The International Journal of Cardiovascular Imaging (2019) 35:1429–1433
https://doi.org/10.1007/s10554-018-1489-8

REVIEW PAPER

Imaging and Surgical Treatment of Primary Pulmonary Artery Sarcoma

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Received: 6 July 2018 / Accepted: 30 October 2018 / Published online: 8 December 2018

Abstract
Primary pulmonary artery sarcoma is a rare tumor originating from the pulmonary artery tree. Given the low incidence, few centers have reported on more than a handful of cases. Because of its rarity it is also commonly misdiagnosed as pulmonary embolism. Multi-modality diagnostic imaging and recognition of specific imaging characteristics along with a high index of suspicion is required to make the correct diagnosis and expedite treatment. The primary imaging modality for most cardiac tumors such as primary pulmonary artery sarcoma is now MRI. It provides superb spatial resolution as well as functional assessment of the heart and pulmonary circulation. CT imaging also is part of routing imaging and remains as the most pertinent imaging modality to evaluate the lung parenchyma and presence of metastatic disease. Here we review the pertinent imaging modalities and tissue characteristics that facilitate recognition of primary pulmonary artery sarcoma. We also provide a short overview of surgical resection and reconstruction, which is the mainstay therapy, for this rare tumor.

Keywords primary pulmonary artery sarcoma · Cardiac tumor · Surgical treatment · Imaging · Cardiac MRI

Introduction

Primary pulmonary artery sarcoma is a rare entity and part of the larger group of primary cardiac sarcomas making up roughly 15% of all primary cardiac tumors. To date only about 300 cases of primary pulmonary sarcoma have been described in the medical literature. The clinical presentation of these patients is determined primarily by tumor size and anatomic location. Symptoms may include constitutional changes, evidence of cardiac obstruction, distal embolization, and other cardiopulmonary manifestations such as atrial or ventricular tachyarrhythmia, conduction abnormalities, pericardial effusion and cardiac tamponade. As with all other cancers, symptoms resulting from local invasion and distant metastases may also be present and vary with the extent and anatomic location of disease.The mainstay therapy for sarcoma is a surgical resection, and in the case of primary pulmonary artery sarcoma removal of the affected pulmonary artery segment in its entirety with reconstruction (Fig. 1). We have previously reported on a classification system and proposed staging system of primary pulmonary artery sarcoma that determines the leading symptoms, urgency of intervention and available surgical options based on the anatomic distribution of sarcoma in the pulmonary artery [1].

Appropriate initial diagnosis and precise definition of tumor burden primarily rely on imaging. Imaging modalities available for diagnosis of primary pulmonary sarcoma include echocardiography, computed tomography (CT) and magnetic resonance imaging (MRI). A multidisciplinary cardiac tumor team review is critical, and patients with primary pulmonary artery sarcoma generally benefit from referral to centers of excellence.

Diagnostic imaging

The main diagnostic imaging modalities are echocardiography including transthoracic (TTE), transesophageal (TEE) and intracardiac echocardiography (ICE), gated, multislice CT and MRI. In some patients, further insights can be gained by using supplemental diagnostic modalities such

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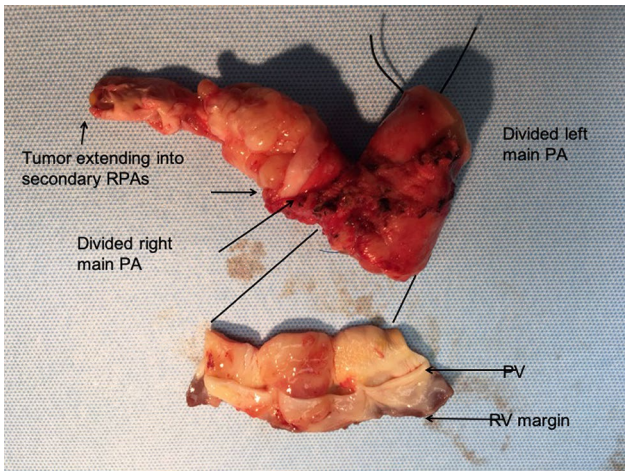

Fig. 1 Surgical specimen from pulmonary artery sarcoma resection. Surgical specimen from a pulmonary artery sarcoma extending from the pulmonary valve to the both main pulmonary arteries and the segmental arteries on the right side. Wide surgical resection can be done safely at experienced centers and is the treatment of choice. Complex reconstruction is often required in these cases

as coronary angiography, and positron emission tomography (PET) (Fig. 2). With advances in technology cardiac MRI has quickly become the preferred imaging modality for primary pulmonary artery sarcoma. In addition to detailed anatomical characterization, cardiac MRI allows for tissue characterization, providing insight into the tumor type and distinguishing from pulmonary embolism, a commonly mistaken diagnosis [2–4].

While no single feature is pathognomonic of malignancy, MRI can leverage different sequences and protocols for detailed tissue characterization (Fig. 3). Pulmonary sarcoma will typically enhance with gadolinium contrast compared to thrombus in the PAs and the degree of enhancement correlates with tumor differentiation. This feature is also particularly helpful during follow up and to evaluate for locoregional recurrence. Spatial resolution is superior to CT and functional assessment is comparable to echocardiography while also overcoming the limitation of acoustic windows encountered with echocardiography. Finally, functional assessment such as flow and gradients across the pulmonary artery vasculature can aid in diagnosing pulmonary sarcoma [5–8].

Multi-slice gated CT remains a useful diagnostic tool that can complement MRI and will provide better resolution for imaging of the lungs. It is also commonly the initial imaging study when other diagnoses may be suspected such as pulmonary embolism. The characteristic finding on CT imaging consists of hyperdense lesions with non-homogenous attenuation from hemorrhage, a beaded appearance of the pulmonary arteries, filling defects within the PAs that have soft-tissue characteristics with distal oligemia, vascular distension and extravascular spread [5, 9, 10] (Fig. 2). PET with fluorodeoxyglucose (18FDG) is used quite commonly for staging purposes in other tumors. This applies for staging purposes in pulmonary artery sarcoma as well; however, tumor burden in close proximity to the RV may be confounded by the frequent high uptake of FDG in the myocardium. Echocardiography (TTE, TEE, ICE) is most helpful for the evaluation of the right ventricular outflow track, pulmonary valve if there is involvement of the proximal pulmonary artery [11–13].

Surgical treatment of primary pulmonary artery sarcoma

Over the last two decades we have operated on 18 patients with primary pulmonary artery sarcoma with a predominance of male patients (80%) and average age of 57 years (range 38–84 years). These patients usually present with symptoms of dyspnea, cough, hemoptysis, or chest pain. The similarity of presenting symptoms along with common imaging algorithms not infrequently leads to an erroneous diagnosis of pulmonary embolism. This may lead to treatment with anticoagulation that often delays definitive diagnosis and treatment of the tumor [1, 14, 15]. Chemotherapy alone typically has poor outcomes [16]. Treatment approaches for primary pulmonary artery sarcoma have included palliative pulmonary artery stenting, pneumonectomy, debulking, tumor endarterectomy and wide surgical excision.

We have shown that wide excision that often includes the pulmonary trunk (Fig. 1) can be safely done and improves survival when tumor biology is favorable [1]. Benefit is gained with neoadjuvant chemotherapy in the appropriately selected patient and this is now our standard approach to all patients with primary pulmonary sarcoma. Initial chemotherapy may permit tumor shrinkage, enhance resectability, and potentially neutralize micrometastatic disease. Most importantly, neoadjuvant chemotherapy will also test tumor biology and identify responders, that benefit most from an aggressive multimodality treatment approach [1]. Patients that have progression of disease in spite of neoadjuvant chemotherapy are not considered surgical candidates as their prognosis is poor regardless of further intervention including aggressive surgical resection. Tissue diagnosis is important to select the appropriate chemotherapeutical regiment and rule out other diagnoses that may present with a similar picture as primary pulmonary sarcoma occasionally. Definitive tissue diagnosis can often be obtained by right heart catheterization biopsy or video-assisted thoracic surgery. Doxorubicin and ifosfamide are the initial drugs of choice for neoadjuvant chemotherapy [17].

In our experience a minority of patients had received preoperative chemotherapy (n = 4), radiation therapy (n = 1) or combination thereof (n = 2). 5 (30%) patients presented with

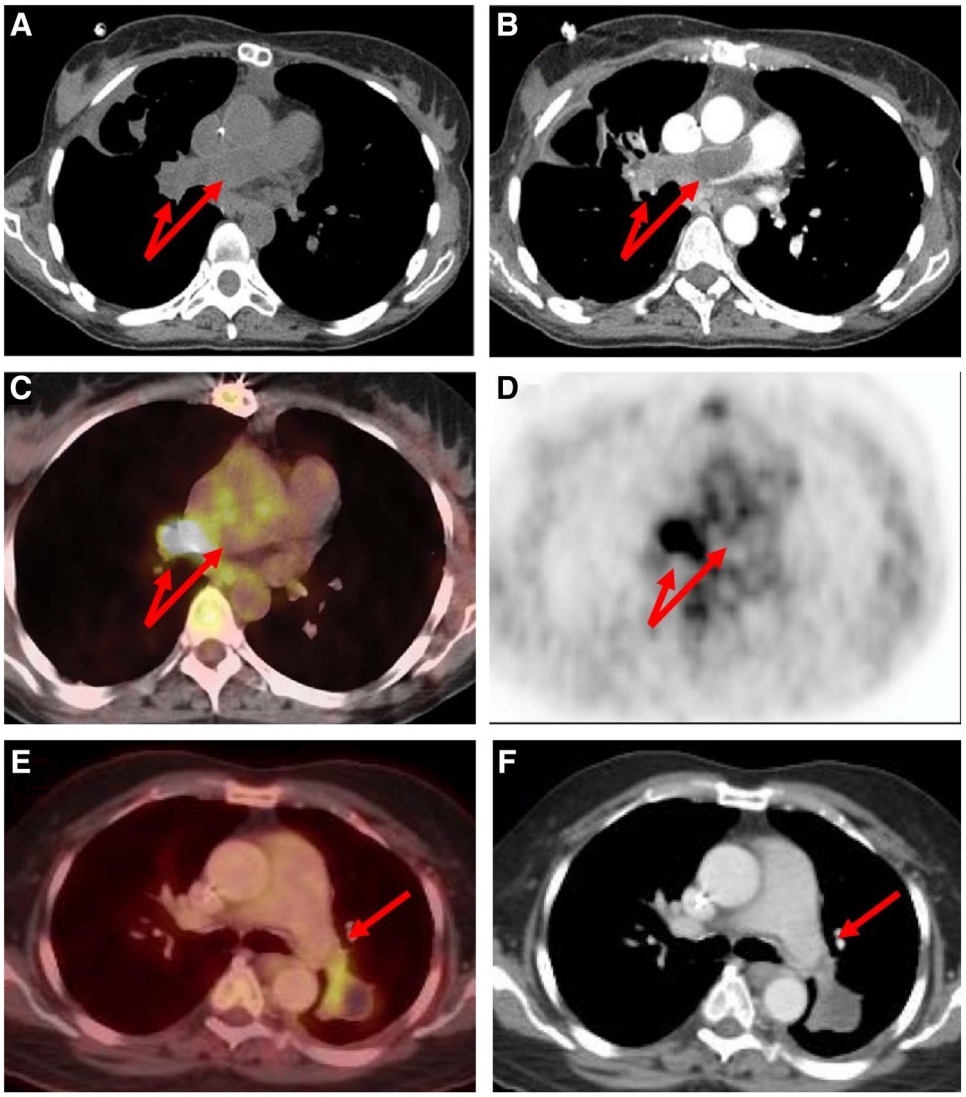

Fig. 2 Computed tomography and PET scan appearance of pulmonary artery sarcoma. Representative computed tomography and PET scan findings from two patients are shown. Panel **a**–**d** show cross-sectional images from PET/CT without (**a**) and with (**b**) contrast. The central PA mass could easily be mistaken for a large pulmonary embolism and is characterized by oligemia of the right lung. The contrast enhanced CT scan further delineates the pulmonary sarcoma with heterogeneous enhancement in and around the distal PA, which is also PET positive (**c, d**). Superimposed thrombus formation due to subtotal occlusion of the right pulmonary artery is also seen. **e, f** Shows similar imaging findings from a different patient with an occlusive left-sided pulmonary artery sarcoma

recurrent disease in the pulmonary artery after undergoing surgery, which commonly consisted of a pulmonary endarterectomy rather than resection of the affected pulmonary artery segment in its entirety.

In our opinion, this observational evidence underlines the importance of a complete, i.e. full thickness resection, rather than an endarterectomy, which may not result in a R0 result. Our surgical approach typically consists of a resection of the affected main pulmonary artery, including the left and right main pulmonary artery depending on the extent of involvement. A Dacron graft is most often used for reconstruction, although pulmonary artery allograft offers an excellent alternative conduit for reconstruction especially when there is involvement of the proximal pulmonary artery and valve. In select cases with involvement of the distal pulmonary artery, a pneumonectomy may offer the only option of complete resection. Patients for this aggressive approach to resection must be selected extremely carefully. For obvious reasons, patient that have disease extending beyond the main pulmonary artery on both sides are not candidates for resection. And although demographics and comorbidities tend to be different from patients with NSCLC undergoing pneumonectomy, the physiological changes of a pneumonectomy can vary depending on the extent of obstruction secondary to the sarcoma. If a patient presents with a complete or high-grade obstruction of the pulmonary circulation, then a 'functional' pneumonectomy has already occurred and changes in right ventricular strain, oxygenation or ventilation may be minimal after pneumonectomy. However, a diligent evaluation of both right-ventricular function and pulmonary reserve are always critical in this setting. This is particularly true if patients previously have had chemo and/or radiation therapy.

At our institution we have adopted a stepwise approach to the patient with primary pulmonary sarcoma requiring a pneumonectomy for extension of tumor into the lungs and

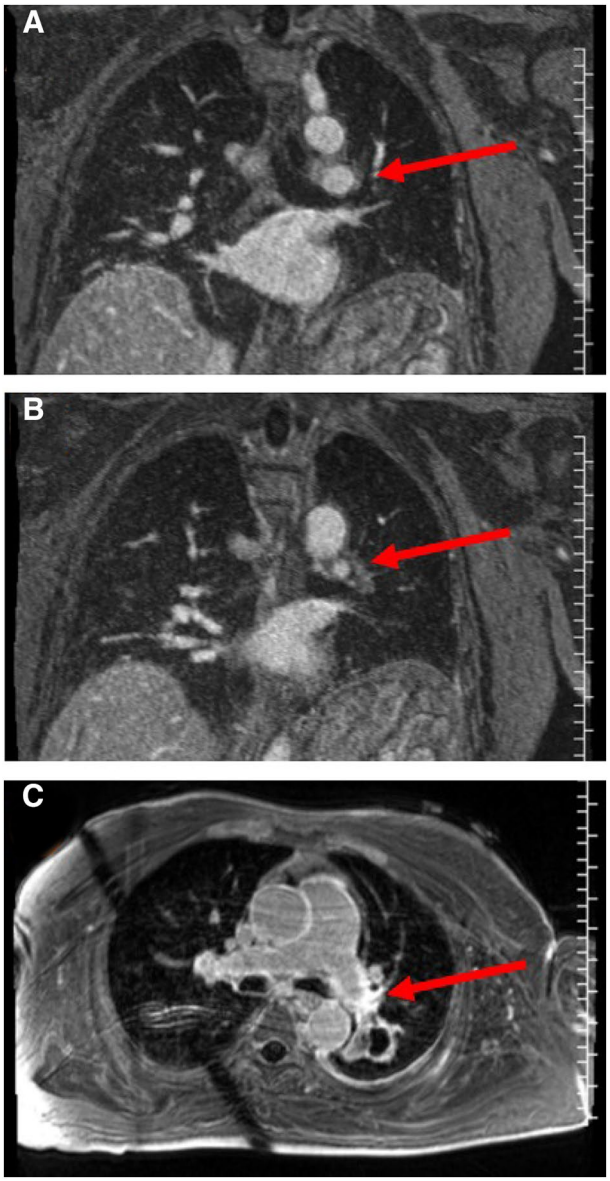

Fig. 3 MRI appearance of pulmonary artery sarcoma. ECG-gated MRI scan of the heart and great vessels demonstrates an occlusive left-sided pulmonary sarcoma. This is the same patient as in Fig. 2e, f. The spatial resolution of the MRI highlights the gradual narrowing of the L main pulmonary artery with a significant increase in wall thickness, representing the sarcoma (**a, b**). **c** Narrowing of the lumen and eventual occlusion along with wall thickening in the L pulmonary artery

lobar pulmonary artery. In a first procedure the main PA and contralateral pulmonary artery are resected and reconstructed as indicated. In a second procedure, the pneumonectomy is completed. We started utilizing this approach after observing a significant mortality with concomitant pulmonary artery resection, reconstruction and pneumonectomy due to high transfusion requirements and pulmonary edema of the remaining lung [17].

In our experience 8/18 (45%) patients also had concomitant distant disease at time of diagnosis. This presents an equally challenging subset of patients where the benefits of a surgical resection, primarily the potential for a survival benefit, must be weighed against the risk of perioperative mortality and morbidity, which in turn may substantially delay other treatment modalities. Given the low incidence of the disease, evidence for guiding management of patients with pulmonary artery sarcoma remains scarce. Treatment algorithms are best formulated at cancer centers with a large experience in the management of complex clinical scenarios and in a multidisciplinary fashion while respecting patient preferences and resources available.

Acknowledgements Dr. Wyler von Ballmoos received funding from the Thoracic Surgery Research Foundation by means of the Davidson Fellowship Award.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individual participants included in the study.

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