Mediastinal Lipoblastoma of Infancy: a case report

Kuo-Tai Li¹  Chao-Ran Wang²  Wan-Chak Lo²  Chuen Hsueh³  Wing-Cheong Chi¹

Department of Radiology¹, St Paul’s Hospital
Department of Radiology², Chang Gung Children’s Hospital at Linkou
Department of Pathology³, Chang Gung Memorial Hospital at Linkou

Mediastinal lipoblastoma is a rare mesenchymal tumor of embryonal fat occurring predominantly in the first three years of life. Less than 20 cases have been reported so far in the Western literature. Lipoblastomas constitute less than 1% of all pediatric tumors. The extremities are the most common location for this tumor. We presented a rare 9-month-old female infant case that differed from previously reported mediastinal lipoblastomas in those patients who were much older. The computed tomography (CT) and magnetic resonance image (MRI) features were discussed. At surgery, the tumor was removed without complication and histology demonstrated an encapsulated and multilobulated lipoblastoma.

Key words: Infancy; Lipoblastoma; Mediastinum

The majority of lipoblastomas arise in the superficial tissue of the extremities but they are occasionally found in the neck or abdomen. They are even rarer in the mediastinum, where less than 20 cases have been reported so far in the literatures [1-6]. This benign tumor has potential of local invasion and rapid growth and presents in two forms: a localized well-circumscribed lesion (lipoblastoma) or a multifocal type (lipoblastomatosis)[10]. The descriptions of “embryonal lipoma” and “infantile lipoma” had been advocated to indicate the benignity of this mesenchymal tumor that exhibited to invade locally but not metastasize [7]. The usual imaging features of mediastinal lipoblastoma are either circumscribed or infiltrative mass with adipose tissue, which can masquerade as germ cell tumors or myxoid variants of liposarcoma of children. Differentiating this rare benign pediatric tumor from other mediastinal lesions is essential since there is difference in surgical planning and prognosis.

CASE REPORT

A 9-month-old female infant presented with croup attacks for one month. She was admitted to a community hospital, where chest film revealed a large, lobulated, homogeneous mediastinal mass occupying the left hemithorax with sharp demarcation from the adjacent lung. The trachea was pushed to the right (Fig.1). Laboratory examination findings, including measurement of WBC, CRP, AFP, β-HCG were all within normal limits. Contrast-enhanced computed tomography (CT)(HiSpeed,GE) of the thorax confirmed the presence of a 15 × 12 cm mediastinal mass filling up the majority of the left thorax. This well-defined mass had a pure fat attenuation without calcification. The great vessels were pushed anteriorly (Fig.2). There were no hilar and mediastinal lymph nodes enlargement. Chest magnetic resonance images (MRI) (1.5T, Magnetom Vision, Siemens) were

Reprint requests to: Dr. Chao-Ran Wang
Department of Radiology, Chang Gung Children’s Hospital.
No. 5, Fu Hsing Street, Kwei-Shan, Taoyuan 333, Taiwan,
R.O.C.
arranged for further preoperative planning. It showed a huge mass lesion occupied most of left thoracic cavity, high signal intensity on T1-weighted images (Fig.3) and heterogeneous low signal intensity on T2-weighted with fat saturation images (Fig.4). The mediastinum was pushed to the right side and the posterior portion of the mass herinated to right posterior mediastinum. The aorta was anteriorly displaced and heterogeneous enhancement within the mass was noted after Gd-DTPA injection. Surgical exploration found a well encapsulated mass at left thoracic cavity compressing on the adjacent lung tissue anteriorly. Pathology showed a 12 × 12 × 6 cm grayish-white multi-lobulated and encapsulated mass without necrosis and hemorrhage. Grossly, the tumor was yellowish and soft. Histological examination revealed lobules of proliferating univacuolated mature and immature adipocytes (Fig.5). The patient did well post-operatively and had no signs of recurrence 6 months later.

**DISCUSSION**

In 1926 Jaffe coined the term lipoblastoma in an article describing recurrent lipomatous tumors of the groin [7]. It is characterized by a distinct histological
hallmark of clusters of mature adipocytes in the central lobule separated by fibrous septa and surrounded by immature lipoblasts, a plexiform capillary pattern and a richly myxoid stroma [9]. Etiology and histopathogenesis are virtually unknown, but they are believed to be related to altered embryogenesis of human adipose tissue, resulting in uncontrolled proliferation of lipoblasts in the postnatal period [1]. Adipose tissue was first recognizable at certain locations in embryos at about 9 cm in crown-to-heel length [8]. Recent advancement of molecular biology reveals that there is a distinct clonal karyotype abnormality involving the long arm of chromosome 8 (8q11-12) [1]. Lipoblastomas are well known as a benign mesenchymal tumor of embryonal adipose tissues with good prognosis despite propensity to local invasion and rapid growth. Local recurrence after resection is more common in the diffuse variant form or lipoblastomatosis. For this reason, the circumscribed form of lipoblastoma carries a better prognosis.

Adipose tissue tumors accounted for 6% of soft tissue tumor in the pediatric population [9]. Lipoblastoma could occur from neonate to the elderly (0-84 years), 90% of the tumor was found in children under 3 years of age, and 40% occurred before the age of 1 year [1]. Boys were more commonly affected with a male to female ratio from 3:2 to 4:1 [10]. Previous report showed the tumor was more prevalent in white children rather than in Asians. The true incidence of the condition is not known; however, recent extensive analysis of computer diagnoses of soft tissue tumors at the Arm Forces Institute of Pathology covering a 10-year-period provides some valuable insight. In this study, Kransdorf looked at the distribution of diagnoses of 18,677 benign soft tissue tumors by age, sex, and anatomic location. In the 0- to 5-year age range, there were 1,064 tumors, of which 79 (7.4%) were lipoblastomas, as compared with three lipoblastomas (0.23%) in 1,305 benign soft tissue tumors in the 6- to 15-year age group. Greater than 70% of all lipoblastomas arose in the superficial layers of the extremities; the rest were deeper and arose in the mediastinum, retroperitoneum, and the axilla [1]. From the imaging point of view, lipoblastoma had variable appearances that were most likely due to the different stages of differentiation of individual lipoblastomas [2]. The most common two appearances of lipoblastoma were well-defined fatty mass or embedded within large masses of fatty tissues. On CT, our case consisted of fat with curvilinear streaks of increased density which showed some enhancement after intravenous contrast medium injection. On T1-weighted MR images, the tumor appeared as relatively heterogeneous high signal intensity without Gd-DTPA enhancement. MRI descriptions often mentioned that the fibrous framework appearing as non-enhanced streak and whorl making the tumor inhomogeneous [11]. If the signal intensity was not that of a characteristic mature fatty tissue, a surgical biopsy became essential for definite diagnosis to exclude the possibility of myxoid liposarcoma [12].

Although the CT and MRI findings can narrow the differential list, they are not sufficient for further classifying the lesion. The main differential diagnoses include other fat-containing lesions such as lipoma, hibernomas, teratoma and liposarcoma. Lipoma and hibernomas demonstrated much slower growth than lipoblastoma and hibernomas were rare in children. The so-called hibernoma was a tumor of adipose tissue, in which they were multivacuolated and contained varying amounts of eosinophilic cytoplasm [8]. Teratoma occurring in the mediastinum may contain gross calcification or ossification intermingled with variable fat and soft tissue components. Liposarcoma, especially the myxoid variant, could mimic lipoblastoma on imaging. It may be almost impossible to distinguish histologically from lipoblastoma. Helpful clues were the lack of lobulation, variable growth pattern and increased nuclear atypia in liposarcoma [15]. However, they usually occurred in the extremity of the children and truncal lesion was exceedingly rare [3]. In fact, among more than 800

**Figure 5.** Histological examination revealed lobules of proliferating, univacuolated, mature (arrow) and immature adipocytes.
liposarcomas on the file at the AFIP, we have not seen an unequivocal example of liposarcoma in a patient less than 5 years of age [9]. Approximately one half of the mediastinal masses were malignant, and of neurogenic or lymphomatous origin. Neurogenic tumors predominated before the age of 4 years, while lymphomas were most common beyond age 4 [14].

Surgery was essential for a definitive diagnosis due to the cytohistological resemblance of lipoblastoma to other fat-containing lesion in particular myxoid liposarcoma. Although the lesion was considered as biologically benign, it could grow locally to impressive proportion leading to significant mass effect. The tumor could cause recurrent chest infection, dyspnea, respiratory distress or even asphyxia due to compression on the trachea or bronchi. Due to worsening hypoxia and aspiration pneumonia, bronchoscopy could not be accomplished [6]. Death may result if expedient surgical removal is not undertaken [4]. Complete resection of the tumor mass to prevent the secondary complication of displacement and compression of adjacent structures is the treatment of choice.

In summary, lipoblastoma, although rare, should be considered in a young child with mediastinal fat-containing tumor. CT and MRI are useful in showing fat density and intensity respectively, ruling out compression of airway or the great vessels and delineating both the size and extent of the tumor.

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縱隔腔脂肪母細胞瘤：病例報告

李國泰¹ 王超然² 劉允澤² 薛 純³ 池永昌¹

聖保祿醫院 放射線科¹
林口長庚兒童醫院 放射線科²
林口長庚紀念醫院 病理科³

縱隔腔脂肪母細胞瘤是一種罕見胚胎脂肪間質瘤，常發生在3歲之前，少於20例已發表在英文的文獻中。在所有小孩腫瘤中，脂肪母細胞瘤佔不到1%的發生率。四肢是它好發生的地方。我們報告此九月大女嬰孩的罕有病例，不同於過往例子，他們都是年紀較大時才被診斷。我們有電腦斷層攝影及磁振造影的影像討論。手術中，腫瘤完全被切除，並沒有併發症。組織報告中証實為包膜葉狀的脂肪母細胞瘤。

關鍵詞：嬰孩；脂肪母細胞瘤；縱隔腔