Clinical Case Report

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A large retroperitoneal lipoblastoma

A case report and literature review

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Abstract

Introduction: Lipoblastoma is a rare benign soft tissue tumor that occurs most commonly in infants and children. However, retroperitoneal lipoblastomas are rare, occurring in <5% of cases. We report a case of large retroperitoneal lipoblastoma and the largest collection of known retroperitoneal lipoblastomas in children in the literature.

Case presentation: A 3-year-old girl presented with left abdominal mass. Magnetic resonance imaging (MRI) revealed a soft tissue mass measuring 12 × 8 × 6 cm in the retroperitoneal region. The mass had a clearly defined margin and a reticular pattern with an interposing fat component. Based on these findings, the mass was suspected to be a soft-tissue tumor, most likely lipoblastoma.

Laparotomy with resection of the retroperitoneal mass was performed. The tumor was easily dissected from the retroperitoneal space without injury to surrounding structure.

A histopathological examination demonstrated the mature proliferation of adipocytes and spindle-shaped cells separated by fibrovascular septa accompanied by myxoid changes. The cells were separated into lobules by septa, and areas of immature adipocytes showing a signet-ring or multivacuolar appearance were present at the periphery. Histopathological diagnosis was lipoblastoma. Follow-up at 6 months revealed no evidence of recurrence.

Conclusion: Retroperitoneal lipoblastoma is rare and tends to be large in size when diagnosed at presentation. Complete resection should not be delayed, as impingement on the surrounding structures is imminent.

Abbreviations: MRI = magnetic resonance imaging, PLAG1 = pleomorphic adenoma gene 1.

Keywords: lipoblastoma, pediatrics, retroperitoneal

1. Introduction

Lipoblastoma is a rare benign soft tissue tumor that occurs most commonly in infants and children.[1,2] The vast majority are detected in children <3 years of age, with >80% of cases occurring before the age of 3 and 40% before the age of 1. Approximately 70% of these tumors occur in the extremities, trunk, head, and neck. However, retroperitoneal lipoblastomas are rare, occurring in <5% of cases.[1,2]

We herein report a case of a large, retroperitoneal lipoblastoma and the largest collection of known retroperitoneal lipoblastomas in children in the literature.

2. Case report

A 3-year-old girl presented with left abdominal swelling. There was no evident congenital abnormalities at birth nor any familial history of disease.

On a physical examination, the child had a soft, moderately distended left abdomen that was not tender when palpated. The hemoglobin, alphafetoprotein, and beta-human chorionic gonadotropin levels were normal. Abdominal ultrasound showed a heterogeneous soft tissue mass measuring 12 × 8 × 6 cm. Magnetic resonance imaging (MRI) revealed a well-encapsulated soft tissue mass lesion in the retroperitoneal region. The mass had a clearly defined margin and a reticular pattern with an interposing fat component showing a reduced signal on fat suppression inversion recovery imaging (Fig. 1A and B). Based on these findings, the mass was suspected to be a malignant soft-tissue tumor, most likely lipoblastoma.

Laparotomy with resection of the retroperitoneal mass was performed under general anesthesia. A well-encapsulated mass was loosely attached to the retroperitoneum. The tumor was easily dissected from the retroperitoneal space without injury to adjacent structures. It was well circumscribed with a thin, fibrous capsule and a yellow, lobulated fatty parenchyma separated by thin fibrous septa with punctate vessels (Fig. 2).

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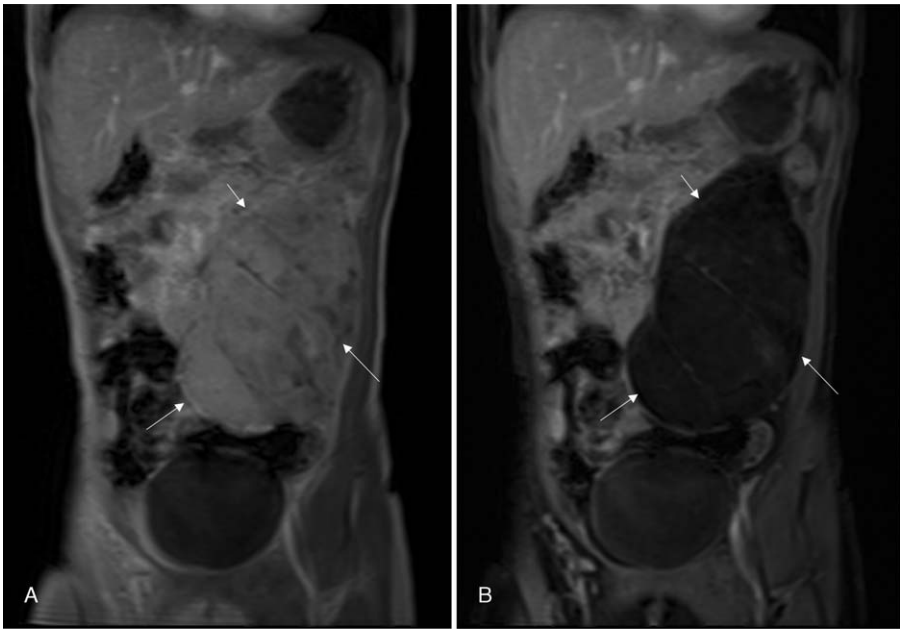

Figure 1. (A) T1-weighted magnetic resonance imaging coronal view images obtained for a large, well-circumscribed mass that appeared to be multilobulated causing a significant mass effect in the left retroperitoneal region (close arrows). (B) The fat component showed a reduced signal on fat suppression inversion recovery imaging (close arrows).

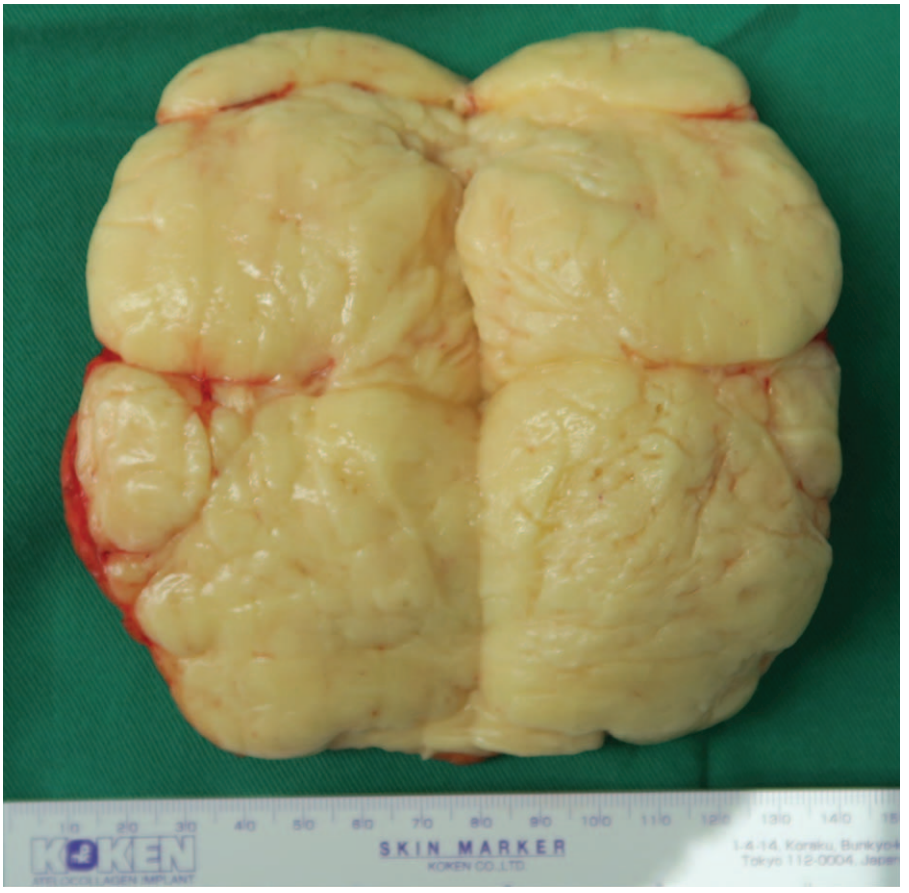

Figure 2. The cut surface of the lipoblastoma revealed a yellow-tan, lobulated mass with no areas of necrosis or hemorrhaging.

A histopathological examination demonstrated the mature proliferation of adipocytes and spindle-shaped cells with bland nuclei separated by fibrovascular septa accompanied by myxoid changes. The cells were separated into lobules by septa, and areas of immature adipocytes showing a signet-ring or multivacuolar appearance were present at the periphery (Fig. 3A and B). A chromosome analysis of the tumor showed no pleomorphic adenoma gene 1 (PLAG1) oncogene rearrangements.

Follow-up at 6 months revealed no evidence of recurrence.

3. Discussion

Lipoblastomas are soft tissue tumors composed of embryonal/fetal fat and characterized by a benign nature, early presentation, male predominance, and rapid growth.[1–4] The long-term prognosis for lipoblastoma is usually excellent.[5] Metastases have never been reported, and the recurrence rates have been reported to range from 9% to 22%, which is attributed to incomplete initial excision of the tumor.[3,6,7] Some reports suggest that lipoblastoma may spontaneously mature or

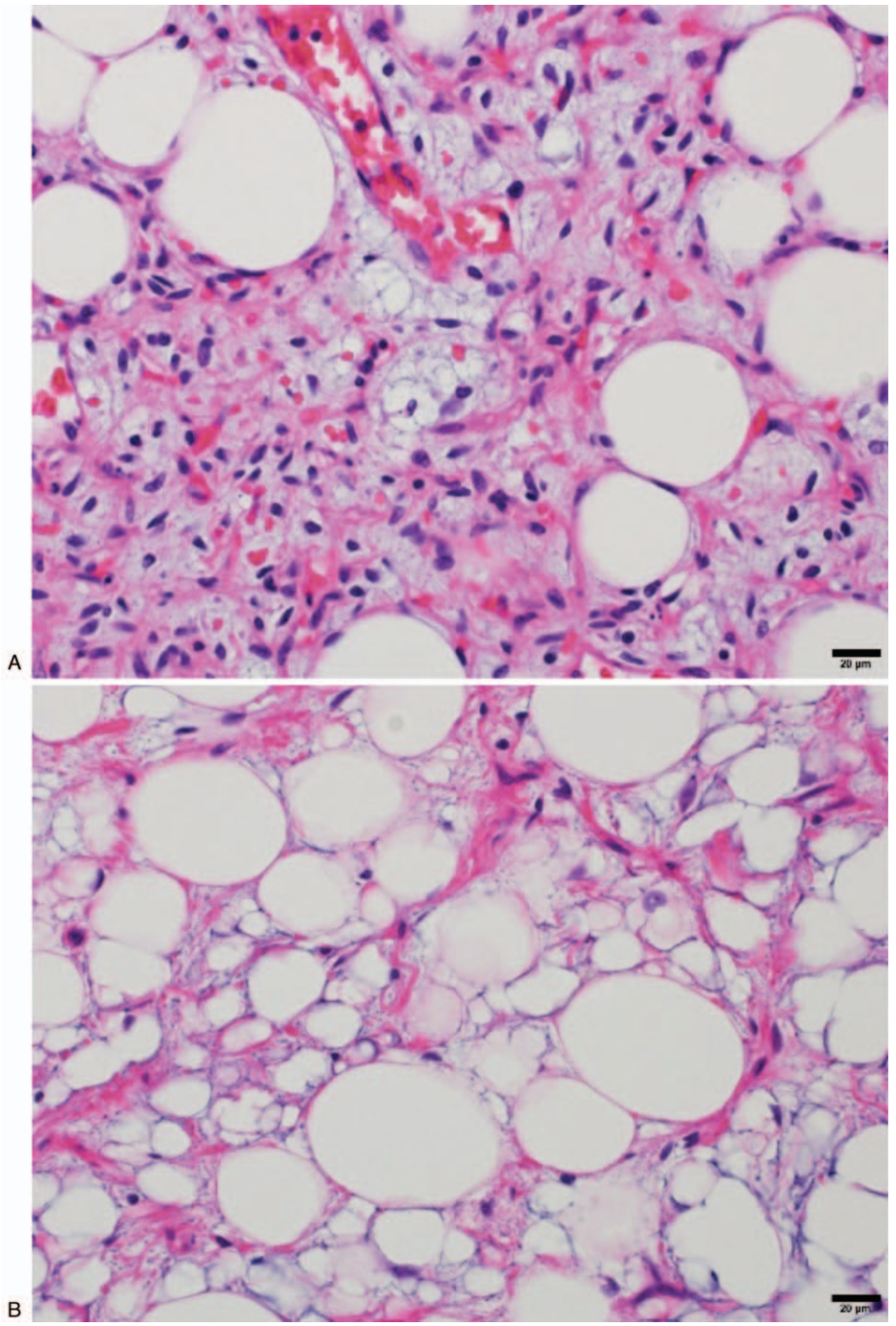

Figure 3. A histopathologic examination revealed areas with myxoid adipocytes and lipoblasts and other areas with more mature adipocytes with clear cytoplasm (bar = 200 μm) (A). The mass was separated into lobules by well-defined fibrous septa. Higher magnification of the tumor cells readily demonstrated the myxoid character of the lipoblasts' cytoplasm (bar = 20 μm) (B).

Table 1

Reported cases of retroperitoneal lipoblastoma (n = 26).

Report	Year	Sex	Age at excision	Onset	Size, cm	Location	Preoperative diagnosis	Pathological diagnosis	Operation	Follow-up	Recurrence	Complication
Chung[3]	1973	NR	NR	NR	NR	NR	NR	Lipoblastomatosis	NR	NR	NR	NR
		NR	NR	NR	14 × 11 × 9	NR	NR	Lipoblastomatosis	NR	NR	NR	NR
Tanyel[14]	1986	F	3 y	L hypochondrial mass	ϕ8	L retroureteropelvic lesion	Lipoma	Lipoblastoma	CR	6 mo	—	—
Jimenez[15]	1986	M	12 y	R abdominal mass, leg venous stasis	19.5 × 12 × 6	R retroperitoneal lesion	Retroperitoneal sarcoma	Lipoblastoma	CR	5 y	—	Chronic venous stasis of both legs
		M	7 mo	R abdominal mass	15 × 10 × 9	R retroperitoneal lesion	Wilms tumor or neuroblastoma	Lipoblastoma	CR	4 y	—	—
Fisher[16]	1981	F	7 mo	R abdominal mass	NR	R retroperitoneal lesion	NR	Lipoblastomatosis	NR	NR	NR	NR
St Omer[17]	1992	M	5 y	Abdominal pain and distension, vomiting	NR	L retroperitoneal lesion	NR	Lipoblastoma	CR	NR	NR	—
Chi[2]	1995	M	1 y	Abdominal distension	16 × 12 × 10	R retroperitoneal lesion	NR	Lipoblastomatosis	CR	NR	NR	NR
Collins[18]	1997	M	34 mo	Vomiting	ϕ21	L retroperitoneal lesion	NR	Lipoblastoma	NR	1 y	—	NR
Huang[19]	1998	M	8 mo	Abdominal distension, constipation	NR	Lower back mascle, retroperitoneum and spinal canal	NR	Lipoblastomatosis	IR	Dead	—	Septic shock
Pollono[7]	1999	M	5 mo	R lower abdominal mass	14 × 12 × 9	R retroperitoneal lesion	Lipoblastoma (FNAC)	Lipoblastoma	CR	5 y	—	NR
		F	19 mo	Urinary tract infection	18 × 9 × 6	L retroperitoneal lesion	Lipoblastoma (FNAC)	Lipoblastoma	CR	11 mo	—	NR
Dilley[9]	2001	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
		NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
		NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
Chun[20]	2001	M	29 mo	Abdominal distension	19.5 × 16 × 12.5	Retroperitoneum	NR	lipoblastoma	CR	9 y	—	NR
Dokucu[21]	2003	M	12 mo	Abdominal mass, R lower extremity swelling	10 × 12	Lower part of abdominal cavity	NR	Lipoblastoma	CR	15 mo	—	Internal R iliac artery and vein reconstruction
McVay[4]	2006	M	17 mo	NR	ϕ17	Midline retroperitoneal lesion	Neuroblastoma	Lipoblastoma	CR*	NR	—	NR
Speer[22]	2007	NR	NR	NR	NR	NR	NR	Lipoblastoma	CR	NR	—	NR
		NR	NR	NR	NR	NR	NR	Lipoblastoma	CR	NR	—	NR
Kok[10]	2010	F	4 y	Abdominal distension	25 × 20 × 7	L retroperitoneal	Cystic mass	Lipoblastoma	CR	36 mo	—	—
Api[23]	2010	F	22 d	Screening for fetal malformation	6.2 × 3 × 3	R retroperitoneal lesion	Hemangioma or lymphangioma	Lipoblastoma	IR	10 mo	—	NR
Burchhardt[1]	2012	F	2 y	Palpable abdominal mass	15 × 11 × 8	R lower quadrant	Lipomatous tumor of retroperitoneal origin	Lipoblastoma	CR	2 y	—	—
Susam-sen[24]	2017	M	11.5 mo	Stomach ache, swelling in the abdominal region, constipation	9 × 5	NR	Lipoma, lipoblastoma	Lipoblastoma	CR	81 mo	—	—
		M	29.5 mo	Stomach ache, abdominal mass, constipation	13 × 10	L retroperitoneal	Lipoblastoma, liposarcoma	Lipoblastoma	CR	2 mo	—	—
Our case	2018	F	3 y	Palpable abdominal mass	12 × 8 × 6	L retroperitoneal adrenal lesion	Lipomatous tumor of retroperitoneal origin	Lipoblastoma	CR	6 mo	—	—

CR = complete resection, F = female, FNAC = fine-needle aspiration cytology, IR = incomplete resection, L = left, M = male, NR = not reported, R = right.

* Staged resection.

regress.[5] Coffin et al[8] reported the maturation of an incompletely excised lipoblastoma into a mature lipoma. The majority of lipoblastomas have been found on the trunk or extremities, with only 5% of cases reported in the reoroperitoneum.[1]

Retroperitoneal lipoblastoma is difficult to diagnose preoperatively. The differential diagnosis of the tumor is broad and includes sarcomas, neuroblastomas, and teratomas. MRI has the highest sensitivity for the pathology of the tumor, as the increased vascularity in lipoblastomas compared with lipomas presents as a lower intensity on T1-weighted images.[9] MRI is therefore the current recommended modality for follow-up, particularly in cases of incomplete resection or prior recurrence.[9] Resection and a pathological examination are ultimately needed for achieving a definitive diagnosis.

The recent use of cytogenetics has proven to be helpful for the diagnosis, as translations involving the long arm of chromosome 8, particularly 8q11-13, with or without PLAG1 oncogene rearrangements, have been found to be associated with lipoblastomas.[10–13] This rearrangement targets the PLAG1 gene and has been reported in 82% of lipoblastomas, only 3% of conventional lipomas and never in myxoid liposarcoma.[12] However, in the present case, PLAG1 oncogene rearrangements were not found.

A literature search was performed using the electronic database "PubMed" for all patients' reports in the English literature with retroperitoneal lipoblastoma using the search term "retroperitoneal lipoblastoma." Relevant data were extracted from all primary reported patients. Patients included in multiple reports were used only once for the analysis. All patient data were combined to create this report. There have been 26 cases of pediatric retroperitoneal lipoblastomas, as shown in Table 1.[1–4,7,10,14–24] No recurrences were reported. Clinical features of the current case have been consistent with those previously reported, including her age, sex, onset, size and location of tumor, preoperative diagnosis, pathological diagnosis operation, and complications.

Nineteen of the 26 cases, including the current case, have been described in detail in the literature. These patients were 12 males and 7 females. The age at presentation ranged from 22 days to 12 years (median 17 months). The tumors ranged in longitudinal length from 10 to 25 cm except for 1 neonatal patient. The retroperitoneal tumors were always large in size and weight[1,2] due to their location. Tumors were located at the right side in 7 patients, at the left side in 7 patients, and at the midline in 2 patients. The others did not report the side of retroperitoneal in details. One patient was diagnosed at a screening for fetal malformations.[23] Five were diagnosed with lipoblastmatosis. In the review, almost all of the patients required complete resection; one patient had intraspinal extension requiring 3 separate surgeries for complete resection.[4] However, 2 patients were resected incompletely due to multiple retroperitoneal lesions of lipoblastomatosis in one and adhesion to the kidney in the other.[19,23] Three cases reported complications associated with tumor resection.[15,19,21] One patient developed chronic bilateral venostasis after excision,[15] one required internal iliac artery and vein reconstruction at resection,[21] and one developed septic shock and died.[19] MRI cannot solely be relied on for the diagnosis of lipoblastoma. However, it was almost used for follow-up and evaluation of recurrent tumors.

Appropriate length of follow-up for lipoblastoma remains controversial. Various lengths of follow-up have been suggested, including 2 years,[9] 3 years,[22] and 5 years.[4] The average time to recurrence was noted to be 3 years (range, 4 months to 10 years).[22]

This report highlighted that there was no recurrence after complete resection. Moreover, complete resection should not be delayed so that the surrounding structures were not injured.

In conclusion, retroperitoneal lipoblastoma tends to be large in size (>10 cm) when diagnosed at presentation. However, the vast majority of all resections were well tolerated with benign postoperative courses. Complete resection should not be delayed, as impingement on the surrounding structures is imminent.

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