An Uncommon Intrafissural Mass: Pleural Primitive Neuroectodermal Tumor

Una masa intracisural infrecuente: tumor neuroectodérmico primitivo pleural

To the Editor:

We report the case of a young asymptomatic patient with a chest mass that was finally diagnosed as an intrafissural primitive neuroectodermal tumor (PNET). A pulmonary mass was detected in this 27-year-old woman on an unrelated chest X-ray. All laboratory test results were normal. Computed tomography (CT) revealed an elliptic, heterogeneous mass with contrast uptake in the left hemithorax, about 12 cm in diameter with smooth borders, located within the oblique fissure of the left pleura (Fig. 1). CT-guided fine-needle aspiration biopsy cytology was inconclusive. Surgical resection revealed an intrafissural mass. Hematoxylin and eosin staining produced a surprisingly uniform pattern of solidly compacted round cells.

Immunohistochemical assays showed tumor cells that were strongly positive for MIC2 (CD99) and for vimentin, but negative for other markers, such as cytokeratin (CK), small cell lung cancer, chromogranin, CK7, CK19 and thyroid transcription factor-1. Molecular genetic studies using fluorescence in situ hybridization showed positive translocation of the long arms of chromosomes 11 and 22 [t(11;22)(q12;q11)]. These results were suggestive of PNET.

No distant metastases were found using standard staging methods, including brain magnetic resonance imaging (MRI), whole body MRI, abdominal CT and bone scintigraphy. After complete surgical resection, the patient received chemotherapy. Since then, she has been monitored in the outpatient clinic and her situation remains unchanged after 2 years.

Chest PNETs, also known as Askin’s tumors, probably originate in the embryonic neural crest cells. They consist of small round cells with neural differentiation, and are associated with translocation of chromosome 22.1,2 They typically appear in the soft tissues of the chest. A primary tumor located inside the lung is very rare, but pleural involvement is common.3 Patients are generally children or young adults, although PNET can develop at any age. It usually presents with pain in the chest wall, and can be associated with pleural effusion and dyspnea.4

The most common CT finding in PNET is a mass with heterogeneous contrast uptake. Occasionally, a central area of low density with no contrast uptake seen inside the mass. The heterogeneous aspect of these large tumors is usually due to focal areas of bleeding or necrosis.5

Diagnosis of PNET is made using various techniques, such as optical microscopy, histological findings of small round cells arranged in a lobular pattern with rosettes or pseudorosettes, and by immunohistochemical and ultrastructural signs of neural differentiation. Reciprocal translocation of the long arms of chromosomes 11 and 22 [t(11;22)(q11;q24)] is currently thought to be characteristic of this tumor family.1 Although there are no

References

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Fig. 1. Chest computed tomography images [axial (A) and sagittal (B)] showing a heterogeneous elliptic tumor with contrast uptake in the oblique fissure of the left hemithorax.

standard treatment guidelines, neoadjuvant chemotherapy and surgical resection, with or without local radiation therapy, followed by adjuvant chemotherapy appears to be the best option. In conclusion, although primary fissural PNET is an extremely rare soft tissue sarcoma, this possibility must be taken into consideration in the differential diagnosis of a primary fissural mass.

References

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Positive Positron Emission Tomography/computed Tomography Scan and Multiple Pulmonary Nodules: An Unusual Diagnosis

To the Editor:

Primary amyloidosis is a rare disease caused by deposits of amyloid fibrils formed by insoluble protein in different tissue types. No genetic or environmental factors that might make an individual more susceptible to amyloid deposits have been identified.

Primary pulmonary amyloidosis, when deposits are limited to the lungs and associated structures with no involvement of other organs, is a very rare manifestation of the disease. Amyloid nodules in pulmonary parenchyma are usually an incidental finding that must be distinguished from other diseases. Pulmonary amyloid nodules with uptake on positron emission tomography (PET) are very rare. To date, only 7 cases have been published in the English literature.

An 80-year-old man, a former smoker, presented with a few months’ history of dyspnea and systemic signs (asthenia, anorexia and loss of weight). Physical examination and laboratory tests results did not reveal any significant changes. Chest X-ray showed multiple disperse nodular lesions in both lung fields. Chest and abdominal computed tomography (CT) found disperse nodular pulmonary lesions with lobulated borders and no calcifications, in both lungs. PET-CT results suggested that some of the lesions in both lungs had significantly higher affinity for FDG than others, and there were no lesions with increased uptake in any other part of the body. Lung function tests showed moderate airway obstruction. Transbronchic and transbronchial biopsies showed amorphous amyloid-type cells. Two years after diagnosis, the patient remains in good clinical condition under treatment with inhaled long-acting beta-antagonist bronchodilator and corticosteroid.

Primary pulmonary amyloidosis occurs in 4 forms: tracheobronchial amyloidosis, nodular pulmonary amyloidosis, infiltrating interstitial amyloidosis and amyloidosis of the lymph nodes. The anatomical and functional evaluation of respiratory tract amyloidosis should be evaluated using standard X-ray, CT, endoscopy and lung function testing.

From a radiological point of view, primary nodular parenchymal pulmonary amyloidosis occurs in the form of single or multiple nodules in any lobe, and must be taken into account in the differential diagnosis of primary or metastatic pulmonary malignancies, pulmonary tuberculosis, pulmonary sarcoïdosis, and connective tissue diseases. Nodules are generally peripheral and subpleural and 4 typical features are observed on CT: well-defined lobulated borders, calcification that is often central or forms in an irregular pattern within the nodule, multiple shapes and sizes, and slow growth, often over years, with no remission.

F-FDG PET-CT is widely used in the detection of cancer. As with other diagnostic procedures, some results may be false positives, and some non-malignant diseases, such as tuberculosis, sarcoidosis, fungal disease, interstitial lung disease, arthrosis, vascular thrombosis, osteoporosis and rheumatoid nodules, can show increased metabolic activity. In our case, multiple pulmonary amyloidosis nodules with moderate F-FDG uptake were observed, an observation also reported by Khan et al. and Zhang et al.

Pathological examination of a tissue sample is the standard reference for diagnosing amyloidosis.

On optical microscopy, hematoxylin and eosin staining shows homogeneous, pinkish, amyloid substances with no cell structure.

Fig. 1. Chest computed tomography showing disperse pulmonary nodules with lobulated borders.