

## Malignant mediastinal and left lung tumour – macroscopically radical resection in an infant with critical respiratory failure

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### Abstract

We present a multidisciplinary surgical treatment of a male 4-month-old infant with malignant mediastinal and left lung tumour who deteriorated rapidly with mechanical ventilation due to symptoms of local compression.

The child underwent life-saving surgical intervention by a multidisciplinary team. Gross total resection of the tumour was achieved via a combination of median sternotomy and “hemi-clamshell” incision. After an uncomplicated postoperative period the child was transferred to the oncology department for further chemotherapy.

Because of emergency hazardous resection the patient could be stabilized and subjected to continued chemotherapy, proving the possibility of multidisciplinary mediastinal tumour excision in emergency settings.

**Key words:** mediastinal tumour, lung tumour, respiratory insufficiency, surgery, malignant teratoma.

Malignant mediastinal tumours may lead to cardiac and respiratory symptoms due to local tumour mass compression. Local expansion of the tumour, including impaction of the heart and lungs with circulatory and respiratory deterioration due to impaired contractility and resultant atelectasis, sometimes leads to critical circulatory or mixed respiratory and circulatory failure. Initial chemotherapy usually leads to regression of the tumour mass and associated symptoms. In selective cases the neoplasm does not respond to chemotherapy. In this case a life-saving surgical intervention, aimed at reduction of tumour mass, might be the last option to save the patient before the final pathological verification of the diagnosis and modification of the chemotherapy.

### Streszczenie

Praca przedstawia raport z leczenia 4-miesięcznego chłopca z rozpoznaniem złośliwego nowotworu śródpiersia i lewego płuca, z gwałtownym pogorszeniem z powodu miejscowej ekspansji guza prowadzącej do krytycznej niewydolności oddechowej, co przyspieszyło decyzję o ryzykownej operacji chirurgicznej przeprowadzonej w wielospecjalistycznym zespole.

Dziecko zakwalifikowano do operacji ratującej życie. Wykonano makroskopowo radykalną resekcję guza z dostępu będącego kombinacją sternotomii pośrodkowej z cięciem typu *hemi-clamshell*. Po niepowikłanym wczesnym okresie pooperacyjnym chłopiec trafił do kliniki onkologii w celu kontynuowania chemioterapii.

Dzięki ryzykownej interwencji chirurgicznej uzyskano stabilizację dziecka oraz możliwość kontynuowania chemioterapii, co dowodzi korzyści wielospecjalistycznej współpracy w sytuacjach zagrożenia życia.

**Słowa kluczowe:** guz śródpiersia, guz płuca, niewydolność oddechowa, chirurgia, potworniak złośliwy.

### Case report

We present a multidisciplinary surgical treatment of a male 4-month-old infant, body weight 8 kg, with malignant mediastinal and left lung tumour. Previous bone marrow biopsy ruled out neuroblastoma. Neoplastic markers were as follows: AFP = 30.48 U/ml, CRP = 15.6 mg/l, VMA and methoxycatecholamines were elevated in 24-hour urine collection.

The child was referred for chest drainage because of the left pleural effusion and left lung atelectasis, but the chest drainage did not improve severe hypoxia caused by dyspnoea at rest and shortness of breath, despite constant oxygen delivery. The patient deteriorated rapidly due to symptoms of local tumour compression and predominant

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tly respiratory failure. Chest X-ray and CT (Fig. 1) showed an extensive mediastinal tumour with infiltration of the pericardium and the left lung. After an open tumour biopsy via left mini-thoracotomy approach the condition of the child deteriorated rapidly, and the boy required mechanical ventilation. Echocardiography findings showed atypical horizontal position of the heart with an impression in the apex of the pericardial sac, with mild decrease of contractility and mild atrioventricular valves regurgitation. The aorta and pulmonary artery were pushed downwards because of the tumour mass impression.

The decision to perform the surgical procedure was undertaken in an atypical, emergency life-saving setting, when the definitive histopathological diagnosis was missing. The strategy was aimed at relieving the symptoms of local tumour compression in order to give a chance for general stabilization of the patient before further chemotherapy.

Surgical intervention was performed by a multidisciplinary team (cardiac surgeon, thoracic surgeon, paediatric surgeon, cardiologist, radiologist, oncologist). The chest was opened via a combination of median sternotomy and “hemi-clamshell” incision. After meticulous dissection, gross total resection of the mediastinal tumour was achieved. The infiltration of the pericardium and the left pleural space was found (Fig. 2). Because of complete left lung infiltration including infiltration of its hilum, left pneumectomy was performed with concomitant resection of the thymus, partial resection of the left portion of the pericardial sac and division of persistent arterial duct (PDA) in the area of invaded aortic arch and pulmonary artery bifurcation. The early postoperative period was uncomplicated, with normal bloody chest drainage. The child was extubated on the second postoperative day. Wound healing was uneventful and the boy was transferred to the oncology department for further chemotherapy on the fifth postoperative day. Only after the surgery was completed the previous open tumour biopsy revealed the histology of

malignant teratoma with undifferentiated primitive neural tissue components. Current chemotherapy consisted of adriamycin, cisplatin, vincristine and cyclophosphamide; next it was modified to a malignant teratoma protocol (VBP scheme). Control echocardiography showed stenosis of the right pulmonary artery that underwent an effective percutaneous balloon angioplasty. Despite the change of chemotherapy an effective response of the tumour was not achieved, and its rapid progression led to death of the child two months after surgery.

## Discussion

Teratomas of the mediastinum, although in the majority benign, are sometimes immature, and may metastasize to regional lymph nodes with a great variety of clinical and radiological features [1]. In such cases the treatment usually starts with chemotherapy [2].

Local mass impression effects might be typical in the course of an aggressively growing tumour [3], but are mostly related to teratomas in adulthood. Symptoms of respiratory failure when the mass compresses great vessels and airways are a common indication for emergency surgery, as is confirmed by the literature [1, 3]. Occasionally mediastinal teratomas cause additional complications, such as massive pleural effusion or haemothorax from its rupture after chest trauma [4, 5].

The presented case is unique because of the age of the child, aggressive growth of the neoplasm with impression of vital organs leading to critical circulatory and respiratory failure, and malignant histology unresponsive to chemotherapy. The youngest similar child reported to our best knowledge was 8 months old [6]. However, circulatory insufficiency due to heart failure was more common in older patients; the youngest of them was a 12-year-old boy with mediastinal teratoma impression of the heart [7].



Fig. 1. CT scan with mediastinal mass protruding into the left pleura with left lung infiltration and atelectasis



Fig. 2. Intraoperative picture: Resection of the thymus, partial resection of the left portion of the pericardial sac and division of the persistent arterial duct (PDA) in the area of the invaded aortic arch and pulmonary artery bifurcation were necessary

In our case the decision to perform an emergency thoracotomy was undertaken in the face of a critical respiratory insufficiency with awareness of the lack of a final diagnosis of the neoplasm in a dying patient. The primary clinical diagnosis, confirmed by imaging, justified the initial presumption that the tumour should respond to adjuvant chemotherapy following emergency surgery [1, 4]. A similar case of a massive immature teratoma aggressively extending into the left pleural cavity was reported in an 18-year-old woman, with a few paediatric cases found in the literature [5-7].

The presented approach proves the possibility of multidisciplinary mediastinal tumour excision in an emergency setting [6, 8].

The good early result enabled early extubation and weaning of the patient from mechanical ventilation, which corresponds with contemporary literature reports [1-8]. This was the turning point for continuation of chemotherapy, as well as for precise histopathological diagnosis and subsequent modification of chemotherapy [2].

Ultimately the patient died two months after surgery despite surgical and oncological efforts. Although rapid

tumour progression due to its unresponsiveness to chemotherapy put such a heroic surgical approach under question, we believe it was justified.

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