CLINICAL CASE

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Pediatric vulvar fibrolipoma
Fibrolipoma vulvar pediátrico

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ABSTRACT
Fibrolipomas have been described as histologic subtypes of lipoma, which are rare and are considered benign mesenchymal neoplasms. They are characterized as tumors that occupy the subcutaneous plane and can appear in different parts of the body and can be round, oval or multilobulated. There is little literature on this clinical entity and its occurrence in pediatric age has not been reported. We present the case of a giant vulvar tumor in a 2-year-old girl who underwent surgery after a thorough study supported by Doppler ultrasound and whose histopathological result reported the presence of mature adipose tissue with fibrous tracts, concluding with the diagnosis of fibrolipoma of the vulva.
Key words: Vulva, fibrolipoma, Girl, prepuberty

RESUMEN
Los fibrolipomas han sido descritos como subtipos histológicos del lipoma, son poco frecuentes y se los considera neoplasias benignas mesenquimales. Se caracterizan por ser tumoraciones que ocupan el plano subcutáneo y pueden aparecer en diferentes partes del cuerpo, pudiendo tener forma redonda, ovalada o multilobulada. Existe poca literatura sobre esta entidad clínica y no se ha informado su ocurrencia en la edad pediátrica. Se presenta el caso de una tumoración vulvar gigante en una niña de 2 años que fue intervenida quirúrgicamente luego de un estudio minucioso apoyado por ultrasonido Doppler y cuyo resultado histopatológico informó la presencia de tejido adiposo maduro con tractos fibrosos, concluyéndose con el diagnóstico de fibrolipoma de la vulva
Palabras clave. Vulva, fibrolipoma, Niña, prepubertad

Introduction

Lipomas are the most frequently occurring benign soft tissue tumors, with the histologic subtype called fibrolipoma being an exceptional presentation[1]. Vulvar lipomas are rarely reported[2]. In adults, lipomatous tumors are the most common mesenchymal tumors, but in the pediatric population they account for less than 10% of soft tissue tumors[3]. The presence of fibrolipomas has been reported in different parts of the body in the pediatric population and generally in areas of adipose tissue of the neck and trunk, being peculiar to find this tumor in areas such as the nasopharynx[4], in the eyelids, ears, lips, in internal areas such as the esophagus or peritoneum and even a giant intrathoracic tumor in a 15-year-old adolescent[5].

Although lipomatous masses are rarely seen in the vulva, vulvar lipomas should be considered in the differential diagnosis of vulvar masses[6].

The clinical diagnosis is confirmed by the histopathological description of the surgical specimen after excision as a considerable collection of fibrous connective tissue with adipose tissue[7].

The etiology of lipomas has not been elucidated, but there are reports that trauma and genetic rearrangements may be involved in their development. They usually have a benign clinical course characterized by slow growth of a localized mass with little symptomatology[8].

Case report

A 2-year-old girl from Zorritos, Tumbes, on the northern coast of Peru, was brought for consultation because she presented with a genital tumor, non-painful and non-bleeding, with a sickness period of about 2 years. She was the product of a third pregnancy with normal delivery and had received complete vaccination. She denied any history of pahology.

On physical examination she was an infant weighing 9,800 g, in regular general condition, nutrition and hydration. In the preferential gynecological examination, she had Tanner stage I breasts, external genitalia with pubic hair in Tanner stage I. She showed a vulvar tumor of 7 x 5 x 4 cm adjacent to the left labium majus, with non-pulsatile vascularity, permeable vaginal orifice, vestibular and perineal erythema. The rest of the organs without alterations. Soft tissue Doppler ultrasound found an isoechogenic image of 7.02 cm x 0.97 cm in length in the left vulvar region. The power Doppler did not show any flow and the color Doppler identified an artery and vein in the lower part which irrigated the lesion with a systolic velocity of 7.7 cm/sec and a resistance index of 0.80.

Figure 1. Vulvar tumor measuring 7 x 5 x 4 cm, adjacent to the left labium majus with non-pulsatile vascularity.

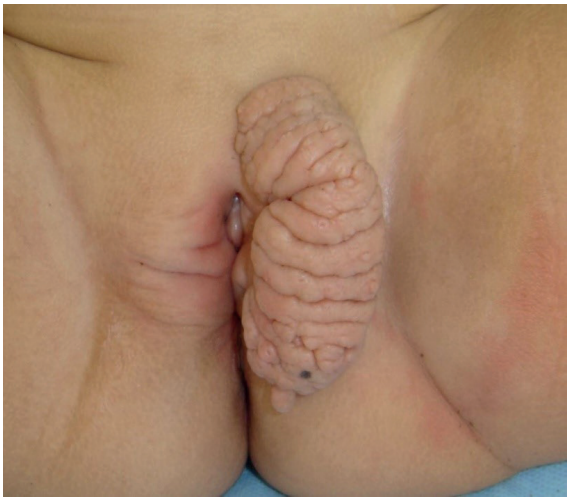
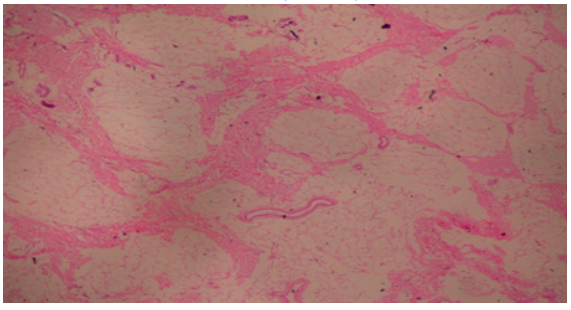


Figure 2. Mature adipose tissue with fibroconnective tissue trabeculae and foci of vascularization (HE 10X).



She underwent surgery with the diagnosis of vulvar tumor of etiology to be determined, and the tumor was excised. She received antibiotics and analgesics after the operation and was discharged two days later, with favorable evolution.

Figure 3. Mature adipose tissue with the presence of fibroconnective tissue tracts that divide it into well-delimited lobules (Masson's trichrome 10X).

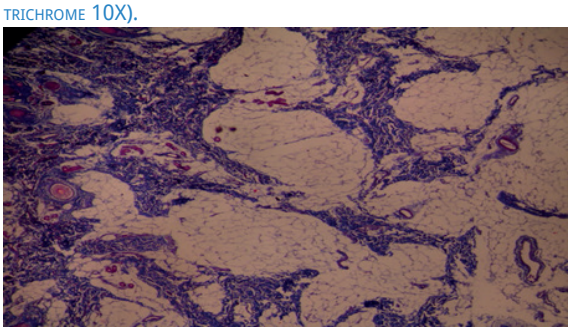


Figure 4. Ultrasonography showing an isoechogenic image of 7.02 cm x 0.97 cm located in the left vulvar region.

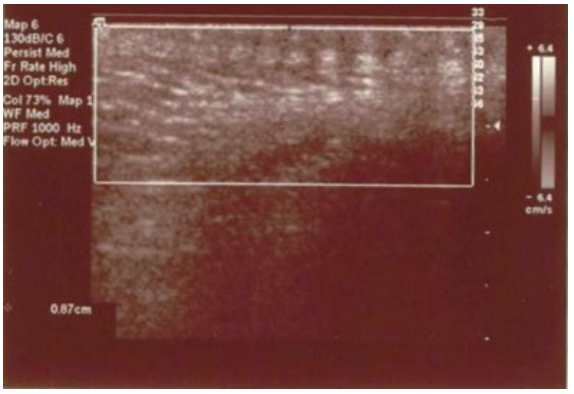
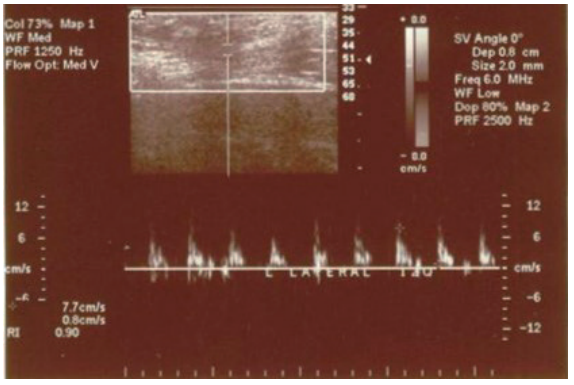


Figure 5. Ultrasonography with color Doppler flowmetry identifies artery and vein in the lower part, which irrigate the lesion with a systolic velocity of 7.7 cm/sec and resistance index of 0.80.



The result of pathological anatomy described a tumor covered by rough skin with folds of 6 x 3 x 2 cm on the surface; its section showed adipose tissue. The histological study was fibrolipoma.

Discussion

Fibrolipomas are very infrequent tumors and being asymptomatic they usually grow without medical attention until they reach a size that deforms the anatomy, as in the present case, which does not differ from those reported in adult women[1]. The presence of vulvar lipomas in their different varieties is rare in infants and there are no reports of vulvar fibrolipomas in girls[3]. In cases of lipomas, the ultrasound study shows the characteristic of the tumor, as in the present case, with normal vascularity and unique Doppler irrigation, which differentiated it from hemangiomas[9]. In this case, the resistance index of 0.8 excluded the possibility of a malignant pathology.

In cases of fibrolipomas in different areas, the surgical indication is decisive to isolate the tissue, even in pediatric patients, and with intrathoracic and intra-abdominal lesions. Following this criterion, surgical exeresis was performed in our patient without major complications and recovery in a short time[5].

Regarding histology, we found the well-known bands of mature fibrous tissue crossing the fat lobules, as mentioned in the literature[4].

In our case, the concentration of adipose tissue aggregates was similar to those of fibrous trabeculae, with true fat lobules found between continuous fibrous bundles along the tumor.

In conclusion, within the differential diagnosis of vulvar tumors in the pediatric age group, the possibility of this tumor should be considered and protocols including imaging, surgical intervention and histopathological study should be followed to confirm the diagnosis.

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