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Cystic Metastasis of a Giant Cell Tumor Causing Recurrent Spontaneous Pneumothorax

Metástasis quística de un tumor de células gigantes causante de neumotórax espontáneo recurrente

Dear Editor:

A 17-year-old male was admitted to the Emergency Department with cough and episodes of hemoptysis. The patient had a history of a giant cell tumor (GCT) in the left tibia, resected 6 months previously. Chest computed tomography (CT) revealed pulmonary nodules, some of which were cavitated (Fig. 1A). Laboratory test findings were unremarkable. The patient's sputum was negative for acid-fast bacilli. He was referred for fiberoptic bronchoscopy with bronchoalveolar lavage. The bronchoalveolar lavage fluid contained a small amount of blood, and was negative for neoplastic cells. Cultures were negative for fungus and bacteria. Video-assisted thoracoscopy was performed, and the biopsy findings from one of the nodules were compatible with GCT metastasis. The patient started a new chemotherapy cycle. Four months later, he had an episode of chest pain associated with hemoptysis. A new CT examination showed a left pneumothorax, and cavitated thick-walled nodules with ground-glass halos (Fig. 1B and C). The pneumothorax was drained. The patient evolved well, with pulmonary re-expansion. Eight months later, he had a new episode of chest pain and dyspnea. CT showed a spontaneous left pneumothorax, and evolution of the cavitated nodules into thin-walled cysts (Fig. 1D). In this phase, the patient presented metastasis to intraabdominal lymph nodes in addition to the pulmonary metastases. He underwent new chest drainage and pleuroscopy with bilateral pleurodesis through the intrapleural instillation of talc. During pleuroscopy, pleural metastases were detected. The patient underwent a chemotherapy regimen with six cycles of doxorubicin and cisplatin, which resulted in regression of some of the lung lesions. He remains in outpatient follow-up with no new complication 1 year after the last pneumothorax.

GCT in the bone is a primary intramedullary tumor; it is generally benign, but can be locally aggressive and even metastatic. Malignant transformation and distant metastasis are extremely uncommon. Malignant transformation may occur as a result of dedifferentiation of the primary tumor or secondary to previous radiation therapy. Metastasis of GCTs most commonly arises in the lungs. Pulmonary metastases are more likely to appear in patients with recurrent GCTs, and often have an indolent course; they are rarely fatal.[1,2] Cavitation of metastases is extremely rare.

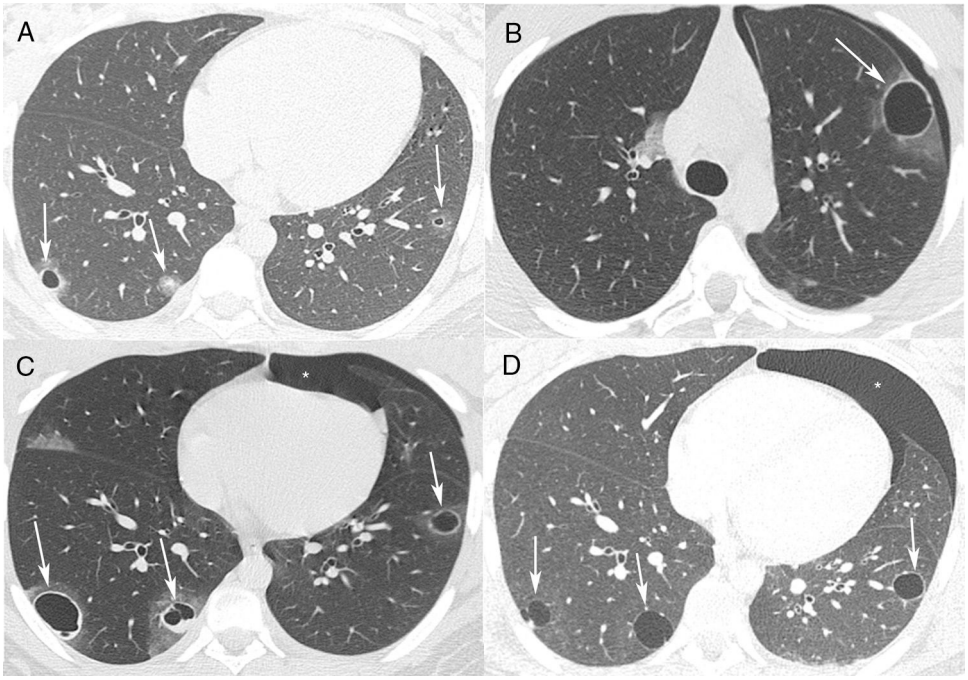

Fig. 1. (A) Axial chest CT with pulmonary window settings shows bilateral small pulmonary nodules, two of which are cavitated (arrows). (B, C) CT performed 4 months later demonstrates a left spontaneous pneumothorax (asterisk) and growth of the nodules, which now present with relatively thick walls and ground-glass halos (arrows). (D) CT performed 1 year after A shows a new left pneumothorax (asterisk) and evolution of the cavitated nodules into thin-walled cystic lesions (arrows).

Pulmonary metastasis initially presents as a solid mass, with an air-filled cavity formed after discharge of the necrotic material inside. The wall of a cavitated metastasis is generally thick and irregular, although thin-walled cavities can be found and may be seen with other lesions at various stages of excavation. The exact mechanism of cavitation is usually difficult to determine, but the cause is presumed to be tumor necrosis or a check-valve mechanism that develops by means of tumor infiltration into the bronchial structure. A potential complication of cystic metastases is pneumothorax, caused by necrosis of subpleural metastases producing a bronchopleural fistula.[3,4] The use of pleurodesis with talc in the treatment of pneumothorax associated with cystic pulmonary metastases has been described in the literature.[5,6] Authors[7,8] have suggested that given the high rates of recurrence, pleurodesis should be performed after the first spontaneous pneumothorax in patients with diffuse cystic lung diseases, rather than waiting for a recurrent episode. To prevent possible right pneumothorax, our patient underwent prophylactic resection of subpleural metastatic lesions and bilateral pleurodesis. The long-term prognosis and survival rate are favorable for patients with pulmonary metastasis, except for those with sarcomatous transformation, who have a worse prognosis.[1,2]

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Cavitary lung nodules associated with IgG4-deposition disease☆

Nódulos pulmonares cavitados con relación a la enfermedad por depósito de IgG4

To the Editor

IgG4-related disease is a multisystem disorder characterized by the formation of fibroinflammatory lesions, causing the affected tissues to malfunction.[1]

The 2 main features of this disease are tissue infiltration by IgG4 and raised IgG4 levels in serum, but these phenomena are not present in all patients.[2]

IgG4-related disease represents a diagnostic challenge as it mimics many other processes, including cancers, infections, and autoimmune diseases. For this reason, in most cases, the affected tissues must be biopsied in order to arrive at a diagnosis.

We report the case of an asymptomatic 79-year-old woman, former smoker, who was referred to our department for the study of lung nodules, detected incidentally on a chest X-ray obtained for a preoperative work-up. Fiberoptic bronchoscopy showed no significant changes. Chest computed tomography (CT) (Fig. 1) was performed, which revealed 2 lesions of thick irregular morphology with large central cavitation and coarse peripheral calcifications lacking interior content in both apical segments, measuring 45 and 40 mm in diameter in the right upper lobe (RUL) and left upper lobe (LUL), respectively.

Clinical laboratory testing showed only positive antinuclear antibodies (ANA), titer 1/160 with a granular pattern, and normal IgG4 levels (53 mg/dl).

Left thoracotomy was performed with a wedge resection of LUL segment 6 and vertex which included a nodular lesion measuring $6 \times 3 \times 1.7$ mm. An inflammatory infiltrate with abundant plasma cells (Fig. 1) was observed that was diagnostic for nodular pulmonary disease due to IgG4-deposition disease (the immunohistochemical study expressed abundant IgG with a IgG4/IgG ratio >40%).

Taking into account that the patient remained asymptomatic at all times, and that her IgG4 subclass levels were normal, we decided to continue with clinical and radiological follow-up. After 3 years of follow-up, the patient remains asymptomatic and her radiological lesions are stable.

IgG4-related disease is a recently described condition that more often affects men (ratio of 1 to 0.7) between 60 to 65 years old, predominantly individuals of Asian race.[1] It is characterized by raised serum IgG4 and IgG4 infiltration in the affected tissue.[2]

In 1995, Yoshida et al.[3] described a form of chronic pancreatitis and postulated that an autoimmune mechanism was the cause of the pancreatic lesion. However, it was not until 2003 that Kamisawa et al.[4] first established the term "IgG4-related autoimmune disease" (formerly also known as "hyper-IgG4 disease" and "sclerosing disease"), while demonstrating that patients who had autoimmune pancreatitis due to this mechanism could also have extensive lesions in other tissues.

Associated symptoms depend on the organ involved and include abdominal pain (40%), respiratory symptoms (13%), pruritus (13%), and diarrhea (6%). Constitutional symptoms such as weight loss,

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