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Case Report

An unusual presentation of vulvar cavernous hemangioma in a 10-year-old premenarchal girl: a rare entity

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ABSTRACT

Vascular tumors of the female genitalia are unusual. Hemangiomas are benign tumors of the blood vessels. Usually they are asymptomatic but may present as discomfort, pain and other symptoms. However, vulvar cavernous hemangioma can be seen in newborns and infants, but rarely occur in children and adult women. A 10-year old premenarchal girl presented with a painless swelling on her right labia majora since birth which was increasing gradually. On examination, a well-defined, soft, mobile and non-tender multicystic swelling was observed involving right labia majora and minora of approximately 5×6 cm in size. An ultrasound of the lesion showed a well-circumscribed, hypo echoic mass with multicystic lesion of size 40×14×44 mm arising from right labia majora with vascularity on color Doppler and venous flow on spectral pattern, suggestive of vascular malformation. Surgical excision of lesion was performed with the utmost care to preserve the surrounding healthy tissue and vulvar anatomy. Histopathological examination (HPE) report confirmed the diagnosis of a cavernous hemangioma with no evidence of malignancy. Vulvar cavernous hemangioma is an uncommon vascular tumor that can be rarely seen in pediatric patients. Imaging studies, such as ultrasound and magnetic resonance imaging (MRI), play a crucial role in confirming the diagnosis and guiding the management. Surgical excision with preservation of healthy tissue is the treatment of choice. Long-term follow-up is essential to monitor recurrence and patient's well-being.

Keywords: Vascular tumors, Vulvar cavernous hemangioma, Premenarchal girl, Labia majora

INTRODUCTION

Hemangiomas are proliferative soft-tumor lesions marked by increased cell turnover. These tumors usually appear after birth, grow rapidly and involute over the years.[1] Mainly (60%), hemangiomas are situated in the cervicofacial region.[2] Female genital tract is an unusual location for hemangiomas, they are seldom found in this site.[3-5] Among them, cavernous hemangiomas are less common then capillary hemangiomas which involute completely by the age of 5 in 80% of patients.[1] These are characterized by abnormal proliferation of blood vessels within the tissues of the vulva. Cavernous hemangiomas specifically refer to lesions composed of dilated, cavernous blood vessels. These vascular tumors can present as painless swellings on the vulva and may gradually increase in size over time. It may be asymptomatic initially but later on sexual dysfunction, pain, and cosmetic problems can occur.

In pediatric and adolescent group, the differential diagnosis of vulvar swellings includes various benign and malignant conditions. Therefore, accurate diagnosis is crucial to guide appropriate management and prevent unnecessary interventions. Imaging studies, such as ultrasound and magnetic resonance imaging (MRI), play a pivotal role in confirming the diagnosis and assessing the extent of the lesion.

This case report highlights the importance of considering vulvar cavernous hemangioma as a potential diagnosis in children presenting with vulvar swellings. It emphasizes the significance of accurate diagnosis through imaging studies and the appropriate management through surgical excision. This case report describes the presentation, diagnosis, and management of a vulvar cavernous hemangioma in a 10-year-old girl. By sharing this case, our aim is to contribute to have better understanding and knowledge of this rare condition in pediatric patients and to help in decision-making for similar cases in the future.

CASE REPORT

A 10-year old premenarchal girl who presented to the outdoor gynecology department with complaints of a painless swelling on her right labia majora. Her parents reported that the swelling had been present since birth but gradually increasing in size which added discomfort in her routine. The patient denied any history of trauma or bleeding from the lesion. Her medical history was unremarkable, and she had no known allergies.

On examination, a well-defined, multicystic swelling was observed involving right labia majora and minora. The swelling was approximately 5×6 cm in size. The lesion was non tender, soft in consistency and not fixed to the underlying tissue and did not involve the clitoris or the vaginal introitus. The remainder of the gynecological examination was unremarkable (Figure 1).

Figure 1: Gross examination of vulvar mass.

Management

An ultrasound of the lesion was performed, which showed a well-circumscribed, hypo echoic mass with multicystic lesion of size 40×14×44 mm noted to arise from right labia majora. Increased vascularity on color Doppler and venous flow on spectral pattern, with no feeding vessel seen at the base of the mass, confirmed a venous vascular channel suggestive of vascular malformation likely hemangioma. After discussing the diagnosis and treatment options like conservative management with steroid therapy, selective embolization and surgical excision with the patient's parents, final decision of excision of lesion was made. Hence, surgical excision of lesion was performed with the utmost care to preserve the surrounding healthy tissue and vulvar anatomy. An excision was done under general anesthesia, by clamping on the base of the pedicle over the labia majora and then raw area was sutured by vicryl no.1-0. The excised lesion measured 4×5 cm was sent for histopathological examination. The histopathological report showed cystically dilated tortuous blood vessels filled with RBCs below the epidermis without any nuclear atypia or abnormal mitotic activity, and was positive for CD34 on immunohistochemistry which confirmed the diagnosis of a cavernous hemangioma with no evidence of malignancy (Figure 2). The patient had an uneventful postoperative recovery and was discharged on the 5th postoperative day. The patient was scheduled for regular follow-up appointments to monitor for any signs of recurrence.

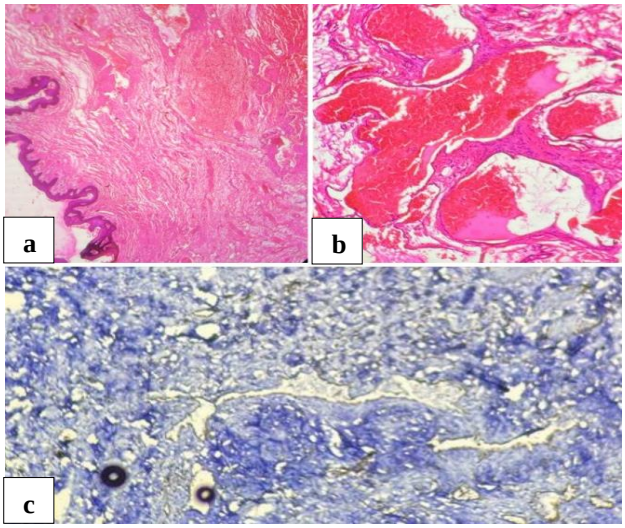

Figure 2: Microscopic findings on (a) 4X, (b) 10X, and (c) immunohistochemistry respectively.

DISCUSSION

Vulvar cavernous hemangioma is a rare vascular tumor that can occur at any age, but it is uncommon in children. This case report describes a 10-year-old girl with a painless swelling on her right labia majora that was eventually diagnosed as a cavernous hemangioma. The patient underwent surgical excision of the lesion, and the diagnosis was confirmed by histopathological examination.

Vulvar swellings in pediatric patients are more likely to be benign than malignant. However, given the rarity of vulvar hemangiomas in children, it is essential to exclude other possible diagnosis such as lymphangiomas, lipomas, angiokeratoma, mixed cutaneous tumor, soft tissue tumor, hernia, hamartoma, embryonic remnants, and other vascular malformations. Preoperative imaging with ultrasonography and MRI can help to differentiate between these conditions and help in confirming the diagnosis of a cavernous hemangioma.

The treatment ranges from monitoring if asymptomatic to radiologic or surgical intervention. Given that cavernous hemangiomas usually regress by age 5 and are asymptomatic. The surgical excision of hemangiomas of the female genitalia is reserved for symptomatic lesions refractory to medical management.[6] The procedure was performed for a curative purpose, to exclude malignancy and to obtain a definite diagnosis. A circular excision and purse-string closure are considered the best surgical approach for hemangiomas.[7] Ridhi et al reported a case of 20-year-old unmarried female with symptomatic vulvar cavernous hemangioma managed successfully with surgical excision.[8] Also, Saffudin et al did an enblock resection of entire mass followed by labial reconstruction in a 10 year old girl with right labial cavernous hemangioma.[9] Hence, surgical excision is the treatment of choice for vulvar hemangiomas, as it is effective in achieving complete removal of the lesion and minimizing the risk of recurrence. However, given the location of the lesion, careful consideration should be given to preserve the healthy tissue and minimize any damage to the surrounding structures.

The long-term follow-up of pediatric patients with vulvar hemangiomas is necessary to monitor for any signs of recurrence and ensure that the patient's quality of life is not affected. Although vulvar hemangiomas are typically benign, there is a small risk of malignant transformation. Therefore, close monitoring of pediatric patients with vulvar hemangiomas is necessary to detect any changes in the lesion and prevent any potential complications.

CONCLUSION

In conclusion, although vulvar cavernous hemangioma is rare in children, it should be considered in the differential diagnosis of vulvar swellings. Ultrasound and MRI can be useful in confirming the diagnosis, and surgical excision is the treatment of choice. Close follow-up is necessary to monitor for any signs of recurrence and ensuring patient's well-being.

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