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10-2021

Intracardiac leiomyomatosis presenting as syncope

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Intracardiac Leiomyomatosis Presenting as Syncope

Primary cardiac tumors are rare, with incidence less than 0.1 percent. Secondary tumors are 20 times more common. Tumor thrombus extending to the right heart is commonly seen with renal cell carcinoma. Less common etiologies include intravenous leiomyoma, gynecological malignancies, Wilms' tumor, hepatoma, and adrenocortical carcinoma.

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46-year-old female with history of uterine fibroid presented with two episodes of syncope, lasting 30 seconds each. Echocardiogram revealed a mobile echogenic mass in right atrium extending from the inferior vena cava (IVC) and moving across the tricuspid valve, causing dynamic obstruction. Cardiac MRI showed the mass originated from the renal vein with similar intra-cardiac extension. Abdominal/Pelvic MRI showed a heterogeneous enhancing mass arising from the fundus of the uterus, and tumor thrombus in the left gonadal vein, extending into left renal vein and through the IVC, into the heart. She underwent exploratory laparotomy with total abdominal hysterectomy/bilateral salpingo-oophorectomy, left gonadal vein resection and tumor thrombectomy with vascular reconstruction, and removal of right atrial and ventricular tumor along with IVC tumor. Pathology of the vena cava revealed tumor thrombus consistent with Intravenous leiomyomatosis (IVL). Uterine pathology consisted of leiomyomata with leiomyomatous differentiation and IVL. Complete resection of the tumor was achieved.

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IVL is a rare uterine neoplasm of benign smooth muscle cells, which by extension grows within the intrauterine and extrauterine venous system. Intracardiac leiomyomatosis (ICL) is present in 10 – 30% of IVL.

Echocardiography revealing a right-sided cardiac mass without attachment to the endocardium or endothelial surface, originating and freely moving in the IVC without a stalk should raise suspicion for ICL. MRI abdomen/pelvis gives precise information about the tumor location and characteristics, and CT with angiography provides detailed information regarding the path of lesion with tumor extension. Complete surgical resection by a multidisciplinary team is the treatment of choice to prevent recurrence.