A 32-year-old nonsmoking woman was admitted to our hospital with progressive dyspnea on exertion and right-sided pleuritic chest pain for the past three months. The patient had been well until 2 years earlier when she developed periodic upper airway infections. She had not lost weight over the past year. On physical examination, the right basal region of the lung manifested dull percussion and at auscultation breath sounds were diminished. The central venous pressure was elevated. Cardiac examination revealed no murmur. There was no peripheral edema, clubbing, or cyanosis. Laboratory tests revealed an elevated erythrocyte sedimentation rate of 43 mm after 1 h and anemia (hemoglobin, 7.8 mmol/L). Pulmonary function tests disclosed no abnormalities.

A chest radiograph showed a markedly enlarged nodular right hilum, a pleura-based nodule in the right upper lobe, and ipsilateral pleural effusion (Fig 1). Vascular markings on the right side were slightly diminished compared with the left. Contrast-enhanced CT scans showed the presence of an intraluminal mass occluding and expanding the right pulmonary artery (Fig 2). In the apical segment of the right lower lobe, a pleura-based nodule was depicted. Small right-sided pleural effusion was present as well. There was no mediastinal lymphadenopathy. The anterior segmental bronchus was displaced laterally without signs of bronchial obstruction (Fig 3). Subsequent coronal T1-weighted magnetic resonance images clearly confirmed the intraluminal localization of the mass (Fig 4). The lesion extended into the right upper lobe in a vascular pattern, but the bifurcation of the pulmonary trunk was not invaded.

Ventilation and perfusion scans showed diffuse hypoventilation and complete absence of perfusion of the right lung. Fiberoptic bronchoscopy revealed no endobronchial pathology. There was extrinsic compression of the anterior segmental bronchus of the right upper lobe. Right-sided thoracocentesis did not show malignancy. At thoracotomy, biopsy specimens of the right pulmonary artery were obtained.
Diagnosis: leiomyosarcoma of the pulmonary artery

A right-sided pneumonectomy was performed. Pathologic examination of the surgical specimen showed that the right pulmonary artery was completely obstructed by a large solid tumor with close connection to the vascular wall. The tumor extended to the right upper and lower lobe and along the intima to the margins of resection. In both lobes, several areas of infarction were present. The patient received additional radiotherapy (total dose, 60 Gy). Postoperative recovery was uneventful. However, at 18 months, she was readmitted because of an epileptic attack secondary to a solitary brain metastasis. The patient was treated by resection of the metastasis, followed by radiotherapy (total dose, 40 Gy). At a 2-year follow-up, a solitary pulmonary metastasis was detected and successfully resected. Three years after the initial diagnosis, the patient is still doing well with no evidence of local recurrence or metastatic disease.

Discussion

Primary sarcoma of the pulmonary artery is a very rare tumor. A variety of histologic types have been reported in the literature, undifferentiated sarcoma and leiomyosarcoma being the most common. There appears a slight female predominance and a wide age range at detection. The tumor almost invariably arises in the intima and the luminal part of the media of the pulmonary artery or the pulmonary trunk. Pulmonary artery sarcoma spreads peripherally along the pulmonary arterial intima. Retrograde spread to the pulmonary trunk is less common. Massive pulmonary infarction is rare even in the presence of complete occlusion of the pulmonary artery. Direct invasion of the mediastinum takes place by penetration of the arterial wall. Lymph nodes also may be involved. Pulmonary metastases occur in half the cases, and distant metastases to most organs have been reported.

Presenting symptoms of pulmonary sarcomas are remarkably similar and in general nonspecific: malaise, dyspnea, thoracic pain, coughing, hemoptysis, and right heart failure. Episodes of syncope also may occur. The most common presumptive misdiagnosis is pulmonary thromboembolism. However, in that case, a more rapidly deteriorating clinical picture can be expected.

A chest radiograph usually shows one or more of the following abnormalities: a hilar mass, solitary or multiple pulmonary nodules, and diminished pulmonary vascular markings in contrast with the normal tapering of the pulmonary artery and its branches in the non-affected lung. Pleural effusion or right-sided cardiac enlargement or both are reported as well. Pulmonary angiography demonstrates a filling defect in or occlusion of the pulmonary artery. Distinction from thromboembolic disease is only possible when the tumor is polypoid and moves during the cardiac cycle. Mediastinal lymphadenopathy and direct invasion of the mediastinum depicted by CT indicates a neoplasm. Ipsilateral pulmonary nodules does not necessarily represent metastatic disease and should be differentiated from pulmonary infarcts. Magnetic resonance imaging by its multiplanar imaging capability facilitates visualization and delineation of the process. Vascular tumor extension to the pulmonary parenchyma from the pulmonary artery into the branches and the pulmonary trunk is nicely depicted. Magnetic resonance images are indistinguishable from occlusion due to thrombus. Contrast enhancement would argue in favor of a sarcoma; however, the enhancement may be difficult to depict.
The prognosis of pulmonary artery sarcomas is very poor. Many of the patients are already suffering from an advanced stage of tumor on admission to the hospital. Most people die within a short time after diagnosis due to right heart failure. Improved survival may be accomplished through early diagnosis and complete surgical resection. As the tumor spreads along the intima, extent of tumor involvement may be underestimated, and invasion of the pulmonary trunk should be taken into account when considering surgical excision. Part of the pulmonary bifurcation may have to be resected and reconstructed with a vascular wall plasty. The rarity of this tumor makes generalizations about chemotherapeutic or radiotherapeutic management or both difficult.

References

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