Simultaneous hemopericardium, hemoperitoneum and hemothorax after complete surgical resection of mediastinal cystic lymphangioma: A case report

**Alexis Mupepe Kumba, Filippo Banchini, Luigi Conti,
Rocco Delfanti, Patrizio Capelli**

ABSTRACT

Introduction: Lymphangioma is an atypical non-malignant tumor. It is most commonly observed in pediatric population and typically found in the cervical region. Its prevalence is estimated between 0.01% and 4.5% of all mediastinal tumors. Complete surgical resection is required to prevent recurrence.

Case Report: We present the case of a 47-year-old male with massive right pleural effusion. Magnetic resonance imaging showed a massive mediastinal cystic lymphangioma. A radical excision of the thymus and lymphangiomatosis cystic mass was performed by sternotomy. The postoperative course was marked by simultaneous hemopericardium, hemoperitoneum and hemothorax, the origin of which, was not well explained due to its presentation in the distal site of surgery. This required transfusions and new thoracoabdominal surgical exploration. The patient developed pulmonary and wound infections and died of sepsis and multi-organ failure.

Conclusion: Our case would be a further demonstration of an unusual complication of simultaneous hemopericardium, hemoperitoneum and hemothorax explained by some authors as a local production of fibrinolysin.

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CASE REPORT

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Simultaneous hemopericardium, hemoperitoneum and hemothorax after complete surgical resection of mediastinal cystic lymphangioma: A case report

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Introduction: Lymphangioma is an atypical non-malignant tumor. It is most commonly observed in pediatric population and typically found in the cervical region. Its prevalence is estimated between 0.01% and 4.5% of all mediastinal tumors. Complete surgical resection is required to prevent recurrence. **Case Report:** We present the case of a 47-year-old male with massive right pleural effusion. Magnetic resonance imaging showed a massive mediastinal cystic lymphangioma. A radical excision of the thymus and lymphangiomatosis cystic mass was performed by sternotomy. The postoperative course was marked by simultaneous hemopericardium, hemoperitoneum and hemothorax, the origin of which, was not well explained due to its presentation in the distal site of surgery. This required transfusions and new thoracoabdominal surgical exploration. The patient developed pulmonary and wound infections and died of sepsis and multi-organ failure. **Conclusion:** Our case would be a further demonstration of an unusual complication of simultaneous hemopericardium, hemoperitoneum and hemothorax explained

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Keywords: Cystic lymphangioma, Hemoperitoneum, Hemothorax, Mediastinum neoplasm, Pericardial effusion

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INTRODUCTION

Lymphangioma is an atypical non-malignant tumor, which occurs during the embryonic development of the lymphatic system. It is most frequently observed in the head and neck, but can occur at any location in the body [1–3]. The exact incidence is unknown, but its prevalence is estimated to be between 0.01% and 4.5% of all mediastinal tumors [4–6]. Lymphangiomas have been classified into three types: simplex lymphangioma (capillary lymphangioma), cavernous lymphangioma, and cystic lymphangioma (cystic hygroma) [7]. Since the mediastinum is hypobaric, most lymphangiomas are cystic. [5]. Largely asymptomatic, cystic lymphangioma can present a diagnostic challenge often requiring lymphoscintigraphy and magnetic resonance imaging (MRI) scan in order to be identified. Treated with complete

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surgical resection, long-term prognosis is excellent [5, 8]. We report the case of a young man with mediastinal cystic lymphangioma who developed hemopericardium, hemoperitoneum and hemothorax of unknown origin one day after complete surgical resection and later died.

CASE REPORT

A 47-year-old male was transferred to our institute from a nearby hospital for massive right pleural effusion. He presented with tachypnea, hypoxia (PO_2 66 mmHg on room air, PCO_2 33 mmHg, SaO_2 94%), without fever. Respiratory movements were absent in the right pulmonary fields. The abdomen was increased in volume with ascites effusion and a slight defensive reaction. Biological examinations were normal, except for CRP (1.17 mg/dl), white blood cells ($16.95 \times 10^3/\mu\text{l}$) and D-dimer ($3200 \mu\text{g/l}$). A chest radiograph, on frontal projection, showed a homogeneous water-density mass in the right pulmonary fields and in the paratracheal area (Figure 1).

An additional thoracoabdominal computed tomography scan revealed abundant pleural effusion associated with collapse of the right lung, and the presence of multiple swollen lymph nodes (7 cm maximum axial diameter) partially confluent in the upper mediastinum and cardiophrenic angle. Multiple hypodense structures of about 12 mm maximum axial diameter were observed around the spleen, lymph nodes (2.5 cm of the maximum axial diameter) were also observed in the obturator region bilaterally, along the internal and external iliac chain and other lymph nodes (1.5 cm of the maximum axial diameter) in the paracaval and para aortic regions.

We decided to perform diagnostic thoracoscopy due to the presence of unknown mediastinal mass that revealed chylous effusion along the mediastinal pleura, diffuse thickening and fibrosis of mediastinal tissue dripping chyle. A biopsy of the mediastinal tissue in the Barety lodge until tracheal plane was carried out. Histologic examination of the biopsy sample showed fibroadipose tissue comprising residues of timing structures, dilated vascular spaces provided with flat endothelial coating positive for CD31, CD34 and Podoplanin (D2-40).

Magnetic resonance imaging (MRI) scan revealed a massive lymphangioma extending craniocaudally 22 cm and 9 cm in transverse diameter involving the entire anterior and posterior mediastinum surrounding the lower third of the esophagus and involving the celiac trunk with hypointensity in T1 and low hyperintensity in T2 (Figure 2). Therefore, a radical excision of the thymus and lymphangiomatosis cystic mass was performed by sternotomy and debulking of the mediastinum without opening the abdomen and pericardium (Figure 3).

Final histology pointed to cystic lymphangioma with overlapping outbreaks of chronic and acute suppurative inflammation. The postoperative course was marked by simultaneous hemopericardium, hemothorax and hemoperitoneum which required transfusions and

Figure 1: A chest radiograph showing a homogeneous water-density mass.

Figure 2: Magnetic resonance imaging scan showing massive lymphangioma.

Figure 3: Radical excision of thymus and lymphangiomatosis cystic mass was performed by sternotomy and debulking.

new surgical exploration. A thoracoabdominal incision enabled both chest and abdomen exploration at the same time, without objectifying the source of the blood effusion. Packing was performed to achieve hemostasis. Pericardial blood effusion was evacuated by left anterior thoracotomy. As a consequence of this complication, the patient developed pulmonary and wound infections and died of sepsis and multi-organ failure despite VAC-therapy and oriented antibiotics.

DISCUSSION

Lymphangioma is more commonly observed in the pediatric population and is typically found in the cervical region, because of its derivation from a primitive lymphatic sac, Ninety percent of cases are diagnosed by two years of age and are more common in females [1, 4, 6]. In a study of 37 cases with lymphangioma, Rajesh et al. found a mean age of 45 years [1]. Higher prevalence of lymphangioma is seen in patients with chromosomal abnormalities like Down syndrome, Turner syndrome, Edwards syndrome, and Patau syndrome. An acquired variant of lymphangioma can occur in patients due to chronic obstruction of the lymphatics. This is seen more commonly in middle-aged patients with a history of surgery, those undergoing radiation therapies for malignancy, and those suffering from a chronic infection [1]. In the pediatric population, lymphangioma is typically found in the cervical region but, in adults, it commonly presents as a mediastinal mass without a cervical component [4].

In this case, the patient was an adult male with a mediastinal mass. Screening for chromosomal abnormalities was not performed as no morphological sign matches were present. There was no history of surgery neither of chronic infection nor radiotherapy. Most lymphangiomas are asymptomatic [9]. Symptomatic patients are often found to have large lesions. The symptomatic manifestation can be variable depending on the site. Dyspnea, respiratory distress and hemoptysis are the most common symptoms that occur when the lymphangioma compresses the tracheobronchial tree, pharynx, or phrenic nerve, or presents as pleural effusion [4, 10, 11]. The patient presented pleural effusion, dyspnea and abdominal distension. Lymphangiomas can affect all compartments, but anterior and superior mediastinum are those most commonly affected as in our case. Some unusual sites described are the hilum, pericardium, axilla, shoulder joint, retroocular region, esophagus and chest [12]. Imaging techniques such as computed tomography (CT) scan and magnetic resonance imaging (MRI) scan may be used to make a clinical diagnosis and for follow-up. They respectively show homogeneous low density and T2 hyperintensity [12]. The latter was not confirmed in our case with only low hyperintensity in T2.

The final diagnosis is based on a combination of clinical, radiological, and histopathological findings. On

CT scan and/or MRI scan, a cystic lymphangioma can be suggested based on multiseptated and loculated mass [12]. The gold standard treatment of lymphangioma is complete surgical resection [1, 2, 13]. However, due to the infiltrative nature of lymphangiomas, this procedure becomes incomplete and hard to use and can only be applied in 10–50% of the cases. Therefore, recurrence represents an important problem, with a prevalence rate between 0% and 27% after total exeresis and between 15% and 53% after partial exeresis. [2, 13, 14].

The patient presented hemopericardium, hemothorax and hemoperitoneum without any obvious cause one day after complete surgical resection. In literature, coagulopathy has been reported in association with lymphangiomatosis, and several potential mechanisms for this association have been advanced. Local production of fibrinolysin by lymphangiomas may be responsible for the coagulopathy [15]. This could explain blood effusion far from surgical site in the patient. In literature surgical complications, which occur in 19–33% of the cases, include formation of hematoma, lymphocele, scar, abscess, infection, wound dehiscence and nerve palsy have been noticed by several authors [14, 16–18].

To the best of our knowledge, simultaneous hemopericardium, hemothorax and hemoperitoneum after mediastinal lymphangioma surgery has not been reported. Various alternative treatments for lymphangioma have been attempted; they include laser therapy, radiation therapy, chemotherapy and the use of sclerotizing agents [2, 19]. The efficacy of these treatments is highly variable in incidental reports. They can be used as a therapeutic weapon against lymphangiomas, especially in patients with unresectable or life-threatening lesions or in cases in which surgery would be mutilating. Spontaneous regression of mediastinal lymphangioma has been seen, although it remains uncommon [20]. Among sclerotizing agents, OK-432 (Picibanil), a lyophilized mixture of *Su*-protein of group A *Streptococcus pyogenes*, incubated with penicillin G, had a response rate up to 92% with minimal side effects and no cicatricial damage to the skin [21].

CONCLUSION

Mediastinal lymphangioma is a rare condition and should be included in the differential diagnosis of patients presenting with pleural effusions. The treatment of lymphangioma is well known to be a complete surgical resection, but this technique is not always possible to perform. Hemopericardium, hemoperitoneum, and hemothorax together are rare but when present, complicate surgery and are difficult to manage. Some alternative treatments have been tried with good outcomes.

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Author Contributions

Alexis Mupepe Kumba – Substantial contribution to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Filippo Banchini – Substantial contribution to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Luigi Conti – Substantial contribution to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Rocco Delfanti – Substantial contribution to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Patrizio Capelli – Substantial contribution to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

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Conflict of Interest

Authors declare no conflict of interest.

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