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Case report (Pages: 15365-15369)

Vulvar lipoblastoma in the utero of a female fetus: A case report

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Abstract

Background: Lipoblastoma is one of the rare masses in childhood and its diagnosis is very uncommon in the perinatal period, so we reported a case with a diagnosis of this mass during pregnancy.

Case Report: In a routine third-trimester ultrasound at 28 weeks, a hyperechoic mass in the size of 27*30mm was detected in between labia majora. However, no abnormal findings were found in perinatal care before this gestational age. Finally, after the birth, at 40 weeks of gestation, the mass was resected and now the baby is in good condition and no recurrence has occurred. There was also an anorectal anomaly associated with this mass in our patient who underwent Anorectoplasty surgery.

Conclusion: Due to the rarity of these tumors, it is important to diagnose them in the perinatal period and to investigate other associated anomalies.

Key Words: Diagnosis, Lipoblastoma, Perinatal ultrasound.

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1- INTRODUCTION

Lipoblastoma is a benign mesenchymal tumor occurring almost exclusively in infancy and early childhood, and is more frequent in boys under 3 years of age (1-4). It was first described by Jaffe in 1926 (5), as a tumor of immature fat cells, but gained acceptance as a distinctive entity in 1958, when Vellios et al. (6) described a benign lipoblastic diffuse tumor of the postnatal period resembling fetal fat and designated lipoblastomatosis. However, Chung and Enzinger (7) in 1973 described the morphologic criteria for the diagnosis of this tumor for the first time and proposed the differentiation between lipoblastoma and lipoblastomatosis. Lipoblastomas tend to occur in the subcutis, contrary to lipoblastomatosis, which is a diffuse form with a deep and infiltrative growth pattern to soft tissue and muscular planes, with a tendency to recurrence, particularly when the excision is incomplete (1). The differential diagnoses of perineal lesions other than lipoblastoma include lipoma, teratoma, infantile hemangioma, hamartoma, and liposarcoma (8). MRI is the modality of choice for evaluating tumor extension and for surgical planning; however, this imaging cannot differentiate between different adipose tumors (9). Diagnosis of these masses in perinatal time has been very rare until now. The choice of treatment for these lesions was complete surgical excision (4). Prognosis after surgery is very good and only relapses when the mass is not completely removed (10). The presence of lipoblastoma in the fetus, its diagnosis by ultrasound in the perinatal period, and its association with the anorectal malformation led us to report this rare case. This article, then, aims to introduce a case of vulvar lipoblastoma in the utero of a female fetus.

2- CASE REPORT

A 27–year-old primary gravid woman with 28 weeks gestational age was coming to Imam Khomeini Hospital in Tehran, Iran, for her prenatal care. All her previous pregnancy care has been normal up to this age and there was no previous medical history in herself or her family. They also had no family marriage and she didn’t use any drugs except supplements.

During routine third trimester ultrasound at 28 weeks’ gestation, a hyperechoic mass in the size of 27*30 mm was detected in between labia majora (**Fig. 1**), which also remained unchanged during the follow up ultrasound examination. The fetus's anatomy seemed normal in a detailed anomaly scan. Finally, the fetus was born at 40 weeks by normal vaginal delivery. The Apgar score was 9 at 1 and 10 at 5 mins after birth (**Table 1**). In the macroscopic view at birth (**Fig. 2**), a red polypoid mass in the lower part of the left labial major was seen. The lymph nodes of the inguinal nodes were not palpable. Furthermore, we objectified malposition anus in the neonate. All tests including blood cell count, renal tests, liver tests and urine tests were normal. After consulting with the parents and obtaining consent, resection of the mass and Anorectoplasty surgery were performed for the neonate, 2 days after birth. After general anesthesia during surgery, a 4-cm soft mass was removed and an invasion of deeper tissues was not observed; then, the specimen was sent for pathology. The histological report informed us of predominantly mature adipose tissue and lipoblasts with prominent paucicellular fibrous septa. At the current time, the baby is 2 months old and no evidence of mass recurrence has been reported.

Table-1: Clinical parameters of the neonate

Gestational age at the delivery time	Apgar score at 1 min	Apgar score at 5 min	Weight/g	Height/cm	Head circumference/cm	Size of vulvar mass/cm
40w+1d	9	10	3100	48	36	4*3*3

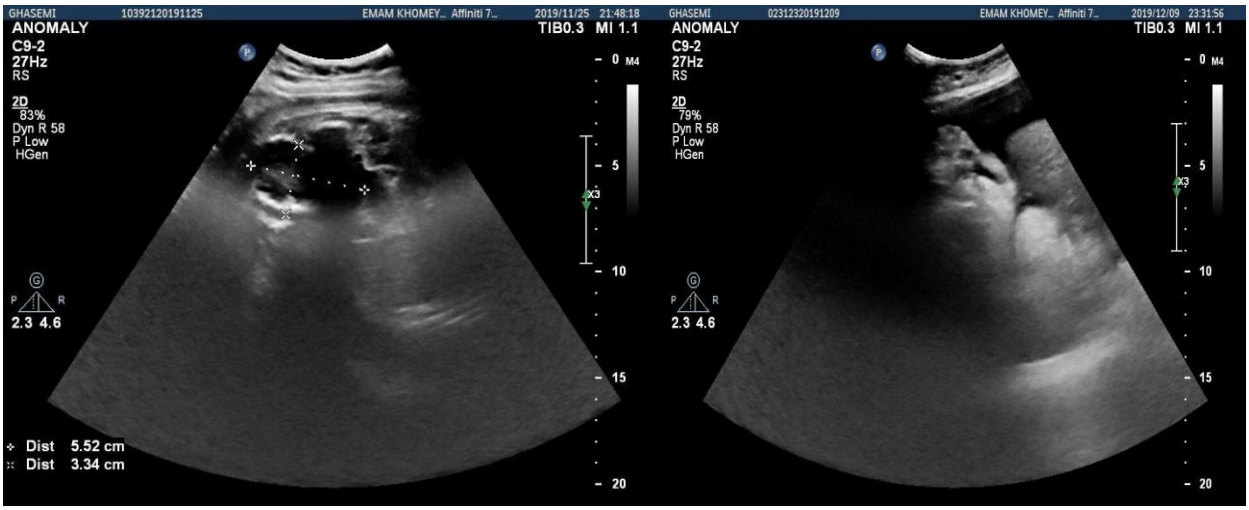

Fig. 1: A hyperechoic mass in the size of 27*30 mm was detected in between labia majora.

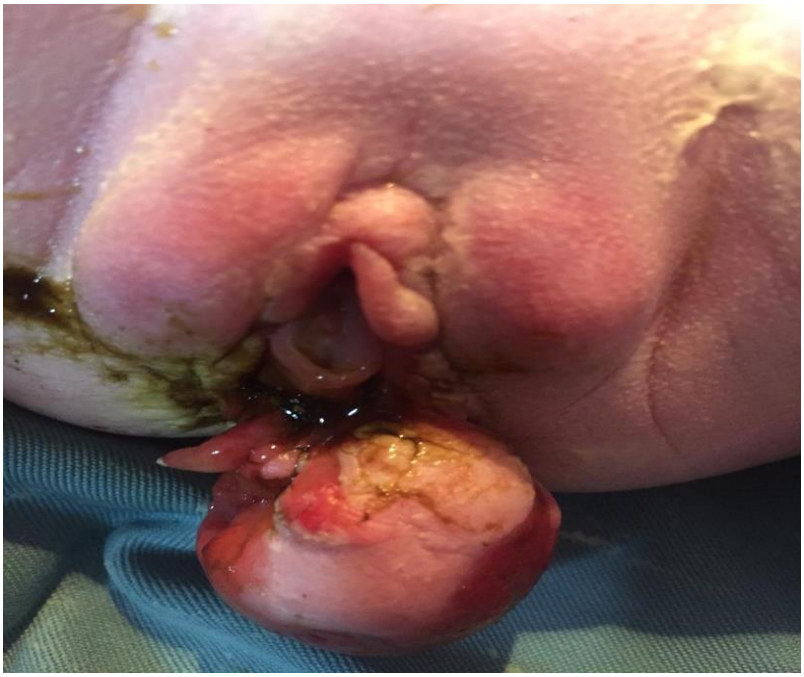

Fig. 2: the first figure of polypoid mass taken in the delivery room.

3- DISCUSSION

Perineal mass is rare and can be diagnosed with accurate prenatal ultrasound. When detected, it is important to have an accurate ultrasound to rule out other pelvic anomalies such as the uterus, bladder, kidney, and anus (5). Due to our patient's association with the anomalous anus, anus examination is necessary to rule out anorectal atresia (6). Lipoblastoma was first described by Jaffe in 1926 as a mass of fat cells in the thigh (5). In 1973, with a better description, the tumor was divided into two groups: the first type with a definite capsule called lipoblastoma and the other with a multifocal and non-capsular mass called lipoblastomatosis (7). Lipoblastoma is a rare tumor of fetal adipocytes that occurs mostly in children under 3 years of age and infants. 50% of this mass occurs in children under one year of age and 90% in children under 3 years of age (11-12). The most common site is the limb (40%) (13). The lowest incidence was seen in the neck, scrotum, retroperitoneum, and pararectal regions (14). Ultrasound and CT or MRI are diagnostic methods. Prognosis after surgery is very good and only relapses when the mass is not completely removed. The recurrence rate in these cases was about 14 to 25% after 84 months of operation (15). Cytological studies have confirmed chromosomal abnormalities in the long arm of chromosome 8 (16). This rearrangement anomaly is in the PLAG1 region on 8q12 and in the absence of PLAG1 is in the polysomal chromosome 8 (17).

4- CONCLUSION

Vulvar masses like Lipoblastoma are rare in neonates. The preferred diagnostic modalities for lipoblastoma are ultrasonography and MRI. Surgical excision is the treatment of choice. Due to the rarity of these tumors, it is important to diagnose them in the perinatal period and to investigate other associated anomalies.

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