



Metastatic Cranial Solitary Fibrous Tumor to the Left Pulmonary Vein and the Left Atrium

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Abstract

Solitary fibrous tumors are a class of mesenchymal neoplasms with rare tendency to undergo malignant transformation. Lesions which have formerly been known as hemangiopericytoma bear exactly the same gene fusion as solitary fibrous tumor and may give rise to distant metastasis many years after. Hilar tumors with atrial involvement are complex tumors which require extensive surgery. Three dimensional models may guide surgical planning in advanced cases. We present a clinical case and surgical approach to malignant solitary fibrous tumor metastatic to the left hilum presenting many years after successful resection of intra-cranial hemangiopericytoma using three dimensional modeling for surgical planning.

Introduction

Solitary Fibrous Tumor (SFT) is group of mesenchymal neoplasms most often involving the pleura, with rare malignant transformation tendency [1]. Lesions diagnosed as Hemangiopericytomas (HPC) bear exactly the same gene fusion as SFT and may give rise to distant metastasis, often many years after. We describe a clinical case and surgical approach to malignant SFT to the left hilum presenting many years after successful resection of Intra-Cranial Hemangiopericytoma (ICHPC).

Case Presentation

After multiple episodes of dyspnea and vague chest discomfort, a 67 years old female presented for evaluation. She had resection of ICHPC 27 years earlier, with repeat resection for local recurrence, followed by cranial irradiation 4 years after her initial surgery.

Chest imaging which included Computed tomography and Magnetic resonance imaging demonstrated heterogeneous lobulated left hilar mass with progressive enhancement extending inferiorly across the interlobar fissure, extending and invading into the inferior Left Pulmonary Vein (LIPV) and the Left Atrium (LA) without invasion to the Left Main Pulmonary Artery (LMPA) (Figure 1A). Echocardiogram demonstrated 20X17mm mass in the LA emanating from the inferior pulmonary vein with preserved PA pressure and LV function. Computed tomography guided core needle biopsy of the hilar mass revealed Mesenchymal neoplasm positive for CD34, BCL-2 and negative for AE1/3, S100 and SMA; compatible with SFT.

The patient underwent extensive pre-operative work up which included Coronary angiography, Carotid Duplex, Trans-esophageal echocardiogram, complete pulmonary function testing and split perfusion ventilation scan and was found to be fit for combined pulmonary and cardiac resection. Distant disease was further excluded by Positron Emission tomography. After discussion at multidisciplinary tumor board; the patient was planned for en-bloc left pneumonectomy with partial left atrial resection via trans-sternal approach with Cardio Pulmonary Bypass (CPB). Detailed Three dimensional printed models were done for preoperative planning in order to assess resectability, evaluate intimate relation to the hilar structures and to guide the surgical approach (Figure 1B).

Surgery was commenced with median sternotomy. After hilar exposure, the LMPA was ligated first and divided intra pericardially followed by CPB initiation via aortic and dual stage venous cannulation with ante grade cardioplegia to avoid systemic tumor embolization from initial direct manipulation of the LA and LPV tumor. This allows the exposure and division of the left main stem bronchus at the level of the carina followed by exposure and division of the LSPV. Lastly, we turned to manipulate and dissect the tumor. The LIPV was opened with Left atriotomy allowing en-block resection of the atrial mass with the left lung (Figure 2). The atrial defect was reconstructed

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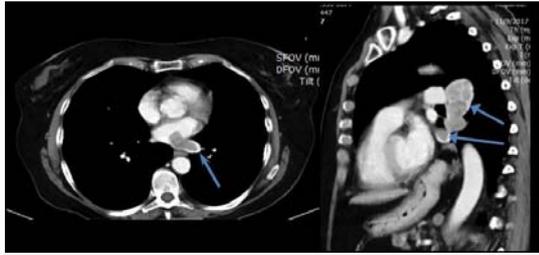


Figure 1A: Computed tomography of the patient, arrows demonstrating the hilar mass.



Figure 1B: Three dimensional models designed for this case without the atrial wall showing the tumor relations to the hilar structures (tumor marked in black).

with previously harvested autologous pericardium. The patient was then rewarmed and easily weaned from the CPB. The patient was extubated the following day and did well in the early post-operative period. Unfortunately, following transfusion of packed RBC in the week following surgery, the patient developed severe Transfusion related acute lung injury which resulted in severe respiratory failure and prompted initiation of veno-venous ECMO. The rest of the patient's hospital course was significant for acute renal failure and sepsis from which the patient did not recover and eventually expired. Pathology showed high-grade spindle and epithelioid cell neoplasm present in the LIPV extending into the LA grossly involving the adjacent parenchyma. Immunohistochemistry was positive for CD34, BCL2 and STAT6 and negative for MelA, desmin, SMA, CD31, s100 and AE1/3. 2/29 lymph nodes completely replaced by tumor with high mitotic rate. Features those were consistent with malignant SFT.

Discussion

SFTs are rare mesenchymal neoplasms frequently appearing in the pleura, meninges and soft tissues. Although historically HPC and SFTs were considered 2 separate entities, recent discovery of mutual underlying inversion on chromosome 12q13 resulting in activation of STAT6 fusion gene recognized them as a single entity [2]. SFTs are mostly benign neoplasms that rarely metastasize. Although there are no definite criteria for malignancy, WHO classification recognized moderate cellular atypia, hypercellularity, >4 mitoses per 10 high power field, necrosis and gross infiltrative margins as signs of malignancy [3]. Our patient presented many years following successful resection of ICHPC with new diagnosis of highly aggressive malignant SFT involving the left hilar structures. Although the initial slides of her previously resected tumor were unavailable for comparison, the current tumor cellular characteristics and immunopositivity for CD34 and STAT6 led us to believe that



Figure 2A: Left atrial appendage resected En-block with the left lung. Note the trans-hilar extension of the tumor (yellow line) from the inferior pulmonary vein to the left atrium.

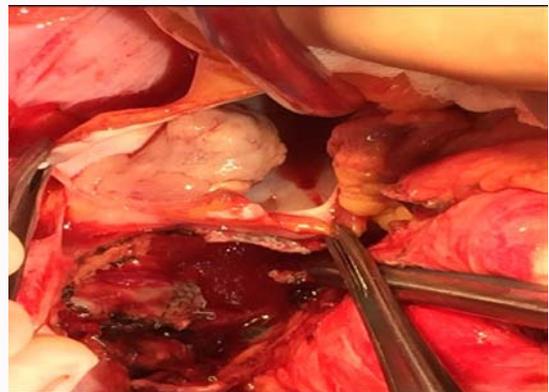


Figure 2B: Left atrium opened demonstrating the left atrial tumor extension.

this is probably a metastatic slowly growing SFT presumably from the previous ICHPC rather than second primary neoplasm.

Left atrial HPC are extremely rare entity. The clinical presentation is usually of dyspnea in the presence of atrial mass which mandated emergent surgery [4]. Given the rarity of malignant HPC with cardiac involvement, limited data is available on neither its oncologic outcome nor the benefit of using neo adjuvant or adjuvant chemotherapy or radiation therapy. Although ligation of pulmonary vein first technique was shown to have less systemic spread in non-CPB bronchoalveolar cancer pulmonary resections, the presence of LA and pulmonary vein mass and the need for CPB prompt us for early ligation of the LMPA and initiation of CPB prior to tumor manipulation, followed by wide en-block resection of the cardiac mass with the lung in-order to avoid shed of tumor cells to the systemic circulation.

Finally, the complexity of combined pulmonary and cardiac resection necessitates team approach with meticulous surgical planning. The advancement of preoperative imaging and three dimensional printed models has gained wide acceptance and enhanced surgical planning [5]. We recommend the use of these models for every complex cases especially if involves highly vitalized structures.

In summary, although limited data available for the proper management of malignant SFT metastatic to the hilum and the heart, surgical resection should be offered when feasible for cure or to avoid emergent hemodynamic collapse in case of cardiac

outflow tract obstruction. Meticulous preoperative planning using all available modalities including printed 3D models should be done and limited tumor manipulation prior to the initiation of CPB should be practiced. To our knowledge this is the first report of combined atrial resection with left pneumonectomy for SFT.

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