A case of angiosarcoma metastatic to bone marrow

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Case Report

Abstract

Cutaneous angiosarcoma is rare and accounts for only around 1% of all sarcomas. It most commonly presents as a rapidly enlarging erythematous-to-violaceous patch on the head, neck or trunk. Many affected patients are elderly or have a preceding history of cancer, mastectomy, radiation or lymphedema. The prognosis for cutaneous angiosarcoma is generally poor; however, survival rates seem to vary substantially based on patient age and primary site of involvement. The most common sites of metastases from cutaneous angiosarcoma are the lungs, liver, spleen and brain. Metastasis of angiosarcoma to the bone marrow is exquisitely rare. We report the case of a 45-year-old woman without any classic risk factors who presented with primary cutaneous angiosarcoma of the upper back that subsequently metastasized to the bilateral breasts, liver, subcutaneous tissues, spine and bone marrow.

Keywords: Angiosarcoma; Cutaneous Angiosarcoma

Introduction

Cutaneous angiosarcoma is rare and accounts for about 1% of all sarcomas. It most commonly presents as a rapidly enlarging erythematous-to-violaceous patch on the head, neck or trunk. Cutaneous angiosarcoma is more common in patients who are elderly or have a preceding history of cancer, mastectomy, radiation or lymphedema. The prognosis for cutaneous angiosarcoma is generally poor; however, survival rates seem to vary substantially based on patient age and primary site of involvement.

The most common sites of metastasis from cutaneous angiosarcoma are the lungs, liver, spleen and brain. Metastasis of this particular tumor to the bone marrow is exquisitely rare. We report the case of a 45-year-old woman who presented with primary cutaneous angiosarcoma of the upper back, which subsequently metastasized to the bilateral breasts and bone marrow.

Case presentation

A previously healthy 45-year-old female without any history of breast cancer, radiation or lymphedema presented to the Dermatology clinic for evaluation of a purplish skin lesion on her left upper back which had been enlarging over the past year. (Figure 1) Biopsy of the lesion revealed atypical vascular spaces lined by plump endothelial cells involving the dermis and subcutaneous tissue, a mitotic rate of 28 per 10 high power fields and foci of necrosis. These findings were consistent with high-grade cutaneous angiosarcoma. (Figure 2) CT of the chest, abdomen and pelvis revealed numerous lesions in the bilateral breasts, concerning for metastases. (Figure 3) Core biopsy of several of the masses revealed complex vascular structures exhibiting endothelial atypia, significant mitotic activity and foci of necrosis, findings consistent with metastatic angiosarcoma. This impression was independently confirmed by a reference pathology center.

The patient underwent radical resection of her cutaneous mass as well as bilateral mastectomy, both with reportedly negative margins. Following surgery, she began chemotherapy with Paclitaxel and Bevacizumab, which she continued for 4 months before follow-up CT revealed multiple subcutaneous nodules in her upper back and left thigh. Paclitaxel and Bevacizumab were replaced with Gemcitabine, which she continued for 3 months before PET/CT revealed multiple liver lesions and interval progression of her subcutaneous nodules. (Figures 4-5) Gemcitabine was replaced by Doxorubicin, which she continued for 1 month before routine lab work revealed anemia and tear drop cells.

PET/CT performed at that time revealed multiple lesions in her spine as well as interval progression of her liver lesions and subcutaneous nodules. (Figure 6) Bone marrow biopsy was performed to evaluate her new cytopenias and revealed dilated vascular spaces lined by atypical endothelial cells, consistent with metastatic angiosarcoma. (Figure 7) Doxorubicin was replaced with Pazopanib. At the time of this report, the patient was still alive and on Pazopanib, 15 months after initial presentation.
FIG 1: Violaceous lesion on left upper back at original time of presentation.

FIG 2: Hematoxylin and eosin stain of left upper back cutaneous lesion at 40x magnification. Demonstration of atypical vascular spaces lined by plump endothelial cells, consistent with angiosarcoma.

FIG 3: CT of the chest, abdomen and pelvis at initial workup. Demonstration of both the primary cutaneous angiosarcoma of the back.

FIG 4: CT of the chest, abdomen and pelvis 8 months after initial presentation. Demonstration of numerous liver metastases.

FIG 5: CT of the chest, abdomen and pelvis 8 months after initial presentation. Demonstration of multiple subcutaneous metastases in the upper back.

FIG 6: CT of the chest, abdomen and pelvis 9 months after initial presentation. Demonstration of multiple lytic metastases throughout the thoracic and lumbar spine.
FIG 7: Factor VIII stain of bone marrow biopsy at 20x magnification. Demonstration of dilated vascular spaces lined by atypical endothelial cells, consistent with metastatic angiosarcoma.

Conclusion

Cutaneous angiosarcoma is a rare form of cancer, accounting for only around 1% of all sarcomas. It most commonly occurs on the head, neck or trunk, but has been reported in a wide variety of anatomic locations. Cutaneous angiosarcoma most commonly presents as a rapidly expanding erythematous to violaceous patch, but enlargement is sometimes insidious. Many patients with cutaneous angiosarcoma are either elderly or have a history of preceding cancer, mastectomy, radiation, lymphedema, vascular malformation or some combination of these factors.¹

The prognosis for cutaneous angiosarcoma is generally quite poor, however, survival rates seem to vary substantially based on patient age, primary site of involvement and extent of disease at initial presentation. In the largest published series to date, older patients with tumors of the scalp and neck fared the worst, while younger patients with tumors of the trunk fared the best. In the same series, patients with disease confined to the skin had 10-year survival rates around 50%, while very few patients with spread to the lymph nodes or beyond survived more than 10 years.² In another series, the overall 5-year survival among all patients was 34%.

The most common sites of metastasis from cutaneous angiosarcoma are the lungs, liver, spleen and brain.³ Our review of published reports to date did not reveal a single case of bone marrow involvement in cutaneous angiosarcoma, suggesting that this is an exquisitely rare event. Our report contributes to the available literature by bringing awareness to the potential of cutaneous angiosarcoma in relatively young patients without any identifiable risk factors and by demonstrating the versatile metastatic behavior that cutaneous angiosarcoma can exhibit.

Conflict of interest

The authors declare that they have no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

References