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Adolescent Paraspinal Lipoblastoma: A Case Report

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Learning Point of the Article:

While most lipoblastomas are diagnosed in infancy, they can occur in adolescents.

Abstract

Introduction: Lipoblastomas are fatty tumors of mesenchymal origin with a peak incidence between the ages of 5 and 6 years. Several cases of lipoblastomas have been reported in the pediatric population, with few in the paraspinal musculature. We present a case of a paraspinal lipoblastoma in an adolescent.

Case Report: An 11-year-old male presented with an asymptomatic thoracic mass. Imaging revealed a 5 cm3 lesion involving the left fourth and fifth ribs with extension into the T4-T5 neural foramen. Biopsy confirmed lipoblastoma. Marginal resection was performed without complication, and there was no recurrence at 2 years.

Conclusion: Lipoblastomas are rare mesenchymal tumors whose differential diagnosis includes lipomas and liposarcomas. While benign, lipoblastomas grow rapidly and can cause mass effect on neurovascular structures. Most cases are diagnosed in infancy; however, this patient was diagnosed in adolescence.

Keywords: Lipoblastoma, adolescent, differential diagnosis.

Introduction

Lipoblastomas are fatty tumors of mesenchymal origin that typically occur in the first decade of life [1]. Their peak incidence is between the ages of 5 and 6 with a male to female predominance of 4:1 [2]. Although more commonly found in the extremities, these tumors can occur adjacent to the axial skeleton [1,2]. Proximity to the spine can result in mass effect on neurovascular structures and possible compromise. A radiographic analysis study showed that of paraspinal cases, 38% involved the neural foramina or central canal [3]. Several cases of lipoblastomas have been reported in the pediatric population, with few in the paraspinal musculature [2,4,5,6,7]. We present a case of a paraspinal lipoblastoma in an adolescent.

Case Report

The patient is an 11-year-old male who initially presented to his pediatrician due to concern for a left upper thoracic hump at the age of 9 years. Radiographs were obtained and reported as thoracic dextroscoliosis measuring 12° without any vertebral body anomalies or paraspinal processes. Given that the patient was neurologically intact and asymptomatic, he was treated conservatively by his pediatrician with observation. The patient then presented 2 years later with cough, lymphopenia, and 6

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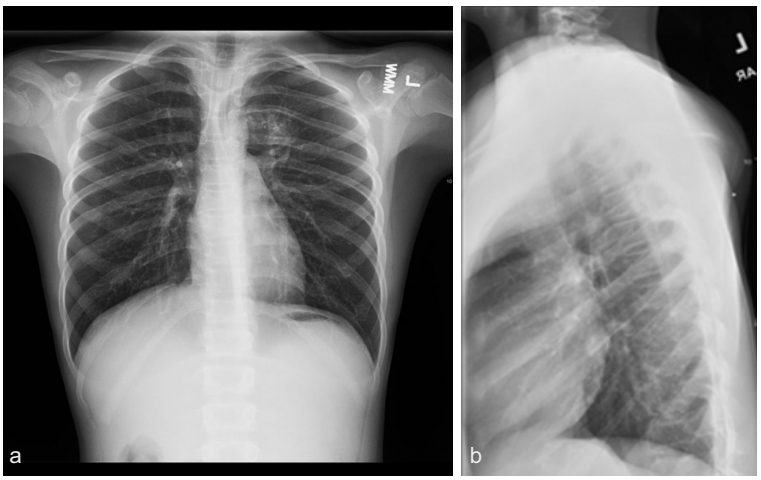

Figure 1: Thoracic spine radiographs (a: Anteroposterior, b: Lateral) performed with an external marker placed on the posterior prominence, demonstrating a calcific-appearing, radiopaque posterior chest wall lesion with 4th and 5th rib thinning and deformity.

weeks of fatigue. Infectious work-up included a chest radiograph, which revealed thinning of the left fourth and fifth ribs with an associated calcified mass. Thoracic spine radiographs were obtained with a marker placed within the mass (Fig. 1), which confirmed radiographic and clinical relationship. Computed tomography and magnetic resonance imaging (MRI) scans of the chest revealed a large heterogeneous, lobulated, and well-defined mass in the posterior mediastinum and paraspinal musculature with extension into the T4-T5 neural foramen. There were both fatty and calcified components. The mass measured approximately 4.6 × 5.2 × 5.0 cm in the anteroposterior, coronal, and craniocaudal dimensions, respectively (Figs. 2 and 3).

On clinical examination, the patient was 147 cm tall and weighed 30 kg. He was well-developed and well-nourished. He did not have any reported limitations with activity, neurologic symptoms, or bowel or bladder dysfunction. His family history was negative for tumors and positive for scoliosis in his father. On exam, gait was normal with plantigrade feet and no deformity. There was no evidence of spinal dysraphism. On both upright and forward bending visual inspection of the spine and thorax, there was no spinal rotational deformity, but he did have a palpable non-tender mass of the left upper back, which was soft and fixed in position. He reported no pain, numbness, or radicular symptoms. He had 5/5 strength, sensation was intact to all dermatomes, and his reflexes were normal.

To obtain a diagnosis, the patient underwent an ultrasound-guided, core-needle biopsy of the mass with interventional radiology. Twelve samples were sent for pathology. Results were consistent with lipoblastoma and orthopedic oncology, pediatric spine surgery, and pediatric general surgery recommended marginal resection of the mass due to proximity to the spinal canal.

The family and patient elected to proceed with surgery. The patient underwent general anesthesia, and neuromonitoring was used. He was placed prone on a spine bed and prepped in routine sterile fashion. An oblique incision was made caudal to the left scapula and over the mass. Dissection was performed, revealing two lobes of the tumor. The fourth and fifth ribs were osteotomized approximately 1.5 cm distal to the lateral margin of the tumor, and the costotransverse joints were disarticulated, allowing for the tumor to be removed en bloc with careful dissection from the left T4-T5 neural foramen with no cerebrospinal fluid leakage encountered. Complete excision was performed,

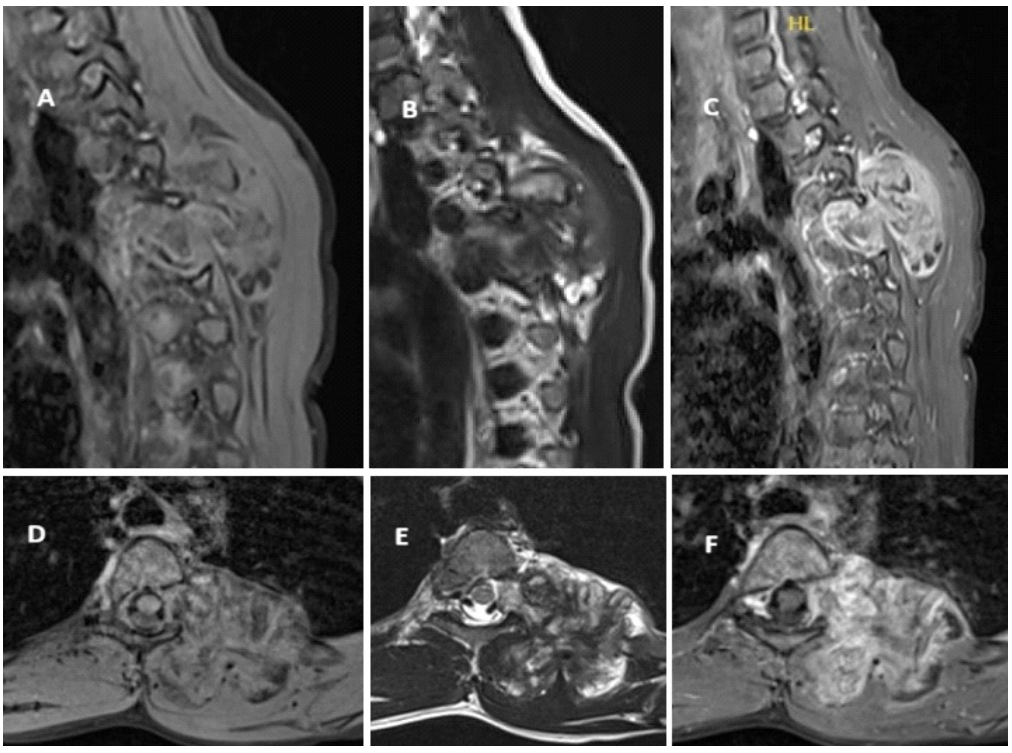

Figure 2: Representative magnetic resonance imaging images, including sagittal (a: T1-weighted, b: T2-weighted, and c: Post-contrast) and axial (d: T1-weighted, e: T2-weighted, and f: Post-contrast) views depicting the calcified T4-T5 lesion with extension into the left neural foramen.

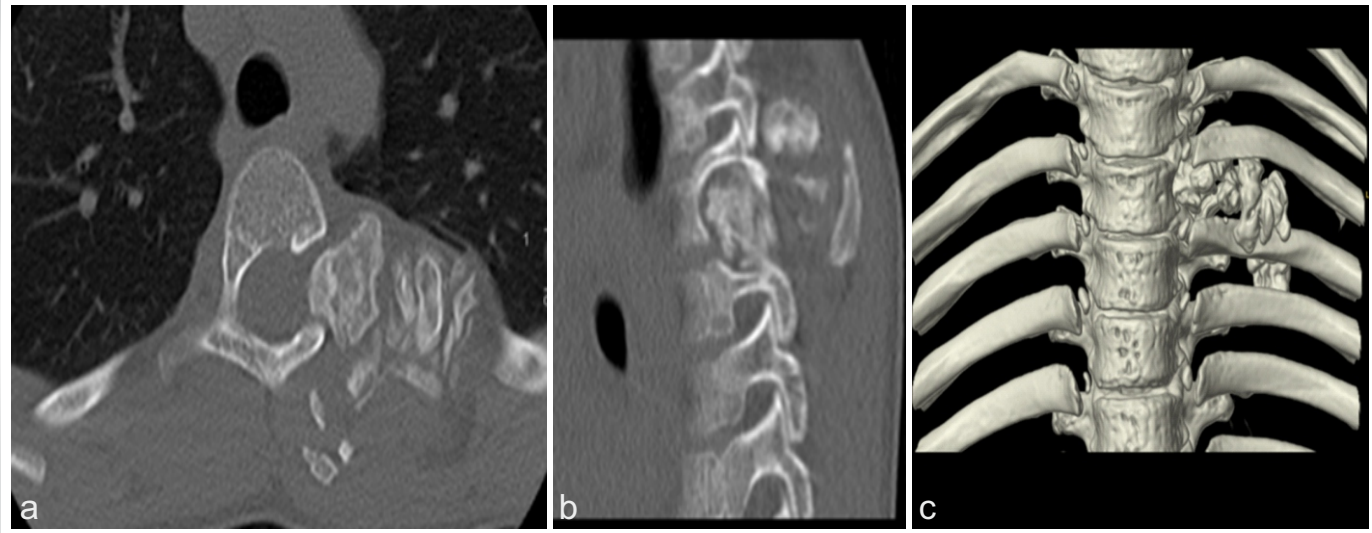

Figure 3: Computed tomography images, including (a) axial, (b) sagittal, and (c) 3D reconstruction depicting a calcified mass with extension into the left neural foramen.

hemostasis was achieved, and the pleural defect was repaired. The posterior chest was reconstructed by the general surgery team with placement of a 5 × 6 cm Gore-Tex patch over the posterior chest wall defect. The wound was irrigated, and layered closure was performed, and intrathecal morphine was injected into the lumbar interspace. Neuromonitoring was stable throughout the procedure, and the patient was awakened and taken to the post-operative holding area without complication. The final pathology of the tumor confirmed lipoblastoma, and margins were clean (Fig. 4).

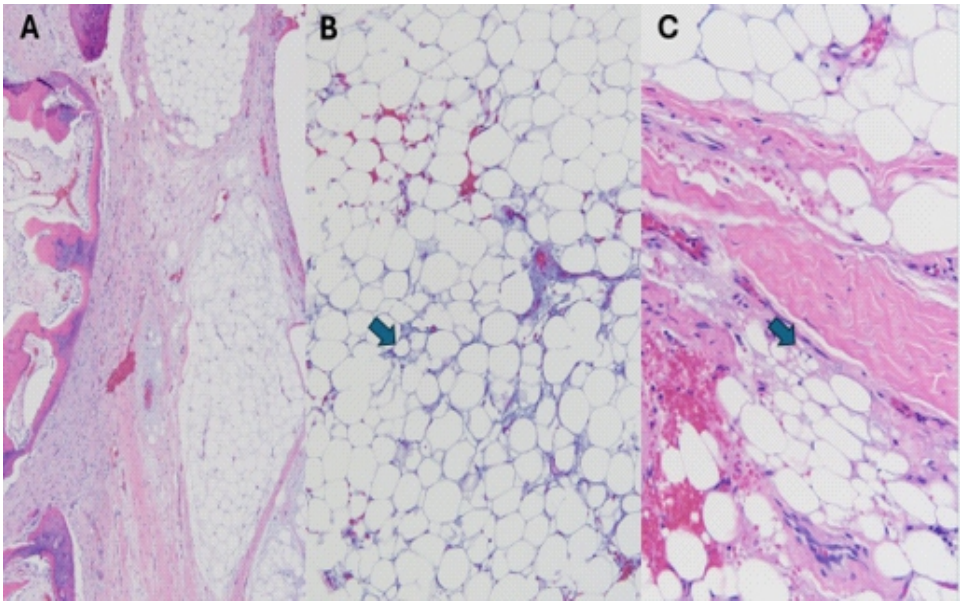

Figure 4: Hematoxylin and eosin-stained sections following decalcification demonstrate lipoblastoma. (a) Lobulated maturing and immature adipose components with myxoid areas, diagnostic of lipoblastoma (×4). The left side of panel A includes areas of the tumor that are ossifying, correlating to the intratumoral calcification seen on imaging. (b) Focal areas of the tumor show lipoblastic features, including signet ring cells (arrow) (×10). (c) Multivacuolated cells with a signet ring appearance (arrow) in another area of the tumor (×20).

The patient followed up at approximately 2 weeks, 6 weeks, 1 year, 2 years, 3 years, and 4 years. Over that time, the patient healed appropriately and was without complication. Each year, the patient undergoes a thoracic spine MRI with and without contrast for surveillance. He developed mild scoliosis (Fig. 5) that required no treatment and is stable 3.5 years post-operatively. There has been no evidence of recurrence.

Discussion

Lipoblastoma, the isolated and circumscribed version of lipoblastomatosis, is considered a tumor of infancy [1]. Presentation, physical examination, and advanced imaging do not reliably distinguish lipoblastoma, lipoma, and liposarcoma. Therefore, a biopsy is required to obtain a diagnosis. One key reason diagnosis based on imaging is challenging is that calcifications often present within fatty tumors secondary to fat necrosis. Dystrophic calcifications accumulate in dead or necrotic fat tissue and appear on imaging studies. Radiographic studies have reported that, while calcifications are not specific, they are more likely to appear in malignant adipocytes. The studies found calcifications in 32% of liposarcomas and 11% of lipomas [8,9]. Additional factors that were found to be significantly associated

Figure 5: Radiographs (a: Anteroposterior, b: Lateral) obtained 3 years post-operatively demonstrate mild scoliosis but no evidence of recurrence.

with liposarcomas included size >10 cm, presence of thick septa, presence of globular and/or nodular non-adipose areas, and lesions <75% fat [8]. Histologically, the key to diagnosis is the presence of lipoblasts at different stages of development that range from spindle-shaped pre-adipocytes to a mature fat cell displaying the classic "signet ring" pattern [1]. Recent studies have shown that abnormalities on chromosome 8 and rearrangements of the PLAG1 gene may be related to lipoblastoma and lipoblastomatosis [10]. The prognosis for lipoblastoma following marginal resection is excellent, with a reported rate of local recurrences ranging from 9% to 22% [11]. If recurrence does occur, it is more commonly seen in infiltrative lipoblastomatosis. The most important factors related to recurrence have been shown to be incomplete resection and the presence of diffuse disease rather than any morphologic feature [1].

Conclusions

Evaluation for pediatric spinal pathology is routine for pediatricians. While adolescent idiopathic scoliosis is relatively common, it is important to consider a broad differential when a patient presents with spinal asymmetry. Our study demonstrates a rare tumor in a unique age group. At 4-year follow-up, there is evidence that treatment of lipoblastoma with marginal resection and surveillance is applicable to adolescent patients. As seen in the patient discussed, a multidisciplinary approach is often required when the mass is adjacent to critical structures in the chest and spine. There is no proven role for adjuvant chemotherapy or radiation therapy. Follow-up is centered around standard post-operative wound healing evaluation and care. Surveillance is a yearly MRI of the post-operative site to evaluate for recurrence.

Clinical Message

Our report suggests that while lipoblastoma in an adolescent is exceedingly rare, the work-up and treatment can be the same as that of lipoblastomas found in all age groups.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/ her images and other clinical information to be reported in the journal. The patient understands that his/ her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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