areola complex and the axillary accessory breast tissue[(1)]. The most important differential diagnosis is with angiosarcoma[(5)].

Excisional biopsy is the treatment and the final diagnosis is histologic[(6)]. Diagnosis, management and esthetic outcome are very important in this age group. These large tumors are deforming and require early surveillance, diagnosis and management in order to avoid radical surgeries that cause large scars, deformity and asymmetry, and may have repercussions on the behavior and emotional state of the adolescent.

Among the most common benign breast tumors in adolescence are juvenile fibroadenoma, juvenile hypertrophy (virginal) and pseudoangiomatous stromal hyperplasia. Phyllodes tumors, lymphomas, angiosarcomas are extremely rare[7-10].

We present the case of a patient who underwent a submammary approach that preserved breast symmetry and esthetics.

Presentation of the clinical case

An adolescent girl aged 14 years and 3 months reported that she noticed a progressive increase in the size of her right breast over 6 months, with asymmetry of the breasts and no pain. Her 42-year-old mother was a housewife and her 43-year-old father was a businessman, both apparently healthy. The teenager was born with a normal delivery, weighing 3,200 g, had complete vaccinations, menarche at 11 years of age, catamenial regimen 4/30, without dysmenorrhea.

At the admission examination she weighed 53 kg, height 1.54 m, body mass index 22.3, cooperative during the evaluation. General, nutritional and hydration status was regular. The right breast, Tanner stage 4, was occupied by a single mass with defined borders of 9 x 6 cm in radius 6 to radius 12, mobile, non-painful, of hard consistency and deforming the nipple and areola. The left breast, Tanner stage 4, showed no alterations. Biochemistry, hematological and urine tests were normal. Right breast ultrasound showed a circumscribed oval mass with acoustic shadow, measuring 9 x 6 cm in radius 11 and 12 (Figure 1).

Surgical treatment consisted of excision of the tumor (Figure 2) through a submammary approach (Figure 3). No complications were evidenced during the postoperative period, and she was discharged after 1 day of hospitalization, with analgesics, antibiotics and breast bandage. She went to the control 72 hours after de postoperative period for removal of the bandage. After one week the sutures were removed. On physical examination, the operative wound was closed and without phlogosis.

The histological report was: Grayish-brown tumor of 11 x 7.7 x 5.5 cm in size. Final diagnosis:

Figure 1 (A, B). In the breast ultrasound before surgery, the findings are variable, but in most cases a solid, circumscribed, oval or rounded mass is observed, as in the case presented.

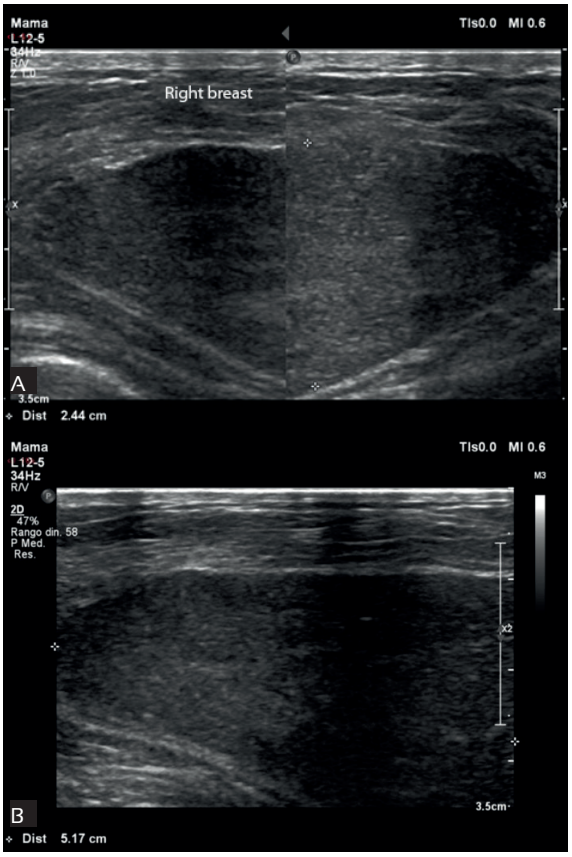
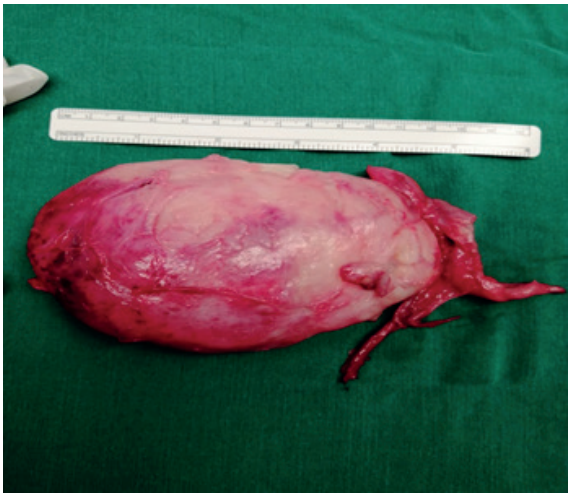
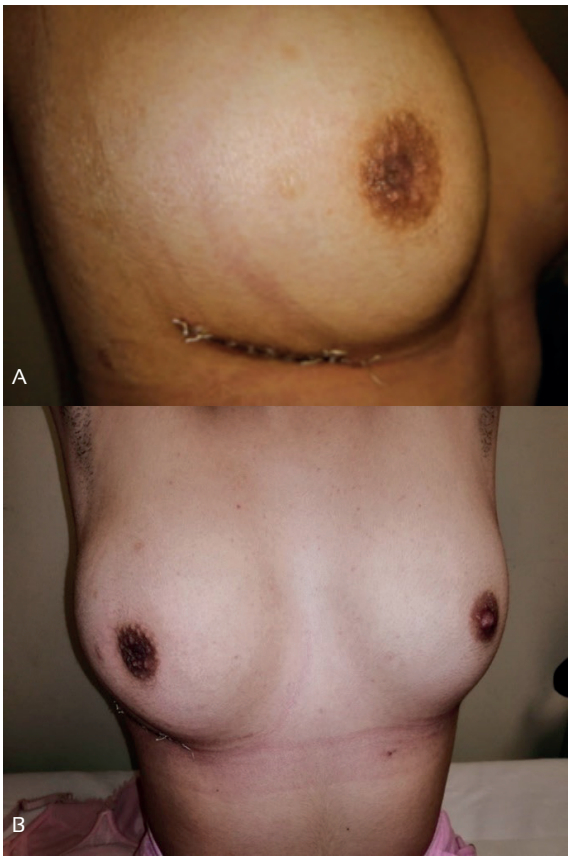


Figure 2. Operative specimen: a solid mass with defined borders, grayish brown, 11 x 7.7 x 5.5 cm, is observed.



Pseudoangiomatous stromal hyperplasia (PASH) (Figure 4). Currently, the patient continues to be monitored and with favorable follow-up. In the last control in 2021, there was no evidence of another tumor growth.

Figure 3. A. Right submammary incision. B. Control at one week.



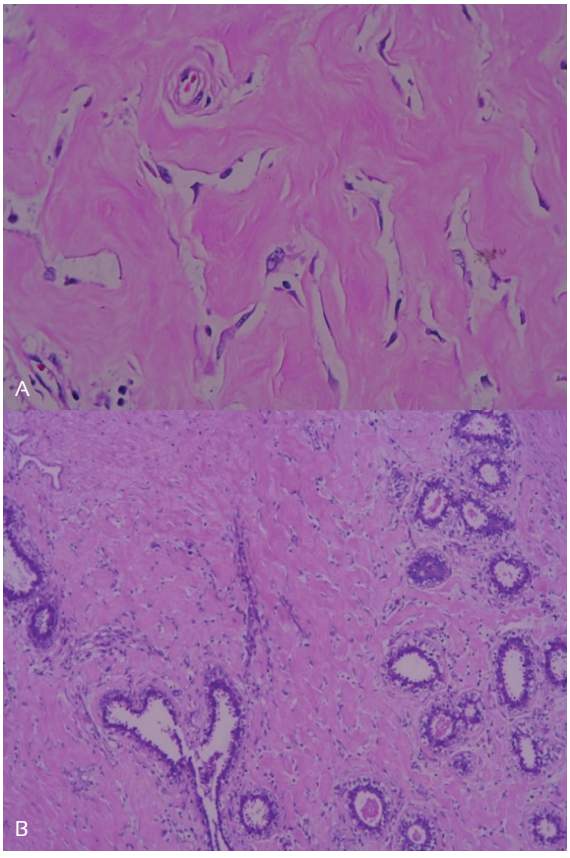
Discussion

We report the first case of PASH in our institution, being important to make known this pathology, frequent between the ages of 12 to 75 years and more prevalent in women of reproductive age. In adolescence, they are usually benign. The etiology of PASH is unknown.

A complete medical history, general physical examination and breast examination should be performed in any gynecologic check-up of an adolescent girl. In these cases, the patient will come for consultation with a well-demarcated, hard, non-painful, deforming and rapidly growing lump.

The differential diagnosis of breast tumors is fundamental for management of fibroadenomas, gigantojuvenile fibroadenomas, phyllodes tumor, lymphoma. Among the diagnostic tests, breast ultrasound[11-13,16] in this type of tumor is non-specific, generally finding a solid, circumscribed, oval or round mass. The diagnosis is anatomopathological; a breast parenchyma with numerous stromal clefts is described. These

Figure 4. Histological description: (A) Histological study confirms pseudoangiomatous stromal hyperplasia: Histological sections show mammary parenchyma with numerous stromal clefts. These clefts are lined by myofibroblasts. The spaces produced by the clefts are empty, simulating complex anastomosed blood vessels. (B) Clefts surround lobules of normal appearance and involve both the interlobular and the intralobular stroma.



clefts are lined by myofibroblasts[15]. The spaces produced by the clefts are empty, simulating complex anastomosed blood vessels. The clefts are present in both the interlobular and intralobular stroma.

In this age group, the surgical approach should be conservative and adequate. Patients should be followed up, because recurrence occurs in around 15 and 22% of cases[8,14,16].

In conclusion, pseudoangiomatous stromal hyperplasia is a lesion that is generally seen in women of reproductive age or as an incidental finding as a result of a biopsy. However, it has also been described in girls and adolescents as a palpable, unilateral, firm, non-painful, mobile mass. Diagnosis is by imaging and histology. Differential diagnosis with gigantojuvenile fibroadenomas and lymphomas is essential. Surgical treatment should be conservative in this age group.

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