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Journal of the Chinese Medical Association 77 (2014) 49–51

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Case Report

Primary pulmonary leiomyosarcoma

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Received August 1, 2011; accepted June 8, 2012

Abstract

Primary pulmonary leiomyosarcoma (PPL) is an extremely rare malignant tumor. In the case presented here, a 52-year-old Chinese female with a lung mass underwent a right upper-middle lobectomy with pulmonary artery sleeve resection and reconstruction, and was thereafter diagnosed with PPL. After 28 months, the patient was well and without local recurrence or distant metastasis.

Keywords: lobectomy; primary pulmonary leiomyosarcoma

1. Introduction

Primary leiomyosarcoma rarely arises in the lung. The first case of primary pulmonary leiomyosarcoma (PPL) described in the literature was presented by Davidsohn in 1903. Since that time, it has been observed that PPL can originate from the smooth muscle of the pulmonary parenchyma, the pulmonary arteries, or the bronchi. Patients may present with symptoms similar to those seen in other primary lung tumors, or they may be asymptomatic. A definitive diagnosis of PPL is made by pathological examination of a tumor specimen. Early detection and complete surgical resection are significant contributors to survival.

2. Case report

A 52-year-old Chinese female presented to our hospital complaining of a nagging cough that had lasted for 9 months and had gradually worsened over the previous 3 months. Her vital signs were normal on physical examination. The lungs were clear to auscultation bilaterally; there were no rales or rhonchi. In addition, there was no chest deformity and no superficial adenopathy. The patient was not a smoker and she had no family history of lung cancer. The review of systems was otherwise nonremarkable. Initial laboratory evaluations, including peripheral blood count, serum chemistry, and urinalysis, were all normal.

An enhanced computed tomography (CT) scan of the chest revealed a 6.4 cm × 5.8 cm × 8.5 cm irregular heterogeneous mass in the right hilar region with mediastinal and hilar lymphadenopathy. The mass extended along the right main pulmonary artery. The radiologic appearance was suggestive of a malignant tumor arising from the pulmonary artery, raising suspicion of sarcoma (Fig. 1). There were no metastatic lesions detected on abdominal CT, magnetic resonance imaging (MRI) of the brain, and a whole-body bone scan by emission CT. The patient's lung biopsy indicated a mesenchymal lesion and the preliminary diagnosis was a non-small-cell tumor of the right upper lobe, with invasion of the superior vena cava and the right pulmonary artery trunk. The patient underwent a right upper-middle lobectomy with pulmonary artery trunk sleeve resection and reconstruction, partial peeling of the adventitia of the superior vena cava, and mediastinal and hilar lymphadenectomy in March 2009. Intraoperatively, the tumor was located in the right hilar region and was adhered to the arch of the azygos vein and the proximal portion of the superior vena cava. It had also invaded the right pulmonary artery trunk. Adhesions were found

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1726-4901/$ - see front matter
http://dx.doi.org/10.1016/j.jcma.2013.10.009

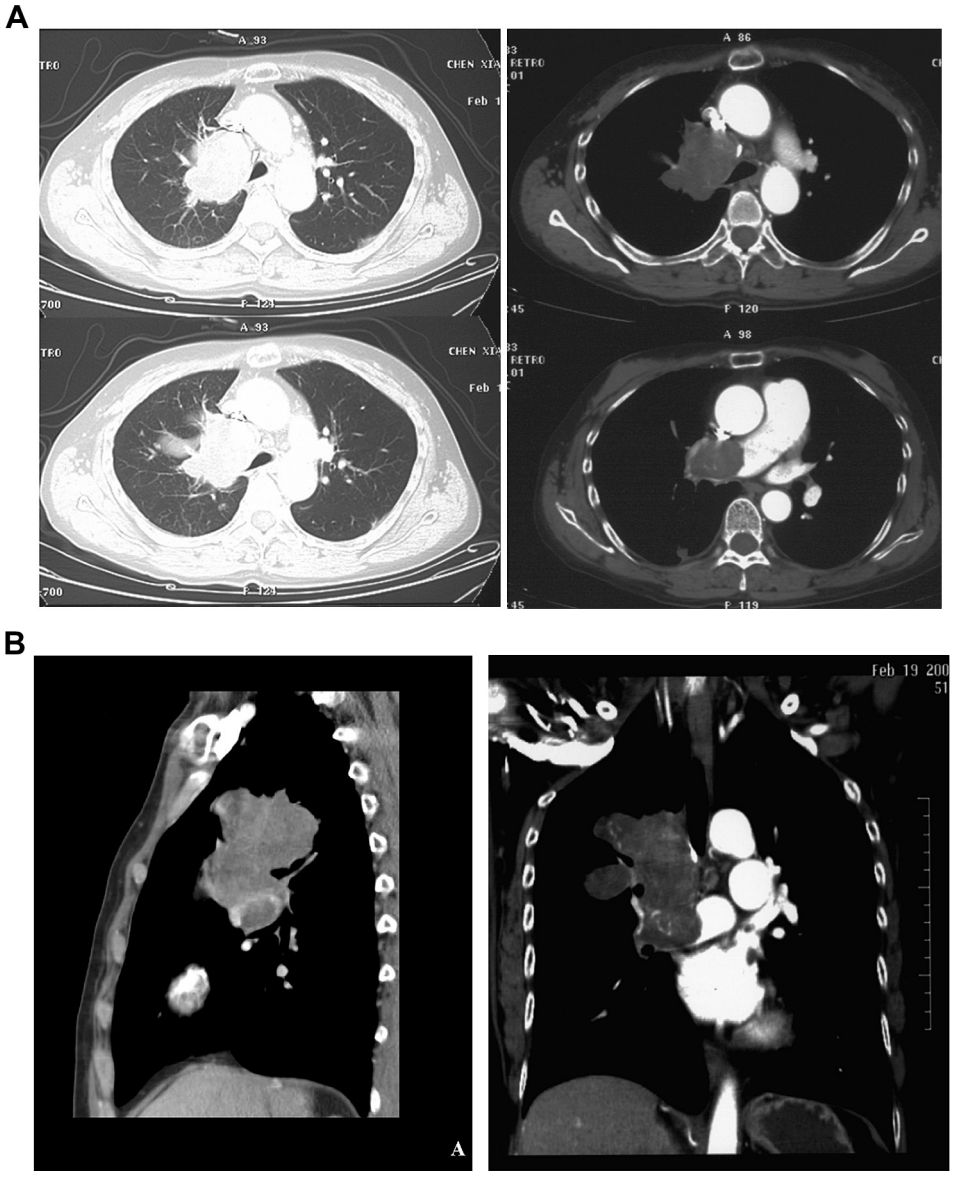

Fig. 1. An enhanced CT scan of the chest showed a large irregular mass extending into the right pulmonary artery trunk and its branches, with circumferential narrowing of the upper lobe and the anterior segmental bronchus. The mass compressed the superior vena cava and invaded the right superior pulmonary vein. (A) Horizontal and (B) axial (left panel) and coronal (right panel) views.

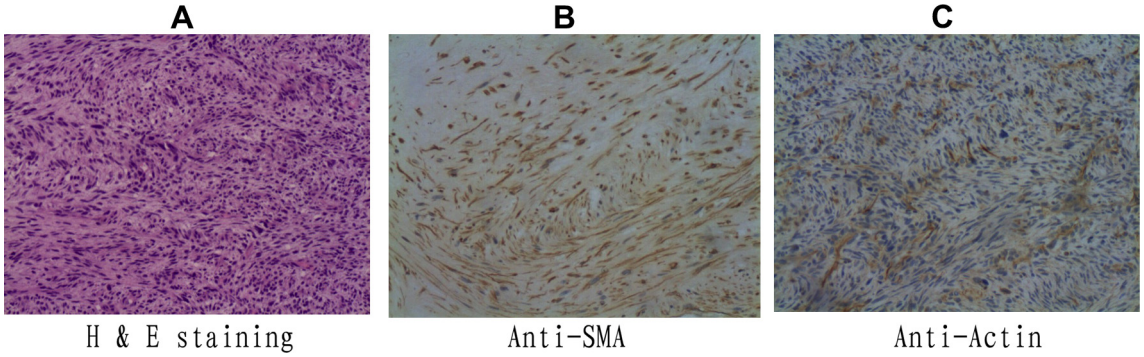

Fig. 2. (A) Hematoxylin–eosin staining of the primary pulmonary leiomyosarcoma showing spindle cells arranged in interweaving bundles (200×). (B,C) Immunohistochemical staining of the primary tumor cells revealed reaction with (B) anti-SMA and (C) actin antibodies, confirming cytoplasmic expression of these markers (streptavidin peroxidase (SP) 200×).

in the right thoracic cavity, but there was no pleural effusion. On gross pathology, the tumor was described as a huge grayish white mass of 7.5 cm × 5.0 cm × 4.0 cm in size, located at the bronchial margin in the right hilar region. Microscopic observation revealed spindle cells arranged in interweaving bundles (Fig. 2A). Nuclear atypia, nuclear differentiation, and prominent vascularity were common, and hemorrhage and necrosis were present. Immunohistochemistry was positive for SMA (Fig. 2B) and actin (Fig. 2C); CD99, CD68, EMA, S-100, and Bcl-2 results were negative. The pathological diagnosis was a well-differentiated leiomyosarcoma of the right lung with invasion of the pleura. No tumor cells were found at the margins of the bronchi. No metastases were found in the 2nd, 3rd, 4th, 5th, 7th, 9th, 10th, or 11th station mediastinal lymph nodes. Thus, the tumor stage was $T_4N_0M_0$ (IIIA). The patient's postoperative course was uneventful. There were no complications and she was discharged from hospital on postoperative Day 18. After 28 months of follow-up, the patient is alive and well and without local recurrence or distant metastasis.

3. Discussion

PPL is a mesenchymal tumor that appears to originate from smooth muscle cells of the bronchial wall or a vessel wall, and accounts for less than 0.5% of all malignant pulmonary tumors.[1] PPL can be classified by type as intraluminal, intrapulmonary, or pulmonary vascular.[2] The intrapulmonary type is the most common. The pulmonary vascular type grows along the vascular wall and occurs in the pulmonary artery, where it may cause stenosis or obstruction.[3] The tumor in this report was the intrapulmonary type.

PPL is commonly a solitary mass that usually affects people over 50 years of age, although some cases have been reported in children, with a 2:1 male to female ratio. From 1980 to 2010, only one patient has been diagnosed with PPL in our hospital. Patients may present with symptoms similar to those seen in other primary pulmonary tumors, including fever, cough, hemoptysis, sputum production, dyspnea and chest pain,[4] or they may be asymptomatic.

It is difficult to differentiate PPL from lung cancer, pulmonary tuberculosis, mediastinal tumor, pulmonary artery sarcoma, and pulmonary artery stenosis. Radiographically, PPL appears as a sharply defined, heterogeneous density that is smooth in contour with calcification in areas of presumed ischemic tissue damage. When a chest radiograph shows a solitary oval or round, well-circumscribed, calcified, and heterogeneous nodule or mass, PPL should be considered along with other types of lung cancer and pulmonary tuberculosis. PPL is usually located in the periphery of the lung, but may be central and adherent to hilar structures. Pulmonary artery sarcoma also appears as a heterogeneous density. The differential diagnosis is mainly through postoperative pathological examination.

PPL is grossly characterized by a firm grey or white surface. Microscopically, the majority of these tumors consist of interlacing bundles of spindle cells with oval vesicular nuclei and varying degrees of nuclear atypia. Hemorrhage and necrosis are common. Preoperative bronchoscopic examination, sputum smear, and lung biopsy are often negative. The diagnosis can be confirmed by intraoperative frozen section biopsy or postoperative pathological and immunohistochemistry examination. When actin, SMA, and desmin immunohistochemical results are positive, the tumor is presumed to originate from smooth muscle. When CD99 is negative, Ewing's sarcoma should not be considered. A negative EMA result indicates that the tumor typically does not originate from epithelial tissue. When S100 is negative, a tumor of the nervous tissue tumor is not suggested.

Metastasis of PPL is uncommon and typically occurs late in the disease process, which highlights the importance of early detection.[5] The tumors are amenable to surgical resection, which is the primary and definitive mode of treatment. Surgical options include lobectomy, pneumonectomy, and bronchial sleeve resection. The role of adjuvant treatment modalities has yet to be defined; radiochemotherapy is recommended in cases of incomplete resection and malignancy.[6]

If an early complete resection is performed, the 5-year survival rate is close to 50%, and there have been reports of survival at 20 years post-resection.[7] Prognostic indicators include tumor size, extent of bronchial invasion, and degree of malignancy.

In conclusion, PPL is rare and grows rapidly. It is difficult to differentiate PPL from other pulmonary tumors because there are no specific manifestations. Preoperative diagnosis via sputum smear, lung biopsy, or bronchoscopic examination is difficult. The most reliable methods for detection and diagnosis include chest radiography and postoperative pathological examination. Increased awareness, early diagnosis, and complete surgical resection with adjuvant therapy in selected cases may improve the prognosis.

Acknowledgments

This study was partly supported by grants from the Science and Technology Support Key Program of Tianjin (09ZCZDSF04100, 09ZCZDSF04000).

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