Case Report • DOI: 10.2478/pneum-2025-0032 • 74 • 2025 • 96-100

Pneumologia

Angiosarcoma – a rare cause of hemoptysis

Maria-Mădălina Vava[1,*], Raluca-Cosmina Mureșan[1], Oana Gocan[1], Adriana Părău[1], Doina-Adina Todea[1,2]

[1]"Leon Daniello" Clinical Hospital of Pneumology, Cluj-Napoca, Romania
[2]"Iuliu Haţieganu" University of Medicine and Pharmacy, Cluj-Napoca, Romania

Abstract

English:

Introduction: *Angiosarcoma is a very rare and aggressive vascular malignant tumor with origin in the vascular and lymphatic endothelium. It can be located in any part of the body, however pulmonary location is very infrequent. Hemoptysis can be the revealing symptom in these cases.*
Presentation of the case: *We present the case of a 74 year old male patient admitted in our clinic for prolonged cough and hemoptysis after being discharged from the maxillofacial clinic where he was admitted for drainage of dental abscesses, the patient being totally edentulous at the time of the presentation. He also presented aggravation of a chronic left shoulder pain in the past 2 months.*
Multiple investigations were required because of the uncertain tumoral aspect shown on the CT scan and the lack of endobronchial expression of the tumor. Following these procedures, an upper right cervico-mediastinal tumor and a left osteolytic scapular tumor were detected. The diagnosis was confirmed by the ultrasound-guided biopsy of the cervico-mediastinal formation. The evolution was towards the appearance of a spontaneous right hemothorax and severe anemia with progressive worsening and death in the Oncology department approximately 2 months after the initial presentation.
Discussions: *We presented the case of a patient with persistent hemoptysis with the initial suspicion of bronchopneumonia, in which, following further investigations, extrapulmonary angiosarcoma was identified, with 2 locations, cervico-mediastinal and bone location (scapular) but without histopathological confirmation of the lung lesion. Given the risk of massive hemoptysis, invasive bronchoscopic procedures were avoided.*
Conclusion: *Angiosarcoma is a very rare pulmonary malignant tumor and a rare cause of hemoptysis with a difficult diagnostic process.*

Keywords

angiosarcoma • haemoptysis • spontaneous haemothorax • ultrasound-guided biopsy

Angiosarcom extra-pulmonar: o cauză rară de hemoptizie

Rezumat

Romanian:

Introducere: *Angiosarcomul este o tumoră malignă vasculară foarte rară și agresivă, cu origine în endoteliul vaselor sanguine și limfatice. Poate apărea în orice regiune a organismului, însă localizarea pulmonară este extrem de rară. În aceste situații, hemoptizia poate reprezenta simptomul de debut.*
Prezentarea cazului: *Prezentăm cazul unui pacient de 74 de ani, internat pentru tuse persistentă și hemoptizie, apărute la scurt timp după externarea dintr-o secție de chirurgie oro-maxilo-facială, unde fusese tratat pentru abcese dentare. La momentul prezentării, pacientul era complet edentat și acuza agravarea unei dureri cronice la nivelul umărului stâng în ultimele două luni. Din cauza aspectului tumoral neconcludent la tomografia computerizată și a absenței expresiei endobronșice, au fost necesare investigații multiple, care au evidențiat o formațiune cervico-mediastinală dreaptă și o leziune osteolitică a scapulei stângi. Diagnosticul a fost confirmat prin biopsie ghidată ecografic a formațiunii cervico-mediastinale. Evoluția a fost rapid nefavorabilă, cu*

*Corresponding author: Maria Madalina Vava
E-mail: madalina.vava@yahoo.com

apariția unui hemotorax drept spontan și anemie severă, soldată cu decesul pacientului la aproximativ două luni de la prezentarea inițială.
Discuții: *Cazul ilustrează o cauză rară de hemoptizie persistentă, inițial interpretată ca bronhopneumonie, ulterior diagnosticată ca angiosarcom extrapulmonar cu localizări cervico-mediastinală și osoasă, fără confirmarea histopatologică a unei leziuni pulmonare. Din cauza riscului crescut de hemoragie masivă, procedurile bronhoscopice invazive au fost evitate.*
Concluzie: *Angiosarcomul este o malignitate pulmonară extrem de rară și o cauză neobișnuită de hemoptizie, caracterizată printr-un proces diagnostic dificil și un prognostic rezervat.*

Cuvinte-cheie

angiosarcom • hemoptizie • hemotorax spontan • biopsie ghidată ecografic

Introduction

Angiosarcoma is a rare and aggressive malignant vascular tumour originating in the endothelium of the blood and lymphatic vessels, with possible localisation in any part of the body, but more frequently in the cephalic extremity. It accounts for <2% of all sarcomas (1). Metastasis of angiosarcoma is common in about half of diagnosed cases, being predominantly in the lungs, with primary tumours being rare at this stage (2). Thus, initial clinical manifestations may present as non-specific respiratory tract symptoms (dyspnoea, cough, haemoptysis).

Case report

We present the case of a 74-year-old male patient, non-smoker, vaccinated against SARS-COV-2, no drug allergies or environmental factors, no occupational exposure to respiratory toxins, with a medical history of high blood pressure, heart attack, ischaemic heart disease, minor valvular abnormalities, two episodes of ischaemic stroke with sequelae tremor in the upper limbs and generalised epilepsy under treatment. Anamnestically, he reports the onset of symptoms 3 weeks ago, with whooping cough and haemoptysis after discharge from the oral-maxillofacial surgery service, where he was hospitalised for the treatment of dental infectious outbreaks; he was totally edentulous at the time of presentation. The patient presented with an episode of hypoxemia objectified at home 2 weeks ago, with a minimum SpO_2 value of 90% in room air and chronic left shoulder pain, which had worsened over the past 2 months.

Physical examination revealed pale skin, grade I obesity with abdominal distribution (body mass index (BMI) 30.42 kg/m^2), functional impotence in the left upper limb, with significant limitation of movement in all planes, slightly kyphotic thorax, present vesicular murmur bilaterally, slightly diminished right basally, rhonchi in the lower half of the right lung and right

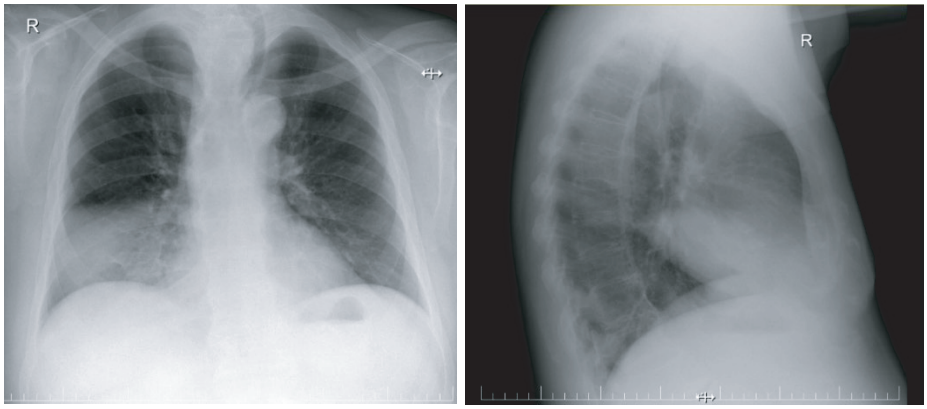

Figure 1: Chest radiograph postero-anterior view (PA) and right lateral view (LLR) incidence: cervical formation with mass effect on the trachea, lung condensation located in the middle lobe.

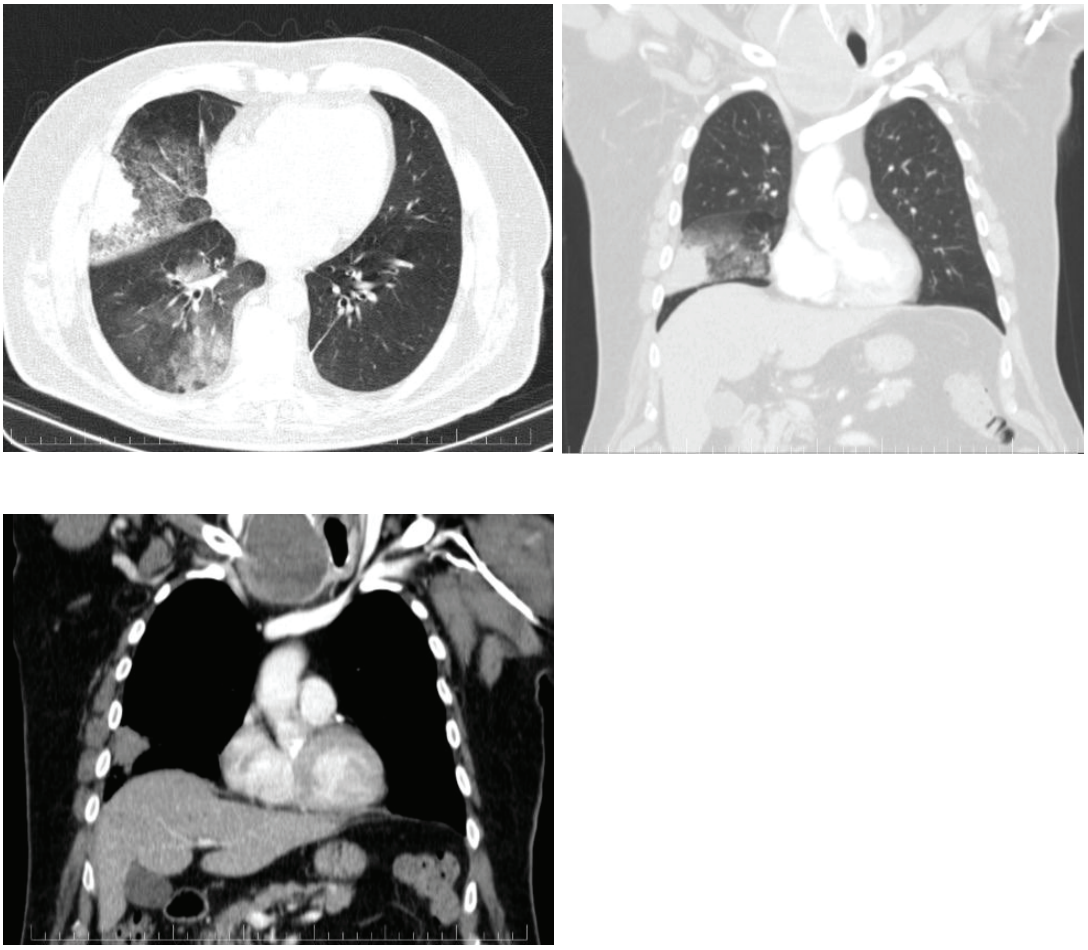

Figure 2: Chest computed tomography: pulmonary condensation of the middle lobe, surrounded with ground glass opacity both at the middle lobe and right lower lobe.

internal supraclavicular formation, firm to palpation, painless, fixed in the deep planes, approximately 5 cm in diameter and without swallowing difficulties.

Clinically, aspiration pneumonia was initially suspected in connection with multiple dental extractions, but a proliferative process could not be excluded (Figure 1).

Mild inflammatory syndrome was identified, with procalcitonin within normal limits and elevated D-dimer. AngioCT examination ruled out pulmonary thromboembolism, but revealed a cervico-mediastinal anterior paratracheal right formation with tissue structure, with postcontrast peripheral iodophilia and mass effect on the structures, pulmonary condensation at the middle lobe, without air bronchogram, plaqued on the pleura, associated with changes in 'ground glass' both at the middle lobe and right lower lobe with suggestive aspect of alveolar haemorrhage in the clinical context presented. We also identified osteolytic lesions

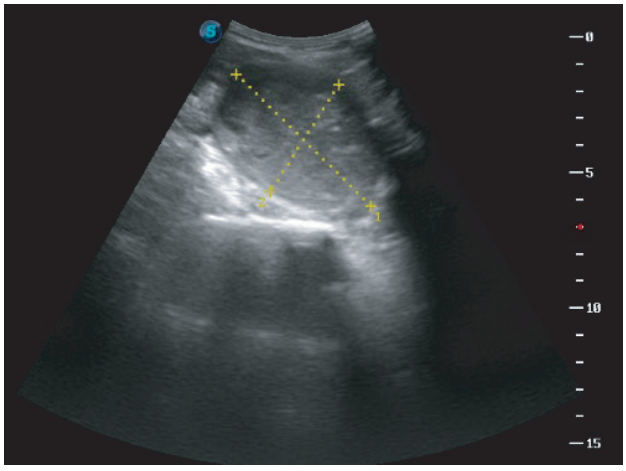

Figure 3: Ultrasonography of the cervico-mediastinal mass: hypoechoic structure with oval shape and well-defined margins.

associated with fracture at the scapula body and left coracoid bone, in the absence of thoracic trauma (Figure 2).

Fibrobronchoscopy showed no evidence of endobronchial lesions, only bleeding and clots from the middle lobar bronchi and contralateral haemorrhagic secretions, probably aspirated. In this context, 'blind' transbronchial biopsy of the middle lobe is avoided. For the differential diagnosis of alveolar haemorrhagic syndrome, pANCA and cANCA are performed, with negative results. Mycologic examination of the bronchial aspirate identifies the presence of Itraconazole-sensitive *Candida glabrata*. In the absence of an endobronchially or ultrasonographically accessible pulmonary lesion, an ultrasonographic biopsy of the right upper cervico-mediastinal formation is performed after an interventional radiology consultation, which temporises the biopsy of the left scapular lesion (Figure 3).

Imaging monitoring reveals atelectatic changes in the middle lobe.

Under intravenous antibiotic treatment with third-generation Cephalosporins and Lincosamide, antifungal, haemostatic, intravenous corticoid in low doses, gastric and hepatic protector, the patient's general condition remained good, and haemoptysis was reduced quantitatively but persisted, with pain in the scapular region and functional impotence of the left upper limb in this context. The patient was discharged pending histopathologic results. He was readmitted by emergency 3 weeks after discharge, with a worsened general condition manifesting as shortness of breath, persistent haemoptysis, altered physical performance, severe anaemia (Hb 4.8 g/dL) with repeated transfusion requirements and hypoxemic (SpO_2 88% in room air). Thoracic-pleuro-pulmonary radiography in postero-anterior view showed the presence of a small right pleural effusion associated with known right basal atelectasis (Figure 4).

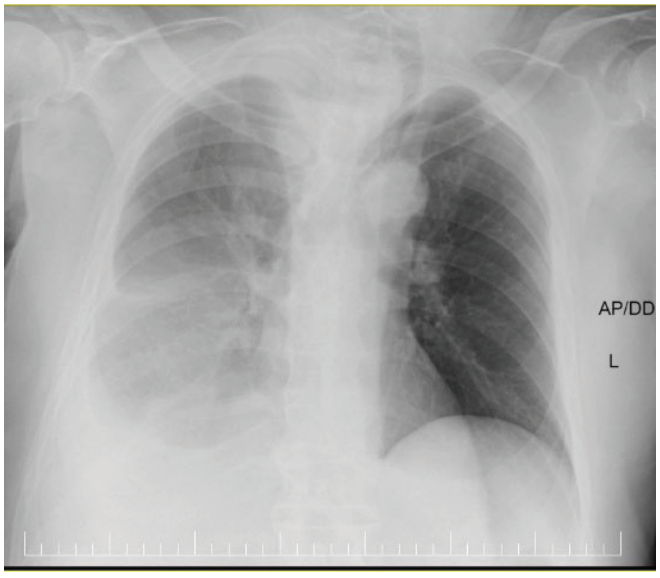

Figure 4: Chest radiograph: right pleural effusion.

Due to the worsening tendency of dyspnoea, the patient was monitored ultrasonographically, with the identification of a growing right pleural effusion in large quantity. A right thoracocentesis was performed at the bed, with evacuation of 1000 mL of intensely haemorrhagic fluid. A small-medium remaining collection was seen, as an intense haemorrhagic exudate (haematocrit 21.9%), without signs of malignancy on cytologic examination. The histopathologic findings from the cervical formation suggestive for angiosarcoma were obtained. Due to the massive necrosis, biopsy from another localisation or excision of the formation was recommended. The patient was transferred to the Oncologic Institute for further evaluation and treatment and was admitted to the ICU. Subsequently, angiosarcoma was confirmed from guided CT biopsy of the left scapular lesion. The patient died approximately 2 weeks after transfer to the Oncology ward.

Discussion

We present a case of persistent haemoptysis in a patient with an initial suspicion of bronchopneumonia, in whom extrapulmonary angiosarcoma with two localisations was identified, the cervico-mediastinal and left scapular bone, but without confirmation of pulmonary lesion, the source of haemoptysis. It could not be specified whether the lesions detected represent the primary tumour or its metastasis. Because of the risk of massive haemoptysis, transbronchial invasive procedures were avoided. Subsequently, the appearance of haemothorax supports the aggressive course with haemorrhagic complications of an angiosarcoma with parenchymal lung localisation. The general condition of the patient did not allow further invasive investigations.

D-dimer are degradation products of fibrin, a protein involved in blood coagulation. They are used as indicators of coagulation processes; elevated levels suggest systemic activation of haemostasis and fibrinolysis. Angiosarcoma, due to its vascular origin, can lead to activation of the coagulation system with clot formation and their degradation, leading to elevated D-dimer levels (3). This has been associated with shorter survival, a higher risk of mortality (4) and increased risk of metastasis (5).

The particularity of the case is the masked 'pseudopneumonic' onset in connection with repeated surgeries in the oral-maxillofacial sphere in a patient with painful symptoms and supraclavicular formation, neglected by the patient and unidentified at previous medical examinations. The time from presentation to death was approximately 2 months.

The most common risk factors are chronic lymphoedema and a history of radiotherapy (6), which were not identified

in this case. It is diagnosed at a higher rate in older Caucasian men, often postmortem (7). The oncological treatment varies depending on the localisation and stage; in advanced or metastatic forms options include chemotherapy with doxorubicin and paclitaxel, as well as immunotherapy (8).

Conclusions

Haemoptysis is an unpatognomonic symptom, common in pulmonary pathology, often raising multiple problems of differential diagnosis, involving complex examinations (imaging, bronchoscopic), for which oncologic pathology is one of the main causes. Angiosarcoma is a rare malignancy and all the more so, because of its pulmonary localisation. For this reason, diagnosis can be difficult. Even in the presence of histopathologic confirmation, the course is aggressive, the prognosis poor and the therapeutic options limited (9).

Conflict of interest

The authors declare there is no conflict of interest.

Informed consent statement

Informed consent was obtained from the subjects involved in the study.

Funding

This study did not receive any funding.

References

1. Lim R, Harper L, Swiston J. Clinical manifestations and diagnostic methods in pulmonary angiosarcoma: protocol for a scoping review. *Systematic Reviews*. 2017;6(1): 136. doi:10.1186/s13643-017-0531-6
2. Grafino M, Alves P, de Almeida MM, Garrido P, Hasmucrai D, Teixeira E, et al. Angiosarcoma of the lung. *Jornal Brasileiro de Pneumologia: publicação oficial da Sociedade Brasileira de Pneumologia e Tisilogia*. 2016;42(1): 68–70. doi:10.1590/S1806-37562016000000193
3. Ay C, Dunkler D, Pirker R, Thaler J, Quehenberger P, Wagner O, et al. High D-dimer levels are associated with poor prognosis in cancer patients. *Haematologica*. 2012;97(8): 1158–1164. doi:10.3324/haematol.2011.054718
4. Raj SD, Zhou X, Bueso-Ramos CE, Ravi V, Patel S, Benjamin RS, et al. Prognostic significance of elevated D-dimer for survival in patients with sarcoma. *American Journal of Clinical Oncology*. 2012;35(5): 462–467. doi:10.1097/COC.0b013e31821d4529
5. Morii T, Mochizuki K, Tajima T, Ichimura S, Satomi K. D-dimer levels as a prognostic factor for determining oncological outcomes in musculoskeletal sarcoma. *BMC Musculoskeletal Disorders*. 2011;12: 250. doi:10.1186/1471-2474-12-250
6. Gaballah AH, Jensen CT, Palmquist S, Pickhardt PJ, Duran A, Broering G, et al. Angiosarcoma: clinical and imaging features from head to toe. *The British Journal of Radiology*. 2017;90(1075): 20170039. doi:10.1259/bjr.20170039
7. Sturm EC, Marasco IS, Katz SC. Multidisciplinary management of angiosarcoma – a review. *The Journal of Surgical Research*. 2021;257: 213–220. doi:10.1016/j.jss.2020.07.026
8. Cao J, Wang J, He C, Fang M. Angiosarcoma: a review of diagnosis and current treatment. *American Journal of Cancer Research*. 2019;9(11): 2303–2313.
9. Mullins B, Hackman T. Angiosarcoma of the head and neck. *International Archives of Otorhinolaryngology*. 2015;19(3): 191–195. doi:10.1055/s-0035-1547520