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Case Report
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Pulmonary Artery Sarcoma Intimal Mimicking Embolism Lung

Raul Fava Alencar[1], Filadélfia Passos Rodrigues[1], José Aurillo Rocha[2], Fábio Rocha Fernandes Távora[3], Victor S.M.de A. Meireles[4] and Paulo Henrique da Silva Leao[4]*

[1]Department of Pulmonology, Messejana Hospital, Brazil

[2]Department of Oncology, Clinical Pharmacology - Federal University of Ceara, Brazil

[3]Department of Pathology, Messejana Hospital, Brazil

[4]Messejana Hospital, Brazil

Abstract

The intimal sarcoma of the pulmonary artery (SIAP) is a rare tumor of mesenchymal origin that primarily affects the large glass. The first report of the disease has been described in autopsy performed by Mandelstamm in 1923. On displays are often nonspecific signs and symptoms, the materials SIAP is commonly diagnosed as TEP, but other diagnoses are recognized as arteritis and pulmonary neoplasia pulmonar.

The disease has a poor prognosis even after the intervention surgery, still considered the treatment of choice of treatment, and a few months to years.

Case Presentation

Female patient 46 years old with no prior medical history, dyspnea initiated efforts for one year, loss of weight and discrete episodes of isolated fever at the beginning there, bringing chest X-rays that period without changes in the lung fields and cardiac ; being attributed to viral infections of the upper airway and sinus disease. Then sought medical attention for presenting atypical chest pain, epigastric pain, and the electrocardiogram showed T wave inversion in anterolateral wall; Cardiac catheterization was performed without visualization of coronary lesions. Chest X-ray of the time without changes.

After three months, the new hospital because of episodes of pre-syncope and dyspnea on moderate and small efforts. X-ray (Figure 1) showed rectification of the pulmonary artery trunk and spiky image contours in the left hilar region later. Transthoracic echocardiogram showed dilated right heart chambers, pulmonary artery systolic pressure of 115 mmHg and pulmonary dilated with hiperrefringente the level of the bifurcation mainly in the left branch, suggestive of thrombus image. Patient was conducted as Pulmonary Embolism (PE), and inciado full anticoagulation and subsequent workup cause to investigate. Ultrasonography of the lower limbs was negative for thrombus. Negative rheumatologic tests.

CT angiography of the chest (Figure 2) showed macrolobulada lesion with soft tissue coefficient fair compromise bronchial vessels in adjacency; ipsilateral pleural involvement with nodular formation; ground-glass opacities in the right middle lobe and image enhancement with the intraluminal contrast to the level of the bifurcation and the left pulmonary artery; suggesting thromboembolism and neoplastic lesion.

Bronchial stenosis was seen on the left by bronchoscopy. Transbronchial biopsy performed (Figure 3) in the left upper lobe with histopathological description of undifferentiated carcinoma with spindle cells, nuclear pleomorphism, atypical mitosis. Immunohistochemical panel showed positive for AE1-AE3 and negative for the other markers (actin, desmin, S-100, TTF-1, bcl-2, CD45, CD34, CK7, estrogen and progesterone receptor, CD99). Completing the diagnosis of intimal sarcoma of the pulmonary artery mimicking pulmonary metastasis and TEP.

Before the results of histopathology and immunohistochemistry, the hypothesis was lung cancer as a risk factor for pulmonary thromboembolism. Patient had significant worsening of dyspnea and underwent radiation therapy in an attempt to reduce the injury causing bronchial stenosis. Patient without conditions at the time of initiation of chemotherapy. Died due to respiratory failure and

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Correspondence:
José Aurillo Rocha, Department of Oncology, Federal University of Ceará, Nucleus of Oncology Studies, Brazil, E-mail: jaurillor@hotmail.com

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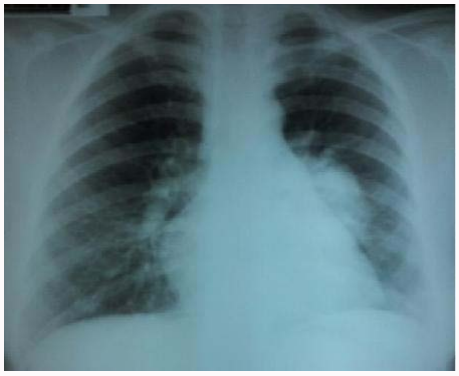

Figure 1: X-Ray showing rectification of the pulmonary trunk and left perihilar hypotransparent.

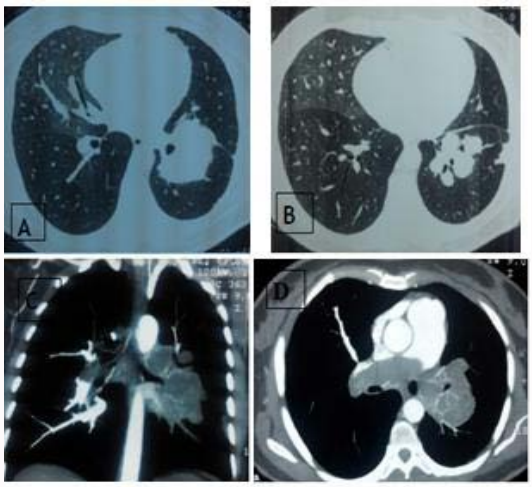

Figure 2: (A,B) Lesion macrolobulada bronchial fair and areas of ground-glass opacification in LMD. (C,D) Sagittal and axial with the intraluminal level of the bifurcation of the pulmonary image with contrast enhancement**.**

cardiovascular failure of the right ventricle.

Discussion

The SIAP is a rare malignant tumor of mesenchymal origin of the cardiovascular system with an estimated incidence of 0.001%. The first description of the case occurred in 1923 by Mandelstamm. Currently, over 200 cases have been reported [1]. The rarity of the condition can be explained by diagnostic difficulty, which often occurs during surgery, aiming endarterectomy or autopsia [2-4]. The prevalence by sex is still uncertain, slight preference being reported by females. The age is located in middle age with the average between 45 and 55 years. Metastases are common in the lungs, but can occur in the pancreas, kidney, lymph nodes, brain and skin in addition to retrograde invasion affecting pulmonary valve and right ventricle. The etiology is suggested from primitive cells in cardiac or arterial bulb bulb, which appears in the human fetal heart ventricle near the primitive, and subsequently gives rise to most of the right ventricle and the outflow tract of aorta and artery pulmonar. SIAP has twice as likely to occur in the dorsal region of the pulmonary artery when compared to aorta, but may still arise in veins cavas. Has growth from the intima of the artery and continues through the pulmonary arterioles forming polypoid mass. In 50% of patients, growth is transmural and is adjacent to the lungs, the bronchial wall, mediastinum and linfonodos.

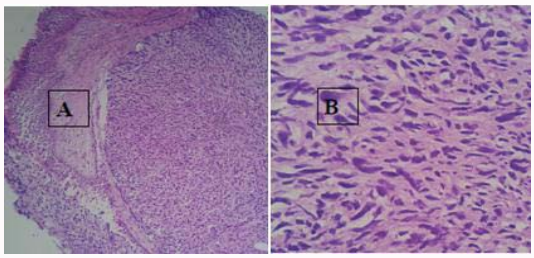

Figure 3: A) Increase 10X, showing respiratory epithelium and neoplastic lesion on the left to the right. B) Increased 40X, showing spindle cells with cellular atypia.

Clinical manifestations often mimic pulmonary embolism, being non-specific, occurring in the final stage of the disease. In the literature review and in isolated cases, the main signs and symptoms are: shortness of breath, chest pain, cough and hemoptise. However, one must pay attention to warning signs such as fever, weight loss and clubbing digital. Laboratory tests can be found elevated ESR, and leukocytosis, polycythemia, thrombocytopenia and evidence of intravascular coagulation disseminada. Cases described, report on changes in the coagulation tests, such as protein C resistance and heparin-induced thrombocytopenia, causing doubt it is by coincidence or by the evolution of advanced disease. Imaging findings depend on tumor size and location on the affected vessel wall, but still, it is a difficult diagnosis between sarcoma and pulmonary embolism by radiological imaging. When the dough is intraluminal and not distend the artery, the chest X-ray may appear to be normal, and a test that helps exclude other causes with similar symptoms. To distend the artery, but remaining intraluminal Unilateral hilar mass protruding into the lung parenchyma in the topography of the arterial branches, there may be areas of distal oligemia; however mass extending from transmural broncogênico may mimic a carcinoma. Computed tomography in four patients analyzed, some signs may suggest SIAP as filling defects with low density with an increase in arterial lumen diameter and extraluminal extension with mosaic attenuation, which may be specific for patients with advanced disease. Therefore, it is a warning sign for the presence of sarcoma, if distension glass. The 18-Fluorodeoxyglucose/computed Positron Emission Tomography (18FDG/PET) has the advantage of capturing the tumor contrast more strongly as compared with the thrombus, and may be a useful test when no surgical impossibility of biópsia. In the literature, studies on the role of 18FDG/PET in the initial stage of the evolution of SIAP are needed, since there are some reports that used the test showed no significant uptake, mimicking the presence of thrombus. MRI shows good ability to identify soft tissue, and after the use of contrast, there is a great variability in uptake and may correlate with different degrees of differentiation of sarcomas or other diseases; having the disadvantage of requiring a long period of inspiratory pause, which may be possible due to the symptoms of patients. The diagnosis of two cases reported in the literature was conducted with the use of transbronchial needle aspiration guided by endobronchial ultrasound (EBUS-TBNA), already used biopsy of mediastinal lymph nodes for lung cancer, but in expert evaluation, was considered a technique with potential complications due to the SIAP patients, mostly having associated pulmonary hypertension.

Macroscopically, the tumor has a gelatinous appearance and brancacento 3. Under microscope, it is noted to be an undifferentiated neoplasm; predominantly spindle cells with nuclear polymorphism, Mitotic activity variable and well vascularized

Table 1: Characteristics of case studies in the literature of SIAP.

Author/year	Sex	Age	Symptoms	Picture	IHQ	Handling post-SIAP
Perez, [21]	F	73	Fever Hemoptysis	Condensation perihilar	Vimentina Actina Desmina	Death
Favaloro, [13]	M	42	Dyspnea	PET : tumor mass AP		Death
Denine, [10]	F	34	Dyspnea Hemoptysis syncope	Failure branch		Death
Matto, [9]	F	32	Fever Dyspnea Weight Loss	Failure branch filling esq.		Free margin of the tumor
Furest, [5]	F	41	Dyspnea	Mass at the bifurcation AP		Ifosfamida Adriamicina
Alsoufi, [6]	M	76	Dyspnea Adinamia	Thrombus in the trunk AP		chemotherapy
Ni, [4]	M	73	Dyspnea Palpitação	Obstruction of the TP ECO	S100 Desmina	Ciclofosfamida Dacarbazina
Hirose, [25]	F	45	Dyspnea	AP mass by CT	Vimentina Actina	Carboplatina Ifosfamida
Dornas, [1]	M	45	Dyspnea Fever Weight Loss	Failure to fill pulmonary nodule	Vimentina	chemotherapy
Ciledag, [11]	M	30	Dyspnea Fever Weight Loss	Total occlusion AP PET captante	Vimentina	Free margin of the tumor
Lyle, [2]	M	65	Dyspnea Fever Weight Loss	subacute thrombosis		Doxorrubicina
Nozue, [12]	F	39	Dyspnea	Poor contrasted area in AP	Vimentina Actina	Ifosfamida Doxorrubicina
Bhagwat, [22]	M	30	pleuritic pain Fever	Failure branch filling esq		Free margin of the tumor
Vasuri, [20]	F	44	Dyspnea	AP thrombus in the ECO	Runx-1 CD-44	chemotherapy
Vaideeswar, [3]	F	55	Dyspnea Fever chest pain		Vimentina	Death
Lee, [17]	M	58	Dyspnea	Failure trunk filler AP	Actina Desmina	Death after resection

with few infiltrates lymphomononuclears. Out of the vascular bed, the tumor foci can present thrombi. The origin of the tumor is in pluripotent mesenchymal cells, electron microscopy showed that the appearance of myofibroblasts, but do consider the heterogeneous elements multipontencialities these cells. There is a poverty immunohistochemical marker, the presence of vimentin being more common. Endothelial markers such as CD31, CD34 are negative. Some reports express factor VIII, actin and desmina [21]. In working with research into new markers to identify the true cellular origin of SIAP, positivity for RUNX-1, WT1, and CD44, related hemangioblast described in the vascular wall were found; early stage of endothelial cells; and mesenchymal stem cells and mononuclear hematopoietic respectively.

The SIAP shows a poor prognosis with median survival 12 to 18 months after the start of sintomatologia. The treatment of choice is resection still surgery. In reviewing the literature, the median survival after complete resection was 36.5 months (± 20.2) compared to 11 months (± 3) for incomplete; while patients with combined treatment survival was 24.7 months (± 8.5) compared to 8 months (± 1.7) treatment with a modalidade [23]. Bacha et al. [24] in a study of 23 patients with various forms of pulmonary sarcomas showed that tumors smaller than 5 cm and complete resection correlated with greater curve survival. The role of chemotherapy and radiotherapy in the SIAP is still uncertain, with views of the use to be tried in patients with unresectable or recurrent tumor after surgery. Doxorubicin is one of the effective agents in sarcomas of soft tissue in adults with reports that concomitant infusion radiotherapy is effective. The amrubicin, doxorubicin derivative, proved more potent in experimental animals and without presenting cardiotoxicity. Ifosfamide can be regarded as a second-line drug in the treatment of patients with failure with the use of doxorubicin response in around 20%.

Conclusion

The SIAP is a rare tumor with potential to be misdiagnosed due to nonspecific symptoms and similarity to an event of pulmonary thromboembolism, there is often treated with anticoagulation without improvement. The presence of fever, elevated ESR and haematological changes should alert to the possible diagnosis, as well as CT angiography with low image density contrast, dilatation of the artery and tiled areas, taking 18FDG/PET as a method of choice for differentiating thrombotic event. Studies should further evaluate the panel of immunohistochemical markers with the need to increase the knowledge of the etiology and facilitate diagnosis; as well as evaluating other treatment besides surgery.

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