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Case Report
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Ulcerated Pseudoangiomatous Stromal Hyperplasia (PASH) of the Breast- A Rare Form of Presentation in a 13-Year-Old Girl

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Abstract

Pseudoangiomatous Stromal Hyperplasia (PASH) is a benign lesion of the breast characterized by proliferation of stromal mesenchymal cells of myofibroblastic origin and is thought to be hormonally driven. It is mainly found incidentally, but it may manifest as a large solid mass of the breast. Imaging findings of PASH are often nonspecific and it can be misdiagnosed with other benign and malignant lesions of the breast. Definitive diagnosis is mostly given by histologic examination with core-needle biopsy or surgical specimen sample. Surgical excision of the lesion with adequate margins of resection is the usual therapeutic approach, and with so, recurrence is low. We present a clinical case of a 13-year-old girl presenting with what we believe is the first reported case of PASH manifesting as an ulcerated tumor.

Introduction

Pseudoangiomatous Stromal Hyperplasia (PASH) is a benign mesenchymal proliferative lesion of the breast. It was first described in 1986, by Vuitch et al. [1] as a mammary stromal proliferation with complex inter-anastomosing channels lined by slender spindle cells.

Its form of presentation may vary from an incidental microscopic finding to a large palpable mass. PASH can be clinically and radiologically confounded with other benign breast conditions like fibroadenoma. However, it can also mimic a Phyllodes tumor or a low-grade angiosarcoma, and thus, definitive diagnosis is most often made by histologic examination, characterized by interanastomosing slit-like channels, lined by spindle cells and surrounded by collagenous stroma. Although core-needle biopsy may provide us the diagnosis, its accuracy is uncertain and it has also failed to diagnose PASH in a significant number of cases. In most patients, surgical excision is the recommended approach. Recurrence rate is low if excision with adequate margins is performed. We present a unique case of PASH presenting as an ulcerated tumor of the breast [2-8].

Case Presentation

A 13-year-old girl presented with a large ulcerated tumor of the right breast. She has been evacuated from the Democratic Republic of São Tomé and Príncipe to our country with a 7-month history of an enlarged nodule in the right breast, with no symptoms associated. Core biopsy revealed a biphasic tumor, unable to distinguish juvenile fibroadenoma from Phyllodes tumor. The tumor continued to grow in size and 5 months after its initial presentation, it ulcerated and the patient was transferred to our hospital a month after.

On our evaluation, she presented a right breast enlargement with an ulcerated, solid tumor affecting most of the breast and compromising skin and nipple area (Figure 1). She didn't have any relevant personal or family medical history, didn't take any medication and hasn't yet reach menarche. A CT scan was performed, revealing a 9 cm × 9 cm solid mass with ulceration and no pathologic lymph nodes were detected.

After a multidisciplinary team decision, surgery was the treatment of choice and the patient underwent right total mastectomy (Figure 2 and 3). Definitive histologic examination revealed PASH with ischemic foci and skin ulceration [9-11].

Discussion

The exact etiology and pathogenesis of PASH is still unknown, but it is believed that hormonal

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Figure 1: Preoperative view of the patient presenting with an ulcerated right breast tumor.

Figure 2: Gross appearance of the surgical specimen.

Figure 3: Postoperative view of the patient after right total mastectomy.

stimuli may play a role on myofibroblast aberrant and exaggerated response, being progesterone the main hormone implicated, and so, the vast majority of patients are premenopausal women or in postmenopausal women on hormone replacement therapy. Nevertheless, PASH has been reported in both women and men, and the age range of patients is of 3 to 75 years. In the pediatric population, PASH has rarely been reported and the greater part of cases was in female adolescent patients who have already reached menarche.

Imaging with ultrasonography or CT scan may aid and even suggest the diagnosis but histologic confirmation is the only definitive diagnosis.

In the majority of cases, it is seen incidentally as a focal microscopic finding and rarely presents as a clinically palpable well-circumscribed, solid mass.

If incidentally found in an asymptomatic patient close follow-up may be considered. Nonetheless, the usual management of PASH is surgical excision.

A more conservative approach is preferred; however, it couldn't be performed in our patient because of the high tumor-to-breast volume ratio, and lack of skin and nipple sparing by the tumor. Taking into consideration the patients age and morphotype, and the fact that she hasn't reach menarche, a reconstructive surgery was postponed and decided to be made at an adult age [12-15].

Conclusion

We report a case of an aggressive form of PASH in a 13-year-old girl, presenting with a large, rapid growing tumor, compromising the majority of breast tissue and leading to massive ulceration of skin. This form of aggressiveness, in such a young age and thus leading to an invasive surgical approach, turns this case into a rarity. To our knowledge, this is the first case reporting an ulcerated PASH tumor of the breast.

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