Pulmonary Blastoma with Submandibular, Scrotum and Adrenal Metastases: Case Report

Kostas N. Syrigos a, Nikolaos Katirtzoglou a, Vasileia Ntomi a, Aikaterini Pierrakou b, Maria Chorti b, Grigorios Stratakos a, Ioannis Dannos a, Muhammad W. Saif c

a Oncology Unit, 3rd Department of Medicine, Sotiria General Hospital, Athens University School of Medicine, and b Department of Pathology-Anatomy, Sismanogleio General Hospital, Athens, Greece; c Division of Hematology/Oncology, Department of Medicine, Columbia University College of Physicians and Surgeons/Gastrointestinal Oncology Section, Division of Hematology/Oncology, Herbert Irving Comprehensive Cancer Center (HICCC), Columbia University/Pancreas Center, New York Presbyterian Hospital, New York, N.Y., USA

Abstract

A 62-year-old patient diagnosed with pulmonary blastoma with submandibular, scrotum and adrenal metastases was admitted to Sotiria General Hospital in Athens. No other such case has been published to date. The patient started receiving chemotherapy, but the scrotum metastasis grew rapidly and erupted. This led to sepsis despite surgical excision of infected and necrotic tissues and intravenous antibiotics. Treatment strategy in pulmonary blastoma should be defined by a multidisciplinary team, and surgical treatment should be considered as quickly as possible when such a tumor is suspected.

Key Words
Adrenal metastases • Pulmonary blastoma • Scrotum metastases • Submandibular metastases

Established Facts

- Pulmonary blastoma is a rare malignant lung neoplasm with a poor prognosis.
- Liver, central nervous system and bones are the most frequent locations of distant metastases.

Novel Insights

- Pulmonary blastoma may involve very atypical metastatic sites at diagnosis, such as the submandibular glands, the scrotum and the adrenal glands.
- It progresses in a very short time.
- Treatment strategy should be defined by a multidisciplinary team and it is essential that surgical treatment be considered as quickly as possible when such a tumor is suspected.
**Introduction**

Pulmonary blastoma is a rare pulmonary malignancy accounting for 0.25–0.5% of all malignant lung neoplasms. Its histological origin remains uncertain and it is currently considered as one of the main histological variants of pulmonary sarcomatoid carcinomas [1]. The liver, central nervous system and bones are the most frequent sites of metastases, and only few atypical cases have been reported in the literature [2]. We present the case of a 62-year-old patient who was diagnosed with pulmonary blastoma with submandibular, scrotal and adrenal metastases. To the best of our knowledge, no other such case has been reported to date.

**Case Report**

A 62-year-old male presented to the Emergency Department of our hospital following a motorcycle accident and was diagnosed with bilateral hip fracture. A chest X-ray showed increased opacity in the region in the left upper lobe, and chest computed tomography (CT) scans revealed a 10 × 7 cm tumor of the left upper lobe as well as enlarged mediastinal lymph nodes. The patient immediately underwent orthopedic surgery and remained hospitalized for a month. At the same time, three rapidly growing painless nodules appeared: two submandibular nodules and one on the scrotum.

A nondiagnostic bronchoscopy was performed and was followed by a thoracotomy. Biopsies were taken from the lung, the mediastinal lymph nodes and the nodules. The tumor was histologically characterized by the presence of well-differentiated tubular glands set in a cellular stroma with small oval-to-spindle cells undergoing a high rate of mitosis (fig. 1a). These characteristics led to the diagnosis of pulmonary blastoma. Adjacent pleura and 5 lymph nodes were found to be infiltrated. Immunohistochemical staining was positive for vimentin and CD99 (fig. 1b). The mandibular and scrotum nodules were also characterized as metastatic.

The patient was then referred to the Oncology Unit for further evaluation and treatment. New CT scans were performed to determine the exact degree of disease spread. The chest CT scan showed remaining disease in the left lung, scar tissue as well as the already known enlarged mediastinal lymph nodes. The abdominal CT scan revealed a right adrenal mass measuring 3 cm in maximum diameter.

Upon clinical examination, the patient had an ECOG performance status of 3 due to postoperative hip pain and inability to walk without assistance. He had been a heavy smoker (>150 pack-years) and was now receiving analgesic treatment with gabapentin, acetaminophen as well as bronchodilators due to chronic obstructive pulmonary disease. The two submandibular and the scrotum nodules initially measured approximately 2, 1 and 1 cm in maximum diameter, respectively. They were hard on palpation, painless with no signs of inflammation or necrosis. Routine laboratory evaluation was unremarkable. The patient started receiving first-line chemotherapy three times a week with carboplatin area under the curve (AUC) 5, adriamycin (15 mg/m²) and cyclophosphamide (1,200 mg/m²). It was well tolerated with minimal toxicity (grade I anemia, alopecia and diarrhea). In the meantime, the right submandibular nodule was growing quickly and was becoming painful, inflamed and necrotic (fig. 2). The patient was referred to the surgical unit for assessment and the submandibular nodule was successfully removed. Chemotherapy was continued after healing of the incision. After the third cycle, new CT scans were performed and disease progression was detected in both the left lung and the right adrenal mass. The scrotum nodule also began to increase in size (fig. 3) and the patient started complaining of severe scrotal pain and subsequent difficulty in walking. A complete white blood count showed leukocytosis (30 × 10⁹/l) and neutrophilia (28.5 × 10⁹/l). He was prescribed oral antibiotics and was again referred to the Surgical Unit for removal of the scrotum mass. The latter erupted before surgery, causing severe bleeding. The patient was admitted to the Intensive Care Unit in critical condition. He received intravenous antibiotics and fluids and underwent surgery for the excision of the infected and necrotic tissues and drainage of pus. Nevertheless, sepsis and septic shock developed and he died from multiple organ failure 2 weeks later.

**Discussion**

Pulmonary sarcomatoid carcinomas are currently defined as poorly differentiated non-small-cell lung carcinomas with sarcoma characteristics or sarcoma-like (spindle and/or giant cell) features. They consist of 5 major histological variants: pleomorphic carcinoma, spindle cell carcinoma, giant cell carcinoma, carcinosarcoma and pulmonary blastoma. Pulmonary sarcomatoid carcinomas are thought to represent carcinomas ‘in transition’, with diverse pathways of clonal evolution accounting for histological differences of a common ancestor lesion [1, 2]. Pulmonary blastoma is rare and accounts for 0.25–0.5% of malignant lung neoplasms. Originally named ‘embryoma’ by Barnard in 1952, the term blastoma was introduced by Spencer [1] to reflect that the tumors arise from the pulmonary blastoma as other tumors develop from fetal tissue. Adult pulmonary blastomas and childhood blastomas are considered different entities [3]. A clinicopathologic and immunohistochemical study of 75 cases showed that cytokeratin 7 and TTF-1 (as opposed to surfactant protein-A) are useful markers and confirmed that disease stage is currently the only significant prognostic parameter. This study also showed that the prognosis of these tumors is worse than the prognosis of conventional non-small-cell lung carcinomas at surgically curable stage I, justifying their classification as an independent histologic type in the WHO classification [4].
Pulmonary blastoma presents at a younger age than non-small-cell carcinoma \cite{5, 6} and 40% of the cases are asymptomatic and found coincidentally on chest X-rays performed for other reasons \cite{7}. The chest radiography usually reveals a well-circumscribed mass measuring 1.5–13 cm in diameter \cite{8}. Patients may present with non-specific symptoms, similar to those of a respiratory infection. The most common symptoms are cough (30%), hemoptysis (20%), dyspnea and chest pain. This tumor may even rarely present as spontaneous pneumothorax \cite{9}. Some studies have suggested that pulmonary blastoma is associated with smoking \cite{10}. Diagnosis is usually made by bronchoscopy \cite{11}, needle biopsy and thoracotomy as a fair amount of tissue is frequently required due to the biphasic pattern \cite{1, 12}. Overall 5-year survival is approximately 16%. The factors that indicate poor prognosis are tumor recurrence, metastasis at initial presentation, tumor size over 5 cm and lymph node metastasis. Liver,
central nervous system and bones are the most frequent locations of distant metastases [2]. A review of the literature revealed only few atypical cases: a rare case which presented as a solitary upper lip metastasis in a paraplegic male (diagnosis was confirmed by autopsy findings) [2], a case of pulmonary blastoma with endobronchial growth [13], a rare case of classic biphasic pulmonary blastoma in a 40-year-old Hispanic female [14], two atypical cases of Caucasian women, aged 21 and 41 years [7], and an 84-year-old male, the oldest patient reported [15]. It should be noted that the metastasis to the adrenal gland was not histologically proven; however, its enlargement upon disease progression strongly suggests metastasis.

Even though pulmonary blastoma is usually diagnosed at advanced stages, surgical treatment is the standard care for this condition. Chemotherapy can be administered in the neoadjuvant or adjuvant setting as well as in patients not eligible for surgery [16, 17]. Radiotherapy is also used to treat patients unresponsive to other types of treatment [18] and some authors support a multimodality approach [17, 19]. The incidence of pulmonary blastoma is too low for clinical trials to be designed; therefore, the role of each treatment method has yet to be defined and conclusions with respect to therapy are difficult to reach.

Since pulmonary blastoma progresses rapidly in a short period of time, early detection [20] and careful staging are imperative [21, 22]. Treatment strategy should be defined by a multidisciplinary team that includes a pulmonologist, an oncologist, a thoracic surgeon and a general surgeon. It is essential that surgical treatment be considered as quickly as possible when such a tumor is suspected [15], and possibly supported by adjuvant chemotherapy and/or radiotherapy [19, 23].

Written informed consent and authorization to publish this case report and the accompanying images was obtained from the patient’s next of kin. A copy of the written consent is available for review from the Editor-in-Chief of this journal. The study conformed to the terms of the Helsinki Declaration.

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References