Radiographic, CT, and MRI findings in primary pulmonary angiosarcoma

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Abstract

We report a rare instance of primary pulmonary angiosarcoma presenting as a large solitary mass in the left upper lobe with mediastinal invasion. In particular, we emphasize the magnetic resonance (MR) imaging features, which included a markedly heterogeneous pattern consisting of hyperintense areas scattered throughout a background of intermediate signal intensity, rendering the lesion a cauliflower-like appearance especially on T2-weighted images. Being unreported so far in primary pulmonary angiosarcomas, these distinct MR imaging findings may be helpful in the differentiation of these neoplasms from lung cancers. © 2001 Elsevier Science Inc. All rights reserved.

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1. Introduction

Angiosarcomas of the lung are very rare tumors and usually represent metastases from the heart, pulmonary arterial trunk, or extrathoracic organs [1]. Primary pulmonary origin of this tumor has been reported very infrequently [2–10]. In this paper, we describe radiological findings in a patient with a solitary left upper lobe mass, which was diagnosed as angiosarcoma by histopathological and immunohistochemical examinations.

2. Case report

A 50-year-old man presented with a 2-month history of chest pain, malaise, and hoarseness. Physical examination revealed decreased respiratory sounds in the left upper zone and left-sided Horner’s syndrome. Routine laboratory examinations were unremarkable. Chest X-ray showed a large, well-defined homogeneous opacity occupying the upper zone of the left hemithorax (Fig. 1). Computed tomography (CT) of the chest demonstrated a 10-cm diameter hypodense mass with soft tissue attenuation in the left upper lobe. The mass had slightly irregular, lobulated margins and showed inconspicuous peripheral contrast enhancement. The esophagus, left subclavian artery, and trachea appeared infiltrated by the lesion (Fig. 2), while the main pulmonary arteries were unaffected. The mass was heterogeneous on T1-weighted magnetic resonance (MR) images with scattered areas of relatively high signal intensity and several hypointense punctate foci (Fig. 3). T2-weighted MR images better displayed the marked heterogeneity of the lesion caused by regions of high signal intensity interspersed in a background of intermediate intensity, resembling a cauliflower (Fig. 4). After the injection of gadolinium-DTPA, several linear structures probably representing vessels were observed entering the interior of the mass from the enhancing peripheral rim (Fig. 5). Histopathological examination of biopsy material showed vascular spaces lined by malignant endothelial cells, some containing vesicular nuclei, suggesting high-grade angiosarcoma (Fig. 6). Immunohistochemistry, using streptavidine–biotin peroxidase technique, showed positive staining for factor VIII-related antigen and CD31, confirming the diagnosis of angiosarcoma. Due to extensive mediastinal invasion, the tumor was considered inoperable, and chemotherapy was commenced. The patient is alive 9 months after the diagnosis.
3. Discussion

Angiosarcoma is a rare tumor, which constitutes less than 1% of all sarcomas [4]. The most frequently involved sites are the skin and subcutaneous tissue, liver, breast, and heart [1,4]. Angiosarcomas occurring in the lung usually represent metastases from the heart, pulmonary arterial trunk, and extrathoracic organs [1]. Primary pulmonary origin is very uncommon, being reported only in a handful number of cases [2–10]. Predisposing factors include thorotrast, polyvinyl chloride, and phenylethylhydrazine for liver tumors, postmastectomy and postirradiation states for skin and chest wall lesions, and chronic empyema for pleural space angiosarcomas. Our patient had no history of radiotherapy and exposure to the former agents.

Primary pulmonary angiosarcomas may occur either as multifocal lesions or as a solitary nodule. Patients usually present with chest pain, hemoptysis, dyspnea, cough, and...
malaise [5]. When the involvement is multifocal, as has
been described in many of the previous patients, chest
X-rays show bilateral reticulonodular or alveolar infiltrates,
with or without pleural effusion. This pattern of involvement
suggests metastatic cancer and lymphangitic carcinoma [7].

The solitary form of primary pulmonary angiosarcoma
has been described in a few patients. The size of the lesion
may vary from a small nodule to a large mass invading the
mediastinum or chest wall [4,5]. There may be accompan-
ing pleural or pericardial effusions, and the surrounding
lung may contain hemorrhage. Erosion of the adjacent
bronchial structures may cause hemoptysis [7]. Ott et al.
[5] reported the CT appearance of a solitary pulmonary
angiosarcoma as a large, inhomogeneous solid mass invol-
ving all three lobes of the right lung with mediastinal
invasion. In our patient, CT demonstrated a large mass with
peripheral contrast enhancement and a slightly heterogen-
eous hypodense interior without appreciable opacification.
Marked peripheral contrast enhancement and hypodense
central parts may suggest inflammatory lesions including
abscesses and infected hydatid cysts. However, absence of
consolidation in the surrounding lung and fever and labor-
atory findings indicating inflammation may favor an alter-
native diagnosis. Demonstration of mediastinal and/or chest
wall invasion and mediastinal lymph node enlargement may
lead to a preliminary diagnosis of malignancy. Yet, differ-
etiation from the more common bronchogenic carcinoma is
practically impossible.

To our knowledge, MR features of primary pulmonary
angiosarcomas have not been described. Angiosarcomas of
the heart were reported to have heterogeneous signal
intensity on MR images. In cardiac angiosarcomas, Kim

Fig. 5. Contrast-enhanced axial T1-weighted image displays the enhancing
periphery from which a linear enhancing structure is noted to enter the
interior of the mass (arrow).

Fig. 6. Microscopic section showing vascular spaces (asterisks) containing erythrocytes and lined by malignant endothelial cells, some of which had vesicular
nuclei (arrows) (H&E × 100).
et al. [11] described a cauliflower-like appearance consisting of focal nodular areas of increased signal intensity interspersed within areas of intermediate signal intensity on T1- and T2-weighted sequences. Likewise, the tumor in our patient exhibited a very heterogeneous pattern containing focal areas of high signal intensity in an overall background of intermediate intensity on both sequences yet more prominent on T2-weighted images. Such a high degree of heterogeneity is unusual in bronchogenic carcinomas and may be a useful hint in the distinction of angiosarcomas from lung cancers. Further case descriptions and patient series are needed to validate the potential utility of this finding.

Despite the availability of sophisticated radiological methods and peculiar MR signal intensity characteristics, accurate differentiation of pulmonary angiosarcomas from other masses, especially lung cancer, is not possible without biopsy. The distinction between benign and malignant vascular lesions can be quite challenging even on histopathological grounds and requires immunohistochemical methods in most patients. Among the various immunohistochemical markers used for classification, factor VIII-related antigen and CD31 are considered specific for tumors derived from endothelium [7,9].

Pulmonary angiosarcomas are characterized by an insidious growth and a late stage at presentation, when extensive local invasion and hematogeneous metastases have already occurred [5]. They are usually inoperable at the time of diagnosis and respond poorly to chemotherapy and radiation. Thus, prognosis is dismal, with most patients surviving less than 1 year [7].

In conclusion, despite their rarity, primary pulmonary angiosarcomas should be considered in the differential diagnosis of aggressive lung masses. Radiologically, the most helpful sign in the characterization of these lesions seems to be the marked heterogeneity on T2-weighted images with focal areas of high signal intensity interspersed on a background of intermediate intensity, giving a cauliflower-like appearance. Nevertheless, the falseproof diagnosis rests on histopathological and immunohistochemical examinations.

References

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