Case Report

Sonographic Findings in Superficial Angiomyxoma of the Vulva in a Perimenopausal Female

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Abstract

Superficial angiomyxoma (SAM) is an extremely rare soft tissue tumor. It is especially rare in the vulva, with only a few such cases reported in the medical literature. Here, we report a case of SAM of the vulva that was initially suspected to be a Bartholin gland cyst. The patient underwent local excision of the vulvar cyst under lumbar anesthesia. Clinical manifestations and B-scan ultrasonographic features are similar between SAM and cysts. Echoes in the mass are uneven and exhibit low echoes and punctate hyperechoic floating. Thus, increasing sonographers' awareness of the high-frequency ultrasonography findings associated with this rare tumor could broaden their knowledge base.

Keywords: High-frequency ultrasonography, superficial angiomyxoma, vulva

Introduction

Superficial angiomyxoma (SAM) is a rare soft tissue tumor that was first described and named "cutaneous myxoma of Carney's complex" by Allen *et al.* in 1988.[1] The incidence of SAM is extremely low, ranging from 0.008% to 3%.[2] It can occur in many parts of the body but mainly presents in the head, neck, limbs, and trunk.[3,4] While SAM occurs at all ages, it primarily develops in middle-aged individuals (20–40 years) and is slightly more common in males than in females.[5] Most publications on this disease are case reports. As of 2016, fewer than 20 cases of SAM occurring in the vulva had been reported in the English literature, and no case in the English literature had described an SAM that occurred in the vulva of a perimenopausal female.[6] Here, we describe such a case. Surgical resection is the main treatment method for SAM. While recurrence generally does not occur after surgery, the rate of incomplete resection resulting in recurrence is 30%–40%.[7]

Case Report

A 50-year-old woman was referred to our hospital for evaluation of a mass in the vulva that was incidentally detected 2 years prior. The tumor had gradually grown but was associated with only mild tenderness. She had G3P1; abortion twice; last menstrual period, on December 20, 2017; and diagnostic dilation and curettage, four times because of dysfunctional uterine bleeding in the past. A clinical examination revealed a soft, movable, cystic, well-circumscribed mass approximately 4 cm × 2 cm in size in the middle to lower part of the left labium majus. The clinical diagnosis was Bartholin gland cyst. Ultrasonography was performed and showed a 3.6 cm × 1.5 cm unilocular mass that was oval, well-circumscribed, heterogeneous, and mildly echogenic, located under the skin in the middle to lower part of the left labium majus. The echo in the mass was uneven and exhibited low echo and punctate hyperechoic floating with slight posterior enhancement [Figure 1]. Color Doppler flow imaging revealed an abundant blood flow signal inside the lump [Figure 2]. Ultrasonic indications showed a rich subcutaneous blood supply on the tumor. The patient underwent excision of the mass under lumbar anesthesia on February 5, 2018. The gross picture of the tumor was a reddish-gray tissue of size about 3.2 cm × 2.8 cm × 1.6 cm [Figure 3]. Postoperative pathology showed a large number of small

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blood vessels, and spindle-shaped interstitial cells that formed nodules were observed under the microscope. The walls of the small vessels were thick and glassy and contained abundant mesenchymal cells and collagen fibers. The stromal cells displayed unclear cell boundaries and round nuclei, and the ova had loose and edematous partial interstitium and exhibited mucous degeneration [Figure 4]. Immunohistochemistry showed the following: actin (vascular +), CD31 (+), CD34 (3+), CD68 (−), K (−), Des (−), EMA (−), FVIII (+), Ki-67 (2%+), S-100 (−), SMA (vascular +), Vim (3+), and β-catenin (cytoplasm +) [Figure 5]. The pathological diagnosis was (vulvar) SAM.

Discussion

Vascular myxoma can be divided into three types: SAM, invasive angiomyxoma, and fibroblastoma. In 1988, Allen provided the first name for cutaneous myxoma of Carney's complex in Carney.[1] SAM is a rare soft tissue tumor with an incidence of approximately 0.008%–3%.[2] As of 2016, fewer than 20 cases of SAM occurring in the female vulva had been reported in the English literature.[6,8] SAM is observed at all ages but occurs primarily in middle-aged patients mainly between the ages of 20 and 40 years old and is slightly more frequent in males than females.[5]

The pathogenesis of the disease is unknown, but it is characterized by a high concentration of small, thin-walled blood vessels and a mucous-like stroma.[6] SAM clinically manifests as painless masses that are visible to the naked eye; these masses exhibit slow growth and occur in many parts of the body but mainly in the head, neck, limbs, and trunk. However, the tumors can also occur in the external genitalia.[3,7,8] Almost all reported cases of vulvar SAM have been confined to the subcutaneous adipose layer and often exhibit nodular growth and papular skin disease.[4] In such cases, ultrasonography reveals an oval or circular heterogeneous hypoechoic mass that may be accompanied by posterior echo enhancement, a capsule, a clear boundary, and an abundant blood flow signal in the mass.

The main reason for missed diagnoses is that the onset of the disease is hidden and associated with no obvious symptoms.

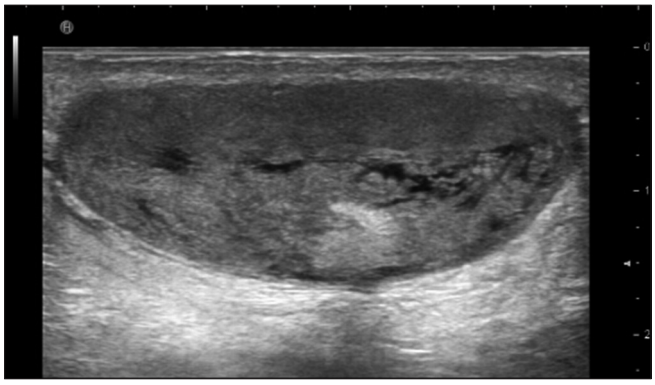

Figure 1: Two-dimensional ultrasonography showing a well-circumscribed mass with inhomogeneous hypoecho

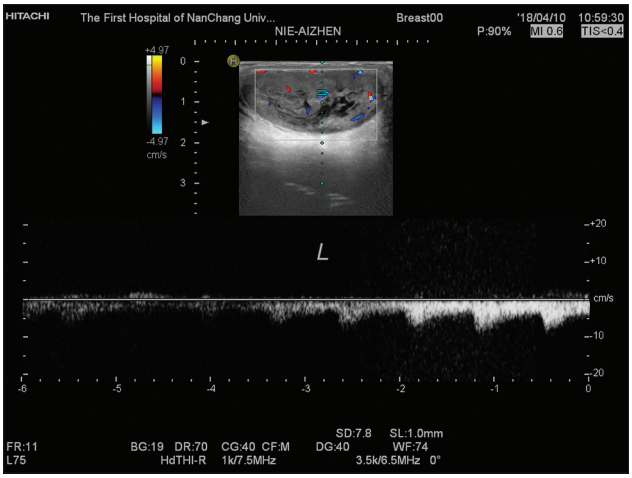

Figure 2: Color Doppler flow imaging showing abundant blood flow signals in the mass

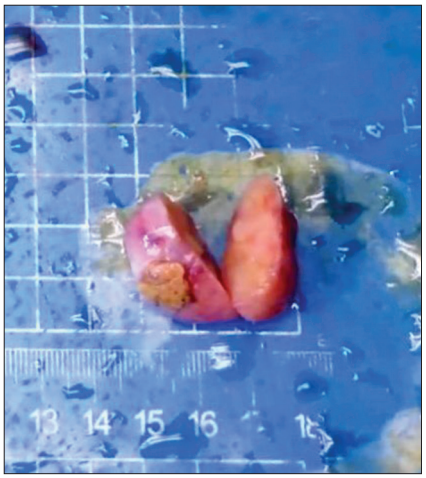

Figure 3: The gross picture of the tumor: reddish-gray tissue of size about 3.2 cm × 2.8 cm × 1.6 cm

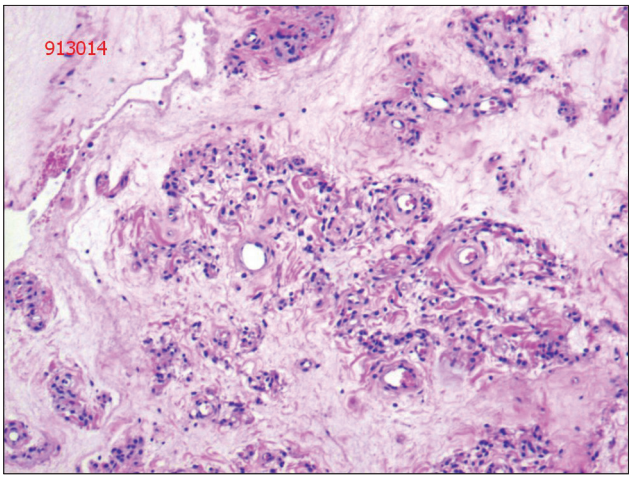

Figure 4: A pathological diagram showing multiple small blood vessels and a flaky distribution of spindle-shaped interstitial cells, a partially loose interstitium, edema, and mucus degeneration (H and E, ×100)

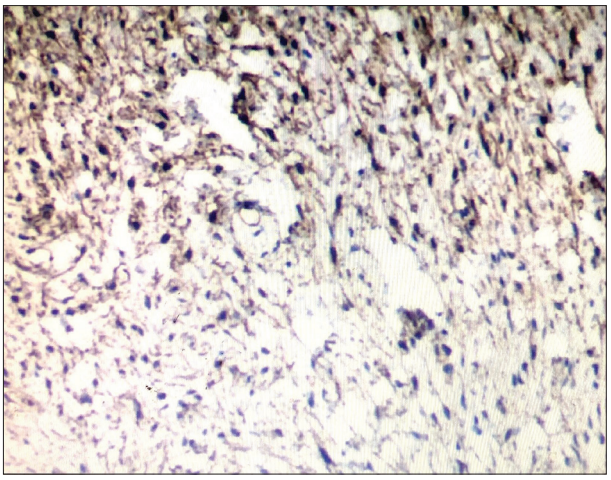

Figure 5: A immunohistochemistry pathological diagram (CD34, ×200) showing actin (vascular +), CD31 (+), CD34 (3+), CD68 (−), K (−), Des (−), EMA (−), FVIII (+), Ki-67 (2%+), S-100 (−), SMA (vascular +), Vim (3+), and β-catenin (cytoplasm +)

Most affected patients do not pay attention to those symptoms that do appear and refuse to be examined. Ultrasonography shows no obvious specificity, such as a heterogeneous low echo or abundant blood flow. The lack of SAM tumor cell components and tumor cell permutation around blood vessels leads to a loss of specificity.[7] In addition, because this is a rare disease, it has been described in few reports, and clinicians are therefore not aware of it.

Differential diagnoses mainly include epidermoid cysts, which have clinical and two-dimensional grayscale ultrasonographic manifestations similar to those of SAM, which is revealed as an uneven echo in the mass, a low high-point echo, and an enhanced echo in the rear. SAM can occur in the labia of the vulva, but such cases are easily misdiagnosed as vestibular large gland cysts because of rich lymphatic characteristic.[4] Some scholars have argued that the abundant vessels and the extremely loose myxomatous stroma observed beneath the epidermis in affected patients might promote the spread of infection.[9] Circulatory nodular fasciitis mainly occurs in the deep layer of the fascia and subcutaneous fat and often lacks capsules. Most of the tumors have no obvious blood flow signal, but small amounts of blood flow signal have been observed in a few masses.[6,10] The abundant blood flow associated with SAM can therefore be identified and diagnosed. Differential diagnosis of vascular fibroblastoma, superficial fibroblastoma, or low mucous spindle cell lipoma should be made in cases occurring in the inguinal region.[7] In addition, the features most useful for differentiating SAM from other myxoid tumors include its superficial location, stromal inflammatory infiltrate, lack of atypia, and frequent association with an entrapped epithelial component.[4]

Conclusion

A diagnosis of SAM can be made based on histopathology and immunohistochemistry, but this rare tumor should be considered in differential diagnoses.[11] Surgical resection is the main treatment method for SAM. Recurrence does not generally occur after surgery, but incomplete resection results in a rate of recurrence of 30%–40%.[6,7] In this case, it showed that increasing doctor's awareness of this rare tumor can rise the diagnostic rate.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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