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CASE REPORT

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Intrathoracic lipoblastoma in a 4-year-old male. A case report

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Abstract

Lipoblastoma is a rare fatty tissue tumor with characteristic histological and imaging findings, known to present in young children. We report a case of a 4 year 9 month old male who presented with a cough and a large intrathoracic soft tissue mass was seen incidentally on chest radiograph. The mass was further imaged with ultrasound, computed tomography, and magnetic resonance imaging, underwent an ultrasound guided biopsy and was subsequently surgically excised. We present the characteristic imaging findings on all four modalities. Additionally, we describe the clinical and histological features of the tumor. Understanding these features can aid the radiologist in distinguishing lipoblastoma from similar fatty tissue tumors including lipoma.

Case report

A 4 year 9 month old boy presented to the emergency department with a two week history of cough and 4 day history of fever. His past medical history was significant for congenital hydrocephalus with agenesis of the corpus callosum, a ventricular septal defect, bilateral vesicoureteral reflux (vur), undescended testes, gastroesophageal reflux disease (gerd) and tracheomalacia of the newborn. A chest radiograph was taken in the which revealed a large rounded density that appeared to involve the right mid to lower hemithorax. The mass resulted in secondary widening of the 5th to 10th posterior right ribs. There was also associated blunting of the right costophrenic angle consistent with a small pleural effusion. The left lung, heart and mediastinum were normal (Figure 1). This mass was not present on his most recent previous chest radiograph taken approximately 3 years prior. The following day an ultrasound of the chest was performed to further assess the opacity. The ultrasound showed a heterogeneously hyperechoic mass measuring 8.2 x 7.7 x 6.3 cm with no calcifications or cysts. Colour doppler examination showed no significant flow within (Figure 2).

The day following the ultrasound, a chest ct with intravenous contrast was obtained. A well-defined intrathoracic, extra-pulmonary, predominantly hypodense rounded mass measuring 8.7 x 7.9 x 7.2 cm was seen. The overall hounsfield units were isodense to subcutaneous fat (Figure 3a and 3b). There were small to moderate volumes of peripheral linear and nodular enhancing foci within the mass but no associated calcifications or cysts. It occupied the posterior half of the right hemithorax at the chest wall contiguous with the pleural surface just deep to the 5th through 10th posterolateral ribs and right lateral aspect of the corresponding vertebral bodies. The mass was confined to the intrathoracic space with no chest wall invasion, neural foraminal or spinal canal encroachment, or mediastinal involvement. The mass resulted in mild widening of the corresponding rib interspaces with anterior beaking/focal remodelling of the eighth right rib but with no associated aggressive osseous features. The bony thorax otherwise appeared normal (Figure 3).

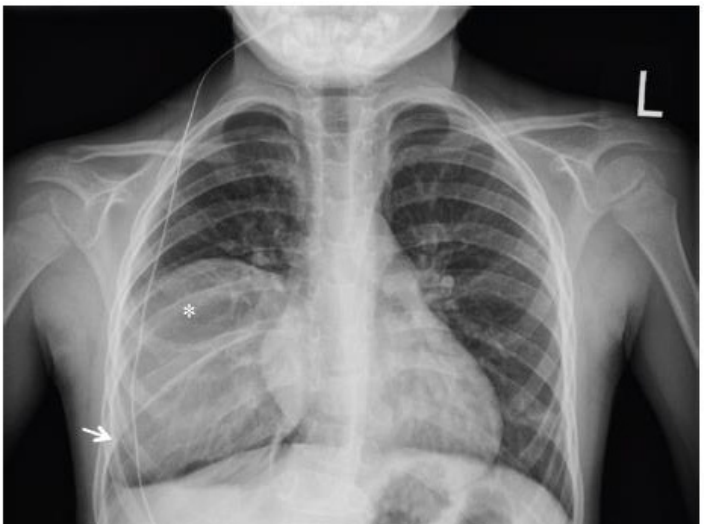

Figure 1. *4 year 6 month old boy with an intra-thoracic liposarcoma. Findings: Frontal radiograph of the chest shows a large rounded density of the right middle to lower hemithorax (star) with secondary widening of the 5th to 10th posterior right ribs and a small pleural effusion (arrow). Note is made of a VP shunt catheter tube traversing the right lateral chest wall.*

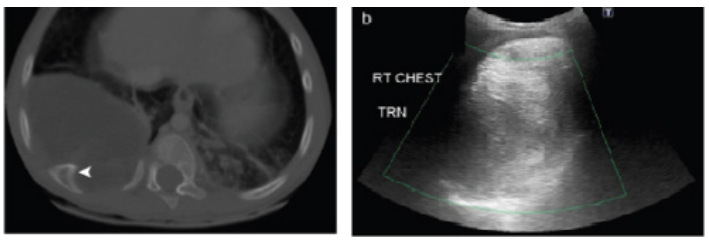

Figure 2. *4 year 6 month old boy with an intra-thoracic liposarcoma. Findings: Ultrasonography of the intrathoracic mass (star). A transverse gray-scale image of the right chest (a) shows a heterogeneously hyperechoic mass with no calcifications or cysts . Colour doppler (b) shows no significant flow within the mass.*

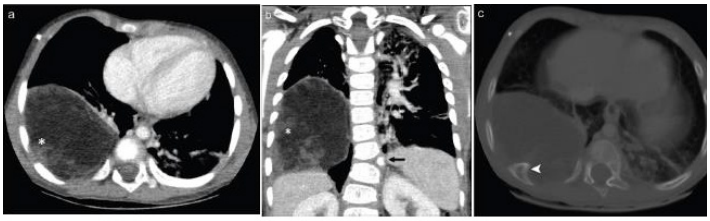

Figure 3. *4 year 6 month old boy with an intra-thoracic liposarcoma. Findings: contrast enhanced ct images of the chest. Axial (a) and coronal (b) images show a well-defined hypodense rounded mass with an attenuation isodense to subcutaneous fat, contiguous with the pleural surface. Small volume enhancing foci are seen within the mass (star) but there are no associated calcifications or cysts. The mass is confined to the intrathoracic space with no chest wall invasion, neural foraminal or spinal canal encroachment, or mediastinal involvement. Note is made of a lower thoracic spine hemivertebrae (arrow). Axial bone window reconstruction (c) shows secondary anterior beaking/ focal remodelling of the eight right rib (arrowhead).*

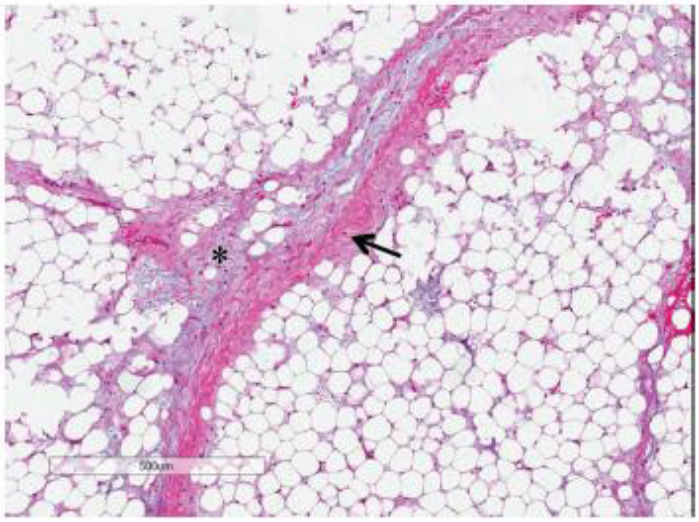

Figure 4. *4 year 6 month old boy with an intra-thoracic liposarcoma. Findings: histomorphology shows lobules of mature and immature adipose tissue separated by fibrous septa (arrow) containing blood vessels and areas of myxoid stroma (star). This myxoid stroma is usually located at the periphery of the fat lobules.*

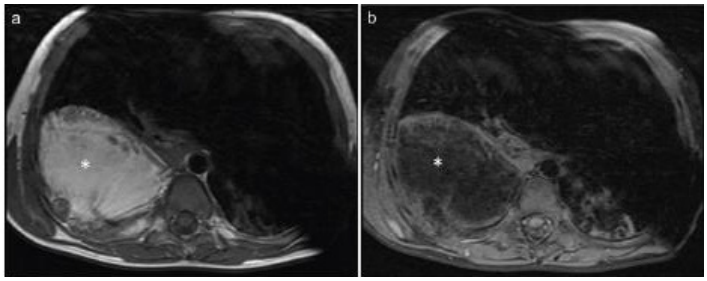

Figure 5. *4 year 6 month old boy with an intra-thoracic liposarcoma. Findings: Axial mri images. T1 non-fat saturated sequence (a) shows hyperintense t1 signal intensity and t1 fat saturated sequence (b) shows hypointense signal of the mass (star), confirming it to be a fat-containing mass.*

Figure 6. *n4 year 6 month old boy with an intra-thoracic liposarcoma. Findings: serially sectioned specimen showing adipose tissue as the main component and mimicking a lipoma. The lesion appears well circumscribed and partially lobulated at the periphery (arrow), which is more common than the diffuse/infiltrative type, distinction that is not.*

Several days later the patient underwent an ultrasound guided biopsy of the mass to differentiate between it being a lipoma, liposarcoma, teratoma and lipoblastoma, which would then guide management. Histology showed predominantly benign adipose tissue with no cellular atypia, pleomorphism, necrosis or abnormal mitotic activity. It was consistent with a benign adipose lesion (Figure 4). The differential was a lipoma or lipoblastoma, but the presence of myxoid changes and slightly increased vascularity was in keeping with a lipoblastoma. To confirm there was no spinal involvement, the patient underwent a full spine MRI exam.

The exam confirmed a largely fatty mass, which was predominantly hyperintense on t1 weighted images and hypointense on t1 fat saturated images, with no spinal involvement (Figure 5). After confirmation that the lesion was benign with no spinal invasion, the patient went to the operating room for complete surgical excision. The mass was grossly removed along with portions of the right 5th and 6th ribs, with several smaller components removed in piecemeal (Figure 6). The tumour was confirmed histologically as a lipoblastoma. At one and six months after the procedure the patient was reported to be doing remarkably well with no residual discomfort and full mobility of his arm. His cough had been incidental and not related to the mass.

Discussion and conclusion

Lipoblastoma is a rare benign tumor comprising only 5-30% of all soft-tissue neoplasms reported in children [1,2]. This tumor presents in two forms: a localized well-circumscribed lesion (lipoblastoma), or a multicentric type lesion (lipoblastomatosis). The differential diagnosis includes lipoma, liposarcoma, and teratoma. These tumors are most often diagnosed by pathology after excision however their radiographic features are unique and can direct a diagnosis. Lipoblastoma has a predilection for males and 90% are seen in children less than 3 years of age. They are commonly found in the axilla, neck, chest wall, and pre-vertebral soft tissues [3]. Surgical excision is usually curative although local recurrence rates between 14% and 25% have been reported [2]. Clinically, these tumors are fast growing but asymptomatic. However, because of the mass effect on adjacent structures the patient often presents with nonspecific symptoms depending on the location of the tumor [4]. Additionally, our patient had several other comorbidities (congenial hydrocephalus, bilateral VUR, undescended testes, GERD and neonatal tracheomalacia). Coffin et al. [1] report on a series of 59 patients with lipoblastoma where 17% of patients had associated developmental delay, congenital malformations (including cleft lip, dextrocardia, hydrocephalus,

brain malformation (not otherwise specified), macrocephaly, and scaphocephaly), seizures, or family history of lipoblastoma. The authors believe that lipoblastomas have a consistent rearrangement of chromosome 8. Others point to the possible genetic role of leptin expression, which influences endocardial cushion development. Deutscher et al. [5] report on a retroperitoneal lipoblastoma in an 8-year-old girl with ventricular septal defect and postulate that leptin may play a role in lipoblastoma growth.

Histologically, lipoblastoma is comprised of embryonic fat cells that continue to proliferate in the postnatal period. Within any individual tumor, a mix of cells may be seen including mature lipocytes, lipoblasts and more primitive mesenchymal cells with a richly vascularized connective tissue septae between the cells dividing them into lobules. They are usually encapsulated with a myxoid stroma [6]. Currently, a histological sample is needed for a definitive diagnosis [4].

Radiologically, lipoblastoma presents as a fatty mass with heterogenous septae. Depending on the maturity of the tumors cells and the extent of septations, these tumors can have slightly varied radiographic findings. However, these primarily fatty masses can be distinguished from similar tumors by radiographic and clinical features. Lipoma always presents as a homogenous mass and any detection of a fibrovascular network on contrast-enhanced CT necessitates considering a differential diagnosis [6]. Liposarcoma presents radiologically and histologically similar to lipoblastoma, however, displacement of normal structures rather than tumor invasion argues against a malignancy. Additionally, liposarcoma is extremely unusual in children younger than 10 years of age [1] while lipoblastoma has virtually never been described in children over 8 years of age [6]. Teratomas have gross calcification or ossification intermingled with variable fat and soft tissue components which is not seen with lipoblastoma [3,7].

Magnetic resonance imaging (MRI) is considered the best method to diagnose these benign tumors. They have characteristics that resemble subcutaneous fat: high intensity on T1-weighted imaging (WI), intermediate on T2-WI, and loss of signal intensity on fatsuppressed T1-WI [8], which was well demonstrated in our case. However, the presence of mature and immature lipoblasts can cause areas of intermediate to high signal intensity on T1- WI [4,8]. Additionally, their fibrous framework appears as streaks and whorls, which are non-enhancing, making the tumor inhomogeneous [6]. CT findings of

lipoblastoma show a hypodense (less than 0 Hounsfields units) mass, which may contain septa and whorls of soft tissue density within. There may also be adjacent bone remodeling and enlargement of underlying ribs, as seen in our case, but the tumour itself does not contain calcifications. These features help differentiate it from a lipoma. Lipoblastomas exert mass effect and displacement of normal structures, such as the ribs, rather than invasion, which distinguish it from liposarcoma [7]. CT is most helpful in determining tumor margins, extension of the tumor, and invasion aiding differentiation between an infiltrative or well-localized mass [3]. On chest radiography, lipoblastomas present as well defined opacities. They have soft tissue densities because of the cellular differentiation and fibrous framework [3]. Ultrasound shows a well-defined primarily homogenous hyperechoic mass with pronounced echogenic streaky foci at the periphery.

These features correspond to fatty tissue with fibrous septae and a myxoid stroma [3,7]. As seen in this case, these tumors often do not have significant vascular flow. Treatment for lipoblastoma is complete but conservative surgical resection. Complete excision is important to prevent local recurrence, but recurrence is uncommon and more frequently observed with deep infiltrative forms. No spontaneous resolutions or reductions in mass have been reported in the literature, nor have metastases or malignant transformation [3,4]. Recurrences most commonly occur within 12 months but have been seen up to 84 months post operatively, so follow up is recommended for a minimum of 5 years [2].

In conclusion, lipoblastoma is a rare benign fatty tumor which mostly presents in males under the age of 5. On imaging, they present as fatty masses with heterogenous septae. The presence of a fibrous framework appearing as streaks and whorls differentiates it from a lipoma, the age group and non-invasive features helps differentiate it from liposarcoma and the lack of calcifications differentiate it from a teratoma. Although a biopsy is still essential for definitive diagnosis, the age at presentation and unique imaging features aid in establishing a radiological diagnosis of lipoblastoma.

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