INTRODUCTION

Cardiac tumors, benign or malignant, are rare, with an estimated prevalence of barely 0.002% to 0.3% in postmortem studies. Even when cardiac tumors are benign, they may adversely impact cardiac hemodynamics. Nevertheless, most cardiac masses are intracardiac when cardiac tumors are benign, they may adversely impact cardiac function. Malignant cardiac masses may extend beyond their primary location or cause metastasis to other structures. Malignant tumors of the brain may metastasize extracranially, with the latter occurring in 2% of cases. Intracranial meningiomas are the second most common primary central nervous system tumors, second only to gliomas, and account for 25% of all spinal tumors and 15% of all intracranial tumors. As per the World Health Organization classification, meningiomas are graded as I (benign), II (atypical), or III (malignant), occurring at a rate of 80.6%, 15.1%, and 4.3%, respectively. Meningiomas rarely (1) extend beyond their primary location or (2) metastasize extracranially, with the latter occurring in 2% of cases. Intracranial meningiomas are more common in women, whereas extracranial meningiomas are more common in men.

Here we describe a rare case of malignant meningioma that was extending as an extracranial metastasis to the right ventricle (RV) through the neck veins.

CASE PRESENTATION

A 14-year-old boy presented to the hospital with a history of recurrent headaches and vomiting. He had multiple emergency room visits over a period of 2 months with the same complaints. A brain computed tomography scan was performed and demonstrated a tumor located in the posterior fossa. This was consistent with aggressive meningioma extending from the left transverse and sigmoid sinuses to the right transverse sinus. The patient was referred to our hospital for further management and workup.

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Conflicts of Interest: None.

This case report was approved by the ethical research committee within the Institutional Review Board (IRB) at King Abdullah International Medical Research Centre with IRB research protocol number RC20/036/R. There was no need for a consent form since our data collection method consisted of chart and imaging data review. Patient confidentiality was maintained at all levels as only the principal investigator and coinvestigators had access to the data and were able to collect it.

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VIDEO HIGHLIGHTS

Video 1: Two-dimensional TTE, pediatric apical 4-chamber view showing a large cystic mass in the RA and protruding across the TV in diastole.

Video 2: Two-dimensional TEE midesophageal window, 89° vertical display with color Doppler, demonstrates the large, heterogeneous tumor mass extending into the RA from the markedly dilated SVC. Flow acceleration around the mass within the SVC can be seen.

Video 3: Two-dimensional TEE midesophageal window, 0° horizontal display, demonstrates the large cystic mass in the RA extending to RV through the TV (arrow).

Video 4: Two-dimensional TEE midesophageal window, 0° horizontal display, with color Doppler demonstrates the large cystic mass in the RA creating diastolic flow acceleration and color turbulence around the mass at the level of the TV annulus.

Video 5: CMR coronal view showing the tumoral thrombus extending to the entire course of the right-sided SVC to the junction with the RA.

Video 6: CMR, steady-state free perfusion sequence, oblique axial view (4-chamber aligned), demonstrating the large complex multilobulated (predominantly fluid-filled) cystic mass occupying the entire RA cavity and entering the RV cavity through the TV in diastole. The mass is mobile and not attached to the RA wall.

Video 7: CMR, first-pass rest perfusion, sagittal (SVC-aligned) view demonstrates that the solid mass is heterogeneous with a slightly perfused as well as a nonperfused portion. The tumoral thrombus is noted to completely fill the visualized portion of the dilated right SVC, the contrast is seen entering the RA via the IVC, and the SVC is almost entirely obstructed and contrast free.

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the posterior fossa. Following this diagnosis, the patient underwent immediate craniotomy for tumor removal with insertion of a ventriculoperitoneal shunt. The histopathology report confirmed a grade III rhabdoid papillary meningioma. He received a course of chemotherapy and radiotherapy. Two weeks after surgery, his parents noticed increasing swelling on the occipital area, and a brain magnetic resonance imaging scan revealed a left extracranial, hypervascular lesion within the posterior fossa. This was consistent with aggressive meningioma extending from the left transverse and sigmoid sinuses via the left jugular vein to the superior mediastinum.

Metastatic Malignant Meningioma Extending Into the Right Ventricle

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One month later he underwent neurosurgery again, this time a suboccipital craniotomy for excision of recurrent posterior fossa rhabdoid meningioma along with embolization of occipital artery and middle meningeal artery branches. One day later, a neck magnetic resonance imaging was done and showed signs of at least 2 nodules suspicious for residual tumor lesions or thrombosis extending from the left sigmoid sinus up to the left subclavian vein.

A transthoracic echocardiogram (TTE) was obtained to exclude cardiac involvement, but this demonstrated a large cystic mass within the right atrium (RA) measuring 44 mm × 33 mm (Figure 1A–D, Video 1). The mass was protruding across the tricuspid valve (TV) and causing partial obstruction (Figure 2A, Video 2).

A transesophageal echocardiogram (TEE) was performed to more fully document the origin and extent of the mass and confirmed that the large tumor mass entered the RA from the superior vena cava (SVC). The mass appeared large (measuring 30 mm × 35 mm) with significant flow obstruction through the TV along with cystic changes suggesting tumor necrosis. The mass was freely mobile within the RA without evidence of wall attachment (Figure 2B–D, Videos 3 and 4).

A cardiovascular magnetic resonance (CMR) imaging scan was performed for tissue characterization, and it demonstrated a giant mass invading a dilated left jugular vein, left brachiocephalic vein, and severely obstructed right SVC (24 mm × 29 mm) with only a small remaining peripheral rim of flow. A large, multilobulated cystic mobile mass was also seen attached to and nearly filling the entire RA. This mass protruded across the TV to the RV with evidence for inflow obstruction (Figure 3A–D, Videos 5 and 6).

The mass did not have a fat component based on precontrast T2-weighted and T1-weighted black blood fat saturation images. A T2* sequence revealed that signal intensity of the cystic mass is higher than the inferior vena cava (IVC) mass, while the solid mass exhibited lower signal intensity than the IVC mass. A first-pass perfusion sequence showed that the solid mass is slightly perfused, while the cystic mass did not show any perfusion. Postcontrast T1-weighted black blood images showed no change in signal intensity of either mass. Early gadolinium enhancement images showed low signal intensity in the solid mass and moderate signal intensity in the cystic mass. No late gadolinium enhancement in either mass and no ventricular myocardial scar or fibrosis was seen (Figure 3E, Video 7). There was no pleural or pericardial effusion noticed.

Based on these images, the cardiac masses were interpreted as a tumoral thrombus from metastasis of the malignant meningioma. Furthermore, the cyst was thought to be secondary to internal bleeding within the mass, and multiple thrombi were also present within the cystic mass.

After another multidisciplinary meeting including ear, nose, and throat, neurosurgery, pediatric surgery, oncology, pediatric cardiology,
and cardiac surgery. After opening the RA and the SVC to the junction with the innominate vein, a dilated azygous vein and large, partially cystic tumor filling the entire RA were observed (Figure 3F). The tumor was resected, and the SVC was ligated just proximal to the innominate vein. The pathology of this tumor was identical to the tumor that was excised 3 years before from the posterior fossa, a rhabdoid meningioma.

Serial echocardiography studies were performed every 6 months for 3 years of follow-up and revealed no recurrence of the intracardiac masses.

DISCUSSION

Cardiac masses may be associated with symptoms, but the majority are noticed incidentally during an imaging examination performed for another indication. Our patient had a rare condition of a pediatric malignant meningioma extending as extracranial metastasis to the RV through neck veins. Earlier patient case reports did not show any involvement of such a tumor in the RV in the pediatric population. Sartor et al.\(^8\) reported the case of a 63-year-old woman with World Health Organization grade 1 extracranial meningioma extending from the intracranial space through the internal jugular vein into the right SVC. Yu et al.\(^9\) described a 60-year-old woman with metastatic meningioma extending to the left atrium through the pulmonary veins.

In our case, TTE was very helpful as the initial diagnostic tool. It confirmed the large intracardiac mass with an extension into the SVC. The left innominate vein to left SVC connection was best imaged through the suprasternal notch view in the coronal plane, right parasternal longitudinal view, and modified subcostal view; however, due to a poor acoustic window as a result of increasing body mass index, for this patient a TEE was readily available and performed on the same day for further delineation of the intracardiac mass and better assessment of RV inflow using color flow and Doppler. The SVC was imaged in the midesophageal position by rotating the transducer angle from 90° to 110° from the neutral position with clockwise rotation until both SVC and IVC were visualized. Additional views of the SVC may be obtained with a transducer angle of 50° to 70° and clockwise (rightward) rotation of the probe. The real-time three-dimensional TEE provides more accurate evaluation of intracardiac mass for anatomy, size, and shape.\(^10\) Use of ultrasound-enhancing agents can help in assessing tumor vascularity by evaluating the relative perfusion of a cardiac mass.

When a tumor extends into a vessel, it may be referred to as a tumor thrombus, as in our case. Doppler echocardiography allows
for comprehensive hemodynamic assessment, which confirmed TV obstruction and reduced right ventricular stroke volume. Despite echocardiography being less expensive and more readily accessible, it is operator dependent and offers less tissue characterization and anatomic assessment than CMR.

CONCLUSION

This report illustrates the importance of multimodality imaging and the availability of a specialized multidisciplinary team in reaching a diagnosis and determining the appropriate steps in the management of this rare and challenging case.

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SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2022.07.003.

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