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Primary hemangiopericytoma of the lung as an incidental finding: report of two cases

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Abstract: Intrapulmonary hemangiopericitoma potentially is a rare malignancy, which originates from the pericytes that surround the basal membrane of the capillaries and venules in lung parenchyma. Objective: The aim is to present two cases as an incidental finding in its presentation and also know the behavior and management of this neoplasm. Clinical cases: A male patient of 40 years with clinical picture characterized by persistent dry cough, fatigue with shortness of breath with rapid progression to small effort dyspnea and weight loss and a female patient aged 37 clinical picture characterized by chest pain oppressive kind in lateral region posterior right chest, radiating to the shoulder in the chest film, multiple nodular parenchymal bilateral central and sub-pleural. The second patient so poster anterior chest radiograph where a pattern of tumor mass located in the center of the right lung, which was confirmed by computed tomography of the chest, is observed is performed. In the patient he proceeded to the surgical recession of the injury to biopsy, while in the female patient's behavior was more radical right pneumonectomy was practical. They are evolving satisfactorily. Report biopsy and immunohistochemistry for primary lung: Hemangiopericitoma / SFT. Both patients were referred to medical oncology service for adjuvant therapy. Conclusion: Is a rare tumor, its pathogenesis is still unknown and although a tumor in appearance and benign behavior, should be treated as malignant potential, must be radical in treatment and careful monitoring, continuous and strict patient.

Key words: hemangiopericitoma; primary; lung; tumor; malignant

1 Introduction

The lung can be the site of both benign and malignant tumors; the latter, in turn, can be metastatic or primary [1]. Primary lung tumors are very rare, and the ratio of primary lung tumors to lung metastases and other non-neoplastic lesions is 1:5:60 [1]. Most primary malignant lung neoplasms are of epithelial origin, with only 2% to 5% being mesenchymal, and other diverse neoplasms, such as Sarcomas are rare, especially when they are non-metastatic and of pulmonary origin [2]. For this reason, the vast majority are malignant in nature [1,2,3] and, due to their rarity, they are not usually included in the differential diagnosis of pulmonary masses. Primary tumors of mesenchymal origin in the lung are a rare finding in routine medical practice. Hemangiopericytic tumors are rare and are classified as a sarcoma of the perivascular tissue. Hemangiopericytoma (HPC) is a rare vascular neoplasm, first described by Stolut A and Murray M in 1942 [2,3]. It is a soft tissue tumor considered to be of pericyte origin, and the current trend is to classify it as a solitary fibrous tumor of fibroblastic origin, although the World Health Organization (WHO) [4] continues to classify it as a hemangiopericytoma or

solitary fibrous tumor. The main argument for abandoning this term is that both morphological and immunohistochemical diagnostic criteria lack specificity; for this reason, the trend is to call it: solitary fibrous tumor—cellular variant.

Hemangiopericytoma is an extremely rare mesenchymal neoplasm, accounting for 1% of vascular tumors. It most commonly occurs in the skin, subcutaneous soft tissues, muscles of the extremities, and retroperitoneum, but rarely in the lungs, trachea, or mediastinum, regarding which there are very few references in the literature [4]. Its pathogenetic basis has been evaluated using cytogenetic techniques, revealing similarities to those observed in the solitary fibrous tumor—a soft tissue tumor of uncertain origin and is associated with multiple chromosomal aberrations and the presence of breakpoints on chromosomes 12q-15, 8, and 12q-15, 8, and 9. Thus, many tumors previously documented as hemangiopericytomas may currently correspond to solitary fibrous tumors [3,4]. It is a tumor of primitive mesenchymal cells with ubiquitous localization, surrounding the endothelium of arterioles, capillaries, and venules, with the ability to contract around them [1,2,3,4]. It can appear anywhere in the body; its peak incidence spans a very wide age range, typically occurring between the fourth and fifth decades of life, more commonly in young adults (with a mean age of 45 years), and it does not have a higher prevalence in either sex [5].

Primary pulmonary hemangiopericytoma originates from the pericytes surrounding the basement membrane of capillaries and small venules within the lung parenchyma; it is characterized as a slow-growing tumor with no predilection for any particular lobe, it is usually located in the center of the lung and extends peripherally, forming nodules or masses that are visible on imaging and exhibit a variable and unpredictable biological potential for malignancy. This type of neoplasm does not have uniform clinical or radiological features; and generally presents as a solitary, asymptomatic intrathoracic mass or with clinical signs and symptoms that are often insidious. Since these are slow-growing, painless masses that cause only mild discomfort, the pressure exerted by the tumor mass on adjacent organs can produce symptoms and signs characterized by cough, pain, hemoptysis, night sweats, dyspnea, and superior vena cava syndrome [6].

For diagnosis, imaging findings are limited to signs of compression and displacement. In long-standing tumors, calcifications appear. Ultrasound (US), computed tomography (CT), and arteriography are available tools when necessary. The histopathological behavior of these tumors is well-defined; internally, they exhibit a pericytic nature is established: abundant cells with oval nuclei, pale cytoplasm, oval mitochondria, few ribosomes and microfilaments, as well as pinocytotic vesicles and rare, poorly developed desmosomes. These cells surround thin-walled vascular channels lined with a layer of endothelium. Microscopically, malignant hemangiopericytoma is characterized by rapid growth, four or more mitotic figures per 10 fields, a tumor size of more than 3 cm to 7 cm, and foci of necrosis, hemorrhage, and cellular anaplasia, all of which are indicative of malignancy. This malignant behavior is observed in up to 23% of cases, primarily in those with extrathoracic locations.

In immunohistochemistry, the markers frequently expressed are: CD34 (positive in 44%–95%), CD99 (positive in 64%–91%), and bcl-2 (positive in 50%). On the other hand, smooth muscle actin, desmin, cytokeratins, S100 protein, and CD31 are usually negative [5,6,7].

Differential diagnoses include: lipomatous hemangiopericytoma, and large-cell angiofibroma, among others [8,9,10]. The treatment of choice is wide surgical excision with negative surgical margins, as this is the primary prognostic factor for recurrence; postoperative local recurrence rates reach up to 50%. Hemangiopericytoma is a radiosensitive tumor [11,12], so strict follow-up is required. Although there is no consensus or clarity regarding its efficacy, the use of postoperative adjuvant radiotherapy or chemotherapy has been recommended to reduce recurrences; many consider it nearly ineffective [13,14,15].

We present a primary pulmonary hemangiopericytoma in two patients. We present the radiological and CT findings,

the pathological anatomy, the medical management, and review the literature.

2 Clinical case 1

A 40-year-old male patient, a non-smoker, born and raised in a rural area of the municipality of Baralt (Venezuela), whose only relevant epidemiological history is that he works at an asphalt plant. Until the onset of the illness, he was known to be healthy and had no significant family history of disease. He presented for consultation with a two-month history of symptoms characterized by a persistent nonproductive cough, fatigue with shortness of breath, and weight loss of approximately 10 kg; these symptoms were exacerbated by rapidly progressive dyspnea with minimal exertion.

On physical examination, the patient presented in fair general condition, with moderate pallor of the skin and mucous membranes, tachycardia, and tachypnea. Auscultation revealed decreased vesicular breath sounds in the left lung field without additional sounds; the remainder of the clinical examination was within normal limits. He was therefore diagnosed with acute bronchitis-like pneumonia and received treatment without clinical improvement. Given the lack of response to treatment, laboratory tests and imaging studies were performed.

Laboratory tests were performed, including a complete blood count, coagulation panel, serum biochemistry, urinalysis, protein profile, erythrocyte sedimentation rate, lactate dehydrogenase, and alkaline phosphatase. Tumor markers (alpha-fetoprotein, human chorionic gonadotropin, CA 125, and CA 19-9) were also measured and were normal.

In light of the findings and the persistence of clinical symptoms, a chest X-ray was performed, revealing on the postero-anterior (PA) view multiple bilateral pulmonary nodules at the central and subpleural levels, with an air bronchogram and areas of ground-glass opacity (Figure 1). To complete the workup, a chest computed tomography (CT) scan was performed using a 16-detector BRIGHT SPEED® scanner without intravenous contrast administration. In the pulmonary window, multiple nodular lesions were evident bilaterally at the central and subpleural levels, ranging in size from 0.6 cm to 3.1 cm, with attenuation values between 23 and 43 HU. An increase in pulmonary attenuation with air bronchogram was noted, as well as bilateral peripheral ground-glass opacities and thickening of the interlobular septa with predominantly in the basal regions (Figure 2). No enlarged hilar or mediastinal lymph nodes were observed that would be considered pathological from a CT perspective; likewise, no pleural effusion was observed.

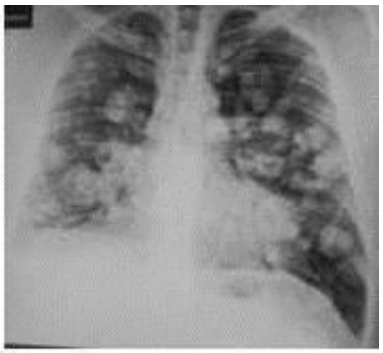

Figure 1. Chest X-ray: multiple bilateral pulmonary nodules at the central and subpleural levels, with air-bronchogram, and areas of ground-glass opacity

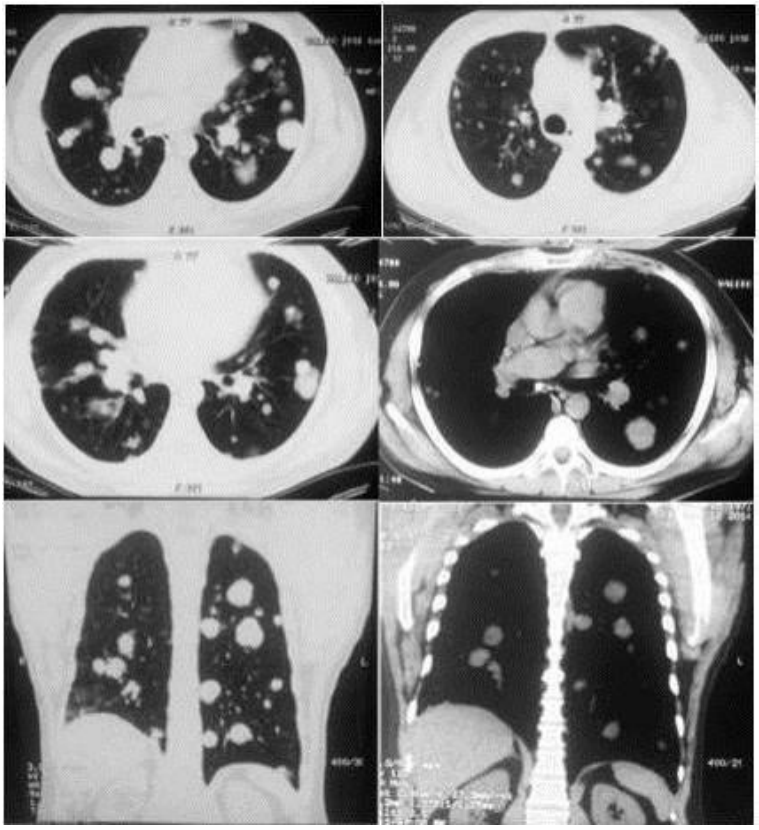

Figure 2. BRIGHT SPEED 16-detector chest CT: multiple nodular images bilaterally in the central and subpleural regions, ranging in size from 0.6 cm to 3.1 cm, with Hounsfield unit (HU) values between 23 and 43. Increased pulmonary attenuation with air bronchogram is noted, as well as bilateral peripheral ground-glass opacities, and thickening of the interlobular septa, predominantly in the basal regions

A left posterolateral thoracotomy is performed through the 5th intercostal space, with surgical resection of a segment of the left lung measuring approximately 5 cm x 3 cm x 2 cm, containing two nodular lesions with a smooth external surface, featuring wine-red areas alternating with yellowish-white areas with fine vascular neovascularization, which is in continuity with the pleura.

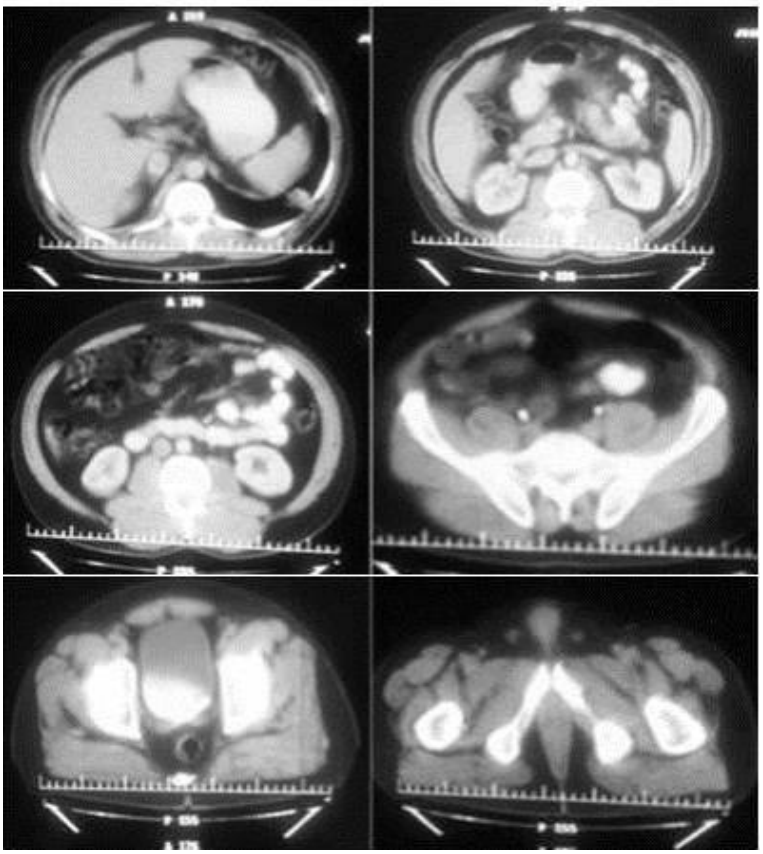

Figure 3. Abdominal-pelvic CT scan: no evidence of neoplastic or inflammatory lesions of the abdominal-pelvic viscera

The specimen is sent to the pathology department, which reports the macroscopic examination: a 6 cm x 4 cm x 3 cm piece of lung tissue, wine-red in color with a with fine vascular tracts, containing two nodular formations measuring 0.9 cm and 0.7 cm, respectively, yellowish-white in color and firm in consistency. Meanwhile, histological examination with hematoxylin-eosin and Masson's trichrome staining revealed: three tumor nodules, consisting of a neoplasm of mesenchymal origin, with the presence of abundant blood vessels of various sizes and branched shapes in a "deer antler" pattern, which are surrounded by rounded cells alternating with sparse spindle-shaped cells with loss of the nucleus-cytoplasm ratio, and with an absence of mitosis and tumor necrosis. Furthermore, in extensive areas of the neoplasm, poorly defined fibrous fascicles are observed, which are Masson's trichrome-positive.

An immunohistochemical study was performed using the avidin-streptavidin technique, and VIM, CD99, HHF-35, Fli-1, and CD34 were investigated using the antigen retrieval method. The immunohistochemical study demonstrated positive reactivity in the tumor cells for VIM, HHF-35, and Fli-1 (Figure 4). These findings, together with the pattern observed in the hematoxylin-eosin and Masson's trichrome staining techniques, confirm the pericytic nature of the neoplasm as a primary pulmonary hemangiopericytoma.

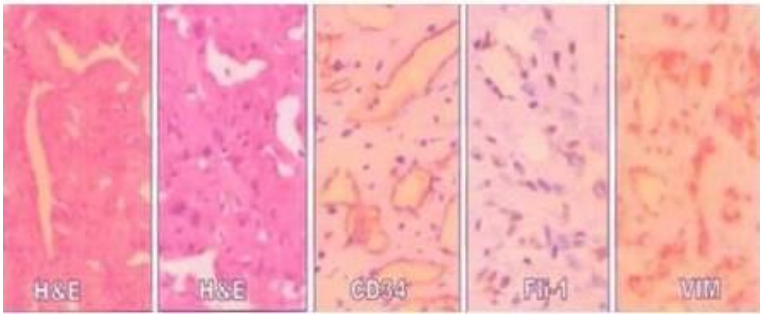

Figure 4. Immunohistochemistry using the avidin-streptavidin technique showed immunoreactivity in the tumor cells with VIM, HHF-35, and Fli-1, consistent with a pulmonary hemangiopericytoma

The postoperative period was uneventful, and the patient was discharged 72 hours after the chest tube was removed. As reported in the reviewed literature, the treatment of choice for this type of tumor is radical surgery followed by follow-up. In this case, radical excision of the lesions was not possible due to extensive bilateral involvement of the lung parenchyma; therefore, the patient was referred to the medical oncology department, which decided to prescribe a postoperative adjuvant polychemotherapy regimen with the following cytostatic agents: cyclophosphamide, adriamycin, and vincristine, administered every 4 weeks for six months. He also received thirty-three sessions of radiation therapy.

3 Clinical case 2

A 37-year-old female patient, an occasional smoker, with a history of surgically corrected valvular heart disease in childhood, currently considered healthy. She presented to the clinic with symptoms that had been present for one month, characterized by oppressive chest pain in the posterolateral region of the right hemithorax, radiating to the shoulder, so she visited the neurosurgery clinic, where she was evaluated and diagnosed with functional back pain due to work-related stress. An intercostal analgesic block was performed without improvement, so the case was reevaluated, and a selective radicular block (SRB) under radiological guidance was planned, to be performed in the operating room under conscious sedation. A posterior-anterior chest X-ray was performed, revealing a tumor mass pattern located in the center of the right lung and extending peripherally, homogeneous with well-defined borders, with a subpleural lobar distribution (right lower lobe), associated with volume loss (Figure 5). For this reason, she was referred to the thoracic surgery clinic, where she was evaluated. The patient appears in good general condition and is hemodynamically stable. On physical examination, the chest is symmetrical with good respiratory expansion and is eupneic; on auscultation, decreased vesicular breath sounds are noted, with basal rhonchi and crackles in the right lung field; the remainder of the clinical examination is unremarkable.

Laboratory results, including complete blood count, coagulation panel, serum biochemistry, urinalysis, protein profile, erythrocyte sedimentation rate, lactate dehydrogenase, and alkaline phosphatase, as well as tumor markers (alpha-fetoprotein, human chorionic gonadotropin, CA 125, and CEA 19-9), were within normal limits.

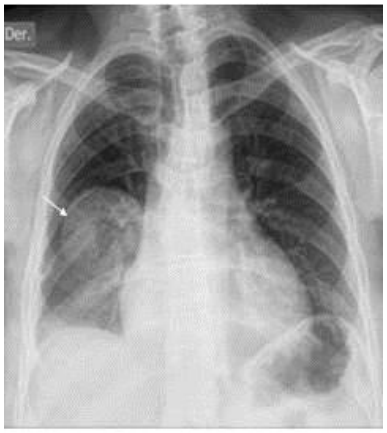

Figure 5. Chest X-ray. Image showing a tumor mass in the right lung field, homogeneous with well-defined borders, with a lobar distribution (right lower lobe), subpleural, and associated with volume loss

In light of the radiological and clinical findings, a chest CT scan was performed to complete the patient's evaluation, using a 16-detector BRIGHT SPEED® CT scanner without intravenous contrast administration. The image shows a mass with well-defined margins and a rounded morphology, measuring 9 cm x 7 cm, with attenuation values between 23–43 HU, homogeneous isodense, with smooth contours in contact with the chest wall and subpleural. No lymphadenopathy was observed in the mediastinal, supracarinal, or perihilar regions (Figure 6).

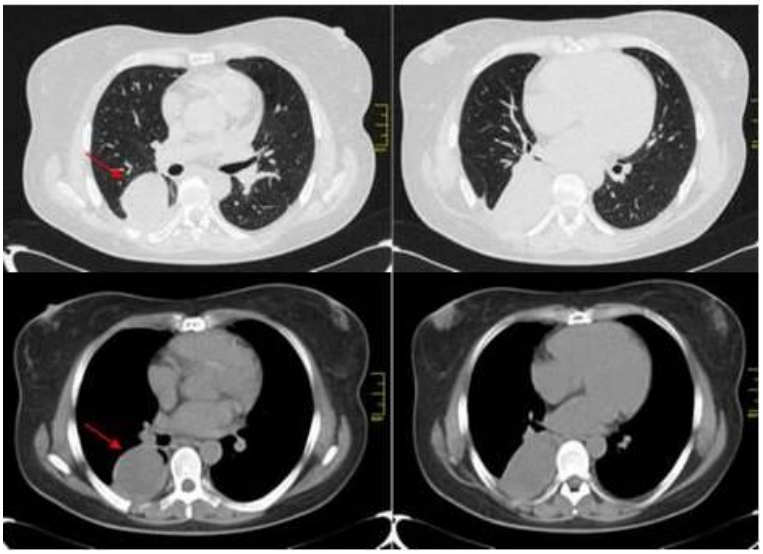

Figure 6. Chest CT: image of a mass with well-defined margins and a rounded morphology, measuring 9 cm x 7 cm, with attenuation values between 23–43 HU, isodense, and smooth contours in contact with the chest wall and subpleural space. Absence of lymphadenopathy in the mediastinal, supracarinal, and perihilar regions

In this case, a radical surgical approach was chosen, and a right pneumonectomy was performed through the 6th intercostal space, with surgical resection of the right lung. The lower lobe, measuring 11 cm x 9.5 cm x 2.5 cm, revealed a nodular lesion with a smooth external surface, featuring wine-red areas alternating with yellowish-white areas with fine vascular neovascularization, which was in continuity with the visceral pleura (Figure 7).

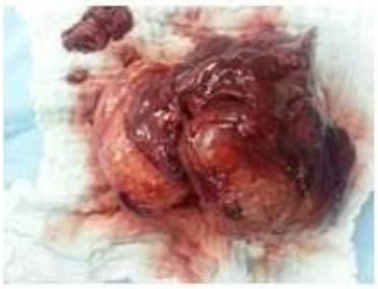

Figure 7. Lung tissue, wine-red in color with a smooth surface and fine vascular tracts, with a tumor-like formation measuring 8 cm x 6.5 cm x 3.5 cm, yellowish-white in color and firm in consistency

The specimen is sent to the pathology department, which reports the following macroscopic findings: lung tissue, wine-red in color with a smooth surface and fine vascular patterns, with a tumor-like formation measuring 8 cm x 6.5 cm x 3.5 cm, yellowish-white in color and firm in consistency. Meanwhile, histological examination with hematoxylin-eosin and Masson's trichrome staining revealed: tumor tissue consisting of a neoplasm of mesenchymal origin, with the presence of abundant blood vessels of various sizes and branched shapes in a "deer antler" pattern, which are surrounded by rounded cells alternating with sparse spindle-shaped cells showing loss of the nucleus-cytoplasm ratio, with no mitosis or tumor necrosis. Likewise, in extensive areas of the neoplasm, poorly defined fibrous fascicles are observed, which are Masson's trichrome positive.

An immunohistochemical study was performed using the avidin-streptavidin technique, and VIM, CD99, HHF-35, Fli-1, and CD34 were investigated using the antigen retrieval method. The immunohistochemical study demonstrated positive reactivity in the tumor cells for VIM, HHF-35, and Fli-1 (Figure 8). These findings, together with the pattern observed in the hematoxylin-eosin and Masson's trichrome staining techniques, confirm the pericytic nature of the neoplasm as a primary pulmonary hemangiopericytoma.

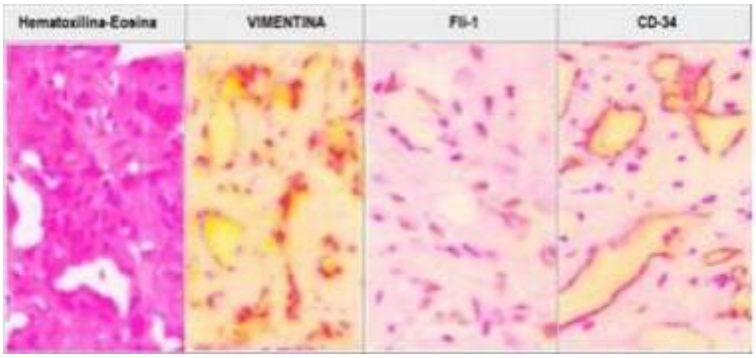

Figure 8. Immunohistochemistry using the avidin-streptavidin technique showed immunoreactivity in the tumor cells with VIM, HHF-35, and Fli-1, consistent with a pulmonary hemangiopericytoma

The postoperative period was uneventful, and the patient was discharged 72 hours after the chest tube was removed. As reported in the reviewed literature, the treatment of choice for this type of tumor is radical surgery; in this case, a right pneumonectomy was performed as treatment.

4 Discussion

Intrapulmonary hemangiopericytoma is a rare, potentially malignant neoplasm that originates from the pericytes surrounding the basement membrane of small vessels (capillaries and venules) within the lung parenchyma. It has a mesenchymal origin, regarding which there are few references in the literature, with fewer than 20 published cases [9]. Its

histogenesis is unclear, with two theories regarding its origin: the first hypothesis proposes an origin from the subpleural mesenchyme found in direct continuity with the connective tissue of the interlobular septa, while the second theory suggests that this tumor derives from submesothelial fibroblasts present in normal lung parenchyma [10,11,12]. Most cases described in the literature have been diagnosed in middle-aged adults, between 40 and 70 years old. The size of the lesions has ranged from 1 cm to 15 cm, with an average of 5 cm [11]. It is a slow-growing tumor that usually does not cause symptoms, so it is generally discovered incidentally [11]. Occasionally, it may cause symptoms secondary to compression of adjacent structures, such as cough, dyspnea, and chest pain [14,15]. In our cases, the presentation was characterized by cough, fatigue, and shortness of breath. Regarding the radiological findings described for this type of tumor, the anteroposterior chest X-ray shows nodular lesions or masses with well-defined margins and a rounded or ovoid morphology [12]. On chest CT, a well-defined lesion with soft-tissue density is observed, appearing homogeneous and without calcifications on the non-contrast study, and showing intense and heterogeneous enhancement following intravenous contrast administration [12]. These findings correlate with our cases, which showed homogeneous lesions on the non-contrast study.

For this reason, during preoperative diagnosis, CT imaging studies and a lung biopsy or radical resection of the affected lung are essential; after excising and analyzing the lesion, which is histologically characterized as a tumor that macroscopically presents as a whitish or grayish mass of firm consistency and well-defined borders [3]. Microscopically, it consists of an encapsulated lesion with a very dense cellular infiltrate, composed of pericytes with hypercellular areas (consisting of multiple spindle-shaped cells with low mitotic activity and few or no atypical features), cells with poorly defined borders, small, ovoid nuclei with loss of the nucleus-cytoplasm ratio, arranged in short, narrow fascicles and surrounded by a large number of capillaries with striking perivascular hyalinization, these vessels exhibit irregular branching, surrounded by vascular channels lined with thin-walled endothelium, giving the vessels a "deer antler" appearance, which is typically observed in hemangiopericytoma [13]. This histological pattern was observed in our patients. Immunohistochemical evaluation is essential for the differential diagnosis with other types of tumors and is characterized by the expression of CD34, bcl-2, and CD99 and negativity for cytokeratins, actin, desmin, and S-100 protein [13]. In our case, the immunohistochemical study demonstrated positive reactivity in the tumor cells with VIM, HHF-35, and Fli-1.

The treatment of choice is radical surgical resection of the lesion with wide resection margins, although there is some controversy regarding the indication for surgery in patients whose diagnosis stems from an incidental finding on a chest X-ray or in asymptomatic patients. Likewise, it is recommended to perform arteriography with prior embolization because it helps reduce intraoperative bleeding and tumor size [14]. The use of radiation therapy and chemotherapy is controversial. Both are used in palliative treatment, for unresectable lesions, or in the presence of metastases, as was the case in our patient where complete excision of the lesions was impossible.

The biological behavior of this tumor is essentially benign, with an excellent prognosis if complete surgical resection with clear margins is achieved. Although there are very few reported cases of recurrence or metastasis, the malignant potential of this tumor is considered uncertain, due both to the lack of long- term follow-up in most reported cases and to the fact that they present a local or distant recurrence rate of up to 10%–15% [15]. As for five-year survival in patients with hemangiopericytoma originating in any organ, it has been reported at 74.2%, and at 10 years, 64.4%, while survival in patients with pulmonary-origin tumors is 30%–35%. However, there are reports indicating that approximately 50% of hemangiopericytomas recur within 5 years [15,16]. It has been demonstrated that recurrent disease generally occurs within 2 years after initial treatment. For this reason, the recommended approach is to perform adequate long-term follow-up to

detect possible recurrences. In summary, intrapulmonary hemangiopericytoma is a rare mesenchymal neoplasm that should be included in the differential diagnosis of a slow-growing solitary pulmonary nodule, and an adequate correlation between radiological and histological findings is essential for its final diagnosis.

Conflicts of Interest

The author declares no conflicts of interest regarding the publication of this paper.

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