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Case Report

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Intimal Sarcoma of the Pulmonary Artery: A Rare and Insidious Neoplasm Often Mistaken for Thromboembolism - A Case Report and Literature Review

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Received Date: 23 Oct 2025;
Accepted Date: 28 Oct 2025;
Published Date: 31 Oct 2025.

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Citation: Filotico M, Plutino FM, Tramacere M. Intimal Sarcoma of the Pulmonary Artery: A Rare and Insidious Neoplasm Often Mistaken for Thromboembolism - A Case Report and Literature Review. Innov Insights Case Rep Rev. 2025; 1(2): 1-4.

ABSTRACT

A case of a rare and aggressive tumor called pulmonary artery intimal sarcoma, which is often mistaken for pulmonary thromboembolism, is presented. We report the case of a 67 year old man with asthenia and a persistent cough. This case highlights the importance of early detection of this rare tumor.

Keywords: *Pulmonary artery, Pulmonary thromboembolism, Intimal sarcoma, Immunohistochemistry, Vimentin*

Introduction

The Intimal sarcoma of the pulmonary artery is a rare tumor. Diagnosis is difficult and often delayed owing to the nonspecific nature of the symptoms. Since intimal sarcoma of the pulmonary artery is so rare and insidious it is often confused with pulmonary thromboembolism and as such treated.

Case Report

A 67 year old man has presented with asthenia and persistent cough since December 2022. In March 2023 hospitalized for pneumonia(?)

From the Patient's Medical Record

» May/08/2023 CT Scan - Presence of extensive endoluminal filling defect of thrombo-embolic nature which affects the common trunk, and extends to extensively affect the right pulmonary artery, and its main branches both for the lower lobe and mainly for the upper one (figure 1a).

» May/19/2023 CT Scan - in the right perihilar site, mostly represented at the level of the upper lung sector, solid tissue with lobulated margins, extended cranio-caudally for approximately 8 cm, which incorporates the bronchovascular branches. Endoluminal filling defects with an occlusive-subocclusive

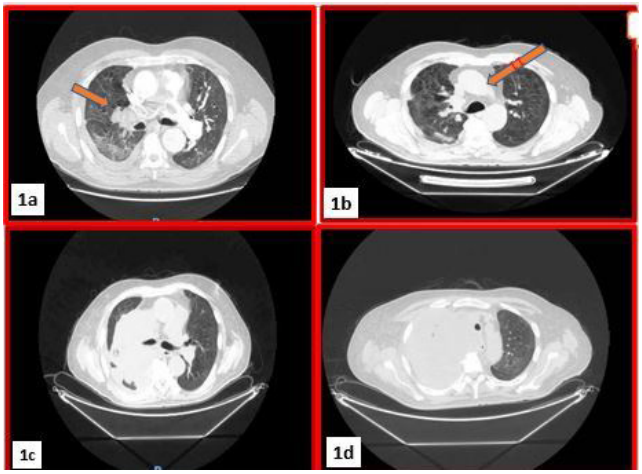

Figure 1. (a) *"CT Scan (05/08/2023) - Extensive endoluminal filling defect affecting the common trunk, and extends to ext ensively to the right Pulmonary Artery(arrow);* ***(b)*** *CT Scan (05/19/2023) - In the right perihilar site, mostly represented at the level of the upper lung sector, solid tissue with lobulated margins(arrow) Endoluminal filling defects with an occlusive-subocclusive appearance affecting the right PA branches, the PA common trunk;* ***(c)*** *CT Scan(08/21/2023) - Compared to the previous CT check, the already known solid tissue appears increased, occupying a good part of the right lung field;* ***(d)*** *CT Scan (08/28/2023) - Further increase in the predominantly hypodense non-homogeneous solid tissue that occupies almost the entire right lung area.*

appearance affecting the right PA branches, the PA common trunk and the proximal section of the main left PA branch (figure 1b).

» On July/25/2023, a transbronchial biopsy is performed from the neoplastic mass.

» August/21/2023 CT Scan - Compared to the previous CT check, the already known solid tissue appears increased, occupying a good part of the right lung field (figure 1c).

» August/28/2023 – CT Scan - 0 where there is almost no longer any visible ventilated parenchyma (figure 1d).

» August/28/2023 - The Angio-CT study demonstrates defective opacification of the left internal carotid artery in its intracranial course from the intracavernous tract, of the middle cerebral artery from its origin to its proximal and distal branches and of the anterior cerebral artery from its origin to the its more distal branches. On the left, extensive hemorrhage is documented, with cortico-subcortical distribution in the parieto-temporo-occipital area with extension to the nucleus-basal region and partial extension to the frontal lobe.

A few days after this last investigation the patient dies.

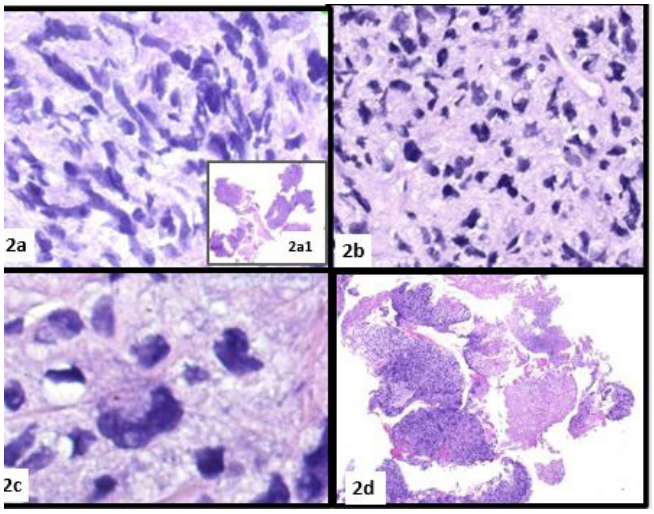

Figure 2. (a) *Spindle cell component of the neoplastic Tissue(HE 250X); a1 the small fragments resulting from the biopsy (insert);* ***(b)*** *Neoplastic cells tend to have a globular shape, indistinct edges, slightly acidophilic cytoplasm(HE 200X);* ***(c)*** *A multinucleated anaplastic cell (HE 250X);* ***(d)*** *Neoplastic fragments mixed with thrombotic material(HE 175X);*

Material and Methods

The biopsy material consists of some small fragments of tissue (figure 2a1) which are fixed in buffered formalin and embedded in paraffin. Sections of 5 µ are prepared from the included material and stained with Hematoxylin-Eosin. Other sections are tested with a large panel of antibodies: Vimentin, α1-Antitrypsin, MdM2, CD31, CD34, CD99, FL1, CD10, CD68, TTF1, P40, Cytokeratin (AE1/AE3), Chromogranin A, Synaptophysin, Actin (smooth muscle), Calretinin, S100, and Ki67.

Histology

It is a disordered and active proliferation of elements sometimes spindle-shaped(Figure 2a), sometimes epithelioid(Figure 2b). with amphophilic cytoplasm with indistinct margins and a hyperchromatic nucleus that is often highly atypical, sometimes multiple(Figure 2c). There is a brisk mitotic activity. The elements tend to be cohesive with each other and little stroma is interposed between them. The fragments of neoplastic tissue are mixed with thrombotic material, often infiltrated by isolated neoplastic cells (Figures.2d,3a,3b).

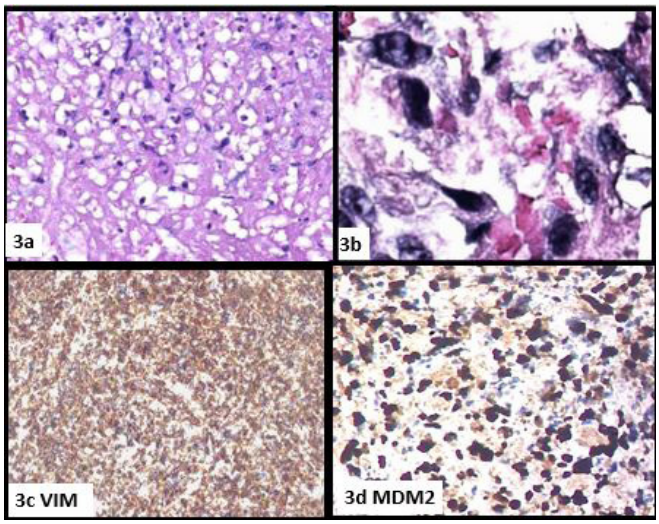

Figure 3. (a) *Thrombotic material in organization(HE 175X);* ***(b)*** *Neoplastic cells infiltrating thrombotic material (HE 220X);* ***(c)*** *Vimentin intense and diffuse expressivity(150X);* ***(d)*** *MDM 2 diffuse nuclear postiviy(220X).*

Immunohistochemistry

The results of the immunohistochemical investigation conducted with the above-mentioned antibodies are shown in Table 1 which demonstrates widespread and intense positivity for Vimentin (Figure 3c), MDM2 (Figure 3d), CD 31 (Figure 4b), sporadic and focal for FL1(figure 4c) and Smooth muscle Actin (Figure 4a).

Table 1.

Vim	MdM2	CD31	CD34	Cd99	Fl1	Cd10	Cd68	TTF1	α1AT	P40	Ck-AE1-Ae3	Chromo	Syn	Act s m	Calr	S100	Ki67
+++	+++	++	–	–	–+	–	–	–	–	–	–	–	–	+–	–	–	60%

Vim = Vimentin, α1AT = α1Antitripsyn, Chromo = Choromgranin A, Syn = Syinaptophysin, Act.SM = Actin smooth muscle, Calr = Calretinin

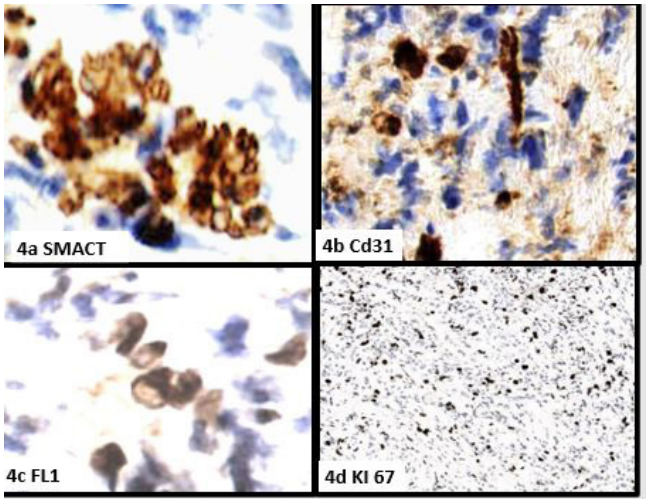

Figure 4. (a) *Smooth Muscle Actin- Isolated cells(200X);* ***(b)*** *CD31 Focal positivity (200X);* ***(c)*** *Fli1 – Isolated cells positivity(200X);* ***(d)*** *Ki67 - Extensive nuclear positivity(150X).*

Discussion

The clinical history of this Patient spans eight months, between the end of December 2022, when he presented with dyspnea and cough, interpreted as pneumonia, and August 2023.

A TC Scan dated 05/08/2023 documents the presence of an extensive endoluminal filling defect of a "thromboembolic" nature which affects the common trunk of Pulmonary artery (PA) and extensively affect also the right branches.

A CT scan performed eleven days later (05/19), in addition to demonstrating a further expansion of the "thrombo-embolic" process, detects the presence of solid tissue in the perihilar area incorporating the bronchovascular structures.

Just over ninety days later(08/21) the right pulmonary field appears almost completely opaque. Five days later the right lung field opacity is complete and the "thromboembolic" process extends to the left carotid artery, the middle and posterior cerebral arteries with associated cerebral haemorrhagic discharge.

After a few days the patient's death occurred

A transbronchial biopsy sample carried out on 07/25 highlighted a undifferentiated neoplasia .The immunophenotypic profile is consistent with an undifferentiated mesenchymal neoplasm such as Undifferentiate Pleomorphic Sarcoma (formerly known as Malignant Fibrous Histiocytoma). In particular, intense expressivity for vimentin and focal for Smact, Fli1 and CD31. In addition, there is significant expressivity for MDM2, high positivity for Ki67. No expressivity, conversely, for antibodies indicative of conventional lung tumors.

The clinical course, the morphological characteristics and, finally, the immunophenotypic expression are consistent for the diagnosis of that peculiar neoplasm which goes by the name of Intimal Sarcoma.

"Intimal sarcomas are malignant mesenchymal tumours arising in large blood vessels of the systemic and pulmonary circulation and also in the heart. The defining feature is predominantly intraluminal growth with obstruction of the lumen of the vessel of origin and seeding of emboli to peripheral organs. Essential and desirable diagnostic criteria : occurrence within the lumen of a large vessel of the pulmonary or systemic circulation or within the heart cavities; primary high-grade sarcoma, with or without heterologous elements.Desirable: MDM2 amplification (in selected cases)"(WHO 2020) [1].

A first report of this lesion appears in the literature in 1923[2].

Burke and Virman in 1993 tackled the problem of sarcomas of the large vessels in an organic way by reviewing 43 cases, 11 involving the aorta, 16 inferior vena cava and sixteen the pulmonary artery. The Authors divide the cases according to the type of growth of the neoplasm: mural or luminal(Intimal). Sarcomas of the Aorta(AS) 8/11 and those of the Pulmonary Artery(PAS) 14/16 are predominantly luminal in growth, while in the Vena Cava (VCS) with mural growth prevail 12/16.

Histologically the luminal tumors were predominantly of the undifferentiated type. The aortic sarcomas and those of the pulmonary artery were histologically substantially similar and called "Intimal". Those of the vena cava, predominantly mural, predominantly histologically differentiated in a leiomyosarcomatous sense [3].

Comparing the various reports, it emerges that these neoplasms, as of undifferentiated sarcomas, do not present well-defined immunophenotypic profiles. The only marker of a certain significance is MDM2, present in about 75% of cases.The biomolecular investigation demonstrated frequent amplifications and gains in the 12q13–14 region combined with overexpression of MDM2 which is systematically expressed by lipocytic neoplasms and some varieties of osteosarcomas [4,5].

From these data it emerges clearly that the term "Intimal" does not indicate a histogenetic derivation of the neoplasia from the structures of the arterial intima, but rather the localization of the neoplastic proliferation at the level of the intimal structures.

Regarding(the Pulmonary Artery Intimal Sarcoma(PAIS), a recent exhaustive review reports that in the just hundred cases found in the literature, there is a slight prevalence of females (1.3:1) and a mean age at diagnosis of 48 years (range13– 86 years).The prognosis of PAIS is extremely poor with estimated survivals of 1.5months without surgical resection and 10 months with resection [6].

Most patients are initially misdiagnosed a pulmonary vascular diseases such as chronic thromboembolic pulmonary hypertension or acute pulmonary emboli [7,8,9].

As in our case, the literature has reported the admixture of thrombotic material with the neoplastic tissue which favors the release of intra- and extrapulmonary emboli [10].

Conclusion

The case we observed fully complies with the diagnostic criteria indicated by WHO, both in the clinical presentation and evolution , in the histological aspects and in the immunological expressivity.

What makes the case in question peculiar is the massive and rapid opacification of the right lung field. Scrolling through the literature we note that these tumors tend to remain endoluminal. We have found only one case in the literature with similar characteristics[11]. The rapid and massive opacification of the right lung field may more likely be attributed to thromboembolic phenomena rather than dissemination of neoplastic tissue. Probably emboli a mixed thrombo-neoplastic composition, as is also evident in the observation of histologic findings (Figures 2d,3a,3b).

Approximately 90 days passed from the detection of the Pulmonary Artery obstruction to death, with the characteristic of a terminal progressive acceleration, occupied the right lung field, reaching the carotid and cerebral circulation.

Conflicts of Interest

The authors declare no conflict of interest and received no specific funding for this work.

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