Bilateral inguinal lipoblastomas presenting as inguinal hernias

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ABSTRACT

Lipoblastomas are rare, encapsulated tumors arising from embryonic white fat. They primarily occur in infancy and early childhood and have a male predominance. Lipoblastomas are usually located on the trunk and extremities although may develop on the head and neck, mediastinum, abdomen, and retroperitoneum. They are seldom encountered in the inguinal region. A complete resection of the tumor followed by diligent postoperative imaging are essential to detect recurrent disease in its earliest stage. Herein, we report the first case in the literature of a 1-year-old boy with bilateral inguinal lipoblastomas which presented as inguinal hernias.

Introduction

Lipoblastomas are benign well-circumscribed neoplasms composed of fetal adipose tissue that are most commonly encountered in infants and children under the age of 3 years.1,2 Lipoblastomas are more frequent in boys than girls.1,3,4 Lipoblastomas are usually observed on the trunk and extremities although have been reported on the head and neck, abdomen, mediastinum, and retroperitoneum.1,3,4 They rarely arise in the inguinal region. Local recurrence may occur in up to 80% of patients, particularly in cases of incompletely resected tumors.1 A complete resection of the tumor offers an excellent prognosis despite the large tumor size and local invasion. Lipoblastomas do not behave aggressively or metastasize although they rapidly grow. We report the first case of bilateral inguinal lipoblastomas presenting as inguinal hernias. The differential diagnosis, surgical treatment, and long-term management of lipoblastomas are discussed.

Case presentation

A 12-month-old boy (length: 31.5 in [80 cm]; weight: 23 lbs 7.3 oz [10.64 kg]; Body Mass Index [BMI]: 16.63 kg/m²) presented with a two-week history of right scrotal swelling. There was no abdominal or genital pain, nausea or vomiting, poor feeding, failure to thrive, unexplained fever, or alteration in bowel habits. Genitourinary review of systems was negative for hematuria, dysuria, foul-smelling urine, or urinary tract infection. Physical examination revealed bilateral transilluminating masses consistent with hydroceles in the inguinal areas, specifically, of the spermatic cord on the left and extending from the scrotum to the inguinal canal on the right. A scrotal ultrasound demonstrated a large right hydrocele and a left inguinal hernia containing fat (Fig. 1). The plan was to wait until the child was 18–24 months old and observe whether the hydrocele resolved.

However, the patient's left inguinal mass significantly increased in size within six months, and he underwent a left hydrocelectomy at the age of 19 months. During the procedure the spermatic cord was isolated which did not appear to have an associated hydrocele. A large mass was noted in the left inguinal area and biopsied. A frozen section demonstrated a benign lipoma. The remainder of the mass was dissected off of the pubic bone where it seemed to originate and removed en bloc (Fig. 2A). The mass measured 1.3 × 1.1 × 1.4 cm and consisted of yellow-tan fatty appearing tissue (Fig. 2B). The histologic examination revealed an adipose tumor with predominantly mature adipocytes with interspersed areas of spindled cells and occasional lipoblasts with thick septae (Fig. 2C). A diagnosis of lipoblastoma was confirmed. A physical examination performed during a follow-up visit in our office seven months later detected an easily reducible right communicating hydrocele. A scrotal ultrasound did not demonstrate any recurrence of the left lipoblastoma.

Two weeks later the boy underwent a right hydrocelectomy and excision of a yellow-tan fatty connective tissue inguinal mass measuring 5.0 × 4.5 × 1.2 cm. There was pathological evidence of mature adipose tissue with areas of increased fibrous septae/fibrosis with a small...
cluster of prominent lymphatic and fibrovascular connective tissue. The final diagnosis was a lipoblastoma. Abdominal and pelvic MRIs with and without Gadolinium contrast performed one and five months respectively following this surgery demonstrated no tumor recurrence.

Discussion

Accurately diagnosing and promptly treating patients with lipoblastoma offer the best prognosis. The most common complaint at presentation is a rapidly enlarging painless mass.1,3 The differential diagnosis includes lipoma, hibernoma, myxoid liposarcoma, fibrosarcoma, and rhabdomyosarcoma.1 Lipomas are the most common pediatric adipocytic tumors followed by liposarcomas. Lipomas lack lipoblasts or primitive mesenchymal cells and are infrequently found on the extremities. Myxoid liposarcomas are rare in children under the age of 10, demonstrate a “pulmonary edema” pattern, are always deeply situated, have a high risk of recurrence, and are malignant requiring aggressive treatment.1 Lipoblastomas are histologically characterized by their variably differentiated adipocytes, primitive mesenchymal cells, myxoid matrix, and fibrous trabeculae. The cytogenetics of lipoblastomas reveal a rearrangement of the 8q11∼q13 region which targets the PLAG1 gene.3

While complete surgical resection is the gold standard for treating lipoblastoma,3,5 the recurrence rate is between 14% and 25%.3 Close follow-up for a minimum of 5 years is recommended.5 MRI imaging every 6–12 months is the preferred method for postoperative surveillance and evaluation of recurrent disease.

The patient in our case presented to our office with the presumptive diagnosis of an inguinal hernia. Interestingly, Sarsu and colleagues reported a case of a 2-year-old girl who had a history of an asymptomatic progressively enlarging swelling at the right inguinal region for 4 months.5 She had undergone a right inguinal hernia repair 8 months earlier. Resection of the mass did not reveal a recurrence of the inguinal hernia but a lipoblastoma. Of the 12 intrascrotal lipoblastomas that have been reported in the literature, only two were accurately diagnosed preoperatively.5

Conclusion

Pediatric urologists and pediatric surgeons should consider lipoblastomas when examining children under the age of 3 years with rapidly enlarging painless masses in the inguinal region. Complete excision of the mass followed by diligent postoperative imaging are warranted to ensure detecting recurrent disease in its earliest stage.

Informed consent

The patient’s parents provided written consent to the publication of the photos and details of the case. The Chair of the University of Louisville Institutional Review Board (IRB) determined that this study does not meet the “Common Rule” definition of human subjects’ research. Therefore, our project did not require IRB review. The IRB number was 18.1356.

Declarations of interest

None.

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