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Case Report

Multiple benign notochordal cell tumors in lung with cystic change ☆,☆☆

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ABSTRACT

Benign notochordal cell tumor (BNCT) is a benign lesion derived from notochordal cells. Although it is relatively common in intraosseous lesion, pulmonary BNCT is extremely rare. We present a case of 54-year-old male with multiple pulmonary nodules, in which were considered to be metastatic chordomas initially. For 20 months follow-up without any therapy, most of the nodules had no remarkable change but some nodules showed cystic change. We consulted with pathologists specializing in chordoma and the final diagnosis of the nodules was considered as BNCT rather than chordoma. We herein report the case of multiple pulmonary BNCTs with cystic change, comparing with previous reports.

Introduction

BNCT is a benign lesion derived from notochordal cells and it is relatively common in intraosseous lesion, being found in 20% of adults at autopsy [1]. Although some cases of BNCT arising in lung has been reported recently [2–7], there is no report of multiple pulmonary BNCTs. We present a case of 54-year-old male with multiple pulmonary nodules showing cystic change and finally diagnosed of pulmonary BNCT.

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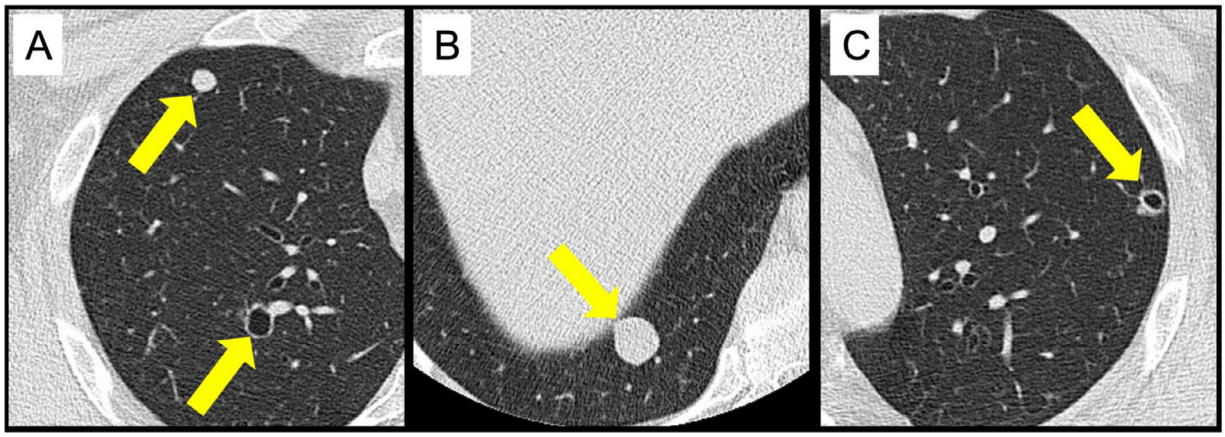

Fig. 1 – Chest CT images (A) the right upper lobe, (B) the right lower lobe, (C) the left upper lobe. CT shows multiple nodules in the bilateral lung fields, in some of which appearing cystic change.

Case report

The patient was an asymptomatic 54-year-old male with a smoking history (20 cigarettes/day for 34 years) and was referred to our hospital for further evaluation of multiple lung nodules that had been incidentally found on a computed tomography (CT) during general checkup for his abdominal pain. Physical examination and laboratory data were unremarkable. Tumor markers including CEA, CA19-9, CYFRA, NSE, proGRP, PSA were within normal limits. CT revealed multiple well-defined, round, solid and partly cystic nodules up to 14 mm in diameter in the bilateral lung fields (Fig. 1), and thus initially suggested multiple lung metastases from unknown origin. Fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography (FDG-PET/CT) showed no significant FDG uptake in the pulmonary nodules and no other abnormalities suggestive of primary malignant tumor (Fig. 2). A CT-guided biopsy of the pulmonary nodule was performed but was not diagnostic. Follow-up CT scans were performed every 2 months for 6 months and showed cystic change in some nodules and no remarkable changes in size. For appropriate follow-up and management, it was decided to obtain a pathological diagnosis by thoracoscopic surgery.

Histopathologically, the tumors consisted of solid sheets of lightly eosinophilic and vacuolated cells that resembled physaliphorous cells. Nuclei were small and had slightly atypia and immunohistochemistry staining revealed that MIB-1 index was 1% and the tumor cells were positive for cytokeratinAE1/AE3, cytokeratin7 and brachyury (Fig. 3).

Based on the above pathological findings and multiple pulmonary nodules on CT, it was initially considered to be metastatic chordoma. Although magnetic resonance imaging including the skull, spine and sacral region was additionally performed and FDG-PET/CT was reviewed again, there were no primary lesion revealed. After the thoracoscopic surgery, follow-up CT examinations were performed every 6 months for 12 months and showed cystic change in some nodules but no remarkable change in other nodules and no new lesion elsewhere (Fig. 4). Based on the above clinical course, repeated consultation with pathologists specializing in chordoma was held. The tumors mainly consisted of vacuolated cells and had central cystic change and little myxoid background although nuclear atypia was slight. As a result of discussion, benign notochordal cell tumor (BNCT) was considered as the final diagnosis rather than chordoma.

Discussion

BNCT is a benign lesion derived from notochordal cells described as a benign counterpart of conventional axial chordomas [8]. BNCT is relatively common in intraosseous lesion being found in 20% of adults at autopsy [1], while extraosseous BNCT is extremely rare and only reported in lung. There are only 6 cases via Pubmed [2–4,6,7] and one in the Japanese journal reported so far [5], after first reported by Kikuchi et al in 2011. Six cases of pulmonary BNCT presented solitary nodule and only 1 case reported by Lee et al. [3] presented bilateral nodules. To the best of our knowledge, there is no reports of multiple pulmonary BNCT in the bilateral lung fields.

Imaging findings of intraosseous BNCTs and chordomas have been previously reported. On CT, BNCTs show mild osteosclerosis with a residual bone architecture part, while chordomas often manifest with bone destruction. On MRI, both tumors show hyperintensity on T2-weighted images, and after contrast enhancement BNCTs show no enhancement, while chordomas show mild to avid enhancement [9]. Although chordomas show substantial inhomogeneous uptake on FDG-PET/CT, there have been no reports for FDG uptake in BNCTs. FDG-PET/CT may be useful for differential diagnosis between BNCTs and chordomas as it provides both anatomic and metabolic information [10]. Pulmonary BNCT is extremely rare and there have been few reports for imaging features. In the previous reports, pulmonary BNCTs have been described as round and regular shape and shown up to 15 mm in diameter. The nodules have mostly located in subpleural area [7]. Similar to the previous reports, the nodules in our case were 14 mm for maximum size and mostly detected in the peripheral

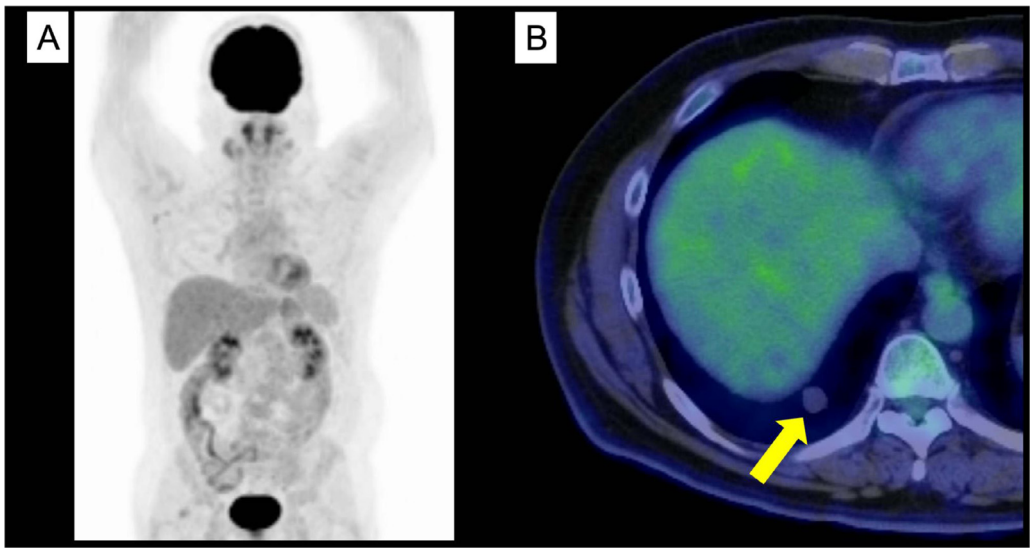

Fig. 2 – FDG-PET/CT (A: MIP, B: fusion): The fusion image shows no significant FDG uptake in the lung nodule (arrow) and MIP shows no abnormal uptake suggesting primary malignant tumor.

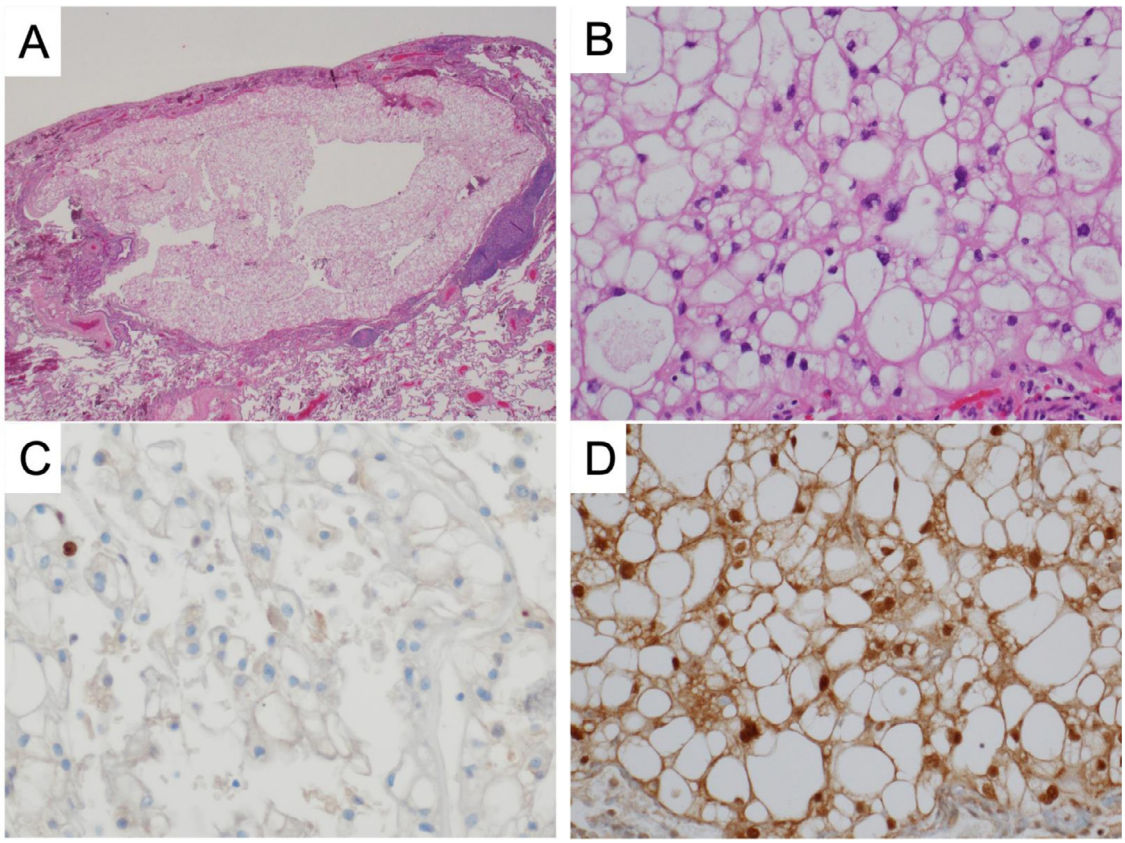

Fig. 3 – Pathological images: (A) HE stain at low-power field, (B) at high-power field, (C) MIB-1, (D) brachyury. The tumor cells are consisted of solid sheets of lightly eosinophilic and vacuolated cells. Nuclei have slight atypia. MIB-1 index is 1% and the nuclei of the tumor cells are positive for brachyury.

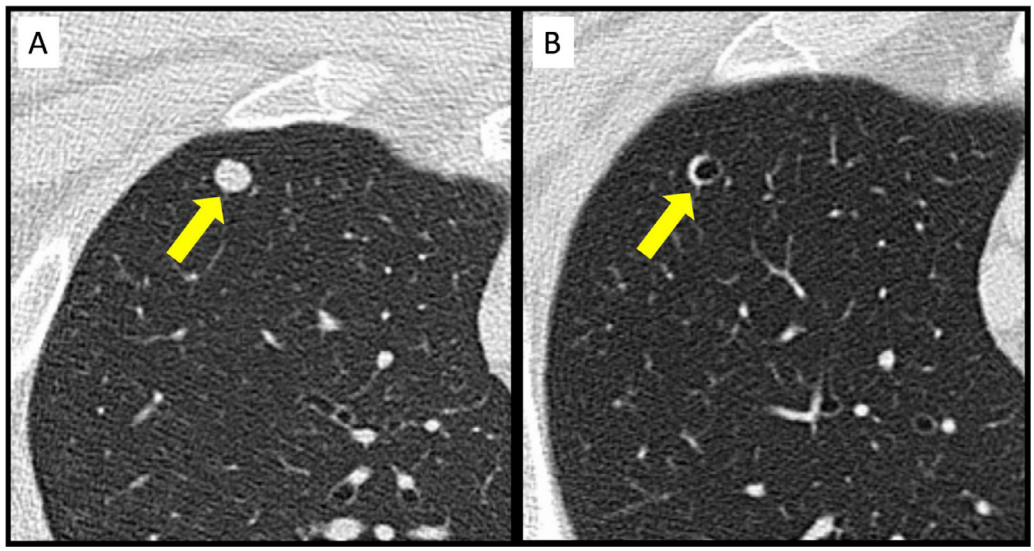

Fig. 4 – The nodule shows cystic change on follow-up (A: initial, B: 20 months later). The other nodules show no remarkable change (not shown).

area, however some nodules have located in the central area and shown cystic change.

In histopathology, BNCT consists of solid sheets of vacuolated cell and eosinophilic and some cystic spaces fill with colloid-like eosinophilic material. The nuclei are usually bland and mitotic figures are absent. In immunohistochemistry, both BNCT and chordoma are positive for brachyury that is a valuable marker for the diagnosis of notochordal-derived tumors. The differential diagnosis of BNCT and chordoma can be difficult due to overlaps of pathological findings between BNCTs and chordomas [11], especially in the lung, where there are few reports. Kikuchi et al. suggested that the previous some cases were possible to be actually not pulmonary chordomas but BNCTs [12,13]. Unlike axial BNCTs, pulmonary BNCTs are often accompanied by mucous retention and show cystic change, and these findings could be useful for differentiating BNCTs from chordomas. Indeed, in our case, some of pulmonary nodules showed cystic change on CT and might reflect the above pathological findings.

Although the differential diagnosis of BNCTs and chordomas may be difficult at initial examination, it is important to distinguish BNCTs from chordomas because of malignant behavior of chordomas but benignity of BNCTs. Recently, atypical notochordal cell tumor (ANCT) is described entity and may reflect a transition from BNCT to chordoma, however the criteria distinguishing ANCT from BNCT and chordoma are ill defined [11]. Because of the rarity of pulmonary BNCTs, further studies with more case series are required to clarify the relationship of these notochordal tumors originated in the lung.

Patient consent

Informed consent for the publication of this case report was obtained from the patient.

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