

Clinical Challenge

Surgery required to remove painless lump in a 7-year-old's arm

CASE AND ANALYSIS BY PAMELA HORN, MSN, CRNP, RNFA

With the help of an expert orthopedist and pathologist, a definitive diagnosis was reached in this unusual case.



FIGURE 1. The growth removed from Gina's forearm measured 2.7 cm x 2 cm.

CASE #1

Gina was an active, healthy, playful 7-year-old. One day her mother noticed a slightly “bunched up” lump in the girl's left forearm just distal to the antecubital fossa on the medial side. Gina was unaware of the painless growth. It was immobile, and no redness or warmth was present. Gina had not had any recent fever, illness, or rash. Finding no other lumps, the girl's mother considered the forearm lump unremarkable and forgot about it.

Three months later, Gina visited her primary-care clinician for her annual physical. She and her mother were getting ready to leave when, as an afterthought, her mom asked the clinician to look at the painless lump. The clinician and his associate examined the girl's arm and found nothing on which to base a definitive diagnosis. Differential diagnoses included an edematous lymph node or perhaps cat-scratch fever. Gina was subsequently referred to an orthopedist with whom she met the next day.

1. DIAGNOSIS

A plain radiograph of the left upper extremity was unremarkable. Ten days later, an MRI suggested the lump was most likely a benign tumor. Biopsy two days after identified the lump as a benign juvenile capillary hemangioma. Surgery was scheduled for three weeks later.

In the meantime, the orthopedist had the pathology slides reviewed by an expert who specialized in soft-tissue tumors. When Gina and her mother arrived for the preoperative meeting with the surgeon, he had just received the results. Gina was diagnosed with synovial cell sarcoma, a malignant soft-tissue tumor.

2. TREATMENT

The patient was eventually referred to our facility, where our oncology team administered neoadjuvant chemotherapy.

Because of the nature and location of the tumor, Gina was then seen by a surgeon who spe-

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cialized in oncologic orthopedics. After the procedure and its risks and benefits were explained to Gina's mother, our young patient was taken to the operating room. Thanks to the chemotherapy, the tumor had decreased in size. It encased her ulnar artery, which was transected. Since the sarcoma circumferentially surrounded her left brachial artery, it was decided that she would need post-resection arterial and soft-tissue reconstruction to help with future arterial patency. Gina would also need postoperative radiation therapy.

The patient was given a general anesthetic and prepped for surgery in the supine position using strict sterile technique. Once she was administered the correct doses of IV antibiotics, the forearm tumor was excised with an appropriate soft-tissue cuff to obtain negative margins. Careful attention to the median and radial nerves found no evidence of nerve involvement (*Figure 1*). The tumor measured 2.7 cm × 2 cm. Inked margins were free of tumor, and no vascular invasion was evident.

Following the excision, Gina's fingers were noted to be very cool and dusky, with capillary refill >3 sec. Because the tumor surrounded the brachial artery, the surgeon completed a radial forearm flow-through flap reconstruction of the artery using an operative microscope. Once the arterial anastomoses were successful, the limb regained a pink color, and Gina's capillary refill was <1 sec. A drain was placed away from the harvested vessels to wick away fluid and decrease edema. The flap was completed, her tissues were sutured closed, and the hand remained pink and viable throughout the case (*Figure 2*). With a sterile dressing in place, Gina was taken to the recovery room.

Since her surgery, our patient's wounds have all healed wonderfully, and she has excellent function. Once Gina completes her postoperative chemotherapy, she will undergo radiation therapy.

3. ANALYSIS

Synovial cell sarcoma is a rare soft-tissue tumor that is unrelated to synovial fluid and can occur in almost any part of the body.¹ The majority of tumors occur in para-articular structures in the extremities; most are localized at diagnosis.² These malignant, high-grade, soft-tissue neoplasms account for 7%-8% of all malignant soft-tissue tumors. They are the most common non-rhabdomyosarcoma soft-tissue sarcomas in pediatric patients, representing 5%-10% of all soft-tissue sarcomas.³ An estimated 800 new cases occur annually in the United States.² The median patient age is 30 years, with an almost equal distribution between males and females. Approxi-



FIGURE 2. A drain was used to remove fluid and decrease edema.

mately 70% of cases occur in patients aged 20 years or older. Patients younger than 20 years are mainly affected in the second decade of life.² Presence of this tumor in children younger than age 10 is very rare.⁴

Synovial sarcoma patients usually present with an asymptomatic swelling or mass. As the tumor grows, patients may complain of pain, discomfort, or loss of function in the area. These symptoms and fatigue or fever can be related to metastases and/or mass effects.⁵ Most tumors are localized at diagnosis,² which is based on histology demonstrating one of three major subtypes: monophasic, biphasic, and poorly differentiated. Synovial sarcoma is defined by the presence of the *SYT-SSX* fusion gene.⁵

Effective treatments include surgery, radiation, and/or chemotherapy. Complete resection is vital for survival of patients with localized synovial sarcoma.⁶ The literature is mixed with regard to the use of surgery alone (only in the case of very small, early tumors), radiation alone after surgery, chemotherapy alone (before and/or after surgery), or a mixture of all three approaches.⁶⁻⁹

One study noted an excellent overall survival rate (87%) with surgery and radiation. Chemotherapy did not appear to have the same impact, although this is a relatively new approach, and additional larger trials are needed.⁶ Radiation therapy can have latent side effects. Some children required surgery to correct a late effect, and others sustained fractures in the irradiated bone. Very few sustained radiation-induced bone malignancy.¹⁰

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One theme in the literature is recurrent. Competent early imaging and immunologic, histologic, and genetic testing will improve patient outcomes. Combined treatment can achieve full remission in the majority of patients with soft-tissue sarcomas localized to the limbs. If the tumor is <5 cm, total resection may be the only treatment necessary (radiation could follow). In patients with large tumors (>5 cm), the treatment should be started with inductive chemotherapy before wide-excision surgery. Tumors >5 cm were associated with a worse prognosis. In comparison with adults, the prognosis of primary, localized non-rhabdomyosarcoma soft-tissue sarcomas in children appears to be more favorable. The newer methods of adjunctive and neoadjuvant treatments can result in outcome improvements for patients with synovial cell sarcoma.⁷⁻⁹

These types of malignancies are extremely rare among the pediatric population. Patients should be referred to a specialist if at all possible. At the very least, initiate imaging studies to be sure.

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Typical angina with an atypical diagnosis

CASE AND ANALYSIS BY MARK A. McELHANNON, MD

The patient continued her aerobic workouts despite persistent chest pressure with an unknown etiology.

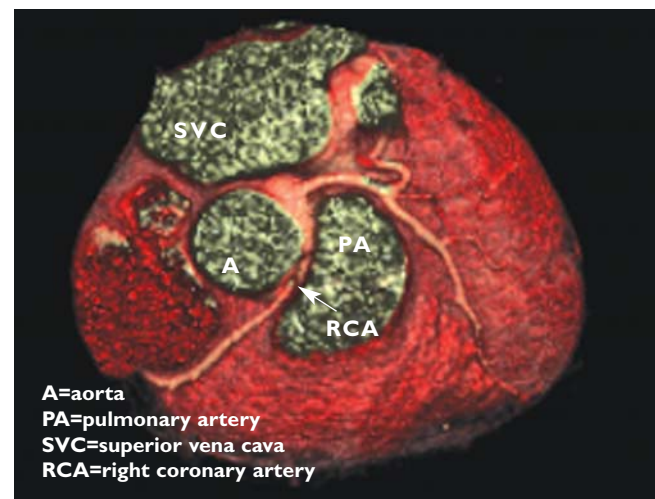


FIGURE 1. Coronary angiography revealed an unexpected finding (arrow).

CASE #2

A 39-year-old woman complained of exertional substernal chest pressure. The pressure occurred in a typical pattern after 20–25 minutes of aerobic exercise and resolved within several minutes of rest. She denied any palpitations, syncopal episodes, or chest pressure at rest. Despite her symptoms, she continued her thrice-weekly aerobic workouts.

Earlier, another physician had diagnosed costochondritis and prescribed a trial of anti-inflammatory medications. The woman's symptoms persisted, however, and after several months, she sought another evaluation in our clinic.

Her past medical history was most notable for breast augmentation surgery. There was no family history of coronary artery disease (CAD) or sudden cardiac death. Social history was positive for ongoing tobacco use, approximately one pack per day for 20 years. She took no chronic medications.

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