IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 23, Issue 10 Ser. 3 (October. 2024), PP 43-46
www.iosrjournals.org

A Rare And Mysterious Case Of Lung Cancer

Rajae Azzeddine, Laila Herrak , Asmae Jniene, Ibrahim Elghissassi, Hanane Inrhaoun, Hasnae Guerrouj, Ayat Mouaden, Mustapha Elftouh, Leila Achachi

Department Of Pulmonology, Ibnsina University Hospital Center , Faculty Of Medicine And Pharmacy, University Mohammed V , Rabat, Morocco.
Department Of Medical Oncology, National Institute Of Oncology, Ibn Sina University Hospital Center, Faculty Of Medicine And Pharmacy, University Mohammed V, Rabat, Morocco.
Department Of Nuclear Medicine, Ibn Sina University Hospital Center, Faculty Of Medicine And Pharmacy, University Mohammed V, Rabat, Morocco

Date of Submission: 01-10-2024 Date of Acceptance: 10-10-2024

I. Introduction

Primary pulmonary sarcomas (PPS) are rare types of non-epithelial malignant tumors of the lungs , representing only 0.013–1.1% of all malignant lung tumors. [1,2] Its diagnosis and treatment are challenging, with a poor prognosis [3]. Undifferentiated round cell sarcomas are an extremely rare form of this cancer and have been rarely described in the literature .We report a rare case of undifferentiated round cell primitive pulmonary sarcoma in a young 30-year-old patient.

II. Case Report

We report the case of a 30-year-old young woman who was neither treated for tuberculosis and has not had recent tuberculosis contagion, and without significant pathological history or chronic smoking.

The patient has been reporting for 3 months a productive cough bringing mucous sputum, sometimes hemoptoic, associated with dyspnea stage II mMRC and atypical right chest pain, evolving in a context of apyrexia and deterioration of general condition the initial Clinical examination finds a conscious patient, eupneic at rest, saO2 = 97% at ambient air, with a pleuropulmonary examination without particularities.

The posteroanterior chest roentgenogram showed homogeneous right hilar opacity with elevation of the right diaphragmatic dome (Figure 1).

The thoracic computed tomography (CT) showed a pulmonary process of the right upper lobe well limited measuring 68x72x68 (Figure 2)

Given this radioclinical presentation, a pulmonary neoplasm was initially suspected, prompting the performance of bronchoscopic examination . this examination revealed infiltration with thickening of the spur of the upper lobe, with biopsy results suggesting nonspecific chronic remodeling .

We followed up with a second bronchoscopic examination and two transparietal biopsy of the process, guided by CT, which also yielded inconclusive results on histopathological study and negative for BK culture.

The progression was marked by clinical worsening with the onset of a superior vena cava syndrome and radiological deterioration ranging from excavation of the pulmonary process(figure 3) to an increase in its volume, occupying the entire right lung , associated with excavated pulmonary nodules in the left upper lobe, multiple cervical-mediastinal lymphadenopathies, and thrombosis of the superior vena cava and innominate trunks (Figure 4)

An abdominal-pelvic CT scan, a brain CT scan, a bone scintigraphy, and a gynecological examination with breast ultrasound and mammography were performed to search for other locations, all of which returned normal results.

A lymph node biopsy of the left supraclavicular adenopathy was performed, revealing findings consistent with reactive adenitis.

We conducted a third bronchoscopic examination and two additional transparietal biopsies, which also suggested nonspecific chronic remodeling.

Faced with this mysterious clinical and radiological presentation, the case was discussed in a multidisciplinary meeting, and a decision was made to complement with 18F-fluorodeoxyglucose positron emission tomography-computed tomography (18F-FDG PET-CT).

The patient's 18F-FDG PET-CT showed a hypermetabolic mass with necrotic center involving the entire right lung (SUV: 12), two hypermetabolic pulmonary nodules on the contralateral side in the left upper lobe (SUV: 5.5) and left lower lobe (SUV: 9.1), along with pathologically hypermetabolic cervical-mediastinal lymph node foci.(Figure5)

Another CT-guided biopsy targeting the hypermetabolic mass while avoiding the necrotic center was performed as the ninth biopsy in total for the patient. After a year of investigation, the histopathological study finally revealed the presence of tumor cells, with immunohistochemical analysis showing diffuse expression of PS100 antibody, intense and diffuse membranous staining with CD99 antibody, and a high Ki67 proliferation index of more than 70%. These cells did not express melanocytic differentiation markers (Melanoma, MelanA, SOX10), neuroendocrine markers (chromogranin A, Synaptophysin), epithelial markers (Cytokeratin, CK7, EpCAM, P40, EMA), and also did not express antibodies against NKX2-2, FLI1, ERG, WT1, CD34, Myogenin, CD1a, CD68, LCA, TdT, CD5, CD117, Beta-catenin, TTF1, or ACE.

This histopathological study concluded the diagnosis of a primary undifferentiated round cell lung sarcoma .

After the confirmation of the diagnosis, the patient was referred to the oncology center for chemotherapy.

III. Discussion

Primary pulmonary sarcomas (PPS) are rare types of non-epithelial malignant tumors of the lungs (0.013–1.1% of all malignant lung tumors). [1,2]. The PPS can originate from mesenchymal elements of the bronchial wall, vessels or pulmonary stroma.

The most common sarcomas include leiomyosarcoma, malignant fibrous histiocytoma, and synovial sarcoma [4], Undifferentiated round cell sarcomas (URCS) are an extremely rare form of cancer, comprising a heterogeneous group of bone and soft tissue tumors occurring primarily in young adults , and representing the second most frequent category of round cell sarcomas (11%), following Ewing Sarcomas (80%) ,but lung involvement is usually due to metastasis. URCS originating primarily in the lungs is exceptionally rare, and it has rarely been reported since the first case was published in 1991 [5].

Symptoms and radiologic appearances are similar to those of more common lung carcinomas. The radiological features of the sarcomas are variable, not lesion-specific, and not sufficient to suggest specific diagnosis . Presentation of the disease depends more on tumor localization than histopathological features [6].

The diagnosis of pulmonary sarcoma poses a real problem, it is hard to identify on either small biopsy samples from bronchoscopy, core or fine needle aspiration or even intraoperative investigation.

After epithelial malignancy is ruled out, the most important differential diagnosis of primary pulmonary sarcoma is metastatic spread from an extrapulmonary sarcoma. Other differential diagnoses include pleomorphic lung carcinoma, pulmonary sarcomatoid carcinoma, and malignant melanoma. Therefore, a detailed medical history and appropriate diagnostic examination is necessary to specify that the tumor has primary pulmonary origin.

Since the introduction of immunohistochemical and molecular diagnosis, a significant improvement in description and classification of sarcomas has been achieved. Nonetheless, sarcomas still remain a diagnostic and clinical problem. [7]

This can be explained by the fact that patients diagnosed with pulmonary sarcoma are quite rare, and therefore it is difficult to define a reliable management protocol for these patients

This perfectly aligns with our observation, which clearly illustrates the diagnostic challenge in our patient, who benefited from multiple radiological investigations and underwent a total of nine histological samples before the diagnosis was established on the basis of the ninth transthoracic lung biopsy.

The URCS, is a highly aggressive malignant tumor , Its diagnosis and treatment are challenging, with a poor prognosis . The standard strategy of treatment in this type of tumor is similar to lung cancer. Lobectomy and pneumonectomy are still a gold standard in surgical treatment of PPS [8], For unresectable and metastatic forms, chemotherapy has recommended for decades, although with limited efficacy [1]. Some clinical trials have indicated that targeted therapy (anlotinib) [9] or immunotherapy [10] are potential treatment options in chemotherapy-resistant patients However, a standard treatment for URCS is still unavailable. A multi-disciplinary discussion including surgeons, medical oncologist, pulmonologist, radiologist, pathologist and radiation oncologists is currently recommended.

IV. Conclusion

Primary pulmonary sarcomas are a rare type of lung cancer, characterized by difficult diagnosis and poor prognosis.

we report an exceptionally rare case of superior vena cava syndrome due to an undifferentiated round cell primitive pulmonary sarcoma in a young woman, which took one year to diagnose after multiple investigations.

Through our work, we emphasize the diagnostic challenges of this histological type, which delay diagnosis and consequently delay therapeutic management, in order to focus on this entity and sensitize practicing physicians to consider primary pulmonary sarcoma when encountering a lung mass in young patients.

Conflicts of Interest

There are no conflicts of interest between the authors and between the authors and the patient.

References

[1] Nascimento Ag, Unni Kk, Bernatz Pe. Sarcomas Of The Lung. Mayo Clin Proc 1982; 57: 355-359.

[2] Porte Hi, Metois Dg, Leroy X, Conti M, Gosselin B, Wurtz A. Surgical Treatment Of Primary Sarcoma Of The Lung. Eur J Cardiothor Surg 2000; 18: 136-142.

[3] Le Loarer F, Baud J, Azmani R, Et Al. Advances In The Classification Of Round Cell Sarcomas. Histopathology 2022 ; 80 : 33-53.

[4] Keel Sb, Bacha E, Mark Ej, Et Al. Primary Pulmonary Sarcoma: A Clinicopathologic Study Of 26 Cases. Mod Pathol. 1999;12:1124–31.

[5] Sánchez Heras Ab, López Rodríguez A, Pastor Borgoñón M, Et Al. Undifferentiated Small Cell Tumor Of The Thoracopulmonary Region. An Med Interna 1991;8:448-50.

[6] Etienne-Mastroianni B, Falchero L, Chalabreysse L, Loire R, Ranchère D, Souquet Pj, Et Al. Primary Sarcomas Of The Lung: A Clinicopathologic Study Of 2 Cases. Lung Cancer. 2002;38:283–9.

[7] Coşgun T, Tezel Y, Akyıl M, Kolbaş İ, Şen A, Tezel Ç. Primary Pulmonary Malignant Fibrous Histiocytoma. Turk Thorac J. 2017 Apr;18(2):54–6.

[8] Gronchi A, Miah Ab, Dei Tos Ap, Et Al. Soft Tissue And Visceral Sarcomas: Esmo-Euracan-Genturis Clinical Practice Guidelines For Diagnosis, Treatment And Follow-Up. Ann Oncol 2021;32:1348-65.

[9] Kyriazoglou A, Gkaralea Le, Kotsantis I, Et Al. Tyrosine Kinase Inhibitors In Sarcoma Treatment. Oncol Lett 2022;23:183

[10] Gordon Em, Chua-Alcala Vs, Kim K, Et Al. Saint: Results Of An Expanded Phase Ii Study Using Safe Amounts Of Ipilimumab (I), Nivolumab (N), And Trabectedin (T) As First-Line Treatment Of Advanced Soft Tissue Sarcoma [Nct03138161]. J Clin Oncol 2022;38:Abstr 11520.

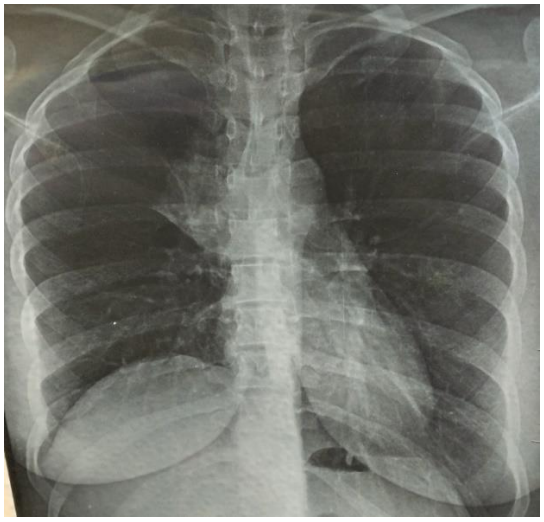

Figure 1 : Posteroanterior Chest Roentgenogram Showed Homogeneous Right Hilar Opacity With Elevation Of The Right Diaphragmatic Dome

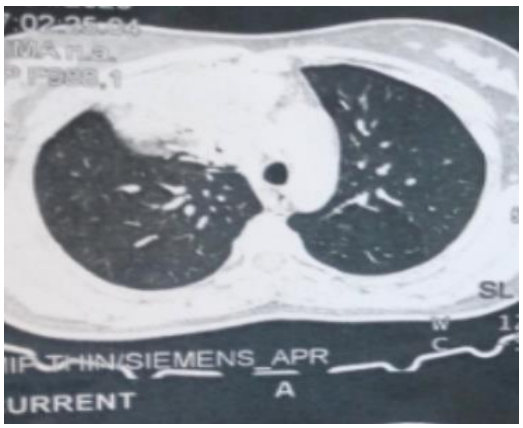

Figure 2 : The Thoracic Computed Tomography (CT) Showed A Pulmonary Process Of The Right Upper Lobe Well Limited Measuring 68x72x68

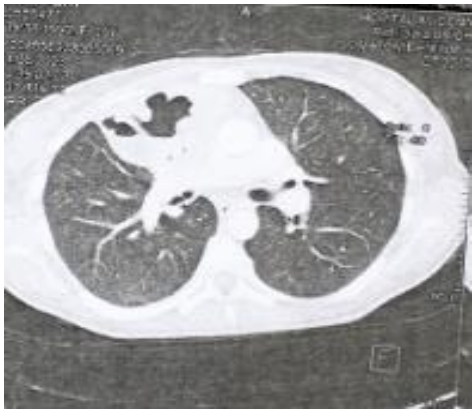

Figure 3 : The Thoracic Computed Tomography (CT) Showed A Radiological Deterioration With Excavation Of The Pulmonary Process .

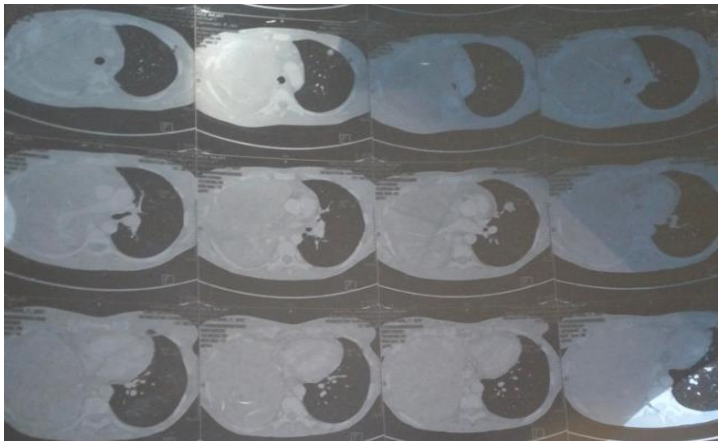

Figure 4 : The Thoracic Computed Tomography (CT) Showed An Increase In Volume Of The Pulmonary Mass Which Occupyies The Entire Right Lung

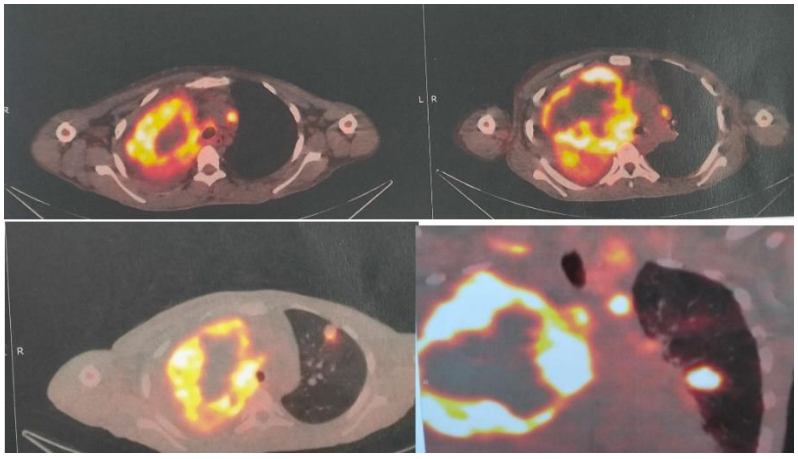

Figure 5 : PET-CT Showed A Hypermetabolic Mass With Necrotic Center Involving The Entire Right Lung (SUV: 12), Two Hypermetabolic Pulmonary Nodules On The Contralateral Side In The Left Upper Lobe (SUV: 5.5) And Left Lower Lobe (SUV: 9.1), Along With Pathologically Hypermeabolic Cervical-Mediastinal Lymph Node Foci.