**Case Report**

**Mediastinal Lipoblastomatosis Invading the Trachea in a 4-month-old Girl: A Rare Cause of Airway Obstruction**

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An infant presented respiratory failure led by a mediastinal tumor invading the trachea, further diagnosed as lipoblastomatosis. Lesion removal and trachea reconstruction were performed successfully.

CT image showing the mediastinal lesion (*) and its infiltration (#) into the trachea.
Abstract
We report a rare occurrence of mediastinal lipoblastomatosis infiltrating the trachea in a 4-month-old female, which led to critical airway obstruction and recurrent cardiopulmonary arrest. The lesion was surgically removed, and tracheal reconstruction was performed, followed by an uneventful postoperative course. Histological and cytogenetic analyses confirmed the diagnosis. We highlight the importance of considering lipoblastomatous tumors in children presenting with a rapid-growing fatty neoplasm, even though it happens in uncommon locations. Also, an adequate period of follow-up is demanded as a consequence of the notable rate of recurrence.

Keywords
Mediastinal tumor; endotracheal tumor; lipoblastoma; lipoblastomatosis; lipoblastomatous tumor.
Case Report

A 4-month-old female weighing 6.5kg presented with progressive wheezing and irritative cough for one month. Physical examination revealed nasal flaring, laryngeal strider, and three-concave sign.Computed tomography (CT) (Figure 1A,1B, Video 1) demonstrated a large hypodense tumor in the superior anterior mediastinum, which infiltrated the trachea and caused critical airway obstruction. Bronchoscopy (Figure 1C, Video 2) confirmed the presence of the 10-mm endotracheal mass. Laboratory examinations were mostly negative. The initial diagnosis was suspected hemangioma, while the size seemed not to decrease after empiric treatment with propranolol and methylprednisolone.

Due to respiratory failure, the patient was intubated and treated by mechanical ventilation. However, cardiopulmonary arrest took place three times. Even though successfully resuscitated, surgical intervention was urgently necessitated.

A sternotomy strategy was decided considering the location and size of the tumor. The venous-artery extracorporeal membrane oxygenation (V-A ECMO) was set up via the right atrium and ascending aorta. The ventilator was turned down during ECMO support. We mobilized the vessels and exposed the tumor (Figure 1D), which was adjacent to the tracheoesophageal groove and invaded the anterolateral trachea wall.

Bronchoscopy guiding the endotracheal margins, the neoplasm, together with involved trachea segment including eight rings, was resected. The trachea was reconstructed by interrupted end-to-end anastomosis (Figure 1E, Video 3). The mass was partly mucous, with a cut surface of lobulated, yellow-gray parenchyma (Figure 1F).
The patient maintained chin-to-chest position and was extubated one week postoperatively. She is currently under follow-up in good condition without anastomotic dehiscence for ten months.

The pathological examination indicated the lesion as a lobulated lipoblastomatous tumor. Immunohistochemical staining detected the presence of MDM2, CDK4, and S100. The Ki-67 index was 5%. PLAG1 was detected by fluorescence in situ hybridization (FISH) (Figure 2). The diagnosis was corrected as lipoblastomatosis, in accordance with its invasive but benign nature.

Discussion

Lipoblastomatous tumors, first reported in 1920s, almost exclusively affecting children under 3, are benign fast-growing mesenchymal neoplasms deriving from fatty tissue. Two subtypes exist: lipoblastoma, localized well-circumscribed form, and lipoblastomatosis, diffuse multicentric form. Around half of lesions are located superficially in the extremities, followed by the trunk, head, neck, groin, and perineum. Rare counterparts, mediastinal lipoblastomatous tumors are typically symptomized by dyspnea or hemiparalysis, resulting from compression of the airway, lung or spinal cord. Specifically, lipoblastomatosis tends to infiltrate surroundings, but no malignant degeneration or metastasis has been documented. Torre et al reported a lipoblastomatosis of the neck with localization in tracheal and esophageal walls that required laryngotracheal and esophageal resection. To our knowledge, this is the first case of lipoblastomatosis invading the trachea.
CT demonstrates the soft-tissue character and fatty origin; however, differential diagnosis from other fatty tumors is challenging. Lipoblastomatosis resembles liposarcoma in terms of fast infiltrative growth, irregular shape and obscure margin, which shows contrast enhancement on CT, and predominantly occurs in adults. Histologically, lipoblastomatous tumors are lobulated by connective tissue, and present with myxoid changes with lipomatous cells at different stages of maturity, while liposarcoma features nuclear atypia. Moreover, PLAG1 generated by rearrangements of region 8q11-13 is the hallmark of lipoblastomatous tumors, whereas a characteristic translocation t(12;16) (q13;p11) is present in myxoid liposarcoma.

Regardless of site, surgical resection is mostly recommended. In our case, the endotracheal part had led to irreversible airway obstruction, and surgery was the only strategy. Generally, postoperative prognosis is favorable, but the local recurrence rate is 14% to 25%, which occurs more frequently in lipoblastomatosis. Sometimes, even though the resection appeared macroscopically complete, relapse still occurred as shortly as one month from the initial surgery. Therefore, careful follow-up of 5 years is recommended.

In conclusion, pediatric mediastinal lipoblastomatous tumor invading the trachea is rare but should be considered in the differential diagnosis of a rapid-growing fatty neoplasm. Surgical resection is recommended, and the prognosis tends to be favorable due to the benign nature. Nevertheless, follow-up of adequate period of time is demanded due to the relatively high recurrence rate.
The patient's parents provided written informed consent for the publication of the study data; IRB approval was not required.
References


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**Figure legends**

Figure 1: Radiologic, endoscopic, and intraoperative manifestations of the mediastinal tumor. (A) CT image demonstrating the mediastinal tumor and its infiltration into the trachea. (B) Three-dimensional reconstruction of CT showing the critical airway obstruction. (C) Bronchoscopy confirming the proximal and distal margins of the endotracheal mass. (D) Intraoperative view after mobilizing the surrounding structures and exposing the mediastinal neoplasm. (E) Intraoperative view after resecting the neoplasm with the involved trachea segment, and reconstructing the trachea. (F) Removed two parts of the lesion.

★️: mediastinal part of the lesion; #: endotracheal part of the lesion.

IV: innominate vein; IA: innominate artery; LCCA: left common carotid artery; AAO: ascending aorta.

A: aortic cannulation via the ascending aorta; V: venous cannulation via the right atrium.
Figure 2: Histopathology and immunohistochemical expression of the specimen. (A) Cryo-section image with hematoxylin and eosin stain presenting myxoid background and lobular structure with adipose tissue. (B) Immunohistochemical stain as positive for MDM2. (C) Immunohistochemical stain as positive for CD34. (D) Immunohistochemical stain as positive for S100. (E) Immunohistochemical stain as the Ki-67 index was 5%. (F) FISH detecting PLAG1 rearrangement.

PLAG1: pleiomorphic adenoma gene 1.

Video legends

Video 1: Three-dimensional reconstruction of CT showing the critical airway obstruction led by the mediastinal tumor and its infiltration of the trachea.

Video 2: Bronchoscopy revealing the location and size of the endotracheal mass.

Video 3: Intraoperative video recording of the mediastinal structures and surgical technique of complete tumor excision in this case.