Case Report: Angiosarcoma in the Upper Extremity Related to a Nonfunctioning Arteriovenous Fistula

HAND
2017, Vol. 12(5) NP132–NP135
© The Author(s) 2017
Reprints and permissions:
sagepub.com/journalsPermissions.nav
DOI: 10.1177/1558944717702466
hand.sagepub.com

Andrea L. Gale^{1,2}, Rex A. W. Marco^{1,2}, Shari R. Liberman^{1,2}, Dmitry Zavlin^{3,4}, and Anthony Echo^{3,4}

Abstract

Background: Angiosarcoma is a rare but aggressive malignancy with a high predilection for the head and neck. There have been few case reports of the development of angiosarcoma in a nonfunctional arteriovenous fistula in a hemodialysis patient. **Methods:** After institutional review board approval, a retrospective chart analysis is conducted to present the case of a patient who received plastic surgical treatment at our hospital. **Results:** We present the case of a 44-year-old male who presented with ischemic changes and paresthesias of the upper extremity associated with the development of angiosarcoma in an arteriovenous fistula. **Conclusions:** Angiosarcoma remains a rare but important diagnosis to include in the differential diagnosis for upper extremity pain and paresthesias in chronic renal failure and nonfunctioning arteriovenous fistula.

Keywords: angiosarcoma, arteriovenous fistula, ischemia, chronic renal failure, radial artery

Background

Angiosarcoma is a rare but aggressive malignancy that is present in the adult population. It represents approximately 1% of all soft tissue sarcomas.¹ Angiosarcoma can arise in any tissue of the body but has a high predilection for the head and neck region.¹ Angiosarcoma has been reported to present in the extremities in areas of chronic lymphedema such as post mastectomy (Stewart-Treves syndrome), areas of local radiation, and exposure to foreign materials with chronic inflammation.² To our knowledge, there have been 22 adult patient cases in the English literature that have demonstrated the development of angiosarcoma at the site of a nonfunctional arteriovenous (AV) fistula for hemodialysis access.³

We present the case of a 44-year-old man who presented with hand ischemia and paresthesias in the left upper extremity for 3 months, who was found to have an angiosarcoma in a nonfunctional AV fistula.

Case Report

This is the case of a 44-year-old left-hand-dominant male with a medical history significant for end-stage renal disease secondary to focal segmental glomerulosclerosis status post cadaveric kidney transplantation. He additionally had a left-sided radiocephalic fistula formation performed approximately 6 years prior for planned hemodialysis; however, his fistula

had never been accessed. He reported to the emergency department with a 2-day history of necrotic change to his left ring and small fingers. A duplex Doppler performed at that time showed evidence of a radial artery thrombosis. He was admitted to hospital, and a heparin drip was initiated. A hypercoagulability workup was performed and found to be negative. Intraoperative angiogram (Figure 1) demonstrated a long segment filling defect in the radial artery which necessitated the radial artery ligation. The ulnar artery was found to be patent; however, there was incomplete filling of the deep palmar arch. Stenosis of the brachial artery was noted as well, and a left brachial artery path angioplasty was performed to enhance the flow to the ulnar artery. He tolerated the procedure well and was discharged from the hospital. The preoperative ischemic changes were stabilized following this procedure with no progressive levels of necrosis noted.

¹Department of Orthopedic Surgery, Houston Methodist Hospital, TX, USA

²Department of Orthopedic Surgery, Weill Cornell Medicine, New York, USA

³Department of Plastic Surgery, Houston Methodist Hospital, TX, USA

⁴Department of Plastic Surgery, Weill Cornell Medicine, New York, USA

Corresponding Author:

Anthony Echo, Institute for Reconstructive Surgery, Houston Methodist Hospital, 6560 Fannin Street, Suite 2200, Scurlock Tower, Houston, TX 77030, USA.

Email: aecho@houstonmethodist.org

Figure 1. Angiogram of the left upper extremity, proximal to distal, demonstrating a long segment filling defect of the radial artery. (a) View of elbow. (b) View of forearm. (c) View of hand.

Figure 2. Coronal magnetic resonance image that demonstrates extension of mass along the forearm to the level of the radiocarpal joint. (a) T1 sequence. (b) Short tau inversion recovery.

Note. Arrows show extent of pathological region.

Approximately 2 weeks following his radial artery ligation and brachial artery patch angioplasty, the patient presented for routine postoperative follow-up with a new mass in his antecubital fossa. At this time, he endorsed pain and paresthesias in his left wrist and forearm in the median, ulnar, and radial nerve distributions that had been present for the previous 3 months. The patient had not endorsed these symptoms of paresthesias at his previous hospital admission but reported them at his first postoperative follow-up appointment as being chronic in nature. Electromyogram was attempted at an outside facility; however, it was unable

to be completed due to the significant swelling of the forearm. Magnetic resonance imaging of the left upper extremity (Figure 2) was performed and demonstrated loculated fluid collections along the course of the radial artery from the level of the antecubital fossa distally to the level of the radiocarpal joint. It was felt that this fluid collection likely represented hematoma formation in the acute postoperative period which was producing increasing paresthesias through a mass effect.

The patient was then taken to the operating room for evacuation of the presumed hematoma. Upon exploration of the surgical bed, it was determined that no significant hematoma could be identified. Instead, the AV fistula site was found to be an enlarged firm mass with irregular margins that was densely adherent to the surrounding tissues with poorly demarcated tissue planes. Given this unexpected appearance of the fistula, the fistula was resected for pathological evaluation. Further exploration of the radial artery demonstrated the distal segment to be filled with a large amount of presumed thrombus within the lumen, and there was very little back flow in the remaining vessel. The distal radial artery was ligated, as the patient did have a complete palmar arch and some back flow filling from the ulnar artery, and specimen sent for culture and pathology (Figure 3). Pathology demonstrated angiosarcoma within the nonfunctional AV fistula.

Staging was completed with a computed tomography of the chest, abdomen, and pelvis. The patient was found to have nonmetastatic disease and subsequently underwent a forequarter amputation for tumor control (Figure 4). Pathologic examination of the complete upper extremity revealed extension of the tumor from the antebrachial fossa throughout the radial volar compartment to the level of the radiocarpal joint along the course of the radial artery with

Figure 3. Gross specimen from exploration of the arteriovenous fistula. (a) Large sample. (b) Smaller samples.

diffuse extension within the volar musculature. Three months following amputation, he was found to have metastatic disease to the right upper lung and the left iliac wing. The metastatic lesions were treated with chemotherapy. He underwent embolization of the right upper lung lesion prior to completion of chemotherapy due to significant hemorrhage within the lesion.

Discussion

Angiosarcoma is a rare cancerous lesion of the vascular system. A recent review of the literature found 22 reported cases of angiosarcoma developing within a nonfunctional AV fistula for dialysis.³ Nineteen of 22 patients were status post renal transplant, and 18 of 22 patients were on long-term immunosuppressive therapy (greater than 12 months duration).³ Oskrochi et al also noted that the average time to symptoms following AV fistula formation was 118.9 months and 96.9 months after transplantation, while the mean age of presentation was 54.9 years, and 20 of 22 patients were male.³ The most common presenting symptoms were pain with or without formation of a mass, and the initial diagnosis was most often thrombosis of the fistula with a delay in correct diagnosis between 2 to 40 weeks.³ Amputation was

Figure 4. Gross images of the angiosarcoma extension throughout the volar compartment of the forearm (left: distal, right: proximal).
Note. Arrows show extent of pathological region.

the most common treatment, and survival was an average of 8.8 months.³ Historically, angiosarcoma has a 5-year survival rate of 10% to 20%.²

Our patient differs from this population in the timing of his presentation. His AV fistula was established 72 months prior to his presentation and had never been accessed. His transplantation occurred 8 months prior to his presentation, and subsequently, he had only been on immunosuppression for less than a year at the time of diagnosis. While he did present initially with ischemic changes and paresthesias, he never displayed motor impairment of the radial, ulnar, or median nerves. Similar to the patients that were included in this review, our patient was initially misdiagnosed with a thrombosis of his fistula and hematoma. Misdiagnosis of thrombosis, infection, or hematoma has led to delay in oncologic diagnosis in all reported cases of angiosarcoma arising in an AV fistula.

Immunosuppression has been shown to be an independent risk factor in the development of cancers in the transplant population.⁴ Oskrochi et al found that there were 4 reported cases of the development of angiosarcoma in an AV fistula in patients who were not on long-term immunosuppressive therapy.³ This would be the fifth reported case in the English literature of the development of angiosarcoma in a patient who had not been on long-term immunosuppressive treatment, as this patient had only been treated with immunosuppressive therapy for 5 months prior to developing symptoms.

Angiosarcoma is a rare condition but an important consideration in the differential diagnosis in a hemodialysis patient presenting with wrist pain and paresthesias. Hand surgeons should be aware of this potential etiology. These patients can present with forearm pain or paresthesias as

well as discoloration of the skin. The development of a mass is variable. This patient presented with paresthesias and pain in the median, ulnar, and radial nerve distributions. He also presented with decreased range of motion at the elbow. The involvement of all 3 major nerves of the forearm should raise suspicion for an alternate etiology, especially in the presence of a palpable mass. This patient developed enlargement of the entire forearm which precluded electrodiagnostic evaluation of his neurologic symptoms. In a patient who presents with polyneuropathy and swelling of the upper extremity in the presence of a nonfunctioning AV fistula, angiosarcoma should be included in the differential diagnosis, and all operative interventions should include histological evaluation. In the setting of nonmetastatic disease, amputation is the treatment of choice; however, this aggressive tumor uniformly has a poor survival rate.

Ethical Approval

The work described in this article was approved by our institutional review board.

Statement of Human and Animal Rights

All procedures were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008.

Statement of Informed Consent

Informed consent in writing was obtained from all patients for being included in the study, and no patient is identifiable in this article.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

References

1. Bardwil JM, Mocega EE, Butler JJ, Russin DJ. Angiosarcomas of the head and neck region. *Am J Surg*. 1968;116(4):548-553.
2. Demey K, Reyns LM, Schepers S. Angiosarcoma arising in an arteriovenous fistula in a patient without kidney transplant. *Acta Chir Belg*. 2014;114(1):75-78.
3. Oskrochi Y, Razi K, Stebbing J, Crane J. Angiosarcoma and dialysis-related arteriovenous fistulae: a comprehensive review. *Eur J Vasc Endovasc Surg*. 2016;51(1):127-133.
4. Qureshi YA, Strauss DC, Thway K, Fisher C, Thomas JM. Angiosarcoma developing in a non-functioning arteriovenous fistula post-renal transplant. *J Surg Oncol*. 2010;101:520-523.