

Follicular Dendritic Cell Sarcoma Arising in the Aortopulmonary Window: A Case Report of Complete Resection Using Proximal Pulmonary Artery Control

Nanako Yokota

Jichi Medical University

Akie Horikiri

Jichi Medical University

Yukina Niwa

Jichi Medical University

Takafumi Taki

Jichi Medical University

Yoshiaki Takase

Jichi Medical University

Yoshihiko Kanai

Jichi Medical University

Kihara Atsushi

Kimitsu Chuo Hospital

Toshiro Niki

Kotobiken Medical Laboratories (Japan)

Hiroyoshi Tsubochi

`h-tsubochi@omiya.jichi.ac.jp`

Jichi Medical University

Case Report

Keywords: Aortopulmonary window, Follicular dendritic cell sarcoma, Mediastinal tumor, Surgical resection, Thoracic surgery

Posted Date: October 18th, 2026

DOI: <https://doi.org/10.21203/rs.3.rs-10578992/v1>

License: © ⓘ This work is licensed under a Creative Commons Attribution 4.0 International License.
[Read Full License](#)

Additional Declarations: No competing interests reported.

Abstract

Background:

Follicular dendritic cell sarcoma is a rare malignant neoplasm, and its occurrence in the aortopulmonary window is exceptionally uncommon. Tumors in this location are technically challenging because of the limited operative space and their close relationship to the aortic arch, left pulmonary artery, left main bronchus, and left recurrent laryngeal nerve. We report a case of follicular dendritic cell sarcoma arising in the aortopulmonary window that was completely resected using a strategy for proximal pulmonary artery control.

Case presentation:

A 46-year-old man was referred to our institution after an abnormal mediastinal shadow was incidentally detected on chest radiography. Contrast-enhanced computed tomography showed a 48 × 29 mm well-circumscribed mass in the aortopulmonary window, broadly contacting the aorta, left pulmonary artery, and left main bronchus. Positron emission tomography showed moderate fluorodeoxyglucose uptake without distant metastasis. Endobronchial ultrasound-guided transbronchial needle aspiration suggested a neuroendocrine tumor but did not establish a definitive diagnosis. Surgical resection was performed through median sternotomy combined with a left anterior second intercostal thoracotomy. Because direct exposure and encirclement of the tumor-adjacent left pulmonary artery were considered hazardous, proximal vascular control was established through the transverse pericardial sinus before tumor manipulation. Division of the ligamentum arteriosum increased pulmonary artery mobility and improved exposure of the operative field. The tumor was completely separated from the left pulmonary artery, aorta, left main bronchus, and recurrent laryngeal nerve without combined organ resection. Histopathological examination showed spindle-shaped tumor cells arranged in whorled and rosette-like patterns. Immunohistochemical staining was positive for CD21, CD23, D2-40, and somatostatin receptor 2, confirming follicular dendritic cell sarcoma. The postoperative course was uneventful, and the patient remained free of recurrence 3 years after surgery.

Conclusions:

Complete resection of a tumor arising in the aortopulmonary window can be facilitated by secure proximal pulmonary artery control and mobilization of surrounding structures. These technical principles may also be useful for other tumors in this anatomically complex region when direct manipulation of the tumor-adjacent pulmonary artery is considered hazardous.

Background

Follicular dendritic cell sarcoma (FDCS) is a rare malignant neoplasm derived from follicular dendritic cells and was first described by Monda et al. in 1986 as a distinct pathological entity with dendritic reticulum cell differentiation [1]. Histologically, FDCS is characterized by spindle to ovoid tumor cells arranged in fascicular, storiform, or whorled patterns, often admixed with small lymphocytes, and immunohistochemically by the expression of follicular dendritic cell markers, including CD21, CD23, and CD35 [2].

Although FDCS most commonly arises in lymph nodes, approximately 30–40% of cases occur at extranodal sites, including the liver, spleen, gastrointestinal tract, and soft tissues [2, 3]. Mediastinal FDCS is particularly uncommon and is frequently misdiagnosed because of its rarity and nonspecific radiological appearance [4, 5]. Consequently, preoperative differentiation from thymic epithelial tumors, lymphoma, neurogenic tumors, and other mediastinal neoplasms is often difficult.

The aortopulmonary window (APW) is a narrow anatomical space bounded by the aortic arch, left pulmonary artery, and left main bronchus. Tumors arising in this region present unique diagnostic and surgical challenges because of their close relationship to major cardiovascular structures and the left recurrent laryngeal nerve [6]. To our knowledge, FDCS arising specifically within the APW has not been well documented, and the surgical management of this entity has rarely been described.

Herein, we report an extremely rare case of FDCS arising in the APW that was successfully treated by complete surgical resection. In addition to the clinicopathological findings, we highlight the surgical strategy used to obtain safe proximal control of the left pulmonary artery and discuss its potential applicability to other tumors arising in the APW.

Case Presentation

A 46-year-old man was referred to our institution after an incidental mediastinal shadow was detected on a routine chest radiograph. He had no significant past medical history and had a smoking history of 20 cigarettes per day for 22 years. He was asymptomatic, and physical examination revealed no abnormalities. Routine laboratory investigations, including serum soluble interleukin-2 receptor, alpha-fetoprotein, carcinoembryonic antigen, and cytokeratin 19 fragment, were within normal limits.

Contrast-enhanced computed tomography (CT) demonstrated a well-circumscribed 48 × 29 mm mass in the aortopulmonary window (APW), adjacent to the ascending aorta, left pulmonary artery (LPA), and left main bronchus (Fig. 1A–C). The tumor showed heterogeneous enhancement and was broadly attached to the LPA and adjacent great vessels, raising suspicion of vascular invasion. ¹⁸F-fluorodeoxyglucose positron emission tomography/CT demonstrated moderate uptake (maximum standardized uptake value, 5.4) without evidence of distant metastasis (Fig. 2).

Endobronchial ultrasound-guided transbronchial needle aspiration suggested a neuroendocrine tumor; however, a definitive diagnosis could not be established. Because malignancy could not be excluded, surgical resection was planned.

The operation was performed through a median sternotomy combined with a left anterior second intercostal thoracotomy. The tumor was broadly attached to the LPA and displaced it anteriorly. Although preoperative imaging suggested possible vascular invasion, the lesion appeared well encapsulated intraoperatively. Because the tumor obstructed direct access to the proximal LPA, direct encirclement of the tumor-adjacent segment of the artery was considered hazardous because of the risk of vascular injury. Therefore, proximal control of the LPA was established before tumor manipulation. After encircling the ascending aorta, the right pulmonary artery (RPA) was exposed between the ascending aorta and superior vena cava and encircled with a vascular tape. Subsequently, through the transverse pericardial sinus, the pulmonary artery trunk and ascending aorta were encircled en bloc. By sequentially passing the right limb of the tape beneath the RPA and then the ascending aorta, the proximal LPA was secured without direct manipulation of the tumor-adjacent arterial segment. Following division of the ligamentum arteriosum, gentle traction on the tape increased the mobility of the LPA and enabled careful stepwise dissection of the tumor from the pulmonary artery wall, extending from the pulmonary artery bifurcation to the distal LPA. Additional dissection was performed along the aortic side while preserving the left recurrent laryngeal nerve. The tumor was also separated completely from the left main bronchus without bronchial resection. Complete macroscopic resection was achieved. The operation time was 270 minutes, and the estimated blood loss was 700 mL.

Gross examination demonstrated a well-encapsulated tumor without extracapsular extension. Histologically, the lesion consisted of spindle-shaped cells arranged in whorled and rosette-like patterns admixed with small lymphocytes (Fig. 3A). Immunohistochemical analysis demonstrated positivity for CD21, CD23, D2-40, and somatostatin receptor 2 (SSTR2), confirming the diagnosis of follicular dendritic cell sarcoma (Fig. 3B–E). The Ki-67 labeling index was approximately 20–30%.

The postoperative course was uneventful, and the patient was discharged on postoperative day 12. He remains alive without evidence of recurrence 36 months after surgery.

Discussion

FDCS is an uncommon malignant tumor with intermediate-grade behavior, most frequently arising in lymph nodes of the head and neck region [2, 3]. Extranodal FDCS accounts for approximately one-third of reported cases and has been described in a wide variety of anatomical locations [2]. Mediastinal FDCS is rare and often underrecognized, contributing to diagnostic delay or misdiagnosis [4, 5]. In the present case, the preoperative biopsy findings suggested a neuroendocrine tumor, illustrating the diagnostic limitations of small biopsy specimens in this uncommon neoplasm.

Radiologically, FDCS typically appears as a well-defined soft tissue mass with heterogeneous enhancement [4], findings that overlap with those of thymoma, lymphoma, neurogenic tumors, and other mediastinal neoplasms. Histopathological examination combined with immunohistochemical analysis therefore remains essential for definitive diagnosis. Classical follicular dendritic cell markers, including CD21, CD23, and CD35, are considered diagnostic hallmarks [2]. More recently, SSTR2 has emerged as a

sensitive and specific marker for follicular dendritic cells and their related tumors [7]. In the present case, expression of CD21, CD23, D2-40, and SSTR2 supported the diagnosis of FDCS and helped distinguish it from neuroendocrine and other spindle cell tumors.

Complete surgical resection is regarded as the cornerstone of treatment for localized FDCS. Previous studies have demonstrated that incomplete resection is a significant risk factor for local recurrence and poor prognosis [4]. A clinically relevant comparison can be made with a previously reported mediastinal FDCS case by Hu et al., in which surgical resection was not feasible and radiotherapy alone was administered [8]. That patient developed distant metastases and died within one year, highlighting the potential aggressiveness of FDCS and the limitations of non-surgical treatment.

Complete surgical resection is generally regarded as the cornerstone of treatment for localized FDCS. Previous studies have suggested that incomplete resection is associated with an increased risk of local recurrence and unfavorable outcome [4]. A clinically relevant comparison can be made with the previously reported mediastinal FDCS cases described by Hu et al. [8]. One patient in whom surgical resection was not feasible and radiotherapy alone was administered developed distant metastases and died within one year. Although the clinical behavior of FDCS is heterogeneous and firm conclusions cannot be drawn from individual cases, these findings support attempting complete resection whenever technically and oncologically feasible.

The APW presents distinct surgical challenges that are not encountered to the same degree in more accessible mediastinal compartments. It is a narrow anatomical space bordered by the aortic arch superiorly, the left pulmonary artery inferiorly, and the left main bronchus posteriorly. The left recurrent laryngeal nerve and ligamentum arteriosum are also closely related to this region. Consequently, even a well-circumscribed tumor may restrict visualization and access to the adjacent pulmonary artery, while traction or blind dissection may result in major vascular injury or recurrent laryngeal nerve damage. Furthermore, when the tumor broadly contacts the left pulmonary artery, direct dissection around the vessel for proximal taping may itself be hazardous because the presence or absence of vascular invasion cannot always be reliably determined preoperatively.

In the present case, median sternotomy provided access to the central great vessels, while an additional left anterior second intercostal thoracotomy improved lateral exposure of the APW. This combined approach allowed visualization of both the proximal vascular structures and the distal aspect of the tumor. However, because the tumor broadly contacted and displaced the left pulmonary artery, direct exposure and encirclement of the tumor-adjacent segment were considered unsafe. We therefore established proximal control of the left pulmonary artery before manipulating the tumor. After encircling the ascending aorta and right pulmonary artery, a tape was passed through the transverse pericardial sinus and sequentially beneath the right pulmonary artery and ascending aorta, allowing the left pulmonary artery to be secured at its most proximal portion without direct handling of the segment adjacent to the tumor.

This strategy provided several advantages. First, it allowed immediate vascular control in the event of pulmonary artery injury during tumor dissection. Second, gentle traction on the tape helped define the course of the proximal left pulmonary artery and improved orientation within the confined APW. Third, proximal control could be obtained before determining whether the apparent tumor–vessel interface represented simple contact, adhesion, or true invasion. These advantages were particularly important because the preoperative CT findings suggested extensive contact with major vascular structures and did not completely exclude invasion. The tumor was ultimately found to be encapsulated and separable from the pulmonary artery, but this could only be confirmed safely after vascular control had been secured.

Division of the ligamentum arteriosum was another important component of the procedure. This maneuver increased the mobility of the left pulmonary artery and widened the operative space between the aortic arch and pulmonary artery, facilitating stepwise dissection from the pulmonary artery bifurcation toward the distal left pulmonary artery. It also improved access to the aortic side of the tumor, where meticulous dissection was required to preserve the left recurrent laryngeal nerve. Thus, the operative strategy involved not only obtaining proximal vascular control but also mobilizing the anatomical boundaries of the APW to create a safer and more accessible dissection plane.

Although the present tumor was an FDCCS, these technical principles are not disease-specific. Similar difficulties may be encountered with other tumors arising in or extending into the APW. When such a tumor broadly contacts the left pulmonary artery and direct encirclement is considered unsafe, proximal control through the transverse pericardial sinus may provide an alternative route for securing the vessel. Likewise, division of the ligamentum arteriosum may improve pulmonary artery mobility and exposure in selected cases. The indication for these maneuvers should nevertheless be individualized according to tumor location, suspected vascular invasion, and the feasibility of obtaining safe dissection planes. If true arterial invasion is strongly suspected, preparation for pulmonary artery resection and reconstruction, with cardiopulmonary support available when necessary, should also be considered.

The role of adjuvant therapy in FDCCS remains controversial. Chemotherapy and radiotherapy have been used in selected patients, particularly for unresectable tumors, positive margins, recurrent disease, or tumors with aggressive pathological features; however, outcomes have been inconsistent [7]. In the present case, no adjuvant therapy was administered because complete macroscopic and pathological resection was achieved and no extracapsular invasion was identified. Nevertheless, the Ki-67 labeling index was approximately 20–30%, and FDCCS may recur after a prolonged interval. Long-term surveillance is therefore warranted despite the absence of recurrence during 3 years of follow-up.

This case is notable not only because of the rare pathological diagnosis but also because it illustrates a reproducible surgical strategy for tumors in the APW. Appropriate selection of the surgical approach, establishment of proximal pulmonary artery control before tumor manipulation, division of the ligamentum arteriosum to improve mobility, and careful identification of the recurrent laryngeal nerve

collectively enabled complete resection without vascular or neurological complications. These principles may contribute to safer resection of other tumors located in this anatomically complex region.

Conclusions

FDCS arising in the APW is exceedingly rare. Tumors in this region pose specific surgical risks because of the limited operative space and close relationship to the aortic arch, left pulmonary artery, left main bronchus, and left recurrent laryngeal nerve. In the present case, a combined surgical approach, proximal control of the left pulmonary artery through the transverse pericardial sinus, and division of the ligamentum arteriosum enabled safe and complete resection. These technical principles may also be applicable to other APW tumors in which direct manipulation of the tumor-adjacent pulmonary artery is considered hazardous.

Declarations

Ethics approval and consent to participate

Written informed consent was obtained from the patient.

Approval by the Institutional Review Board of Jichi Medical University was waived due to the nature of this single case report.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Funding

None.

Conflicts of interest / Competing interests

The authors declare that they have no competing interests.

Data availability

No new data were created or analyzed in this study. Data sharing is not applicable.

Authors' contributions

NY and HT conceptualized the study.

NY drafted the initial manuscript.

HT critically revised the manuscript for important intellectual content.

All authors contributed to data acquisition and interpretation, read and approved the final version of the manuscript.

References

1. Monda L, Warnke R, Rosai J. A primary lymph node malignancy with features suggestive of dendritic reticulum cell differentiation. *Am J Pathol.* 1986;122:562–72.
2. 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.
3. 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.
4. Wu YL, Wu F, Xu CP, Chen GL, Zhang Y, Chen W, et al. Mediastinal follicular dendritic cell sarcoma: a rare, potentially under-recognized, and often misdiagnosed disease. *Diagn Pathol.* 2019;14:5.
5. Perez-Ordóñez 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.
6. Jolles PR, Shin MS, Jones WP. Aortopulmonary window lesions: detection with chest radiography. *Radiology.* 1986;159:647–51.
7. Tao LL, Huang YH, Chen YL, Yu GY, Yin WH. SSTR2a Is a Useful Diagnostic Marker for Follicular Dendritic Cells and Their Related Tumors. *Am J Surg Pathol.* 2019;43:374–81.
8. Hu J, Dong D, Jiang Z, Hu H. Clinicopathological characteristics of mediastinal follicular dendritic cell sarcoma: report of three cases. *J Cardiothorac Surg.* 2016;11:56.

Figures

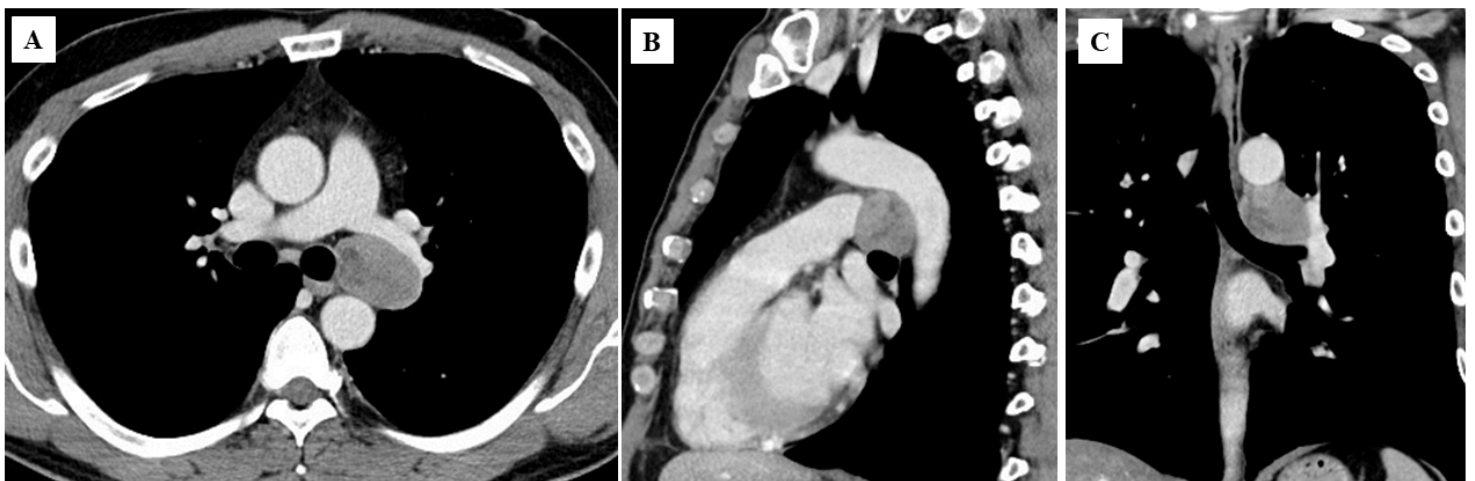


Figure 1

(A, B, C) Contrast-enhanced CT scan revealed a 48 × 29 mm mass surrounded by the aorta, left pulmonary artery, and left main bronchus. The lesion had well-defined borders, extensively contacting the aorta, left pulmonary artery, and left main bronchus, and contained a slightly heterogeneous area of mildly low attenuation.

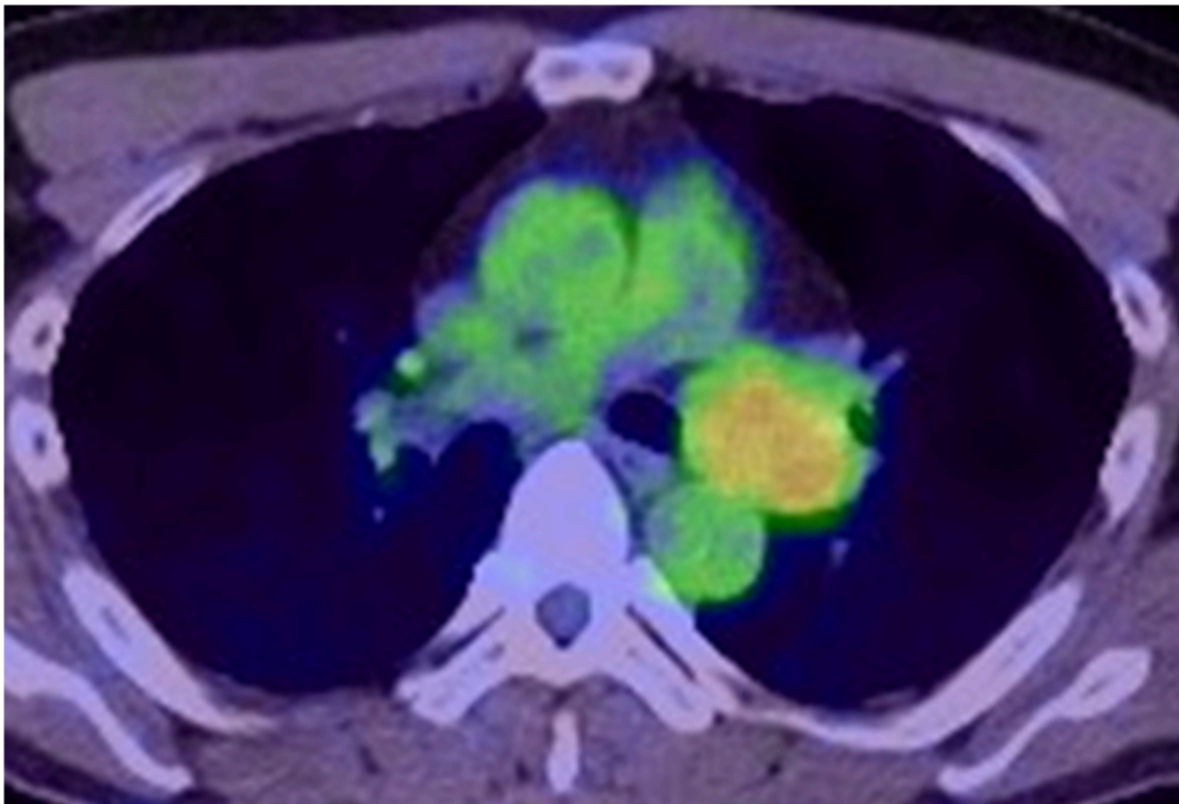


Figure 2

FDG-PET revealed uptake in the nodule with an SUVmax of 5.40.

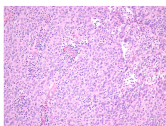


Figure 3(A)



Figure 3(B)

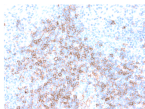


Figure 3(C)

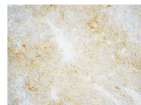


Figure 3(D)

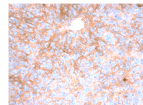


Figure 3(E)

Figure 3

Pathological examinations. (A) Microscopic examination (hematoxylin-eosin staining, original magnification: ×400) revealed dense proliferation of round or spindle-shaped cells, along with indistinct rosette-like structures in some areas. Immunohistochemical staining revealed membranous and cytoplasmic positivity in the tumor cells for CD21 (B) (original magnification: ×200), CD23 (C) (original

magnification: $\times 200$), D2-40 (D) (original magnification: $\times 200$), and SSTR2 (E) (original magnification: $\times 400$)