Journal of Pediatric Surgery Case Reports 27 (2017) 7–11

Contents lists available at ScienceDirect

Journal of Pediatric Surgery Case Reports

journal homepage: www.jpscasereports.com

Intrathoracic lipoblastoma presenting with severe respiratory distress

Joseph Motshedi Sekgololo*, Risenga Frank Chauke, Peter Shere Ramoroko, Khondker Mofazzal Hossain

Cardiothoracic Surgery Department, Sefako Makgatho Health Science University, Dr George Mukhari Academic Hospital, Pretoria, South Africa

ARTICLE INFO

Article history:
Received 29 August 2017
Accepted 2 September 2017
Available online 6 September 2017

Keywords:
Lipoblastoma
Intrathoracic tomour
Lipoblastomatosis

ABSTRACT

Lipoblastoma is a rare benign tumour which originates from an adipose tissue. In this study we report the case of a three year old boy who presented with a large intrathoracic tumour occupying the whole of the left hemithorax. He presented in severe respiratory distress. A chest X-ray showed total opacity of the left hemithorax, and CT-scan showed a low attenuation mass inkeeping with fat in the left hemithorax. A complete resection of a tumour was undertaken, with histopathology report confirming the diagnosis of lipoblastoma. The relevant literature review was done. At three and six months follow up, there was no recurrent tumour on imaging and the child had started thriving well.

1. Introduction

Lipoblastoma/lipoblastomatosis is a rare benign tumour that originates from an adipose tissue [1]. There are two distinctive forms of a lipoblastoma which have been identified. These are localized encapsulated type, which is referred to as lipoblastoma and a diffuse infiltrative type which is called lipoblastomatosis [2]. Lipoblastoma has male predominance, with a male to female ratio of 3:1 [3]. The incidence of 1–2% among all childhood benign tumour has been reported [4]. The tumour commonly affect children aged three years or less [5]. In this case, the child was under three and male. The tumour may arise from anywhere in the body, where there are mesenchymal cells. However, it is reported to be more common in the trunk and extremities. It is uncommon in the neck and mediastinum [5]. Some reports suggest left-sided tumour predominance, as opposed to the right [1]. Our case was that of left-sided tumour. A complete tumour excision is definitive treatment for these tumours. It is important to note that there is local recurrence rate of 14–25% [1]. Follow-up is thus very important, but the appropriate interval for it is unknown. Some authors suggest a minimum period of three years or more for follow-up. In our case, there was a complete excision with no recurrence at 2 weeks, 3 months and 6 months follow-ups.

2. Case report

We report a three year old male patient presented with a history of recurrent respiratory infections, episodes of vomiting and weight loss. The symptoms started at the age of 1 year and at times required hospital admission. The patient was referred to us by our medical paediatric team with provisional diagnosis of left massive pleural effusion, and he was in respiratory distress. On examination, the child was in respiratory distress breathing at the rate of 48 breaths per min, saturating 94% on oxygen via nasal cannula, with heart rate of 156 beats per minutes, and was using accessory respiratory muscles. Examination also revealed evidence of failure to thrive. He had chest asymmetry, increased left chest anteroposterior diameter, decreased vocal fremitus of the left chest, and stony dullness on percussion of left hemithorax and breath sounds were decreased on the left. A chest X-ray showed a total opacity of left hemithorax. However, the opacity was less dense in the upper third (Fig. 1). A thoracentesis was done by paediatrician at our hospital with no fluid aspirated. Urgent CT-scan was then arranged which, showed a well-circumscribed non-enhancing fat density tumour filling about 75% of the left hemithorax, with non-aerating lung tissue compressed (Figs. 2–3) The tumour was lobulated, with no evidence of invasion to the adjacent structures. Echocardiography suggested normal cardiac anatomy and function.

The patient was admitted in Intensive Care Unit and intravenous antibiotics and oxygen supplementation without mechanical ventilation were continued. He was prepared and taken for an urgent operation, the next day. A muscle sparing left posterolateral thoracotomy approach was undertaken. The large yellowish-grey

* Corresponding author.
E-mail address: motshedi.sekgololo@gmail.com (J.M. Sekgololo).

http://dx.doi.org/10.1016/j.epsc.2017.09.005

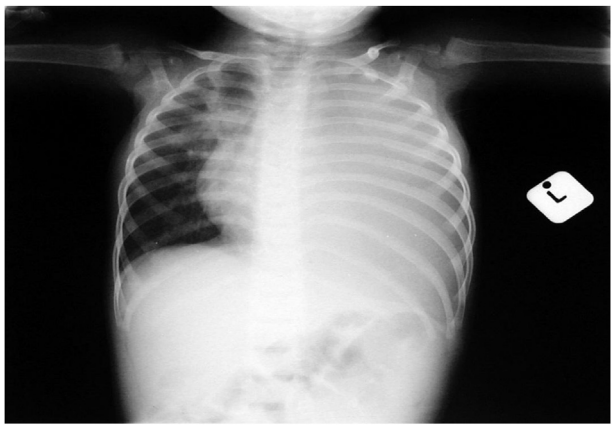

Fig. 1. Pre-operative Chest X-ray: showing the tumour occupying the entire left hemithorax.

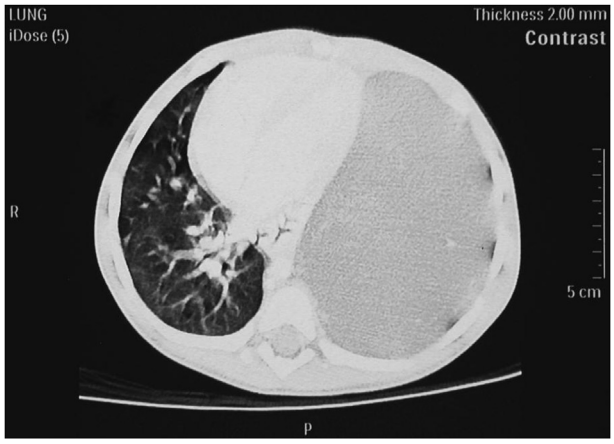

Fig. 2. Pre-contrasted CT chest (pulmonary window).

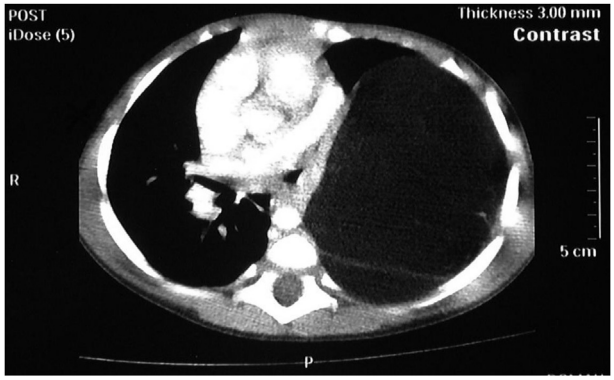

Fig. 3. Post-contrasted CT-scan (mediastinal window).

mass measuring 14 × 10 × 5 cm, weighing about 500 g was found, displacing and compressing the lung (Figs. 4–5). The lung tissue looked normal but atelectatic. The tumour was completely encapsulated with no invasion to the adjacent structures. The small adhesions were found on the left hemi-diaphragm and lateral chest wall. There were no feeding vessels/pediclels supplying the tumour. The postoperative care was uneventful and the child stayed two days in ICU and two days in the wards, and was discharged home on his third day in the ward.

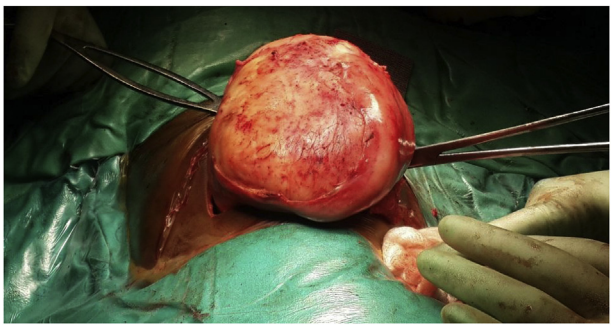

Fig. 4. Lipoblastoma being exteriorised.

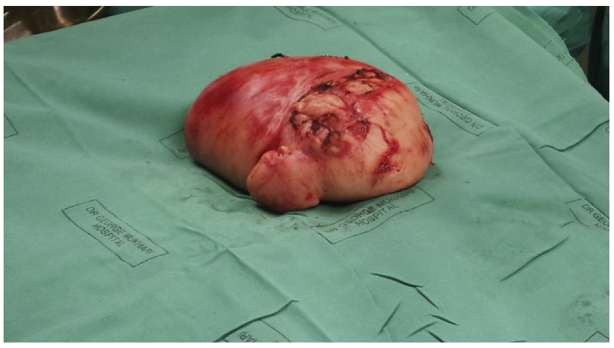

Fig. 5. Lipoblastoma removed from chest cavity.

On follow-up 2 weeks later, the histopathology report confirmed lipoblastoma. This lipoblastoma was characterised by multiple lobules of immature and mature adipose tissues separated by thick and fibrous septae (Fig. 6d). Adipocytes had hyperchromatic nuclei showing intranuclear and intracytoplasmic vacuoles. The myxoid areas were seen on further sectioning of the tumour (Fig. 6b–c). The patient recovered from operation and was thriving well. The X-ray taken after two weeks showed the lung that was fully expanded (Fig. 7).

3. Discussion

Lipoblastoma is a rare soft tissue tumour common during infancy and childhood. It was first described in 1956 by Jaffe [5]. In 1973 Chung and Enzinger suggested two names: lipoblastoma (well circumscribed and localized tumour) and lipoblastomatosis (multicentric and diffuse tumour) [5]. Lipoblastoma have been noted to prefer sites with primitive adipose tissue such as axilla, neck and chest wall in newborn [6]. Lipoblastoma rarely occurs intrathoracically.

Keeley classified intrathoracic lipoblastoma into two groups and these are: pure intrathoracic type and hourglass type [6]. A Pure intrathoracic type has intrathoracic location with or without pleura involvement and Hourglass is characterised by intra- and extrathoracic involvement [6].

3.1. Morphology (histogenesis, macroscopy and pathology)

It is made up of mesenchymal cells showing various stages of adipocytes development. It has cellular pleomorphism with absence of the basement membrane and intercellular tight junctions [6]. Lipoblastoma on macroscopic examination is yellowish-grey, encapsulated and lobulated, with septae [1,6]. It has no myxoid areas, which are usually suggestive well-differentiated