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CASE REPORT

Follicular dendritic cell sarcoma of the mediastinum: CT and ^{18}F-fluoro-2-deoxyglucose PET findings

Hyungjin Kim[1], Chang Min Park[1], Yoon Kyung Jeon[2], Jin Chul Paeng[3], Jin Mo Goo[1] & Hyun-Ju Lee[1]

1 Department of Radiology, Seoul National University College of Medicine, and Cancer Research Institute, Seoul National University, Seoul, Korea
2 Department of Pathology, Seoul National University College of Medicine, Seoul, Korea
3 Department of Nuclear Medicine, Seoul National University College of Medicine, Seoul, Korea

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Correspondence
Chang Min Park, Department of Radiology, Seoul National University Hospital, 101, Daehakno, Jongno-gu, Seoul 110-744, Korea.
Tel: +82 2 2072 0367
Fax: +82 2 743 7418
Email: cmpark@radiol.snu.ac.kr

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Abstract

Mediastinal follicular dendritic cell sarcoma (FDCS) is an exceedingly rare malignant tumor. We report, herein, detailed imaging findings of mediastinal FDCS, appearing as a large, well-circumscribed, strongly enhancing mass with central coarse calcification on computed tomography, with moderately increased metabolic activity on ^{18}F-fluoro-2-deoxyglucose positron emission tomography.

Introduction

Follicular dendritic cell sarcoma (FDCS) is a rare malignant tumor derived from follicular dendritic cells, which serve the function of antigen presentation, generation, and regulation of germinal center reactions.[1,2] The mediastinum is known to occupy only 8% of all FDCS locations,[3] although it is unclear whether the tumors are of lymph node origin or not.

We report detailed imaging features of FDCS including contrast-enhanced computed tomography (CT) and ^{18}F-fluoro-2-deoxyglucose positron emission tomography (FDG-PET) findings with review of recently reported cases of mediastinal FDCS.

Case report

A 68-year old man who had undergone right hemicolectomy for the treatment of ascending colon cancer two years ago, presented to our hospital for follow-up. On routine follow-up chest radiograph, a contour-bulging mass was found in the right paratracheal area (Fig. 1a). The patient was asymptomatic, and physical examination, as well as laboratory tests, were unremarkable. Tumor markers, such as the carcinoembryonic antigen, were also within normal limits. Contrast-enhanced chest CT (LightSpeed Ultra; GE healthcare, Milwaukee, WI) demonstrated a 4.5 cm well-circumscribed mass containing central coarse calcification in the right paratracheal area (Fig. 1b). This mass abutted the trachea, slightly compressing the tracheal wall, and showed strong contrast enhancement of 120 HU on contrast-enhanced CT (Fig. 1c). Considering the patient's history of colon cancer, contrast-enhanced abdomen CT (Brilliance 64; Philips Healthcare, DA Best, the Netherlands) was performed consecutively to evaluate local recurrence or intra-abdominal metastasis, however, no evidence of local recurrence or distant metastasis from colon cancer was observed. On FDG-PET scan (Gemini PET/CT; Philips Healthcare, DA Best, the Netherlands), this mediastinal mass showed increased metabolic activity with a maximum standardized uptake value (SUVmax) of 4.1 (Fig. 1d), with no other abnormal hypermetabolic lesions.

Under the impression of metastatic lymphadenopathy from colon cancer or Castleman disease, the patient underwent video-assisted thoracoscopic mass excision. Gross specimen

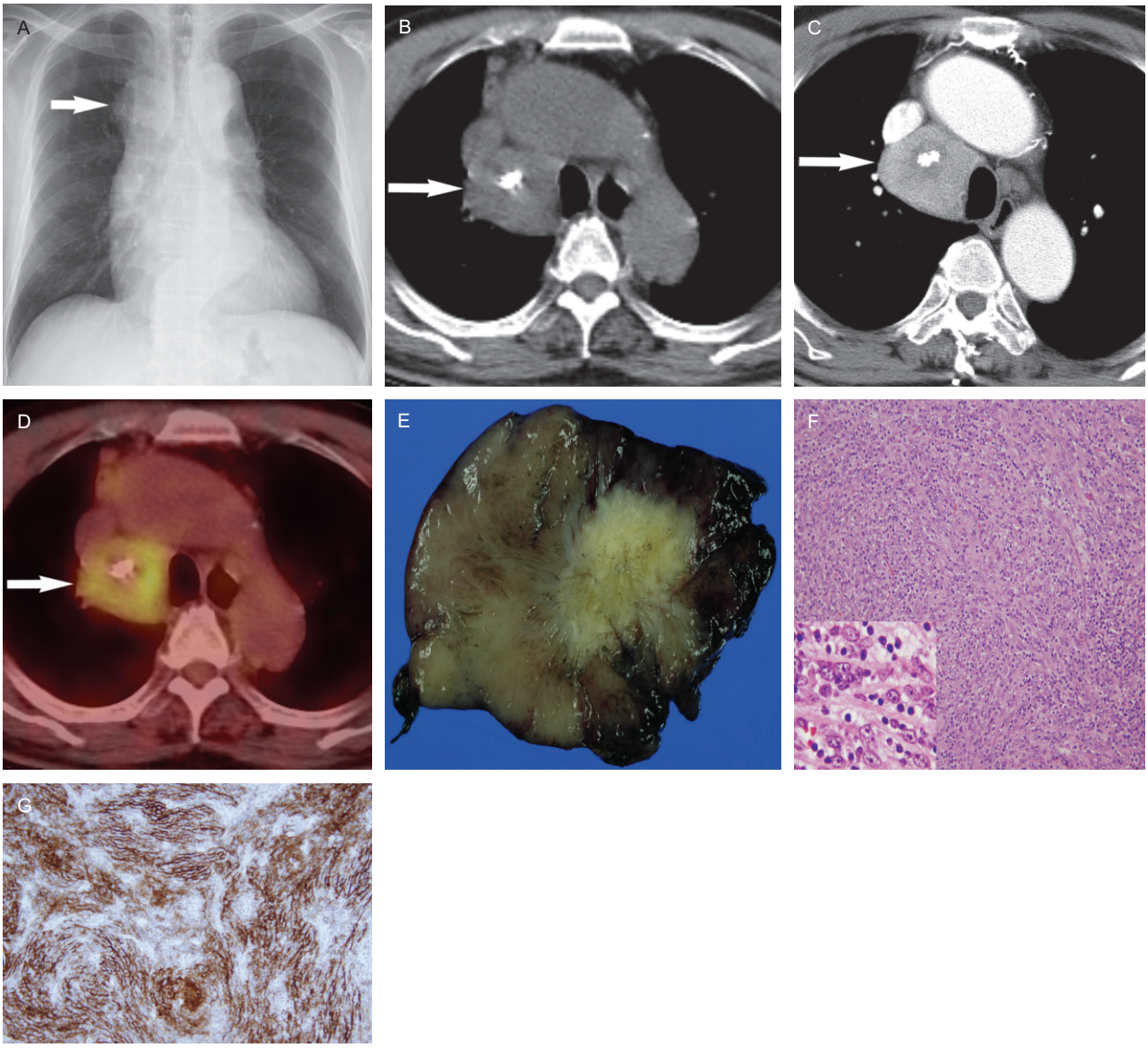

Figure 1 Follicular dendritic cell sarcoma of mediastinum in a 68-year-old man.
(**a**) Posteroanterior chest radiograph shows a right paratracheal mass (arrow).
(**b**) Non-contrast CT image with mediastinal window setting shows a right paratracheal mass (arrow) with a well-defined margin and lobulating contour. The mass contains central coarse calcification and is of soft-tissue-attenuation.
(**c**) Contrast-enhanced computed tomography (CT) image with mediastinal window setting shows homogeneous strong enhancement of the mass (arrow) of 120 HU.
(**d**) On ^{18}F-fluoro-2-deoxyglucose positron emission tomography (FDG-PET) examination, this mass shows increased glucose metabolism with a maximum standardized uptake value of 4.1 (arrow).
(**e**) On gross examination, the mass is encapsulated and has yellowish solid parenchyma with a central fibrotic portion.
(**f**) Microscopically, atypical spindle cells form fascicles with a whirling pattern, and are intermixed with many small lymphoid cells. (H-E staining, original magnification ×200); The neoplastic spindle cells have vesicular nuclei and small distinct nucleoli. (inset; H-E staining, original magnification ×1000)
(g) The spindle cells are positive for CD21 on immunohistochemistry (original magnification ×200).

revealed an encapsulated, yellowish, solid mass with a central fibrotic portion (Fig. 1e). Necrosis or hemorrhage was not evident. Microscopically, atypical spindle cells formed fascicles with a whirling pattern, and were intermixed with many small lymphoid cells (Fig. 1f). The spindle cells were positive for CD 21 on immunohistochemistry (Fig. 1g). These pathologic findings were diagnostic of FDCS with a hyaline-vascular Castleman disease background. On two-year follow-up, the

patient is alive without evidence of local recurrence or distant metastasis.

Discussion

FDCS was first described in a report of a non-lymphomatous primary lymph node malignancy in 1986 as a painless, slowly growing cervical lymphadenopathy, in most cases.[4] Thereafter, several case reports and case series regarding FDCS of nodal and various extranodal sites have been published.[1–8] However, its clinical and radiologic features have not been well established thus far due to the rarity of this tumor.

Mediastinal FDCS is even more uncommon and only constitutes 8% of all FDCS.[3] Leipsic *et al*.[3] first reported the non-contrast CT findings of mediastinal FDCS as a large, lobulating, soft-tissue-attenuated mass with coarse chunk-like calcification, similar to the findings in our case. They underlined the importance of chunk-like coarse calcification and suggested that FDCS can share some of the radiologic features of hyaline-vascular type Castleman disease,[3] in which calcification is typically coarse and centrally located.[9] Other recently reported cases of mediastinal FDCS[10–12] also showed central coarse calcification, which were arborizing or chunk-like, although a Castleman disease background was evident in only one case.[12] Thus, we can cautiously suspect that central coarse calcification may be one of the imaging findings of mediastinal FDCS, albeit with a possible imaging overlap between FDCS and Castleman disease. Calcification is known to often occur within the fibrous tissue, and central fibrosis has been recognized as an important feature of Castleman disease.[13] We assume that central fibrotic portion in FDCS can also cause calcification as in Castleman disease.

The consideration in the imaging overlap between FDCS and Castleman disease arose from a hypothesis that stromal cells in hyaline-vascular Castleman disease could transform into follicular dendritic cell neoplasms.[5] This hypothesis was later supported in a study by Chan *et al*.,[8] who presented the results of sequential biopsies of hyaline-vascular type Castleman disease, and showed the actual transformation of stromal cells in Castleman disease into FDCS. In our case, we were able to identify the Castleman disease background within the tumor, also supporting the hypothesis of Lin and Frizzera.[5]

In addition, mediastinal FDCS in our patient showed very strong enhancement on contrast-enhanced CT. Long-Hua *et al*. also described intense homogeneous enhancement of mediastinal FDCS.[11] However, we cannot hastily conclude, with only two case reports thus far, that mediastinal FDCS is particularly hypervascular. Moreover, previously reported cases of primary pulmonary[14] and intra-abdominal FDCS[7] did not show marked contrast enhancement. Further studies are needed to determine the vascularity of the disease.

The FDCS in our patient showed increased glucose metabolism with a SUVmax of 4.1 on FDG-PET examination. According to a report by Kröber *et al*., mediastinal FDCS showed only moderate uptake, indicating low glucose consumption, although the exact SUVmax value was not available.[6] However, another recent report showed a contradictory finding with a SUVmax of 11.4 in mediastinal FDCS.[10] Due to the rarity of this disease, little is known about the degree of glucose metabolism in FDCS. We assume that the degree of glucose metabolism in FDCS would be variable as FDCS has diverse histologies from low to high-grade histologic features.[15]

The differential diagnoses for the CT and FDG-PET findings in our case were metastatic lymphadenopathy and Castleman disease. Hyaline-vascular type Castleman disease would have been at the top of the list when only considering the radiologic features of this tumor. In this context, we do not believe that one can definitively diagnose FDCS with these radiologic findings. However, we do believe that clinical radiologists should be aware of the association between FDCS and Castleman disease, and the imaging features of mediastinal FDCS.

In this case report, we described the detailed imaging findings of mediastinal FDCS, appearing as a large, well-circumscribed, strongly enhancing mass with central coarse calcification on CT and moderately increased metabolic activity on FDG-PET.

Disclosure

No authors report any conflict of interest.

References

1 Perez-Ordonez B, Erlandson RA, Rosai J. Follicular dendritic cell tumor: report of 13 additional cases of a distinctive entity. *Am J Surg Pathol* 1996; **20**: 944–55.
2 Chan JK, Fletcher CD, Nayler SJ, Cooper K. Follicular dendritic cell sarcoma. Clinicopathologic analysis of 17 cases suggesting a malignant potential higher than currently recognized. *Cancer* 1997; **79**: 294–313.
3 Leipsic JA, McAdams HP, Sporn TA. Follicular dendritic cell sarcoma of the mediastinum. *AJR Am J Roentgenol* 2007; **188**: W554–6.
4 Monda L, Warnke R, Rosai J. A primary lymph node malignancy with features suggestive of dendritic reticulum cell differentiation. A report of 4 cases. *Am J Pathol* 1986; **122**: 562–72.
5 Lin O, Frizzera G. Angiomyoid and follicular dendritic cell proliferative lesions in Castleman's disease of hyaline-vascular type: a study of 10 cases. *Am J Surg Pathol* 1997; **21**: 1295–306.
6 Kröber SM, Marx A, Aebert H, Dohmen BM, Kaiserling E. Sarcoma of follicular dendritic cells in the dorsal mediastinum. *Hum Pathol* 2004; **35**: 259–63.

7 Kang TW, Lee SJ, Song HJ. Follicular dendritic cell sarcoma of the abdomen: the imaging findings. *Korean J Radiol* 2010; **11**: 239–43.
8 Chan ACL, Chan KW, Chan JKC, Au WY, Ho WK, Ng WM. Development of follicular dendritic cell sarcoma in hyaline-vascular Castleman's disease of the nasopharynx: tracing its evolution by sequential biopsies. *Histopathology* 2001; **38**: 510–18.
9 McAdams HP, Rosado-de-Christenson M, Fishback NF, Templeton PA. Castleman disease of the thorax: radiologic features with clinical and histopathologic correlation. *Radiology* 1998; **209**: 221–8.
10 Subesinghe M, Smith JT, Chowdhury FU. F-18 FDG PET/CT imaging of follicular dendritic cell sarcoma of the mediastinum. *Clin Nucl Med* 2012; **37**: 204–5.
11 Long-Hua Q, Qin X, Ya-Jia G, Jian W, Xiao-Yuan F. Imaging findings of follicular dendritic cell sarcoma: report of four cases. *Korean J Radiol* 2011; **12**: 122–8.
12 Choi SY, Kim JM. Follicular dendritic cell sarcoma arising in hyaline-vascular Castleman disease of mediastinum: a case report. *J Med Cases* 2011; **2**: 143–6.
13 Ota T, Mitsuyoshi A, Zaima M, Mizuno M, Morita R. Visualization of central stellate fibrosis in hyaline vascular type Castleman's disease. *Br J Radiol* 1997; **70**: 1060–2.
14 Kim H, Park CM, Jeon YK *et al*. Computed tomography and ^{18}F-fluoro-2-deoxyglucose positron emission tomography findings of primary pulmonary follicular dendritic cell sarcoma: case report and a literature review. *J Thorac Imaging* doi: 10.1097/RTI.0b013e318220309f. [Published Online].
15 Soriano AO, Thompson MA, Admirand JH *et al*. Follicular dendritic cell sarcoma: a report of 14 cases and a review of the literature. *Am J Hematol* 2007; **82**: 725–8.