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Case study

Extraosseous benign notochordal cell tumor presenting as bilateral pulmonary nodules

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Summary Intraosseous benign notochordal cell tumors are rare and the likely precursors of chordoma. Extraosseous benign notochordal cell tumors have been reported in only 2 patients, and both presented as solitary pulmonary nodules. Here, we report a 53-year-old woman with an incidental finding of small nodules bilaterally in the lungs. The clinician suggested the tumors were metastases; however, histologic examination of both tumors showed benign notochordal cell tumor, characterized by adipocyte-like vacuolated cells with bland nuclei and lacking an intercellular myxoid matrix. Although extraosseous benign notochordal cell tumors are extremely rare, the diagnosis should be recognized by pathologists to avoid overtreatment of patients.

1. Introduction

Intraosseous benign notochordal cell tumors (BNCTs) are a relatively recently discovered entity that in many cases is a chance finding at autopsy [1]. Further evidence has revealed the tumor to be a precursor of chordoma, thought to be the intraosseous remnant of the notochord. Surgical operation should be prevented until clinical signs of malignant transformation are detected [1]. In the literature, only 2 cases of extra-axial pulmonary BNCTs have been reported, and both cases demonstrated histologic and immunohistochemical features of BNCTs [2]. This report presents a case of bilateral simultaneous extra-axial BNCTs of the lung.

2. Case report

2.1. Clinical history

The patient was a previously healthy and asymptomatic 53-year-old woman. A computed tomographic (CT) image revealed 2 well-demarcated tumors, each 15 mm in size, located just beneath the visceral pleura of the right upper lobe and left lower lobe (Fig. 1). The findings suggested that the tumors were metastatic lesions, and a wedge resection was performed for the tumor of the right upper lobe. Before the operation, she received a whole-body CT scan and a whole-body positron emission tomography scan, but these failed to reveal a primary lesion.

Two years after the first operation, she underwent a second wedge resection for the tumor of the left lower lobe. During this interval, 5 CT scans and 1 abdominal sonogram were performed, all with no remarkable findings. The tumor of the left lower lobe exhibited no obvious changes in this span of time. After the second operation, follow-up examinations and CT scans were performed every 6 months

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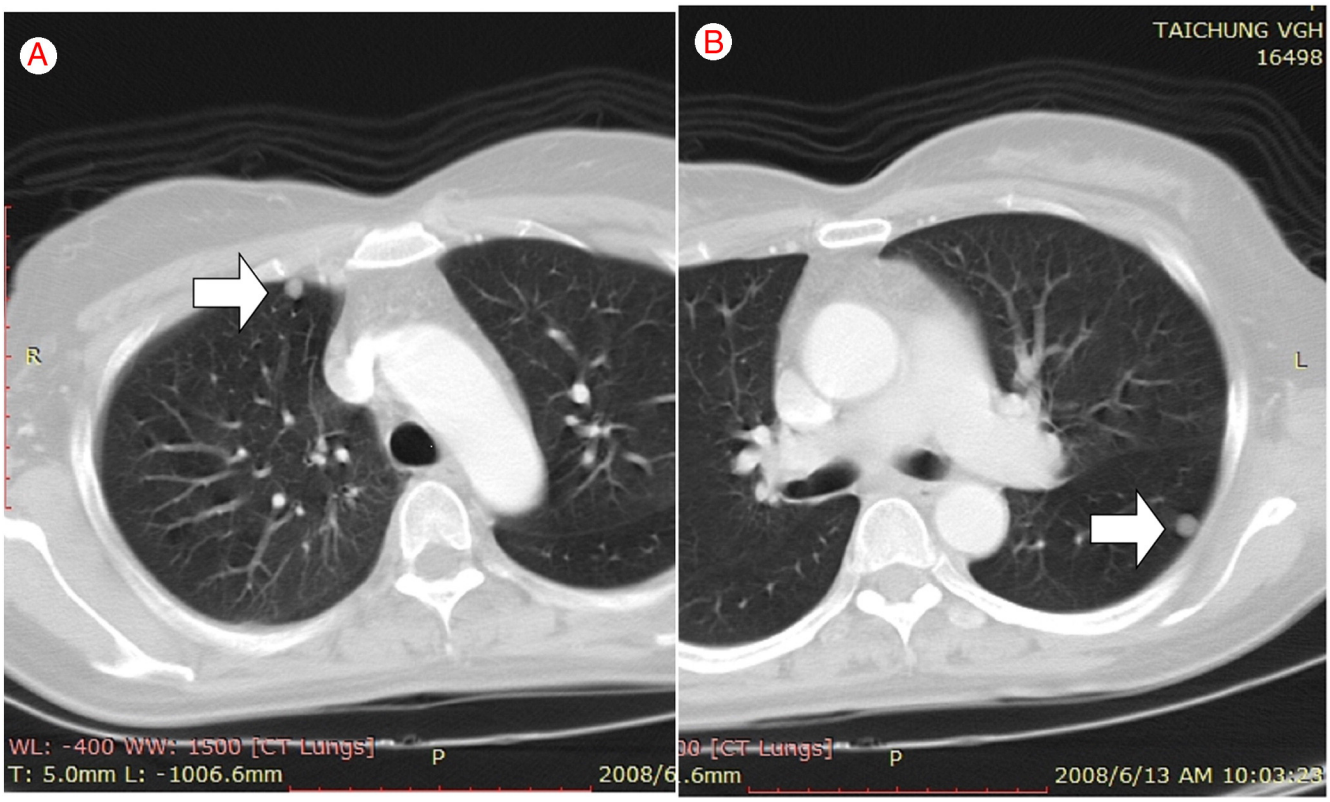

Fig. 1 Computed tomographic image reveals 2 well-demarcated tumors located beneath the pleura of right upper lobe (A, arrow) and left lower lobe (B, arrow).

for 18 months, and no evidence of disease, metastases, recurrence, or vertebral lesion was found by 2 years from the date of the second operation.

2.2. Pathologic findings

The tumors of the right upper lobe and the left lower lobe had similar gross and histologic features (Fig. 2). Both tumors measured 10 mm, were beneath the visceral pleura, and exhibited central cystic changes. Both tumors were characterized by predominantly vacuolated or slightly eosinophilic cytoplasm and peripherally located nuclei, mimicking mature adipocytes. No intercellular myxoid matrix was evident (Fig. 2).

Immunohistochemistry staining was performed on paraffin-embedded sections using the BenchMark automatic immunostaining device (Ventana XT Medical Systems, Tucson, AZ) and the ultraView DAB detection kit (Ventana). Sections were automated for antigen retrieval, and commercial antibodies were used. The tumor cells were strongly immunoreactive for cytokeratin (monoclonal, 1:100; Dako), S-100 (polyclonal, 1:3000; Dako, Carpinteria, CA, USA), vimentin (monoclonal, 1:200; Dako), and brachyury (monoclonal, 1:200; Abcam, Cambridge, MA, USA) (Fig. 3) but negative for CD15 (monoclonal, 1:50; Cell Marque, Rocklin, CA, USA), renal cell carcinoma marker (monoclonal, 1:100; Thermo, Fremont, CA, USA), Pax-8 (polyclonal, 1:100; Cell Marque), Pax-2 (polyclonal, 1:50; Invitrogen, Carlsbad, CA, USA), CD10 (monoclonal, 1:30; NeoMarkers, Fremont, CA, USA), HMB-45 (monoclonal, 1:50; Dako), calretinin (monoclonal, 1:100; Dako), and TTF-1 (monoclonal, 1:50; Thermo). No cytoplasmic glycogen granules were detected using the periodic acid–Schiff technique. There were no additional tumor components, and the resection margins were free of tumor.

Initially, metastatic carcinoma was suspected because of the clear cell features and immunoreactivity for cytokeratin. However, clinical survey and further immunohistochemical studies failed to detect any primary tumor. The features of the tumor cells, such as their sheet-like pattern and the presence of uniform, adipocyte-like vacuolated and less vacuolated eosinophilic cells without lobulated structure or myxoid background raised the suspicion of physaliferous cells and the possibility of notochord-derived tumor, which eventually was confirmed by the brachyury immunohistochemical stain.

Clinically, after the second wedge resection, she received CT scans as a follow-up every 6 months, and the results showed unremarkable change without vertebral lesions or recurrence. Based on the histologic and immunohistochemical findings, the diagnosis of extraosseous benign notochordal cell tumor was made.

3. Discussion

Most BNCTs are intraosseous and intra-axial arising from remnants of the notochord that may undergo malignant