The Heart Surgery Forum #2011-1159
15 (5), 2012 [Epub October 2012]
doi: 10.1532/HSF98.20111159

Online address: http://cardenjennings.metapress.com

Primary Intravascular Synovial Sarcoma: Case Report

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ABSTRACT

Synovial sarcoma (SS), a mesenchymal spindle cell tumor, displays variable epithelial differentiation, including glandular formation, and features a specific chromosomal translocation, t(X;18)(p11;q11). SS accounts for 5% to 10% of soft-tissue sarcomas. These tumors occur mostly in the joints, especially near the knee, but they also occur in other locations. Primary intravascular SS (IVSS) are extremely rare; only 6 well-documented cases have been reported in the English literature. We describe a new case of primary IVSS of the superior vena cava (SVC) in a 16-year-old boy. A transthoracic echocardiogram confirmed a large (4.8 × 4.6 cm) circumscribed mass filling the right atrium, as well as a moderate pericardial effusion. The mass extended from the SVC to the tricuspid valve but did not prevent valve coaptation. Surgery via a transatrial approach revealed a huge mass (8 to 12 cm) attached to the SVC via a 5-mm pedicle. The tumor was excised, and the patient experienced an uneventful postoperative course. Fluorescence in situ hybridization analysis revealed the presence of the SS-specific translocation.

INTRODUCTION

Synovial sarcoma (SS), a mesenchymal spindle cell tumor, displays variable epithelial differentiation, including glandular formation, and has a specific chromosomal translocation, t(X;18)(p11;q11) [Fisher 2002]. SS accounts for 5% to 10% of soft-tissue sarcomas. Most SS (85%-95%) occur near joints, bursae, and tendon sheaths of the extremities, particularly near the knee. A significant number of SS, however, are found in anatomic sites with no obvious connection to the synovium, such as the parapharyngeal area, abdominal wall, vulva, intravascularly, the heart, and so on. The diagnosis of SS in these uncommon sites usually requires confirmation by immunohistochemical, molecular, and cytogenetic studies [Fisher 2004; Suster 2005].

Primary intravascular SS (IVSS) are extremely rare; only 6 well-documented cases have been reported in the English literature. We describe a new case of primary IVSS in the superior vena cava (SVC) of a 16-year-old boy.

Received November 1, 2011; accepted January 11, 2012.

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CASE REPORT

A 16-year-old boy presented to an emergency service with a 15-day history of dyspnea and palpitations. He was tachypneic and orthopneic. A physical examination revealed a second-degree murmur along the left sternal border that was present in both systole and diastole. The electrocardiogram revealed sinus tachycardia. A chest radiograph showed cardiomegaly. A transthoracic echocardiogram confirmed a large (4.8 × 4.6 cm) circumscribed mass filling the right atrium, as well as a moderate pericardial effusion (Figure 1). The mass extended from the SVC to the tricuspid valve but did not prevent coaptation of the valve.

With the probable diagnosis of a left atrial myxoma, we performed a sternotomy. With the patient under cardiopulmonary bypass, our transatrial approach exposed a huge mass of 8 to 12 cm that had attached to the orifice of the SVC by a pedicle 5 mm in diameter and had filled the incision site almost completely (Figure 2). The pedicle was excised from the orifice of the SVC as a button. A tumor section was taken intraoperatively, frozen, and confirmed to be malignant. On gross pathologic examination, the tumor had a uniformly shiny, glazed appearance, instead of the hemorrhagic zones and thrombi more commonly found in atrial myxomas. After closing the atrium and deairing the heart, we easily weaned the patient off cardiopulmonary bypass. The patient had an uneventful postoperative course.

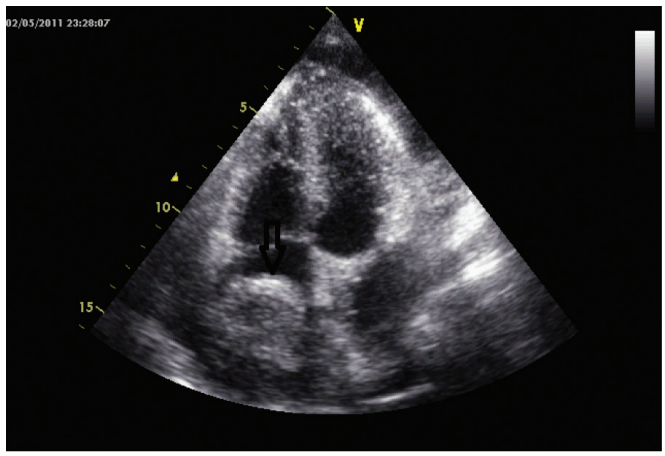

Figure 1. A large circumscribed mass filling the right atrium.

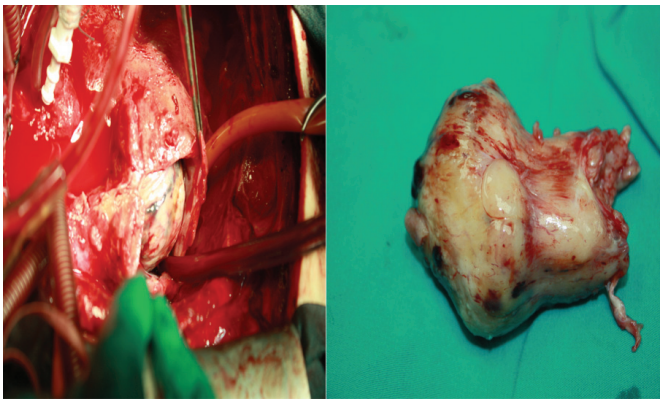

Figure 2. A soft tissue tumor attached to the orifice of the superior vena cava with a pedicle 5 mm in diameter and almost completely filling the incision site.

Pathology

Macroscopically, the tumor was 3 cm in diameter and probably limited. A cut surface was solid, was white in color, and contained foci of hemorrhage. Microscopically, the tumor was biphasic. The epithelial component tested positive in periodic acid–Schiff and diastase PAS histochemical stains and in pancytokeratin, epithelial membrane antigen, and high molecular weight cytokeratin immunohistochemical stains.

The sarcomatous component contained osteoid-like material and calcifications. The spindle cells in the sarcomatous component were immunoreactive diffusely for vimentin and focally for S-100 protein.

We carried out a fluorescence in situ hybridization analysis with the Vysis LSI SYT (18q11.2) Dual Color, Break Apart Rearrangement Probe (now Vysis LSI SS18 Dual Color Break Apart Probe; Abbott Molecular, Abbott Park, IL, USA). This analysis revealed the presence of the SS-specific translocation in the tumor cells.

Computed tomography examinations of the cranium, thorax, and abdomen, as well as magnetic resonance imaging analyses of the heart and lower extremities, were performed to detect the presence of a primary tumor, but no lesions were found. A positron emission tomography evaluation was also performed. No cancer cells were detected.

COMMENT

SS is a rare tumor affecting deep soft tissues of the lower extremities near large joints. It is most prevalent in adolescents and young adults (15-40 years old), and there is a slight male predominance. More than 60% of cases involve the lower limbs, and 20% involve the arms [Robertson 1998]. Unusual sites for primary SS include intra-articular, parapharyngeal, laryngeal, pleural, chest wall, retroperitoneal, cardiac, and genitourinary locations [Fisher 2002]. Primary tumors of the great blood vessels are rare and usually benign, but 25% are malignant. Most such tumors are sarcomas. IVSS is an exceedingly rare tumor.

This case report is of IVSS occurring in a boy. Histology, molecular, and radiologic studies indicated the tumor to be biphasic, and there was no evidence of tumor embolization from another site. The literature describes 6 cases of primary IVSS, 5 of which were in women; only 1 case involving a male patient has been reported [Coen 2008]. The clinical and histologic features of these cases are summarized in the Table. The most common complaints are pain and swelling of the lower extremities of several months' duration. One patient presented with acute abdominal pain and cardiopulmonary arrest. All cases featured extensive thrombus surrounding the tumor. Histologically, all 6 tumors were biphasic in nature, with invasion of the vessel wall without extravascular invasion.

Surgery and radiotherapy for trunk and extremity SS has an overall survival rate of 51% at 10 years and a local-recurrence rate of 3% at 5 years. Tumor size is a key prognostic factor. Tumors <5 cm in diameter are associated with a 10-year survival rate of 88%, whereas patients with tumors >10 cm have an 8% survival rate [Deshmukh 2004]. Other adverse prognostic factors include an age >40 years and areas of poor histologic differentiation. SS is most likely to recur by 2 years after excision, but metastasis can occur after a prolonged interval [Weiss 2001].

Primary SS of the heart, another extremely rare tumor, may resemble IVSS, but has different clinical and pathologic features. It is characterized by a wider age range (13-53 years of age) and a male predominance (2.5 to 1), compared with IVSS [Tong 2006]. Cardiac SS tumors have various myocardial invasion sites, and 60% of cases are monophasic histologically, in contrast with IVSS cases, all of which have been biphasic [Tong 2006].

In summary, IVSS is a rare neoplasm of the large vessels that should be considered in the differential diagnosis of primary neoplasms, leiomyosarcomas, intimal sarcomas, and malignant tumors of peripheral nerve sheaths. Moreover, IVSS should be considered in the diagnosis of venous thrombosis.

Summary of Clinical and Histologic Features of Reported Cases of Intravascular Synovial Sarcoma

Reference	Age, y	Sex	Location	Histology	Follow-up
[Miettinen 1987]	34	F	Left femoral vein	Biphasic	Free of disease at 5 y
[Shaw 1993]	31	F	Inferior vena cava	Biphasic	Died 1 d after surgery
[Robertson 1998]	34	F	Left femoral vein	Biphasic	Free of disease
[White 2005]	56	F	External iliac vein	Biphasic	Free of disease at 2 y
[Tong 2006]	32	F	Superior vena cava	Biphasic	Lost to follow-up
[Coen 2008]	41	M	Right femoral vein	Biphasic	Free of disease at 1 y

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