A Rare Central Thoracic Tumor

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CASE HISTORY

A 51-year-old male former smoker, without relevant medical history, had progressive dyspnea on exertion, cough, minor hemoptysis, and a large suprahilar tumor-like mass on chest radiography. A positron emission tomography–computed tomography scan at the referring center showed a lobulated mass invading the mediastinum and right pulmonary artery, and filling defects suspect for pulmonary emboli (Fig. 1). The lesion had moderate 18F-fluorodeoxyglucose uptake. At bronchoscopy, the right upper lobe was suboccluded by a smooth mass. Forceps biopsies revealed necrosis of unclear origin. The patient was referred for workup.

The absence of venous thrombosis, intracardiac clots, pulmonary hypertension, and arterial hypoxemia reduced the likelihood of widespread pulmonary emboli. After reassessment at our thoracic oncology multidisciplinary meeting, a surgical procedure was proposed based on the suspicion of a tumor mass originating from the pulmonary artery.

A right-sided intrapericardial pneumonectomy and left-sided pulmonary endarterectomy (PEA) through sternotomy was performed on extracorporal cardiopulmonary bypass. The procedure and postoperative recovery were uncomplicated. The pneumonectomy specimen showed a tumor completely obliterating the right pulmonary artery and protruding into the right upper lobe bronchi. Left PEA from pulmonary valve level to the subsegmental branches was able to remove a pale smooth mass (Fig. 2).

Pathology showed a tumor in the pulmonary artery wall with a heterogeneous aspect. Essentially, it concerns a spindle cell process with cellular fascicular to myxoid areas. The tumor cell nuclei were atypical with a mitotic count of 5 of 10 hpf. Immunohistochemistry revealed (nuclear) MDM2 expression and diffuse epidermal growth factor receptor expression (Fig. 3). Stains for desmin, α-smooth muscle actin, CD31, and CD34 were negative. FISH analysis showed strong amplification of the MDM2 and PDGFRα genes, thus confirming an intima sarcoma.

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Disclosure: The authors declare no conflict of interest.

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ISSN: 1556-0864/14/0906-0897

Journal of Thoracic Oncology®  •  Volume 9, Number 6, June 2014  897
Pulmonary artery intimal sarcoma (PAIS) is a very rare thoracic malignancy, first described in an autopsy in 1923, with an estimated incidence between 0.001% and 0.03%. PAIS often presents as pulmonary vascular disease, such as acute pulmonary embolism, chronic thromboembolic pulmonary hypertension, and idiopathic pulmonary artery hypertension, and it can even mimic large-vessel arteritis. Diagnosis

FIGURE 2. Resection specimen showing tumor occluding the right pulmonary artery (A) and a pale pulmonary artery cast after left pulmonary endarterectomy (B).

FIGURE 3. Histopathological examination. Low power view (A) showing the tumor nodules in the arterial branches (arrows), and at higher magnification (B), a moderately cellular spindle cell proliferation is present. Immunohistochemistry showing nuclear MDM2 expression (C) and strong cytoplasmic epidermal growth factor receptor labeling (D).
is challenging, and FISH analysis by experienced pathologists is essential in identifying PAIS.

Less than 300 cases are described in the literature, with only a few small case series focusing on the management of these tumors. Median survival is 45 days, up to 10 months with surgery. Survival is worse in aortic intima sarcoma. In the recently published largest single-center series (31 patients), postsurgical median survival was 17 months. Death is mostly because of local or metastatic recurrence. PEA is often performed for PAIS, solely or in combination with pneumonectomy. Evaluation of completeness of resection is complicated. The role of adjuvant chemotherapy or radiotherapy is unclear because of limited data.

REFERENCES