

Right Ventricular Involvement of an Aggressive Malignant Peripheral Nerve Sheath Tumor

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ABSTRACT

We present a case of a 58-year-old woman who had a painful right thigh mass for a few months. A transthoracic echocardiogram revealed no evidence of an intracardiac mass. She had a whole-body positron emission tomography/computed tomography scan two months later that revealed masses in her right lower extremity and a mass in her right ventricle that had not been initially reported. She had been initially diagnosed with an undifferentiated pleomorphic sarcoma, but this diagnosis was changed to a malignant peripheral nerve sheath tumor with repeat pathology. She was subsequently hospitalized. An echocardiogram showed a mass covering 80% of her right ventricle (RV). Serial cardiac magnetic resonance imaging revealed a 9.4 x 5.6 cm RV mass with vascular and avascular portions and inflow and outflow tract obstruction. Computed tomography showed no other metastases. Due to a delay in diagnosis and a decline in left ventricular ejection fraction, the patient could not undergo palliative chemotherapy or radiotherapy.

CASE REPORT

CASE REPORT

A 58-year-old woman with a history of coronary artery disease and hyperlipidemia presented to an outreach clinic affiliated with our university hospital with a painful right thigh mass that had been affecting her for a few months. Initially, a skin biopsy was performed, which revealed a benign fibrous histiocytoma. A transthoracic echocardiogram (TTE) was performed due to her prior cardiac history; the results were unremarkable, with no intracardiac masses. Due to ongoing pain, she returned for further evaluation five months after her skin biopsy. She underwent a whole-body positron emission

tomography (PET)/computed tomography scan. It revealed lesions in her right foot, calf, and thigh. Biopsies were taken from these sites. The mid-portion of her right sartorius muscle was also excised. Axial PET scan images of the chest revealed uptake in the right ventricle (RV) (Figure 1). However, this uptake was not reported or documented as a concerning finding, which contributed to the failure to diagnose a cardiac mass at the time. The pathology showed a high-grade undifferentiated pleomorphic sarcoma. The immunohistochemical stains for CD68 were positive, with a Ki-67 (tumor proliferative index) of 97%.

She was subsequently referred to the orthopedic oncology clinic at our university hospital. A magnetic resonance imaging (MRI) study of her right thigh revealed a 3.6 x 3.6 x 2.1 cm infiltrating vascular neoplasm of the anterior thigh involving the rectus femoris, vastus medialis, and sartorius musculature that was continuous with the femoral vessels (Figure 2). Right hip disarticulation was performed as a definitive treatment five weeks later. At this time, the surgical pathology revealed a grade 3 malignant peripheral nerve sheath tumor (MPNST) (Figure 3). Two weeks later, the patient had drainage from the wound site, and she was admitted to the hospital for incision and drainage of the surgical site and wound VAC placement.

A TTE was performed due to dyspnea on exertion after the hip disarticulation. The TTE revealed a large mass that occupied more than 80% of the right ventricle (RV), as well as severe dilation of the RV and inferior vena cava (Figure 4). These findings were surprising because the TTE report from another hospital 3.5 months prior was unremarkable, with no mass found in the RV. A cardiac MRI study was subsequently performed. T1-weighted, T2-weighted, first-pass perfusion, and delayed enhancement imaging were performed to characterize the intracardiac mass (Figure 5). The mass appeared isointense on T1-weighted images and hyperintense on T2-weighted images, with heterogeneous contrast uptake on perfusion images and some areas of enhancement on late gadolinium-enhanced images. The mass was 9.4 x 5.6 cm in size, with a broad base and attachments to the inferior and free walls of the RV. The mass was heterogeneous, with some vascular and avascular components, as well as evidence of necrosis and thrombosis. There was also RV outflow obstruction due to the mass extending into the RV outflow tract, as well as RV inflow obstruction due to a portion of the mass herniating across the tricuspid valve. Additionally, the mass pushed against the interventricular septum, compressing the left ventricle. No pericardial space extension was seen. The left ventricular ejection fraction (LVEF) was 53%. A computed tomography scan of the chest, abdomen and pelvis with contrast was performed the next day, which revealed no other metastasis other than the RV mass (Figure 6).

At our center, the diagnosis was changed from an undifferentiated pleomorphic sarcoma to MPNST based on the surgical pathology from the patient's hip disarticulation performed at our hospital [1]. Histologic sections revealed a malignant spindle cell neoplasm with a plexiform growth pattern involving the skin and deep soft tissue. Each nodule consisted of fascicles of relatively uniform spindle cells with varying cellularity. Up to 38 mitoses per 10 high-power fields were identified; geographic necrosis was also identified and averaged 10%. Immunohistochemical staining performed at our hospital showed a Ki-67 index of 70%. Ki-67 is an important prognostic marker in patients with MPNSTs; a Ki-67 labeling index > 25% has been correlated with a reduced survival rate [1]. CD68 staining was negative in the neoplastic cells. Additional immunohistochemical staining was performed. The neoplastic cells were positive for vimentin, BCL-2, CD56 (partial), FLI-1, and TLE-1 (weak and patchy) but were predominantly negative for EMA, CAM5.2, SMA, CD31, desmin, myogenin, CD34, AE1/3, SOX10, S100, CD99, ERG, GFAP, and CD45. INI-1 was retained while H3K27me3 was

lost. The VBG stains were negative. An angiosarcoma was considered less likely due to the negative staining for CD34, CD31, ERG, and VBG around the neoplastic nodules, epithelioid sarcoma and epithelioid malignant peripheral nerve sheath tumor, as well as the retained INI-1. These findings supported the diagnosis of MPNST.

A biopsy of the RV mass was not possible, as it was high risk due to the intense vascularity of the mass noted on the cardiac perfusion images, which prevented a definitive tissue diagnosis of the intracardiac mass. The patient's initial peripheral right thigh mass, imaging information revealing an intracardiac mass not initially seen on TTE images and missed on PET imaging, pathology, and clinical suspicion all led to the final presumptive diagnosis of an aggressive MPNST with cardiac metastases to the RV. The route of extension of this aggressive MPNST was thought to be hematogenous.

Palliative chemotherapy was discussed. However, since the patient was being treated for a surgical site infection, she was not considered an appropriate candidate. Surgical resection of the RV mass was also not an option at the time due to the high vascularity of the mass, potential high risk of bleeding and the patient being a Jehovah's witness. A four-week follow-up cardiac MRI (Figure 7) showed a significant decline in LVEF, from 53% to 35%. Biopsies and surgical intervention were still deemed high risk. Due to a declining EF, the poor prognosis and the patient's wishes, palliative radiation and chemotherapy were not performed. The patient chose hospice care and passed away a few months later. Because the patient's time of death was unknown to our inpatient team, an autopsy was not performed to provide a definitive diagnosis.

DISCUSSION

Etiology & Demographics:

Soft tissue sarcomas are very rare adult malignancies (accounting for ~1% of adult cases) [1]. A malignant peripheral nerve sheath tumor (MPNST) is a rare soft tissue sarcoma that typically develops in peripheral nerves in the extremities [2]. Nearly 40-50% of MPNSTs occur in patients with neurofibromatosis type 1; 40-47% of cases occur sporadically; and the remainder develop due to prior radiation exposure [3]. Cardiac metastases are extremely rare; in one report of 407 cases of malignant tumors with cardiac metastases, 4.2% of the patients had a primary soft tissue sarcoma [4].

MPNSTs have previously been reported using various other names, including malignant schwannomas, malignant neurilemmomas, and neurofibrosarcomas [5]. Metastatic MPNSTs with cardiac metastases have rarely been reported [6]–[8]. Due to differences in the nomenclature and criteria for diagnosing MPNSTs, as well as MPNSTs with cardiac metastases being atypical, our literature search did not reveal many cases of metastatic MPNSTs with cardiac metastases.

Clinical & Imaging Findings:

Patients with an MPNST may be mostly asymptomatic from a cardiac standpoint, necessitating the use of cardiac

imaging based on clinical suspicion and the understanding that an MPNST has the potential for cardiac metastasis. After TTE as the initial imaging modality, cardiac MRI is a good diagnostic tool that should be considered in diagnostic work-ups of these tumors due to its ability to assess location, characterize tissue and detect extension into adjacent tissues [9]. Our patient was initially misdiagnosed. Based on TTE and cardiac MRI images, her metastatic cardiac mass was identified. Based on tissue characterization, cardiac MRI, and reassessment of pathology samples, we diagnosed her with an MPNST with cardiac metastasis. A structured imaging approach used in conjunction with pathology samples from the primary source may be sufficient to make the correct diagnosis, thereby removing the need for intracardiac tissue sampling, which was not feasible in our case and was a limitation for making a definitive diagnosis based on our patient's intracardiac mass.

Treatment & Prognosis:

MPNSTs have a very poor prognosis. The typical median survival time for these patients is 32 months [10]. The overall 2-year and 5-year survival rates are 57% [10] and 15-66% [11], respectively. These poor survival rates emphasize the importance of early diagnosis in order to begin any potential treatments as soon as possible.

Although these patients have a poor prognosis, multiple treatment options have been studied. Complete surgical resection with wide negative margins is the only curative treatment [12], [13]. In cases of unresectable or metastatic MPNSTs, Prudner et al. reported doxorubicin-based cytotoxic chemotherapy as the standard of care [13]. Adjuvant radiation is recommended for marginal excision to improve local control; it is also recommended for intermediate or high-grade lesions measuring > 5 cm in size [12]. Chemotherapy, radiation therapy, and other emerging therapies still need to be studied further. Unfortunately, our patient was referred to palliative care due to a late and accurate diagnosis of her rapidly progressing MPNST with heart metastasis; treatment could not begin since she already had a reduction in LV function and an ongoing infection. The patient opted for hospice care.

Differential Diagnosis:

Cardiac metastases are much more common than primary cardiac tumors. They are most common in patients with advanced tumor disease. Because cardiac metastases of MPNSTs are usually clinically silent, most intracavitary tumors are found on autopsy. Less than 10% of patients present with symptoms of cardiac dysfunction [3]. For example, right-sided metastases can lead to pulmonary tumor emboli with progressive cor pulmonale over weeks to months [6], [7]. The occlusion of the pulmonary vasculature by tumor cells and the associated thrombus would present as subacute and progressive clinical deterioration. The most common tumors with cardiac metastatic potential are malignant melanomas and carcinomas of the lung, esophagus and breast [6]–[8]. MPNSTs are difficult to diagnose because they are less common than the aforementioned metastatic tumors and thus can be confused with other tumors or nontumor lesions. MPNSTs may be clinically silent and are often definitively diagnosed by biopsy and histopathology [14].

Primary malignant cardiac tumors:

Cardiac MPNSTs should be differentiated from other primary cardiac tumors, such as angiosarcomas, leiomyosarcomas, rhabdomyosarcomas, undifferentiated sarcomas, and primary cardiac lymphoma [15]. Cardiac angiosarcomas are large, irregular, and highly vascular tumors that typically occur in the right chambers of the heart, are locally invasive to cardiac tissue, and are broad due to a poorly defined attachment to the myocardium [15]. They can appear as isointense lesions with multiple nodular areas of high intensity on T1-weighted images, and appear with heterogeneous enhancement with possible necrotic cores on late gadolinium enhancement imaging [15]. Cardiac leiomyosarcomas account for less than 1% of malignant cardiac masses; they are typically found in the left atrium and are best diagnosed with echocardiography, CT imaging or MRI [16]. Rhabdomyosarcomas, the most common pediatric cardiac malignancy, are found in the myocardium and present on cardiac MRI as isointense masses on T1-weighted images and hyperintense masses on T2-weighted images; they generally show homogeneous contrast enhancement, with regions of hypointensity due to central necrosis that may be occasionally seen [17]. Undifferentiated sarcomas, the second most common primary cardiac malignancy, often present in the left atrium with no specific histological pattern and can appear similar to angiosarcomas on cardiac MRI as focal or infiltrative masses with necrosis and hemorrhage; however, they can be differentiated from angiosarcomas based on their prevalence in the right atrium [17]. Primary cardiac lymphoma is a rare type of non-Hodgkin's lymphoma that can be confused with angiosarcomas, metastatic carcinomas, and metastatic melanomas; imaging and tissue sampling are needed to make a definitive diagnosis due to the variability in its presentation [18].

Secondary metastatic cardiac tumors:

Secondary metastatic cardiac tumors are considerably more common than primary cardiac tumors and should be considered when diagnosing cardiac MPNSTs. Melanomas and thoracic carcinomas (for example, breast, lung, and esophageal) are more likely to have cardiac involvement; most secondary malignancies have a low signal intensity on T1-weighted images and a high signal intensity on T2-weighted images, except for melanoma, which is hyperintense on T1-weighted images [15].

Conclusion:

Ultimately, many, if not all, of the aforementioned malignant masses may be confused with cardiac MPNSTs. Recognizing cardiac masses using cardiac imaging is crucial in the diagnostic process. A pathological analysis of tissue from the peripheral mass was essential for making the presumed correct diagnosis of a metastatic MPNST since performing a biopsy of the cardiac mass was deemed too risky for the patient while she was alive, and the patient passed away unbeknownst to our inpatient team, preventing us from obtaining a postmortem biopsy during an autopsy. Early detection and diagnosis of the correct type of cardiac mass could potentially allow treatment to begin earlier in patients with MPNSTs.

TEACHING POINT

Malignant peripheral nerve sheath tumors are soft tissue sarcomas that, although rare, can metastasize to the heart. Thus, a high degree of clinical suspicion should be maintained, and appropriate cardiac imaging should be performed. Cardiac MRI is an excellent imaging modality for tissue characterization that can aid in the differentiation of various types of malignant cardiac masses.

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FIGURES

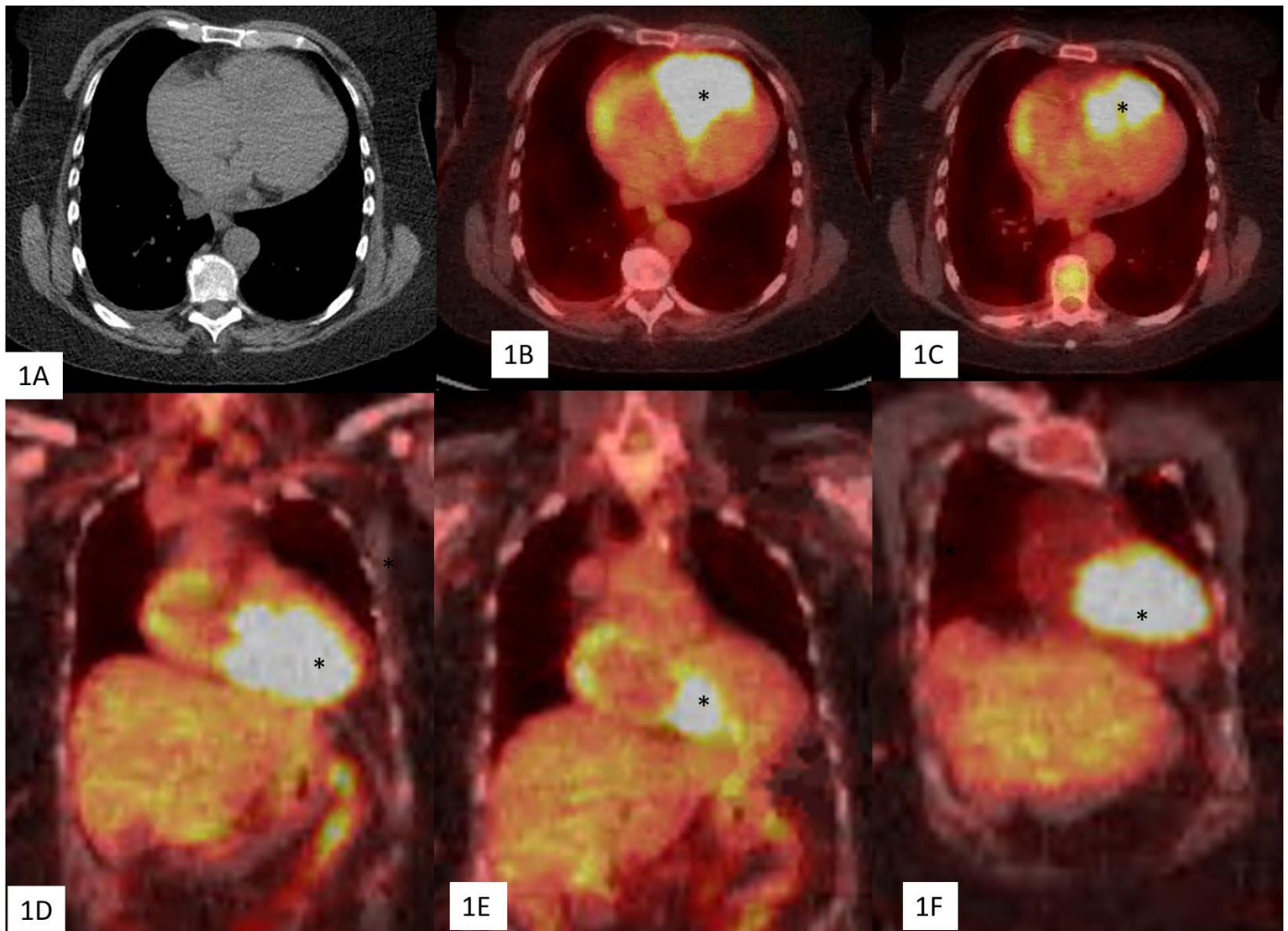


Figure 1: A 58-year-old female with a peripheral malignant nerve sheath tumor of the right femur/thigh with metastasis to the heart.

Findings: Fused PET-CT axial and coronal images revealed an FDG-positive large mass filling nearly the entire right ventricle. However, the mass was not well visualized on the noncontrast CT lesion, indicating a mass in the right ventricle. Evidence of a mass in the right ventricle was seen on both the CT chest image (Figure 1A) and the PET-CT fused image (Figure 1B).

Technique: Positron emission tomography (PET) was performed with 2-[fluorine-18] fluoro-2-deoxy-d-glucose (FDG). Low-resolution noncontrast computed tomography (CT) was performed prior to the PET scan for anatomic assessment. The PET and CT images were then fused for analysis. The slice thickness was 3.3 mm.

Siemens Biograph M CT. FDG dose: 11 mCi, 3.3 mm slice thickness on CT scan, 100 kVp.

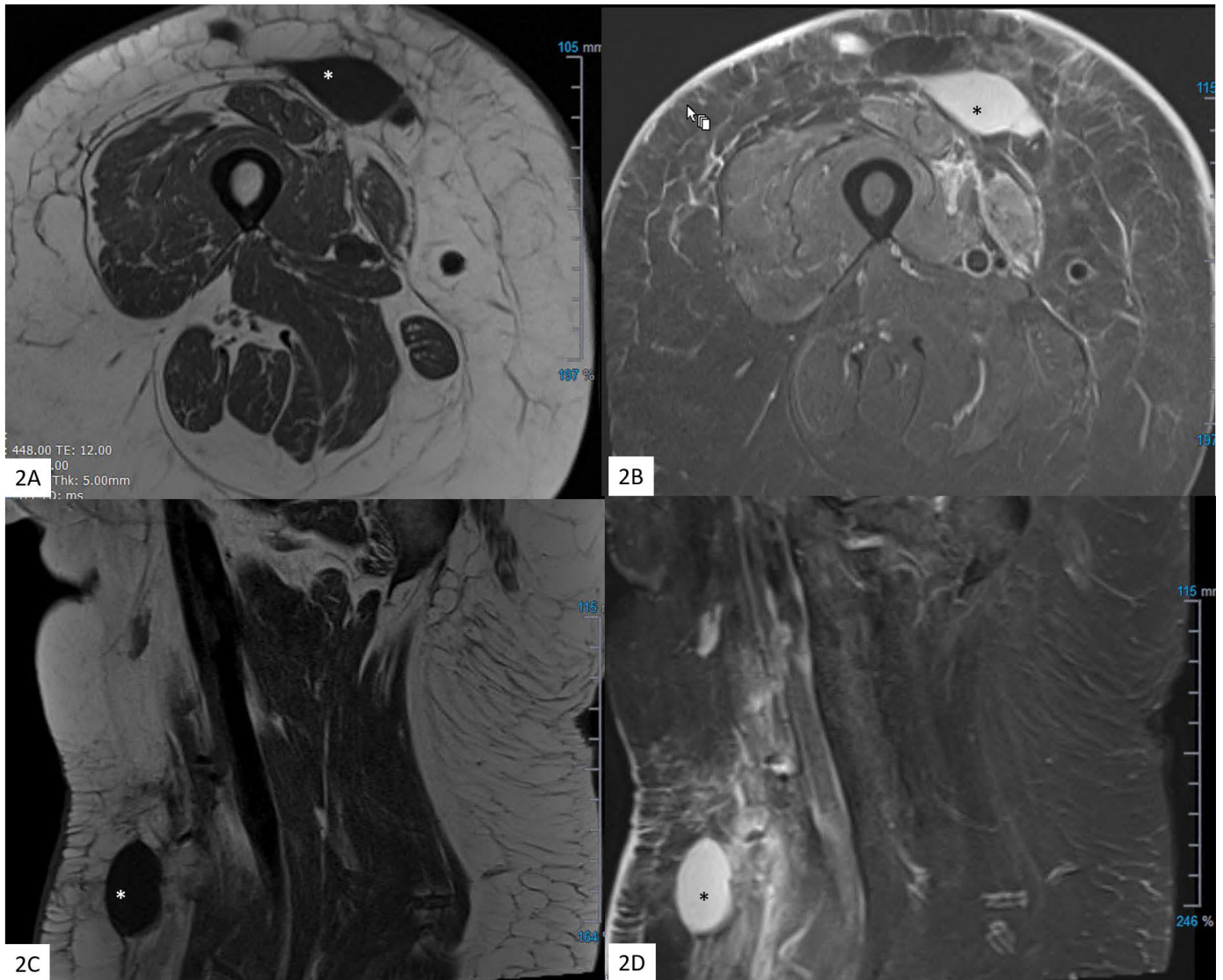


Figure 2: A 58-year-old female with a peripheral malignant nerve sheath tumor of the right femur/thigh with metastasis to the heart.

Findings: MR images without gadolinium show a hypointense mass on T1-weighted images (2A, 2C) and a hyperintense mass on T2-weighted images (2B, 2D) in the anteromedial thigh subcutaneous fat. The mass measured 3.6 x 3.6 x 2.1 cm. Serpiginous tubular structures with the same signal characteristics extend the mass toward the skin surface and the deep fascia, likely representing vessels. The hyperintense signal on the T2-weighted image extends into the adjacent rectus femoris, vastus medialis and sartorius muscle bellies.

(MRI: magnetic resonance imaging. MPNST: malignant peripheral nerve sheath tumor).

Technique: 1.5 Tesla Siemens Magnetom Aera scanner (A and B) axial T1- and T2-weighted images, respectively, TR 470, TE 8.7, 5 mm slice thickness.

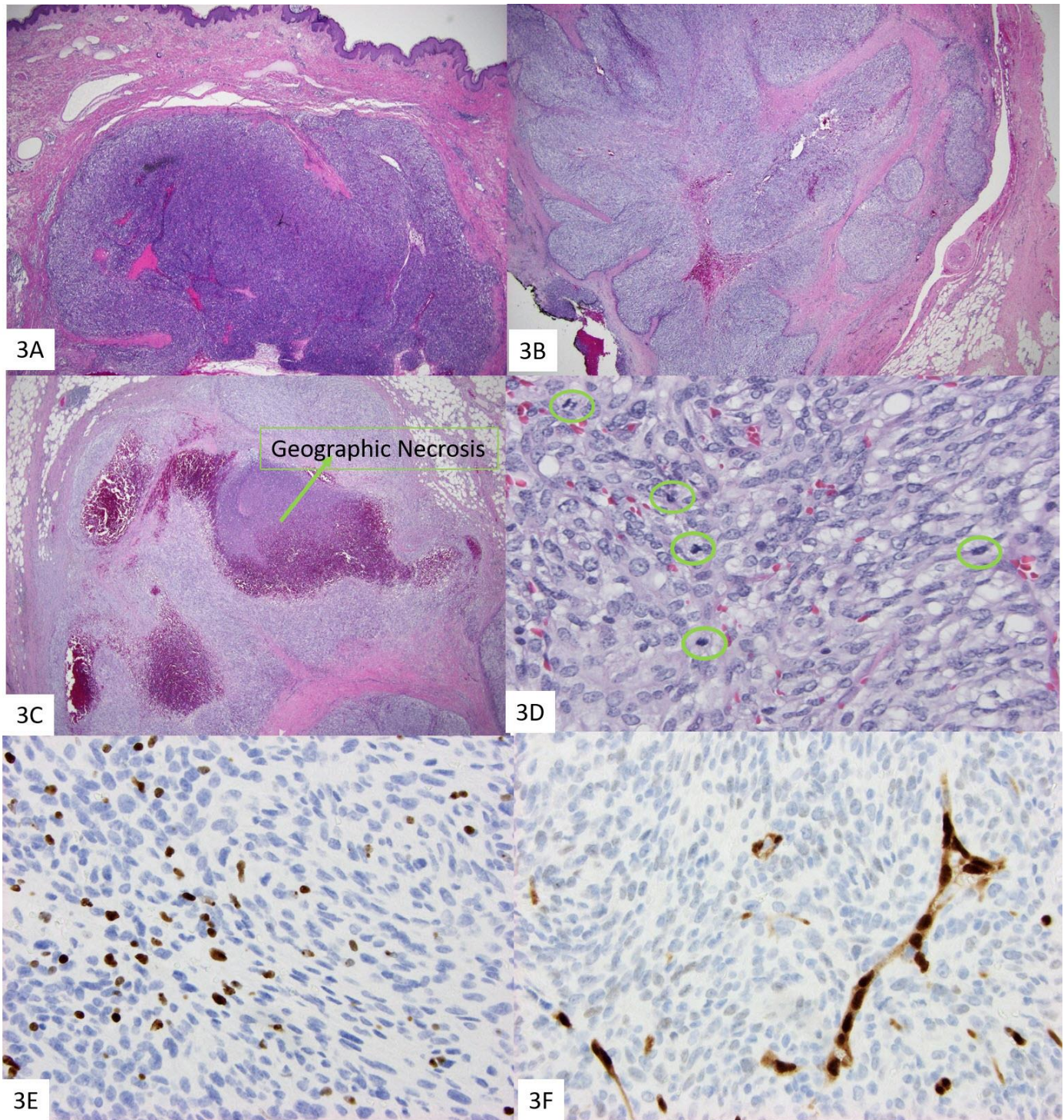


Figure 3: A 58-year-old female patient with a peripheral malignant nerve sheath tumor of the right femur/thigh with metastasis to the heart.

Findings: A skin and subcutaneous tissue segment with deep dermal multinodular and spindled cell neoplasms was found. Each nodule consisted of fascicles of relatively uniform spindle cells with varying cellularity (3A, 3B). The MPNST shows areas of geographic necrosis (circled in green) (3C). Five mitoses were seen in this high-power field (circled in green), and up to 38 mitoses per 10 high-power fields were identified, which indicates a rapidly replicating tumor. The Ki-67 proliferation index is very high (70%) (3D). The loss of nuclear H3K27me3 expression on this H3K27me3 stain (60X magnification) supports the diagnosis of a sporadic MPNST (3E). The negative staining around the neoplastic nodules on this ERG stain (60X magnification) eliminated an angiosarcoma from the differential diagnosis (3F).

(MPNST: malignant peripheral nerve sheath tumor. ERG: Erythroblast transformation-specific [ETS]-related gene).

Technique: Pathologic evaluation of the biopsy from the mass from the right femur/thigh

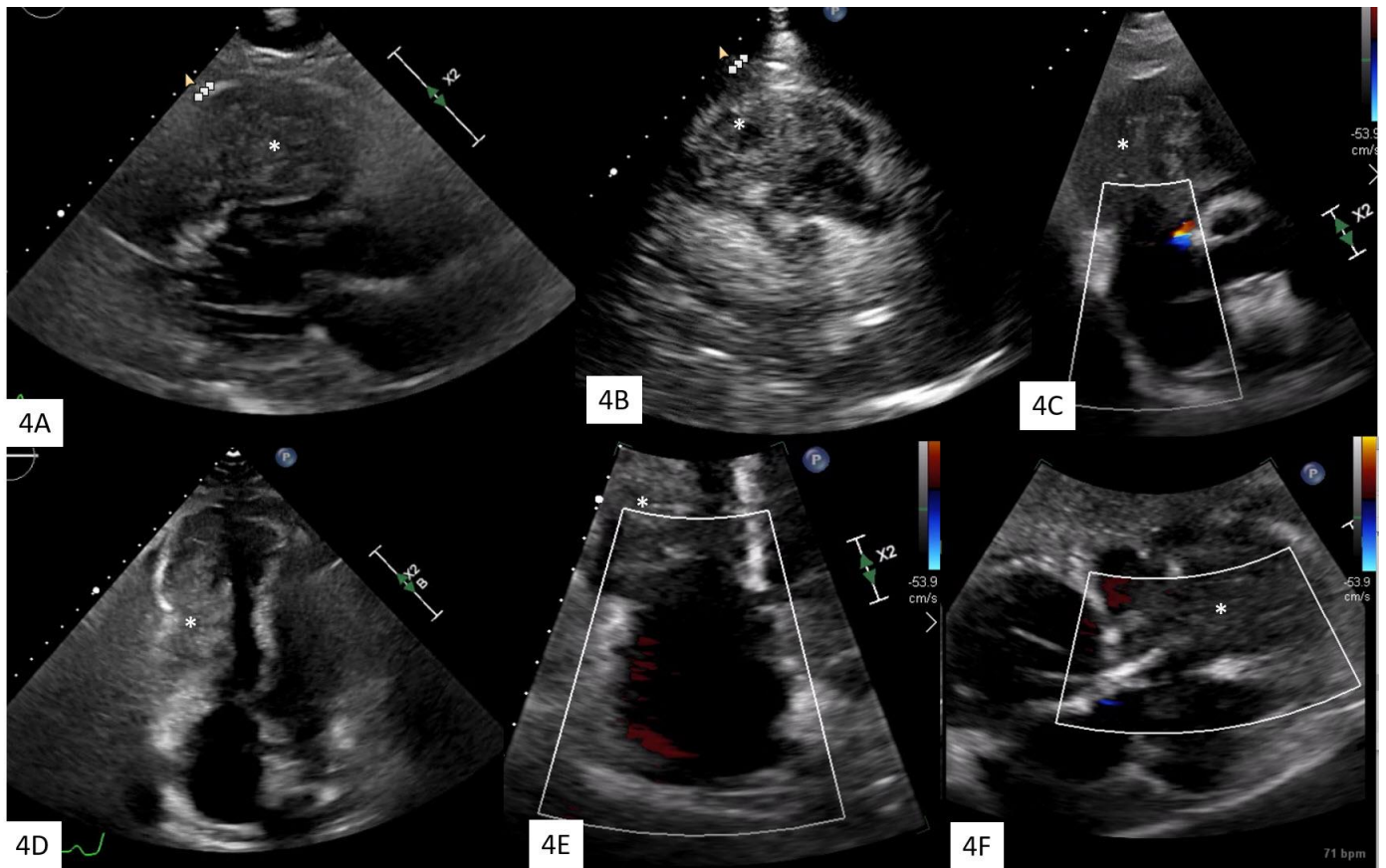


Figure 4: A 58-year-old female with peripheral malignant nerve sheath tumor of the right femur/thigh with metastasis to the heart.

Findings: The parasternal long axis view revealed a large mass filling the RV cavity (4A). This mass showed heterogeneous contrast uptake on definition contrast echo images (4B). The parasternal short-axis view (4C) and right ventricular (RV) focused apical 4-chamber view (4D) show a severely dilated RV, with a mass encompassing >80% of the RV. (4E, 4F) show the presence of a mass near the tricuspid valve obstructing flow from the right atrium toward the RV on color Doppler images. (* RV mass)

Technique: Transthoracic echocardiogram (TTE) on a General Electric (GE) IE-33 machine with 2-dimensional (2D) imaging with and without the use of a definition contrast agent.

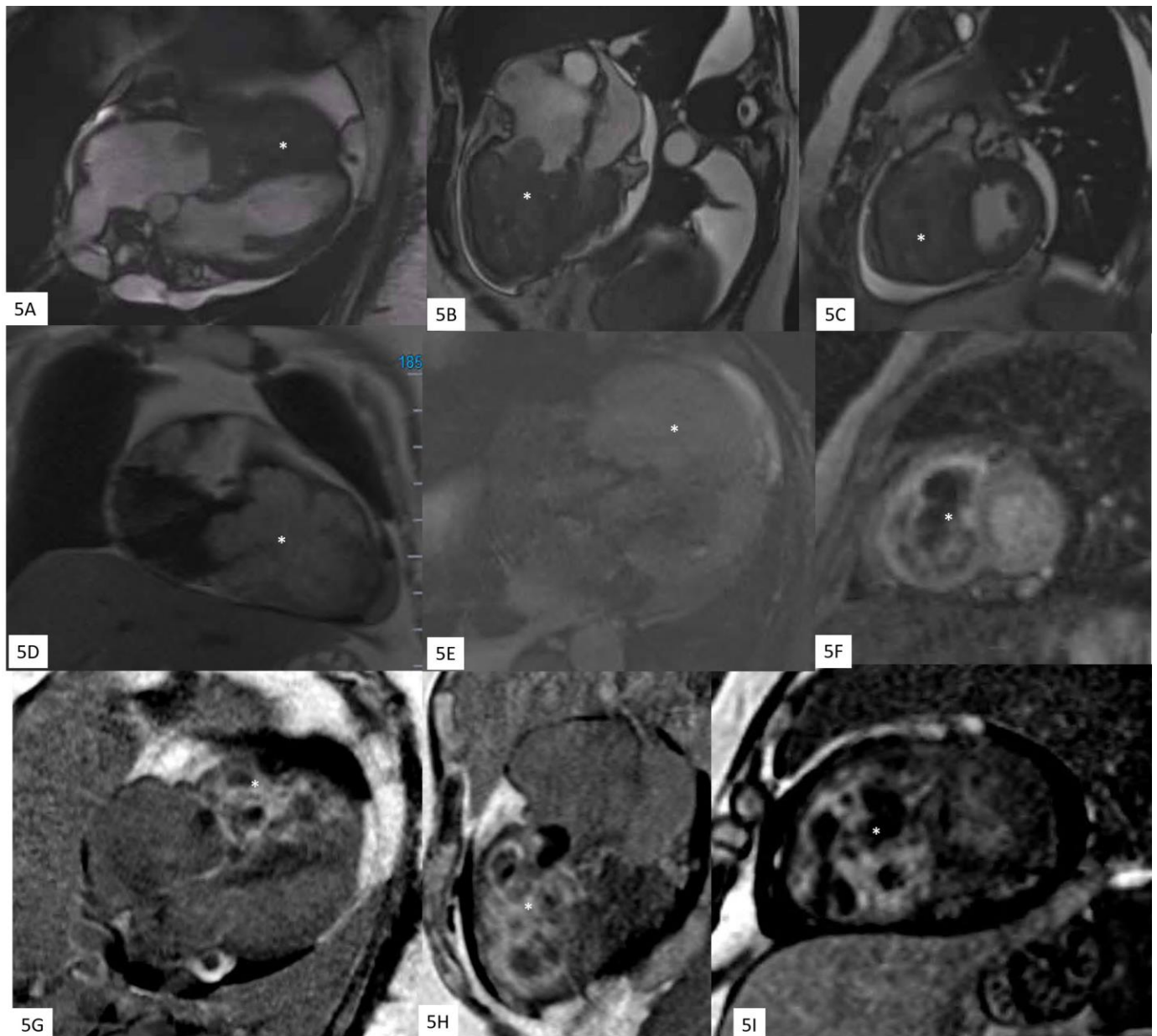


Figure 5: A 58-year-old female with peripheral malignant nerve sheath tumor of the right femur/thigh with metastasis to the heart.

Findings: The initial cardiac MRI images are shown. The SSFP-cine-4-chamber view revealed a right ventricular mass encompassing most of the right ventricular cavity, with extension into the right atrium through the tricuspid valve (5A). The SSFP-cine right ventricular 2-chamber view showed a mass extending into the right ventricular inflow tract (5B), and the SSFP-cine short axis view toward the base (5C) showed a mass extending into the RV outflow tracts, causing flow obstruction. The T1-weighted image in the 4-chamber view revealed the mass to be isointense (5D, 5E). Perfusion imaging showed heterogeneous contrast uptake by the mass (5F). A late gadolinium-enhanced phase-sensitive inversion recovery sequence showed heterogeneous enhancement of the mass, with a probable thrombotic component (5G, 5H). Small circumferential pericardial effusion was identified in all the images.

(MRI: magnetic resonance imaging. SSFP: steady state free precession. MPNST: malignant peripheral nerve sheath tumor). (* Right ventricular mass)

(MRI: magnetic resonance imaging. MPNST: malignant peripheral nerve sheath tumor).

Technique: 1.5 Tesla Siemens Magnetom Aera scanner TR 470, TE 8.7, 8 mm slice thickness. Contrast agent used: Magnevist 0.1 mmol/kg

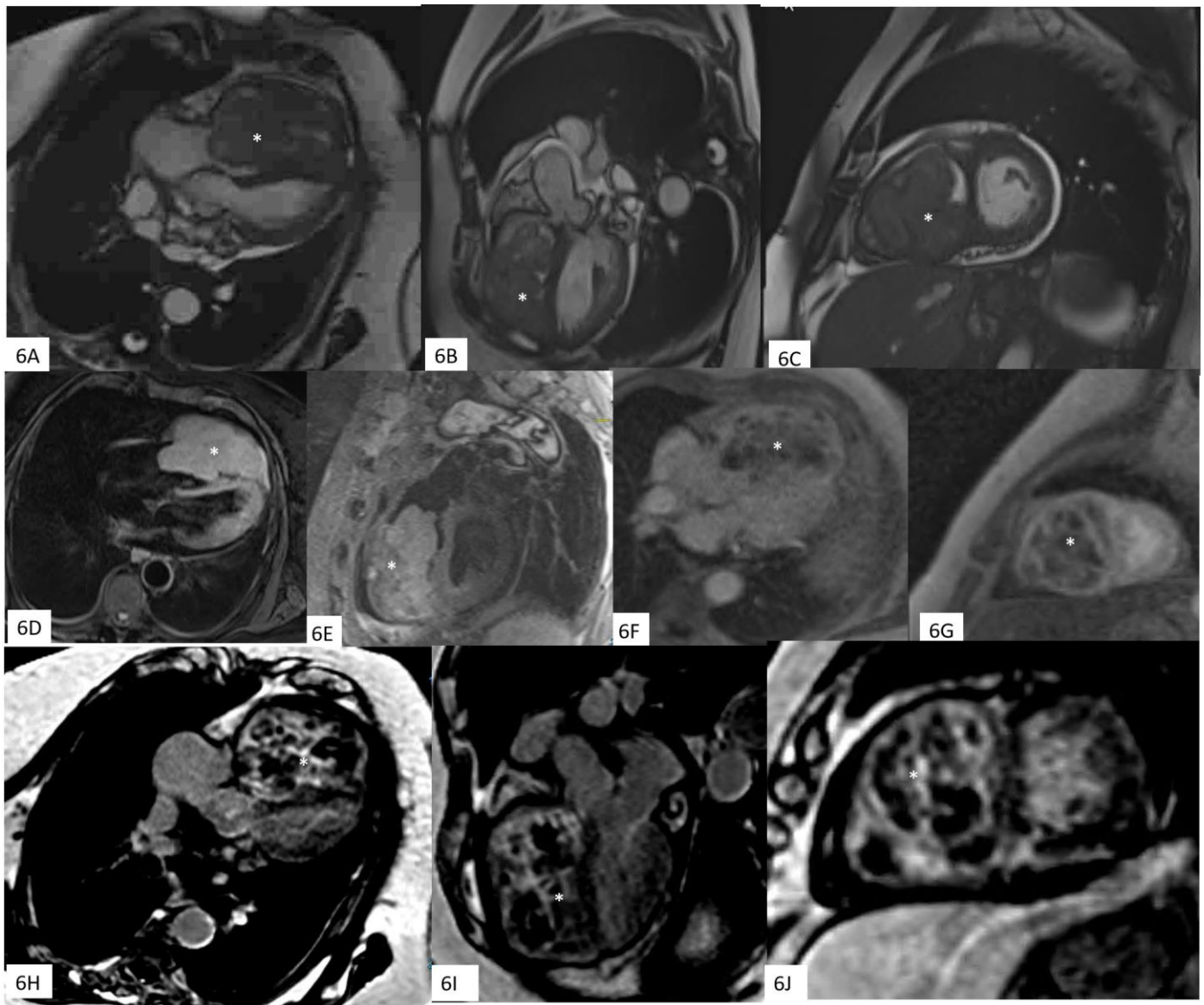


Figure 6: A 58-year-old female with peripheral malignant nerve sheath tumor of the right femur/thigh with metastasis to the heart.

Findings: Follow-up cardiac MRI images show persistence of the RV mass. The SSFP-cine 4-chamber, 5-chamber and short-axis views show a right ventricular mass encompassing most of the right ventricular cavity, with extension into the right atrium through the tricuspid valve (6A, 6B, 6C). T1-weighted imaging in the 4-chamber view revealed the mass to be isointense to slightly hyperintense (6D). T2-weighted imaging in the right ventricular outflow tract view revealed the mass to be hyperintense (6E). Perfusion imaging showed heterogeneous contrast uptake by the mass (6F, 6G). The late gadolinium-enhanced phase-sensitive inversion recovery sequence shows heterogeneous enhancement of the mass (6H, 6I, 6J), with the RV cavity more enlarged than in the baseline MRI scan and a small LV cavity size due to compression of the interventricular septum. Small circumferential pericardial effusion was identified in all the images.

(MRI: magnetic resonance imaging. SSFP: steady state free precession. MPNST: malignant peripheral nerve sheath tumor). (* Right ventricular mass)

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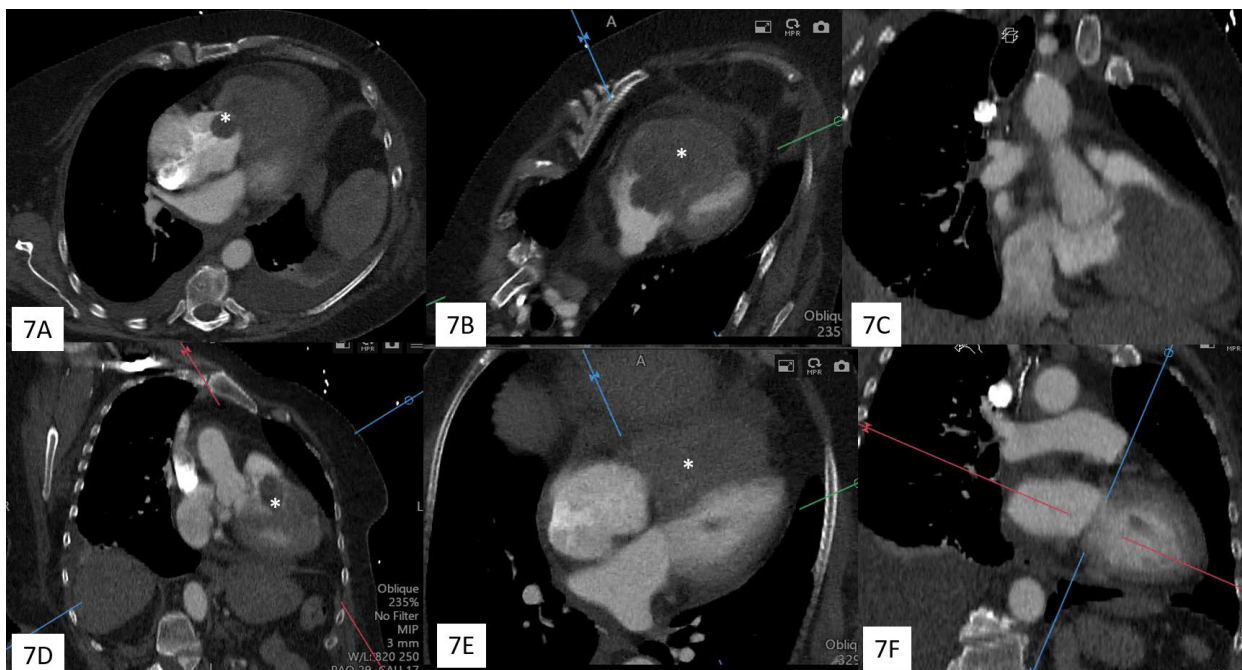


Figure 7: A 58-year-old female with peripheral malignant nerve sheath tumor of the right femur/thigh with metastasis to the heart.

Findings: Chest CT scan with contrast with multiplanar reformat (MPR) reconstruction revealed a bulking mass with irregular borders in the right ventricle extending into the RV inflow and outflow tracts. No other masses were identified in the left cardiac chambers, pericardium or other parts of the chest cavity. (* RV mass)

Technique: Chest CT scan (axial slices) on a Toshiba 320 scanner (non-EKG gated) with IV contrast. 120 kV, 350-650 mA, 3 mm slice thickness, contrast agent used: Iovue 360 @ 5 ml

Etiology	<ul style="list-style-type: none"> Nearly 40-50% of MPNSTs occur in patients with neurofibromatosis type 1; 40-47% of cases occur sporadically Remainder may develop due to prior radiation exposure.
Incidence	In one series of 407 cases with cardiac metastases, 4.2% of these tumors were from a primary soft tissue sarcoma.
Gender ratio	Unknown
Age predilection	Unknown
Risk factors	<ul style="list-style-type: none"> Prior radiation treatment History of neurofibromatosis type I
Treatment	<ul style="list-style-type: none"> Complete surgical resection with wide negative margins is the only curative therapy. In cases of unresectable or metastatic MPNSTs, Prudner et al. reported doxorubicin-based cytotoxic chemotherapy as the standard of care. Adjuvant radiation is recommended for marginal excision to improve local control; it is also recommended for intermediate or high-grade lesions measuring > 5 cm in size. Chemotherapy, radiation therapy, and other emerging therapies still require further study.
Prognosis	<ul style="list-style-type: none"> MPNSTs have a very poor prognosis. Death occurs in 63% of patients within two years of diagnosis. The typical median survival in these patients is 32 months. The overall 2- and 5-year survival rates are 57%¹⁰ and 15-66%, respectively.
Findings on imaging	<ul style="list-style-type: none"> On cardiac MRI with and without contrast. <ul style="list-style-type: none"> The mass is isointense on T1-weighted images and hyperintense on T2-weighted images. Perfusion imaging shows heterogeneous contrast uptake by the mass. Late gadolinium enhanced phase sensitive inversion recovery sequence shows heterogeneous enhancement of the mass. Pericardial effusion was identified, with no left cardiac chambers involved. Chest CT scans reveal mild contrast enhancement of the mass. FDG PET images reveal significant uptake of the mass.

Table 1: Summary table of Malignant Peripheral Nerve Sheath Tumor.

	Transthoracic and transesophageal echocardiogram	Cardiac CT angiogram	Cardiac MRI
Angiosarcoma	<ul style="list-style-type: none"> • Mostly arise in right atrium • Commonly involves the pericardium with pericardial effusion 	Filling defect in right atrium that extends across the adjacent wall of right atrium into the epicardial fat, mostly associated with pericardial effusion.	<ul style="list-style-type: none"> • T1: Heterogeneous • T2: Heterogeneous • Uptake on perfusion imaging: Present • LGE: Heterogeneous
Rhabdomyosarcoma	<ul style="list-style-type: none"> • Large (>5 cm), multiple lesions • Right > Left • Broad base • Modest to intense enhancement on contrast images • Irregular, ill-defined margins • Intra/extracardiac infiltration 	<ul style="list-style-type: none"> • Smooth or irregular low attenuation mass in a cardiac chamber. • Modest to intense enhancement on contrast images 	<ul style="list-style-type: none"> • T1: Isointense • T2: Hyperintense • Uptake on perfusion imaging: Present • LGE: Homogeneous
Undifferentiated Sarcoma	<ul style="list-style-type: none"> • Large, irregular mass or polypoid masses. • Infiltration into the myocardium appear with thickening and irregularity • May manifest as a hemorrhagic mass, replacing the pericardium. 	<ul style="list-style-type: none"> • Large, irregular, low attenuation intracavitary lesion or polypoid masses. • Myocardial infiltration with thickening has been noted. 	<ul style="list-style-type: none"> • T1: Isointense/hyperintense • T2: Hyperintense • Uptake on perfusion imaging: Heterogeneous • LGE: Heterogeneous
Lymphoma	<ul style="list-style-type: none"> • Homogeneous, infiltrating masses that lead to wall thickening and restrictive hemodynamics • Can present as nodular masses intruding into the cardiac chambers, preferentially the right heart chambers and especially the right atrium. 	Mass frequently noted in the right cardiac chambers.	<ul style="list-style-type: none"> • T1: Isointense • T2: Isointense • Uptake on perfusion imaging: None • LGE: None/minimal
Melanoma	Pericardium most commonly involved with presence of pericardial effusion.	Lymphadenopathy with irregular or lobulated appearance and pericardial effusion.	<ul style="list-style-type: none"> • T1: High • T2: Low • Uptake on perfusion imaging: Heterogeneous • LGE: Heterogeneous
Malignant peripheral nerve sheath tumor	Bulky mass on the right side of heart.	<ul style="list-style-type: none"> • May be missed on noncontract CT scans of the chest. • Mass most likely in a right cardiac chamber. 	<ul style="list-style-type: none"> • T1: Isointense/hyperintense • T2: Hyperintense • Uptake on perfusion imaging: Heterogeneous • LGE: Heterogeneous
Metastasis	Direct invasion from a nearby tumor can be seen or extension through inferior vena cava.	<ul style="list-style-type: none"> • Mass noted mostly in right cardiac chambers, with extension into the myocardium and pericardium. • Mass may be noted in the IVC. • A thrombus can be seen associated with the mass. 	<ul style="list-style-type: none"> • T1: Low • T2: High • Uptake on perfusion imaging: Heterogeneous • LGE: Heterogeneous

CT: computed tomography. MRI: magnetic resonance imaging. LGE: Late gadolinium enhanced images. IVC: inferior vena cava.

Table 2: Differential diagnosis table for malignant cardiac masses in adults.

ABBREVIATIONS

LVEF = Left ventricular ejection fraction
MPNST = Malignant peripheral nerve sheath tumor
MRI = Magnetic resonance imaging
PET = Positron emission tomography
RV = Right ventricle
TTE = Transthoracic echocardiogram

KEYWORDS

Malignant peripheral nerve sheath tumor; cardiac magnetic resonance imaging; transthoracic echocardiography; peripheral magnetic resonance imaging; cardiac metastasis

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