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).
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 \(\mu\)m) (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 \(\mu\)m) (B).
| 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 |
| Tanig[14] | 1986 | F | 3 y | L hypochondrial mass | 48 | L retroperitoneal lesion | Lipoma | Lipoblastoma | CR | 6 mo | — | — |
| Jimenez[16] | 1986 | M | 12 y | R abdominal mass, leg venous stasis | 19.5 × 12 × 6 | R retroperitoneal lesion | Retropertoneal 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[7] | 1992 | M | 5 y | Abdominal pain and distension, vomiting | NR | L retroperitoneal lesion | NR | Lipoblastoma | CR | NR | NR | NR |
| Chi[16] | 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 | 421 | L retroperitoneal lesion | NR | Lipoblastoma | NR | 1 y | — | — |
| Huang[20] | 1998 | M | 8 mo | Abdominal distension, constipation | NR | Lower back muscle, retroperitoneum and spinal canal | NR | Lipoblastomatosis | IR | Dead | — | Septic shock |
| Polkono[7] | 1999 | M | 5 mo | R lower abdominal mass | 14 × 12 × 9 | R retroperitoneal lesion | Lipoblastoma (FNAC) | Lipoblastoma | CR | 5 y | — | NR |
| Dilley[8] | 2001 | NR | NR | Urinary tract infection | 18 × 9 × 6 | L retroperitoneal lesion | Lipoblastoma (FNAC) | Lipoblastoma | CR | 11 mo | — | 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 outer quadrant of abdominal cavity | NR | Lipoblastoma | CR | 15 mo | — | — |
| McKay[3] | 2006 | M | 17 mo | NR | 417 | Midline retroperitoneal lesion | Neuroblastoma | Lipoblastoma | CR | NR | — | NR |
| Speer[23] | 2007 | NR | NR | Abdominal pain and distension | NR | NR | NR | Lipoblastoma | CR | NR | — | NR |
| Kok[17] | 2010 | F | 4 y | Abdominal distension | 25 × 20 × 7 | L retroperitoneal lesion | Cystic mass | Lipoblastoma | CR | 36 mo | — | — |
| Api[37] | 2010 | F | 22 d | Screening for fever malformation | 62 × 3 × 3 | R retroperitoneal lesion | Hemangioendothelioma | Lipoblastoma | IR | 10 mo | — | NR |
| Buchharof[9] | 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 lesion | Lipoblastoma, liposarcoma | Lipoblastoma | CR | 2 mo | — | — |
| Our case | 2018 | F | 3 y | Palpable abdominal mass | 12 × 8 × 6 | L retroperitoneal adenome | 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.

[1] Staged resection.
highest sensitivity for the pathology of the tumor, as the increased
includes sarcomas, neuroblastomas, and teratomas. MRI has the
atively. The differential diagnosis of the tumor is broad and
other.[19,23] Three cases reported complications associated with
lipoblastomatosis in one and adhesion to the kidney in the
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 rear-
rangements, have been found to be associated with lipoblasto-
mas.[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.[13,15] 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 “retroperito-
neal 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,
position, 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[2,12]
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
details. One patient was diagnosed at a screening for fetal
malformations.[23] Five were diagnosed with lipoblastomatosis. 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
umor 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,[9,19] 3 years,[22] and 5 years.[4] The average time
to recurrence was noted to be 3 years (range, 4 months to 10
years).[23] 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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