Soft Tissue Sarcomas in Disused Arteriovenous Fistulae

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Dear Editor,

We read with interest the recent paper describing a soft tissue sarcoma associated with a dialysis access site (arteriovenous fistula [AVF]) in a heavily immunosuppressed patient.[1] We refer to a series of similar malignancies in disused arteriovenous fistulae in immunosuppressed transplant patients at our institution.[2] This series included 3 angiosarcomas and a lymphoma; all presented over a 2-year period. Of these cases, 2 of the 3 angiosarcoma patients, died within 2 years.

Our first case, an epithelioid angiosarcoma, developed 2 years after an ABO-incompatible transplant; the patient presented with pain and swelling in a brachiocephalic fistula. In this instance, desensitization was achieved with plasma exchange, rituximab, and daclizumab. Immunosuppression was maintained with tacrolimus. The patient was treated with an above-the-elbow amputation and remains well to date, with a functioning transplant.

The second case in our series was a hemodialysis patient with a right-arm brachiocephalic AVF. Three years after beginning dialysis, she received a live-related renal transplant, with alemtuzumab and methylprednisolone given at induction, and tacrolimus given for maintenance immunosuppression. Three years after the transplant, she presented with pain in her AVF. A magnetic resonance image showed a thrombosed fistula, and importantly, bone edema and abnormal signaling in the ulna. One month later, she developed dusky, purple, palpable, nontender cutaneous and subcutaneous nodules, measuring 1 cm in diameter, all distal to the AVF. A biopsy revealed an epithelioid angiosarcoma. The patient died from lung metastases 1 month after presentation.

Our final case of a sarcoma occurred in a fistula some 14 years after its initial formation, and 8 years after a deceased-donor renal transplant. In this case, induction was accomplished with methylprednisolone, and maintenance was done with mycophenolate mofetil and tacrolimus. The patient had presented initially with warty growths on the fistula, and as he was Epstein-Barr-virus positive, he was first managed with withdrawal of his mycophenolate mofetil, after an initial biopsy proved inconclusive. A repeat biopsy of the fistula revealed an epithelioid angiosarcoma. The patient died from lung metastases within 6 months.

As discussed in our article and others, angiosarcomas are a rare and exceptional complication, occurring within an AVF after renal transplant. There are multiple theories regarding why a malignancy may develop in a fistula given a background of heavy immunosuppression. One postulate is that it could be because of an impaired immune response given the background of a disrupted lymphatic drainage. Other suggested possible factors include oscillatory blood flow causing stress to the endothelial cells, leading to up-regulation of growth factors, or possibly, relative limb ischemia, secondary to the fistula, leading to T-cell suppression and hence—a reduced immune response.

We note that there are only a few similar cases reported, and that all occur in those who are immunosuppressed. Managing this condition surgically is difficult as the only chance of cure is in those cases without metastatic spread. However, it is prudent to make clinicians aware of this important, albeit rare, differential diagnosis, when a patient

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describes new symptoms in a disused fistula after renal transplant. Furthermore, we suggest that prompt imaging and biopsy should be performed to allow for early diagnosis and to minimize morbidity and mortality from this condition.

References

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