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Management of an isolated tricuspid valve chordal leiomyoma

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Abstract

We present the first documented case of an isolated leiomyoma arising from the chord of the anterior tricuspid leaflet.

Keywords

tricuspid valve; leiomyoma valve chord

Introduction

Primary intra-cardiac tumors are rare with an autopsy incidence between 0.001 % and 0.33 % in the general population [1], and nearly seventy five percent of these tumors are benign.

The subgroups of leiomyomas of the heart are extremely rare and are considered benign soft tissue tumors. They may arise from either uterine smooth muscle or from sub-diaphragmatic veins [2]. Case reports and case series describing intra or extra-cardiac leiomyomas have been published, but there have been no reports of chordal leiomyomas.

Case Presentation

A 60-year-old woman had a workup for two episodes of syncope during the preceding three years. She has had no other neurologic symptoms. Nineteen years earlier, the patient had had hysterectomy for fibroid. Her findings on physical examination indicated mild tricuspid regurgitation murmur.

A transthoracic echocardiography (TTE) was notable for a well-circumscribed mass arising from the anterior leaflet of the tricuspid valve measuring up to 2.1x1.5x0.5 cm in dimensions [Figure 1], and resulting in turbulent trans-tricuspid flow with a maximum velocity of 3m/s, and a pressure gradient of 25 mmHg on color Doppler imaging. The left and right ventricles were normal in size and function.

The operation was performed via a midline sternotomy. Under standard cardiopulmonary bypass with aortic and bicaval cannulation for venous return, the mass was exposed and found to be arising from and intertwined with the chordae of the anterior leaflet of the tricuspid valve.

The mass was excised without needing to resect any leaflet tissue or adjacent chordae. The tricuspid valve was repaired with a 30 mm MC3 tricuspid annuloplasty ring (Edwards lifesciences, Irvine, CA). Intraoperative transesophageal echocardiography demonstrated no residual mass and a competent tricuspid valve. The histology and the histochemical staining were consistent with a diagnosis of a leiomyoma [Figure 2, Figure 3(a-c)]. An S100 protein stain that is specific for nerve cells was negative. Her postoperative course was uneventful. She has not shown any evidence of recurrence by TTE or by pelvic computed tomography (CT) during 4.9 years of follow-up.

Discussion

After performing a MEDLINE search, we identified no reported cases of chordal leiomyoma.

Primary leiomyoma should be distinguished from two other categories: intravenous leiomyotosis and benign metastasizing leiomyomas to the heart.[2]

The former is the extension of and arising from a myometrial leiomyoma with vascular invasion through venous channels most commonly the iliac veins or other pelvic veins [3]. It has an insidious process with extension into the right heart occurring slowly over many years and cardiac function can remain uncompromised [2].

Overall, about 10 % of intravascular leiomyomas extend beyond the venous walls within the myometrium and reach the heart [3,4]. The differential diagnosis includes vegetation, other benign tumors, and thrombus. In a summary of a total of 72 cases of intra-cardiac leiomyomas reported between 1900 and 2005, iliac vein only route of extension and history of hysterectomy were reported in 48.6 % and 55.6 %, respectively [5].

Of all leiomyoma tumors, the frequency of primary myocardial leiomyomas remains unknown; many cases go unproven owing to the risk of sudden death by total right ventricular tract obstruction (RVOT). In a case series study involving 194 patients the average age of detection was 48 years, and 2% had preoperative sudden death [6]. In that study, about 30 % had a coexisting uterine leiomyoma and only 2 % were reported with a normal uterus.

In a review of 62 primary cardiac tumors, only one was a leiomyoma that probably represented an intravenous leiomyomatosis [2]. Leiomyomas were more common in women, most of child bearing age, while leiomyosarcomas are more common in men.

Benign metastasizing leiomyoma is a histologically benign growth in distant sites after hematogenous embolization [6], and usually occurs in women with a remote history of hysterectomy for fibroids [1].

In typical leiomyomas, the tumors may be symptomatic or discovered incidentally when they are clinically silent. The symptoms include congestive heart failure, syncope, tricuspid regurgitation, pulmonary embolization, Budd-Chiari syndrome, arrhythmias, or sudden death due to RVOT obstruction.

Cardiac CT has emerged as the preferred diagnostic tool, with cardiac magnetic resonance imaging as an alternative to confirm the diagnosis and to assess the extent to other cardiac structure. Surgical removal is the treatment of choice.

Should this case be labelled as a benign metastasizing leiomyoma? Not definitely, by virtue of its distinguished chordal attachment a feature that makes it a unique entity.

Conclusion

In conclusion, the present case highlights leiomyoma as a cause of chordal mass. Because it was isolated, suggests that it should not be considered an extra-pelvic subtype of leiomyoma and to our knowledge has thus far not been reported. The rarity of the disease and its frequently non-specific nature of the presenting symptoms often result in diagnostic delay; and in some cases the disease is identified only at autopsy. If untreated it could be fatal. As this case shows prompt diagnosis requires complete surgical excision and specific histochemical staining.

Figures

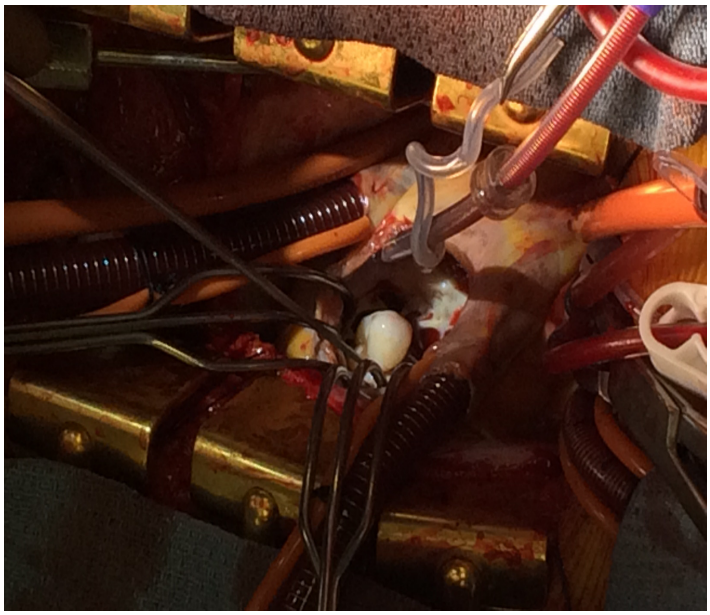

Figure 1: A preoperative transthoracic echocardiogram shows a mass at the juncture of the right atrium and the right ventricle.

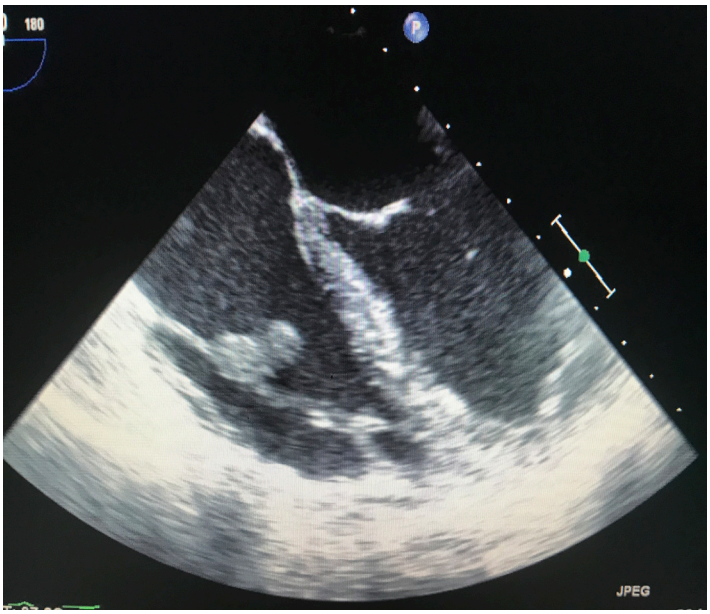

Figure 2: Intraoperative appearance of the tumor which is composed of multi-lobulated mass.

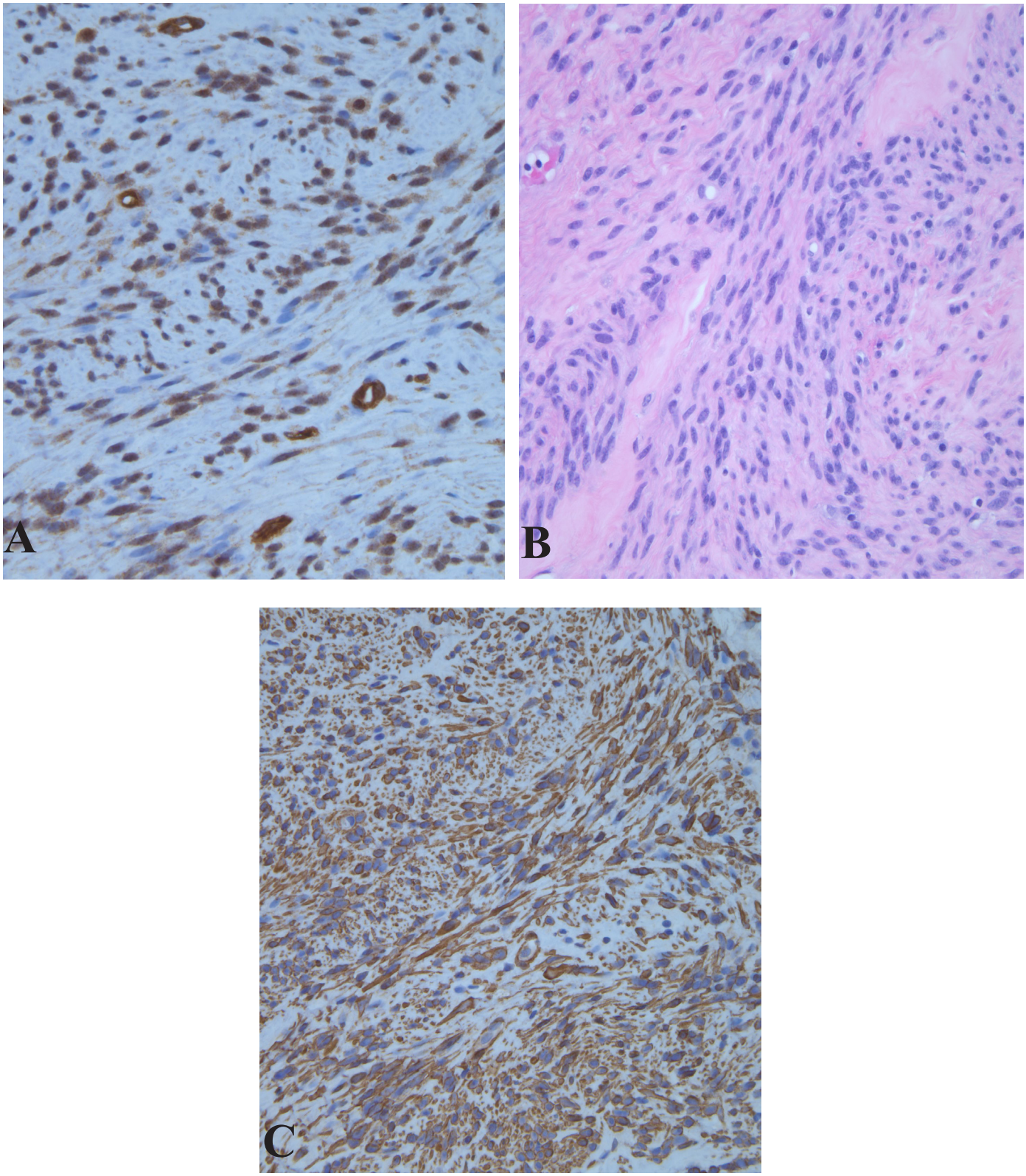

Figure 3: (A) Hematoxylin and Eosin slide at 400X magnification demonstrating fascicles of spindle cells with bland oval nuclei and pink cytoplasm. (B) Actinimmunohistochemical stain at 400x showing diffuse cytoplasmic stain of the tumor cells. (c) Desminimmunohistochemical stain at 400x showing diffuse cytoplasmic stain of the tumor cells

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