CASE REPORT

Mediastinal lipoblastoma: unexpected finding of a chest infection

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SUMMARY

A 2-year-old boy was presented with symptoms of chest infection. The chest radiograph showed a large mediastinal mass, which led to further investigations including biopsy of the tumour. Histopathological analysis revealed a diagnosis of lipoblastoma. We highlight the imaging appearance of the lesion. Although histopathological analysis is required for the confirmation of the diagnosis, cross-sectional imaging is useful in evaluating the extent of the tumour for surgical planning.

BACKGROUND

Lipoblastoma is a rare benign tumour of embryonic fat tissue, which is mostly found in infants and young children. The diagnosis of this lesion after the age of 5 years is unusual and it has male preponderance.¹ A review of 18 677 cases of benign mesenchymal soft tissue lesion revealed only 114 cases (0.6%) of lipoblastoma/lipoblastomatosis, demonstrating its rarity.² The tumour is typically asymptomatic, but has the capability to grow rapidly and cause local mass effect, hence may be presented in different ways depending on its location. Although it can arise almost anywhere within the soft tissue, mediastinum is one of the uncommon locations.³ It is important to differentiate this rare benign paediatric tumour from other mediastinal lesions in view of difference in surgical planning and prognosis. Hence, familiarisation of its imaging appearance is imperative. We report a case of mediastinal lipoblastoma occurring in a 2-year-old boy as incidental findings of mediastinal mass depicted on chest radiograph during admission for chest infection.

CASE PRESENTATION

A 2-year-old boy was presented with a 1-week history of productive cough and fever followed by a 2-day history of shortness of breath. The child was previously healthy with no significant medical and antenatal histories. On presentation to the hospital, he was tachypnoeic (45 breaths/min) associated with mild subcostal recession. Otherwise, he was haemodynamically stable (regular pulse 102 bpm, BP 135/78) and not hypoxic. There was no cardiac related sign or symptom.

INVESTIGATIONS

Routine blood investigations were all normal. The inflammatory markers were also not raised. The frontal chest radiograph demonstrated a large well-defined mediastinal opacity with preservation of the cardiac silhouette suggestive of a posterior

mediastinal mass (figure 1). Subsequently, a contrast-enhanced CT of the thorax was carried out to establish the character, exact location and local extension of the mass. It revealed a large posterior mediastinal mass with both solid and fatty components, displacing and compressing the heart anteriorly (figure 2). Echocardiogram showed that the heart was compressed, but the contractions and functions were normal with a left ventricular ejection fraction of 0.62. In view of the mass, his blood samples were analysed for tumour markers like β -human chorionic gonadotrophin and α -fetoprotein and the results were normal. With regard to the mediastinal mass, a preoperative ultrasound-guided biopsy was performed and the histopathological results showed mainly adipocytes and lipoblasts at varying stages of maturation consistent with mediastinal lipoblastoma (figure 3).

DIFFERENTIAL DIAGNOSIS

Visualisation of fat can narrow down the differential diagnosis, but the presence of fat alone cannot confidently differentiate lipoblastomas from other fatty lesions such as germ cell tumour, myxoid liposarcoma or hibernoma. Intrathoracic germ cell tumour is usually located in the anterior mediastinum and may demonstrate calcification and/or cystic component. On the other hand, liposarcoma usually occurs in older patients as an infiltrating mass with significant enhancing component and thick septation. The myxoid variant of liposarcoma can mimic lipoblastoma on imaging, but usually

Figure 1 Frontal chest radiograph showing large well-defined mediastinal opacity with relative preservation of the cardiac borders.

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Figure 2 Axial (A) and coronal (B) contrast-enhanced CT images show a large posterior mediastinal mass of fat attenuation. The arrow in (A) image demonstrates that the heart is compressed and displaced anteriorly.

occurs in the extremity of children and is exceeding rarely in the truncal region. In addition, it is important to be aware that the infiltrating nature and deep extension of lipoblastomatosis may mimic an aggressive lesion.

TREATMENT

He was initially treated as bronchopneumonia and given antibiotics. Subsequently, the tumour was completely resected via right thoracotomy. Intraoperatively, there was a large well-defined yellowish solid fatty tumour at the posterior mediastinum, pushing the right lower lobe of the lung and the heart anteriorly (figure 4). No immediate complication was encountered during the surgery and ward stay. Further histological analysis of the completely resected tumour was performed which confirmed the initial biopsy results of lipoblastoma.

OUTCOME AND FOLLOW-UP

There was no complication postsurgery and he was discharged home. The child was healthy and asymptomatic at 1-month and 6-month-follow-up clinics.

DISCUSSION

Jaffe⁴ defined lipoblastoma in 1926 as a lipomatous lesion that contained immature fat cells. Lipoblastoma is predominantly well circumscribed, capsular and localised. However, it can be non-capsulated, diffuse and deeply embedded with tendency to extend into adjacent structures, a condition called lipoblastomatosis.⁵ Clusters of mature adipocytes rounded by immature lipoblasts, myxoid and stellate mesenchymal cells are the hallmark of its histological appearance.¹ The absence of pleomorphism,

atypia and hyperchromasia differentiate it from myxoid liposarcoma.¹ Lipoblastoma may arise from anywhere within the soft tissue. The commonest site is extremity in 50% followed by thorax in 18% as well as head and neck in 15% of cases.⁶ Mediastinum is among the uncommon locations for this tumour^{3,7} and the present case is one of the examples.

Clinically, lipoblastoma is typically painless and may rapidly grow into an impressive proportion with propensity to induce mass effect on adjacent structures as demonstrated in our study. Be that as it may, no metastases or malignant transformation of this tumour has been described before. When located in mediastinum, it can cause dyspnoea, chest infection or even life-threatening asphyxia owing to airway compression. Rarely, mediastinal lipoblastoma may cause cord compression through its transforaminal intraspinal extension.⁸

Appearance of lipoblastoma is variable on plain radiograph. A chest radiograph may reveal mass with non-specific soft tissue density, and detection of such mass depends on the size, thickness and site. In the present study, the tumour is demonstrated as a well-defined posterior mediastinal mass of soft tissue density with no calcification or adjacent bony erosions, similar with previous findings of mediastinal lipoblastoma.⁹ Occasionally, lipoblastomas could appear radiolucent owing to fatty components.¹⁰

Lipoblastomas are composed mostly of, but not entirely, fat in its cross-sectional imaging appearance.¹ A CT scan usually shows heterogeneous mass showing areas of low-grade density of fat attenuation. In the present case, the chest CT scan allowed the identification of fatty components through objective

Figure 3 Histological specimen shows lipoblasts with adipocytes in different stages of maturation (H&E, magnification $\times 600$).

Figure 4 Photograph shows a large yellowish fatty tumour with well-defined lobulated border.

measurement in the form of hounsfield unit (HU). Most of the hypodense areas revealed density between -10 HU and -100 HU, consistent with fat. In general, lipoblastoma does not usually enhance after administration of intravenous contrast agent.¹¹ For the present case, non-contrast chest CT was not performed to compare with as it was not necessary and would only add to the total radiation burden. Hence, we could not conclude whether or not there was enhancement shown at post-contrast. However, its density was generally lower than the muscles indicating that even if there was any enhancement it would be minimal. The preoperative CT assessment was very useful to determine tumour margin, deep extension of the tumour and the exact origin of the mass. This enabled good planning by the surgeon prior to surgical resection.

MRI of lipoblastoma usually shows a soft tissue mass with hyperintense signal in both T1-weighted and T2-weighted images; in most instances, the fatty components are isointense to the subcutaneous fat.¹² No significant difference was seen in the sensitivity between CT and MRI in diagnosing the primary lesion, but it is thought that CT may underestimate the tumour extension in cases of recurrence and MRI is recommended for these kinds of cases.¹¹

Cross-sectional imaging is useful in suggesting the diagnosis of lipoblastoma, although definite diagnosis is only achievable by histological examination. Initial histopathological study can be achieved by ultrasound-guided biopsy as exemplified by the present case. Ultrasound has the advantage of monitoring the biopsy needle continuously in real time. It is generally a relatively quick and safe procedure that can be performed at bedside manner, provided that an adequate acoustic window is present.

In general, complete tumour resection is the mainstay of the treatment. However, recurrence of up to 25% has been reported, particularly with those of incomplete resection.¹³

Learning points

- ▶ Lipoblastoma should be considered in the differential diagnosis of rapidly growing fatty mass within the mediastinum in infants or young children.
- ▶ Preoperative CT is a very useful tool to assess the extension and location of the tumour for proper surgical planning.
- ▶ In the selected case with a good acoustic window, ultrasound-guided biopsy is a less invasive and safe procedure to attain tissue sample for the histopathological confirmation of diagnosis.

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Competing interests None.

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