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Case Report

A Rare Case of Retroperitoneal Lipoblastoma with Review of Literature

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Abstract

Lipoblastoma is a rare benign pediatric mesenchymal neoplasm. Common locations include extremities and trunk, while retroperitoneal location is very rare and only <30 cases have been reported worldwide. Retroperitoneal lipoblastomas tend to present as a large, rapidly growing abdominal mass that can be challenging to diagnose preoperatively. Thorough investigations and histopathological examination are imperative to arrive at the correct diagnosis, thus ensuring appropriate treatment to the patient. Here, we report a case of 2-year-old boy who presented with a large abdominal mass compressing the adjacent structures. On resection and histopathological evaluation, a diagnosis of lipoblastoma was made.

Key words: Abdominal mass, Lipoblastoma, Pediatric, Retroperitoneal

INTRODUCTION

Lipoblastoma is a rare benign pediatric neoplasm of embryonal fat seen mostly in infants and children below 3 years of age.[1] The most common sites include trunk and extremities, whereas retroperitoneal lipoblastomas are relatively rare, accounting for <5% of all cases.[2-4] The clinical presentation of retroperitoneal lipoblastomas is similar to malignant tumors, most commonly liposarcoma and teratoma, therefore posing a diagnostic difficulty both clinically as well as radiologically. Lipoblastoma is benign and carries an excellent prognosis on complete excision. Thus, histopathology plays a major role avoiding overdiagnosis and aggressive treatment.

CASE REPORT

A 2-year-old boy presented with gradually progressive abdominal mass for 1 year, associated with abdominal pain, constipation, and vomiting for 1 month along with dribbling of urine and difficulty in micturition for 3 weeks.

The child was initially evaluated in another institute, where imaging studies, biopsy of the mass, and an attempt at surgical resection were made. Ultrasound abdomen showed a large ill-defined irregular echogenic solid mass measuring about 11.7 × 10 cm in the right lumbar region, extending up to right iliac fossa suggestive of neuroblastoma. Computed tomography (CT) abdomen revealed a large almost rounded soft-tissue mass of mixed attenuation in the right lumbar and right iliac region, causing compression of the right hip and displacing bowel loops, suggestive of large germ cell tumor – possibly teratoma. Ultrasound-guided fine-needle aspiration cytology showed fragments of spindle cells, fatty tissue, and fibroid tissue fragments suggestive of spindle cell tumor or nerve sheath tumor. Biopsy of the mass showed atypical epithelial cells with inconclusive diagnosis. Attempt for surgery at the earlier institute resulted only in incisional biopsy as the mass was considered inoperable. HPE showed fibrofatty tissue admixed with few mononuclear cells. No evidence of granuloma or malignancy was noted.

On examination of the child in our hospital, the abdomen was distended by a large palpable mass measuring around

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13 × 12 cm, extending from the right iliac fossa to the right hypochondrium, crossing midline and hard in consistency. Dilated veins and scars were noted in the abdominal wall. Serum alpha-fetoprotein, beta-human chorionic gonadotropin, and urine vanillylmandelic acid were normal.

Positron emission tomography-CT revealed an encapsulated heterogenous mass lesion measuring 18.9 × 13.5 × 15.5 cm in the right anterior pararenal space, with predominantly fat components with interspersed soft-tissue densities. The lesion was seen to cause mass effect on the liver and displacing the right kidney and ureter causing moderate hydroureteronephrosis as well as displacing the bowel loops to the left side, suggesting a diagnosis of liposarcoma [Figure 1].

Exploratory laparotomy was done and intraoperatively, the tumor was found to be adherent to the psoas muscle. The tumor was excised and sent for histopathological examination. The specimen weighed 2.2 kgs and measuring 20 × 19 × 9 cm. External surface appeared gray-yellow and smooth. On cut surface, it appeared pale, gray-white to gray-yellow, and firm in consistency [Figure 2]. On histopathology, multiple sections from the mass showed a fairly circumscribed, partially encapsulated lipomatous lesion showing adipocytes arranged in lobular pattern separated by fibrous stroma composed of oval to spindled cells. Few multivacuolated lipoblasts were noted and focal myxoid areas seen. No evidence of curvilinear blood vessels/atypia/mitoses/necrosis or hemorrhage was seen. A diagnosis of lipoblastoma was made considering the age of the patient [Figure 3a and b].

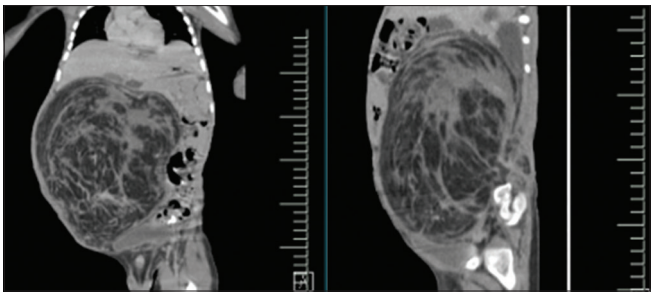

Figure 1: Positron emission tomography-computed tomography images showing a large heterogenous mass lesion in the right anterior pararenal space

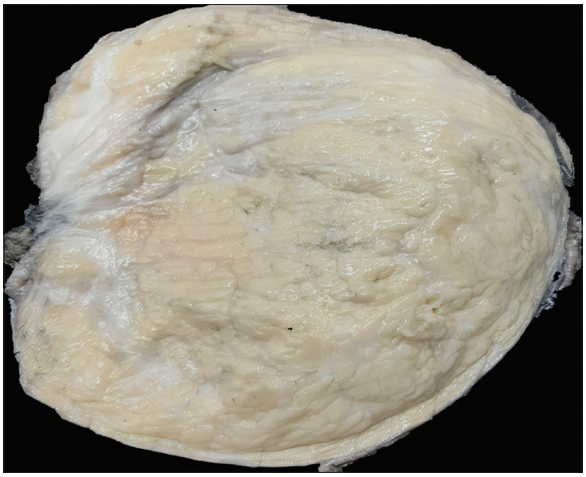

Figure 2: Cut surface of lesion appearing pale, gray-white to gray-yellow and firm

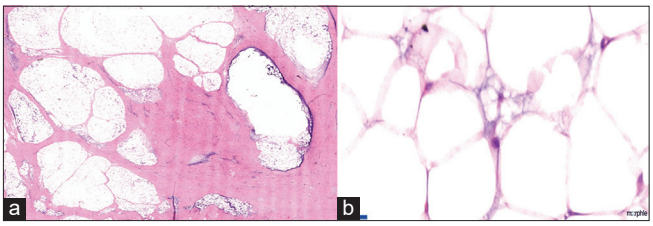

Figure 3: (a) Hematoxylin and eosin (H and E), 40×: Lobular pattern of arrangement of adipocytes, separated by fibrous stroma composed of spindled out cells. (b) H and E, 400×: Lipoblast with multivacuoalted cytoplasm and central round nucleus

DISCUSSION

Lipoblastoma is a rare benign neoplasm of embryonal white fat, which can present as a localized well-circumscribed tumor or as a diffusely infiltrative, multicentric form called lipoblastomatosis, seen predominantly in deeper tissues.[2,5,6] It occurs mostly in infancy and early childhood, with 75–90% of cases occurring before 3 years. Sporadic examples have also been reported in older children and adolescents and very rarely in adults. Most studies have reported a slight predilection for this tumor in males.[2] Most commonly involved sites include the subcutaneous tissues of trunk and extremities. Lipoblastoma may arise in a variety of locations including abdomen, mesentery, retroperitoneum, pelvis, inguinoscrotal or labial region, perineum, mediastinum, and head and neck region.[2,7-9] However, retroperitoneal location is very rare, accounting for <5% of all cases with less than 30 well-documented cases reported worldwide.[3,9] Recent studies by Gerhard-Hartmann *et al*. as well as Sakamoto *et al*. documented a total of 23 and 26 cases, respectively, reported worldwide.[9,10]

Lipoblastomas are typically 2–5 cm in diameter; however, retroperitoneal lipoblastomas are reported to be rapidly growing and commonly attaining an enormous size. Studies by Burchhardt *et al*. and Kok *et al*. have documented cases of retroperitoneal lipoblastomas measuring up to 25 cm in greatest dimension.[2,3,11] Lipoblastomas commonly present as painless superficial soft-tissue masses; however, some large abdominal or mediastinal tumors can impinge on and compress adjacent organs, nerves, or blood vessels, causing pain as well as compromised function and also have a propensity for local invasion.[12] Hence, such a presentation, especially in retroperitoneal location,

can often mimic malignant tumors clinically as well as radiologically. Pre-operative diagnosis of retroperitoneal lipoblastoma can be very challenging and the major differential diagnoses include sarcomas, Wilm's tumor, neuroblastoma, and teratoma. Investigative work-up can include assay of tumor biomarkers, ultrasonography to detect tumors with high fluid content and vascularity, CT scans for mass characterization and lymphadenopathy detection, and magnetic resonance imaging (MRI) for anatomic delineation.[2,3] Although radiological imaging can be helpful in narrowing down the diagnosis to lipomatous tumors, accurate differentiation of immature, and mature adipocytes on any imaging modality is difficult.[5] However, MRI has been reported to be the best radiological method due to its increased sensitivity for lipoblastoma which shows increased vascularity that presents as a lower intensity on T1-weighted images in comparison with lipoma.[13] Due to the various limitations of imaging modalities, an accurate pre-operative diagnosis is rarely made and surgical resection followed by histopathological examination remains the key for the definitive diagnosis of lipoblastoma.[5,10]

On gross examination, lipoblastomas are typically soft, lobulated, yellow-white to tan masses appearing paler than the ordinary lipoma, and may have myxoid nodules, cystic space or fat nodules separated by fibrous septa.[2,11] Histopathologic examination characteristically demonstrates lobular architecture with sheets of adipocytes separated by fibrovascular septa. The adipocytes show a spectrum of maturation, ranging from primitive, stellate to spindled mesenchymal cells to multivacuolated, or small signet ring lipoblasts to mature adipocytes.[2] Prominent myxoid change with a plexiform vascular pattern can also be seen reminiscent of myxoid liposarcoma, making it a close differential diagnosis. However, myxoid liposarcoma shows minimal lobulation with centrifugal loss of maturation and increased cellularity with focal pleomorphic nuclei, whereas lipoblastomas lack nuclear atypia altogether. Other major histological differential diagnosis includes well-differentiated liposarcoma/atypical lipomatous tumor which has prominent spindle cells with large, deep-staining nuclei, and marked nuclear enlargement or pleomorphism. However, both myxoid liposarcoma and well-differentiated liposarcoma are commonly seen in adults and very rare in pediatric age group with cases typically occurring in children older than 5 years. Benign tumors such as lipoma, fibrolipoma, hibernoma, lipofibromatosis, and fibrous hamartoma of infancy share common histological features with lipoblastoma, but they lack lipoblasts.[14]

Recent use of cytogenetic analysis for detection of PLAG1 gene rearrangement is useful for the accurate diagnosis of lipoblastoma in cases with suspicion of myxoid liposarcoma. [7] Three fusion partner genes known in relation to PLAG1 in lipoblastoma are: HAS2 at 8q24.1, COL1A2 at 7q22, and RAD51L1 at 14q24. In addition, two novel fusion genes COL3A1- PLAG1 and RAB2A-PLAG1 have been identified by Yoshida *et al*.[15] In contrast, myxoid liposarcomas show characteristic t(12;16) translocation. Cytogenetic analysis was not done in our case as there was no diagnostic suspicion.

Lipoblastoma has an excellent prognosis after complete surgical excision, despite rapidly enlarging size and tendency for local invasion.[7] In cases where resectability is questionable due to involvement of critical structures, a staged surgical approach has been recommended by Speer *et al*.[3,12] No cases of metastases have been reported and the rate of recurrence is 13–46%, with recurrence predominantly attributable to incomplete excision.[2]

CONCLUSION

Retroperitoneal lipoblastoma is a rare benign infantile and pediatric neoplasm. It can be diagnostically challenging, both clinically and radiologically; thus, histopathological features play a key role at clinching the diagnosis. As it presents as a rapidly enlarging abdominal mass mimicking other soft-tissue tumors, accurate diagnosis and timely management are essential.

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