A 31-year–old white male patient was transferred from a local district hospital for suspected pericardial mass. Two months earlier he had been admitted there for worsening shortness of breath and subsequently was diagnosed with a large pericardial effusion. He had no other pertinent past medical history. After pericardiocentesis (1.5 L) there was no evidence of malignant cells on cytology, and a computed tomography scan did not reveal any abnormality. Thus, the incident was interpreted as (viral) infectious pericarditis and the patient clinically improved under anti-inflammatory medication within 4 weeks. After that, while on a holiday in Tunisia, he again reported worsening shortness of breath. On day 6 of his holiday, he suddenly passed out and was admitted to a local hospital for unexplained syncope, where again a large pericardial effusion was seen. This time, after another pericardiocentesis (1 L), a pericardial mass was suspected by echocardiography and the patient was transferred back to Germany for further workup.

Cardiovascular magnetic resonance on the day of admission to our center revealed a large inhomogeneous mass within the pericardium and the mediastinal space infiltrating the big vessels, as well as the right atrium (Figure 1 and online-only Data Supplement Movies I and II). The mass severely compressed the entire heart, leading to hemodynamic compromise. Therefore, in our multidisciplinary oncology conference, the decision for urgent surgical sampling for...
A histological workup was made, and the patient was started on steroids (1 mg/kg of body weight) under the suspicion of high-grade malignant lymphoma.

Histological workup of the lesion revealed a grade 3 angiosarcoma (Figure 2). Subsequently, the patient reported orthopnea, developed hemodynamic compromise, and was transferred to the intensive care unit. Computed tomography revealed fulminant pulmonary embolism of the right pulmonary artery (Figure 3), which was treated with intravenous heparin.

At this stage, a German transplant reference center was contacted to discuss possible combined heart and lung transplantation, but it was judged that, because of several pericardiocenteses, the tumor might have already infiltrated the incision canal or even the pleural cavity. Therefore, transplantation was deferred, and a chemotherapy according to the vincristine, ifosfamide, doxorubicin, etoposide scheme was commenced. However, after 3 weeks of therapy there was no reduction in tumor size (Figure 4 and online-only Data Supplement Movies III and IV). Therefore, based on rare reports about the efficacy of paclitaxel in angiosarcoma in the literature,1 chemotherapy with paclitaxel was initiated, which led to significant reduction of the tumor over the course of the following 4 months. After 5 months the tumor began to grow again. Therefore, treatment with paclitaxel was stopped and pazopanib was initiated. Under this therapy the patient improved clinically, as well as morphologically (Figure 5 and online-only Data Supplement Movies V and VI). Currently, after 10 months of follow-up, the patient is able to walk without any help and can actively participate in everyday life.

Angiosarcomas are the most common primary cardiac malignant tumors.2 In autopsy series, the cumulative prevalence of cardiac angiosarcoma is 0.0300% to 0.0002%.3 They tend to be more frequent in men than in women, and are found in patients aged 20 to 50 years.4 Angiosarcomas often respond poorly to chemotherapy, and most patients do not survive beyond 6 months of diagnosis. We report here a case of a young male patient with a pericardial angiosarcoma and a favorable course under paclitaxel treatment followed by pazopanib, documented by repeated cardiovascular magnetic resonance imaging for the first time.

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Disclosures
None.

References

Figure 2. Histological findings. A, Hematoxylin-eosin staining shows poorly differentiated sarcomatous cells eventually forming slit-like vascular channels (×400). The tumor cells are positive for CD31 (B) and express nuclear ETS-related gene protein (C) consistent with a vascular differentiation (×400).

Figure 3. Computed tomography of the thorax showing central right pulmonary embolism (red arrow).
Figure 4. Cardiovascular magnetic resonance (CMR) after chemotherapy according to the vincristine, ifosfamide, doxorubicin, etoposide (VIDE) scheme with no reduction in tumor size.

Figure 5. Cardiovascular magnetic resonance (CMR) after paclitaxel followed by pazopanib treatment showing significant reduction in tumor size.
Favorable Course of Pericardial Angiosarcoma Under Paclitaxel Followed by Pazopanib Treatment Documented by Cardiovascular Magnetic Resonance Imaging
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