Case Report

Unsuspected Widespread Cardiac Sarcoma in a Child

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ABSTRACT

The case of a patient with an undifferentiated metastatic cardiac sarcoma is presented. A thallium-201 tumor study was performed to evaluate lung nodules. Thallium-201 chloride was injected intravenously and whole body images, as well as single photon emission computer tomography (SPECT) imaging of the chest, were obtained and reconstructed. They were displayed in three planes and then reconstructed again in cardiac planes. Multiple unsuspected metastases were found in the lower extremities. Viable tumor was demonstrated in both cerebral hemispheres. The lung nodules were not clearly identified. However, cold areas in the myocardium were detected corresponding to the findings on magnetic resonance imaging (MRI). In retrospect, at least one abdominal metastasis was identified. Whole body tumor imaging plays an important role in finding unsuspected primaries and in staging neoplasms to optimize patient care.

INTRODUCTION

A rare case of primary cardiac sarcoma that was known to be metastatic to the brain and suspected to be the etiology for multiple pulmonary nodules is presented. Thallium tumor images demonstrated the metastatic brain lesions and additional widespread disease in both lower extremities that were confirmed on magnetic resonance imaging (MRI) and computed tomography (CT). CT showed multiple abdominal metastases that in retrospect were present on the thallium scan.

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CASE HISTORY

A Caucasian female, 13 years of age, presented to the emergency department with sudden onset of multiple seizures involving the right arm and right side of her face, without loss of consciousness, lasting approximately 1.5 minutes each. The preceding week the patient had a history of morning headaches and vomiting. There were no symptoms of dyspnea, cough, fever, or weight loss. There was no history of trauma, drug abuse, or bleeding diathesis. The family history was not suggestive of seizures, hypertension, connective tissue disease, thrombotic disorders, or stroke. The medical history was unremarkable.

Physical examination

The weight of the patient was 88.8 kg, height 143 cm, blood pressure 120/80, heart rate 80/minute, respiratory rate 16/minute, temperature 97.7°F, and O₂ saturation 97% on room air. On neurological examination the patient was alert, oriented and in no discomfort. There was right-sided hemiparesis and facial palsy. The sensory system was intact. The cardiovascular, respiratory and gastrointestinal systems were normal.

Laboratory data

The results of a complete blood count, chemistry profile, prothrombin time and partial thromboplastin time, were normal at presentation. Bone marrow aspirate and biopsy were negative.

Radiology

The chest radiograph was read as negative (not shown). CT of the head (figure 1) showed multiple lesions in both frontal lobes. The MRI showed a high signal T2WI left cerebellum lesion measuring 3 to 4 cm, all consistent with cerebral metastases. The patient also had a CT of the chest, abdomen and pelvis that were negative, except for the infiltrating cardiac mass (figure 2). Multiple pulmonary nodules were suspicious for, but not biopsy proven, to be metastases. On the right were 2 pulmonary micronodules (submillimeter) and on the left 5 nodules; 3 micronodules, one measuring 14 x 15 mm, and another measuring 13 x 14 mm (not shown).

Echocardiography

Transthoracic echocardiography showed a cardiac mass in the left ventricle arising from the papillary muscle measuring 4 x 3 cm and attached to the left interventricular septum, without involvement of valves or obstruction to blood flow. This was confirmed with an MRI of the heart (figure 3).

Patient course

The patient underwent a partial excision of the cardiac tumor. The biopsy suggested high-grade undifferentiated sarcoma. The initial treatment was chemotherapy consisting of vincristine sulfate (Oncovin, Eli Lilly and Company, Indianapolis, IN), doxorubicin hydrochloride (Adriamycin, Pharmacia & Upjohn, Kalamazoo, MI), cyclophosphamide (Cytosan, Bristol-Myers Squibb Company,

Princeton, NJ), alternating with ifosfamide (Ifex, Bristol-Myers Squibb, Princeton, NJ) and etoposide (Etopophos, VP-16, Bristol-Myers Squibb, Princeton, NJ).

Two months into chemotherapy, the patient developed increasing sizes of the central nervous system (CNS) metastasis and impending brainstem herniation. An emergency resection of the occipital metastatic lesion was performed. A mass developed on the anterior right chest wall. This was resected and the biopsy showed undifferentiated sarcoma. The immediate post-operative phase was complicated by wound sepsis, which was treated with antibiotics. Chemotherapy was delayed due to the infection.

As the metastatic lesion had progressed on chemotherapy, the chemotherapy was changed to a VP-16, ifosfamide, platinum (VIP) regimen used for refractory sarcomas.¹ The patient received two courses of VP-16, cisplatin (Platinol-AQ, Bristol-Myers Squibb, Princeton, NJ) and ifosfamide, and showed a partial tumor reduction in the CNS metastatic lesions. The cardiac MRI showed a significant response in tumor size. Two additional courses of VIP were given. The CT evaluation of the CNS revealed further reduction in the masses, resolution of cerebral edema and improvement in the right-sided hemiparesis. The patient no longer had seizures, but remained on anti-convulsants.

Following chemotherapy, the patient developed Gram-negative *Enterobacter cloacae* sepsis of the central line, which was effectively treated with ceftazidime (Fortaz, Glaxo Wellcome Inc, Research Triangle Park, NC) and tobramycin sulfate (Nebcin, Eli Lilly Co, Indianapolis, IN). Prior to the restart of chemotherapy, the patient was readmitted with a *Staphylococcus epidermidis* and vancomycin-resistant enterococci central line infection. The central line was removed and replaced with a peripherally inserted central catheter (PICC) line. The infection was treated with antibiotics and chemotherapy was postponed.

The persistent infections delayed the chemotherapy for approximately 6 weeks. It was important to define the extent of the disease, before proceeding with further treatment. This could be determined by using either an 18-fluoro-2-deoxyglucose positron emission tomography (18-FDG PET), or tumor imaging with Thallium-201 (Thallous chloride TI-201, Bristol-Myers Squibb Medical Imaging, Inc, N. Billerica, MA), or Cardiolite (Tc-99m Sestamibi, Bristol-Myers Squibb Medical Imaging, N. Billerica, MA) scans.

The patient was referred to nuclear medicine for a PET scan. The scan was not performed, the patient's insurance did not approve coverage, in that PET is not recognized for this diagnosis. Instead, tumor imaging with Thallium-201 chloride was selected because of its proven track record for sarcoma evaluation.

To reduce the chance of vomiting and nausea, the thallium scan was done with the patient's head in a near vertex view. This decreased the visibility of the neck, but clearly demon-

Figure 1. CT of the head shows multiple cerebral metastases.

Figure 2. MRI of the heart shows multiple areas of the cardiac sarcoma.

Figure 3. CT scan of the chest showing an infiltrating tumor of the myocardium.

strated viable tumor in both cerebral hemispheres. The lung nodules were difficult to evaluate because of the normal background activity. Even with the single photon emission computer tomography (SPECT) imaging, the lung nodules were not identifiable. There were cold areas on the planar

and SPECT images (figure 4) where MRI demonstrated cardiac sarcoma. Incidentally, there were multiple unsuspected abnormal foci of increased abnormal activity in the hips and legs (figure 5A and 5B).

Figure 4. SPECT myocardial perfusion images demonstrate cold apex where MRI shows tumor.

Figure 5. Whole body thallium scan shows multiple metastases in the hips and lower extremities and in the lower abdomen. **A.** Light copy for cardiac lesion. **B.** Dark copy for the abdominal and extremity lesions.

Figure 6. MRI of the right leg shows multiple metastases.

An MRI of the right leg demonstrated multiple lesions (figure 6) corresponding to the thallium scan. An abdominal CT scan showed multiple metastases, of which at least one was seen in retrospect on the thallium scan (figure 7). Normal gut activity decreased thallium sensitivity of the abdominal metastases. In retrospect, the largest lesion can be easily identified with its necrotic center.

Approximately 3 weeks later, the patient presented with abdominal pain, nausea, bilious vomiting and a partial bowel obstruction. An abdominal x-ray showed marked dilatation of the small bowel and a relatively collapsed colon. The combination of findings suggested that there was a bowel obstruction, probably in the distal small bowel. An upper endoscopy revealed no obstruction but found gastritis and duodenitis. The colonoscopy was normal. Limited upper gastrointestinal series showed no obstruction through the area of the duodenum.

An abdominal CT scan revealed several endoluminal, probably mucosal metastases within the jejunum and ileum, dilated small bowel down to the right lower quadrant, and a relatively large, bulky mass lesion. At this point, the obstruction was believed to be a metastatic tumor and surgery was considered. The patient had multiple dilated loops of bowel at surgery. The small bowel had intraluminal metastases distal to the ligament of Treitz, as well as extraluminal small bowel metastases. The largest site was near the distal ileum and the ileocecal valve, where there was a large pelvic mass incorporating the mesentery of the small bowel, pelvic and abdominal wall. It was not resectable and was surgically bypassed. Because of extensive disease in the abdomen involving the jejunum, mesentery and terminal ileum, no treatment was pursued and palliative care was done. The patient died because of disseminated disease within 2 weeks.

Figure 7. CT of the abdomen shows multiple metastases. The most inferior one is necrotic. In addition, there are multiple hip metastases.

DISCUSSION

In summary, this report is a rare case of disseminated, poorly differentiated cardiac sarcoma, where thallium tumor imaging played an important role. Thallium imaging was introduced in the early 1970s. At least one comment in the literature suggested it might be good for tumor imaging.² In 1976 the first incidental tumor was detected using thallium imaging, which confirmed the original belief.³ As part of our reason to choose thallium, some reports have discussed cerebral metastases similar to this case. The lesion presented by this patient could be used as a control.^{4,5} It is the experience of the authors that most insurance companies will cover thallium tumor imaging.

PET is the first study of choice for most tumor imaging, including sarcoma evaluation. However, in this case an application to the Centers for Medicare and Medicaid Services for soft-tissue sarcoma approval was denied on the basis of lack of evidence. Preauthorization is obtained for all PET requests. In this case, the insurance company would not cover indications beyond the approved list of the Centers for Medicare and Medicaid Services.

CT and MRI have yet to offer on a large scale whole body imaging, which is the strongest point of nuclear medicine. Sometimes unsuspected primaries or metastases are identified.⁶⁻⁸ For example, a gallium tumor scan showed a single sacral metastasis that changed an operable carcinoma into an inoperable carcinoma (unpublished), for a patient with lung carcinoma. Other cases have detected the unknown primary tumor from a bone scan in renal soft tissue that was unsuspected.

The myocardial perfusion images were incidental and not primary in this case. The apical cardiac sarcoma was cold on the myocardial perfusion images, while the metastases were hot. Recently, a case of adult cardiac rhabdomyosarcoma was published that showed a relatively cold perfusion defect.⁹ However, this was not a benign lesion nor was it present in a young child. Another case report using Thallium-201 described a primary myocardial sarcoma in the right ventricle that thallium showed extended directly into the region of the pulmonary artery.¹⁰ An autopsy proven liposarcoma metastatic to the left ventricle showed up on a thallium myocardial perfusion study as a cold defect.¹¹ Neither of these cases describe primary cardiac sarcoma with widespread metastases.

Cardiac sarcomas have been described in the literature, but not widespread metastases, as in this case. The 7 pulmonary nodules were not demonstrated in this case probably due to their small size. However, other more important metastatic lesions were demonstrated, which have not been previously described in the literature.

Nuclear medicine tumor imaging plays an important role in the staging of most neoplasms, even though there is an increasing role and remarkable anatomic images from ultrasound, CT and MRI. Whole body MRI is still not practical for all, or most patients. Until whole body MRI is practical for the majority of patients it is useful to remember that once a radioisotope is injected, multiple images can be obtained with no additional radiation exposure. Remember to look for the incidental findings, they may be more important than the primary reason for the examination.

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