Angiosarcoma at Dialysis Fistula Site in a Patient on Continuous Ambulatory Peritoneal Dialysis

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Arteriovenous fistula is the best permanent access for hemodialysis. Swelling and pain due to thrombosis and infection is common at fistula site. Angiosarcoma is one of the rare but important differential diagnoses of these signs. We present a patient on continuous ambulatory peritoneal dialysis with angiosarcoma at the arteriovenous fistula site.

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Case Report

INTRODUCTION

Angiosarcoma, a rare aggressive neoplasm with a dismal prognosis, is a high-grade malignant neoplasm of endothelial derivation. It can occur in various anatomic sites but has a predilection for skin and superficial soft tissue, accounting for 60% of cases. Cutaneous angiosarcoma was first described in 1945 by Caro and Stubenrauch.[1] Jones described that cutaneous angiosarcoma primarily affected scalp and face of the elderly in 1964.[2] About 50% of cutaneous angiosarcoma affects head and neck area in the older men (approximately a 2:1 male:female ratio), particularly the scalp.[3] Clinical presentation in primary cutaneous angiosarcoma varies from early ecchymosis-like patch to advanced violaceous nodules with hemorrhage and ulceration.[4] Accession of this malignancy at hemodialysis fistula is rare. We present a case of angiosarcoma at this site.

CASE REPORT

A 79-year-old woman who had end-stage renal disease due to nephrolithiasis and hypoplastic right kidney was under continuous ambulatory peritoneal dialysis (CAPD) from 9 years ago. Cholecystectomy due to stone and thyroidectomy of unknown reason was positive in her past medical history. An arteriovenous fistula (AVF) had been created at the left arm on the brachial site 11 years ago, but 1 year after initiation of peritoneal dialysis, the AVF was failed spontaneously without any use. One year before her last operation, swelling and redness at the AVF redounded to surgical intervention and the pathologic findings of the excised lesion was normal. After 1 year, again a reddish-purplish discoloration appeared at the suture line accompanied with severe pain and swelling, and the patient was a candidate for surgery with suspicion of infected or thrombotic aneurysmal fistula. Color Doppler ultrasonography showed an ill-defined lobulated hypoechoic and highly vascular mass without calcification or cystic component (Figure 1). On operation, the mass did not have any pulsation or thrill and the

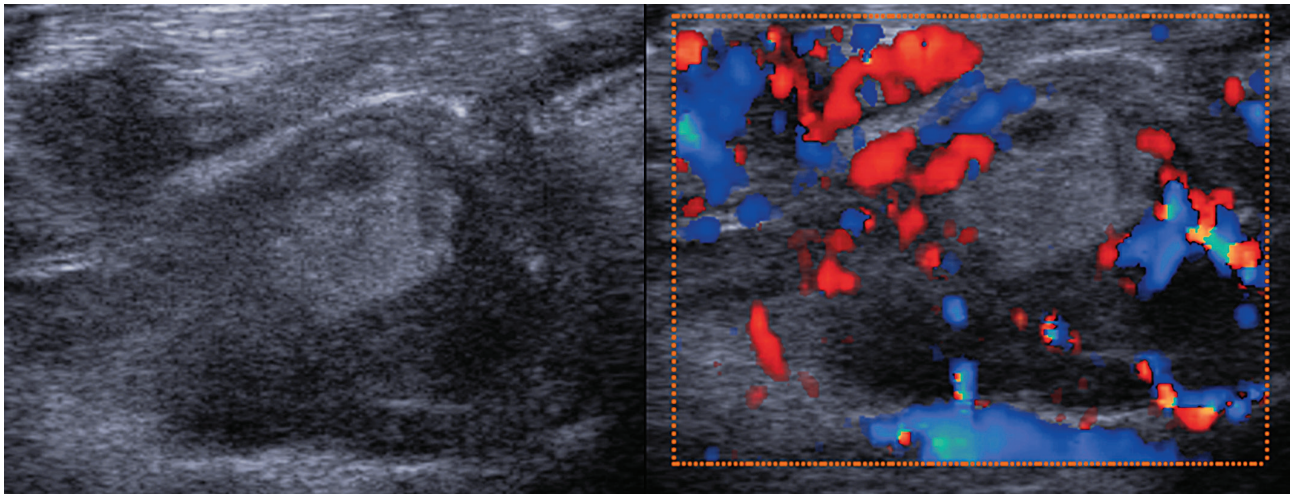

Figure 1. Left, Color Doppler ultrasonography showed an ill-defined lobulated hypoechoic mass. **Right,** The mass was highly vascular without calcification or cystic component.

distal pulses were normal. The skin over the mass was necrotic. The origin of lesion was the venous side of fistula far enough from the arteriovenous anastomosis. With an elliptical incision and a 5-cm safe margin, the mass was totally excised. After surgery, the hand was warm without any sign of ischemia or distal arterial hypoperfusion.

On histologic examination, the tissue was a creamy-colored mass with 8 × 6 × 3.5-cm diameter and lobular appearance. Microscopic examination showed neoplastic proliferation of pleomorphic spindle-shape cells elongated to oval-shape nuclei, small prominent eosinophilic color, 7 to 8 mitoses per 10 high-power field that arranged in anastomosing and slit-like spaces and intraluminal papilation. Hemorrhage and necrosis were also seen in less than 50% of tumoral surface area (Figure 2). Immunohistochemistry staining for CD31 and CD34 were positive (Figure 3). According to the histologic findings, the diagnosis of angiosarcoma at the fistula site was confirmed. Secondary workup for ipsilateral axillary lymph node involvement and distant metastasis to lung and liver was negative. Because the patients did not consent to further major surgery, left arm amputation was not done, and she was referred to radiotherapist for local irradiation.

DISCUSSION

Angiosarcomas, which represent approximately less than 1% of all sarcomas, are malignant neoplasms characterized by quickly proliferating extensively infiltrating cells derived from the vascular system.[5] Unlike most sarcomas, angiosarcomas commonly

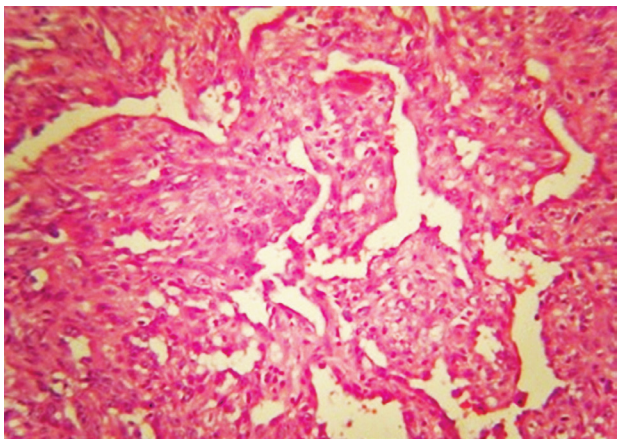

Figure 2. Hemorrhage and necrosis were also seen in less than 50% of tumoral surface area.

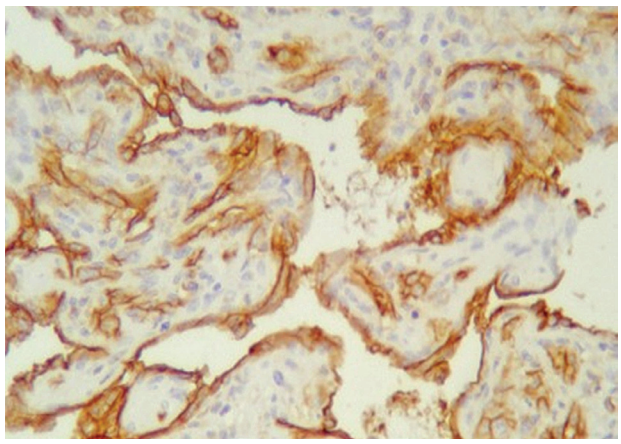

Figure 3. Immunohistochemistry staining for CD31 and CD34 were positive.

occur in the skin or superficial soft tissues, presenting as ill-defined (nodular or ulcerated) lesions with purplish discoloration. Pain is the usual presenting symptom and metastasis is often to the lung and liver. Most angiosarcomas rapidly become metastatic because their vascular origin permits tumor dissemination without the need for initial recruitment of new blood vessels. Angiosarcomas generally metastasize directly to the lungs via the vascular system.[6,7] These tumors are usually poorly differentiated, biologically aggressive, and consistently express endothelial antigens (CD31, CD34, and factor VIII-related antigen). The median survival rate of patients with this tumor is very short, generally less than 6 months.[8]

To our knowledge, angiosarcoma arising in the dialysis fistula reported in 10 cases to date. All of those patients were men with ages from 35 to 71 years. In 2 cases, the lesion occurred in the thrombotic aneurysmal fistula site,[5] and in another 3 cases, angiosarcoma developed in a patent but not used AVF.[9-12] In 4 cases, angiosarcomas was shown in previously ligated fistulas; their fistulas were ligated either due to asymptomatic aneurysmal dilatation (2 cases)[13,14] or after kidney transplantation and immunosuppressive treatment (2 cases).[15,16] Management of malignancy in 3 of 10 cases was amputation at first,[10,11,16] and one case after radiochemotherapy.[9] In 5 cases, like our patient, only radiotherapy with or without chemotherapy after wide excision of lesion were chosen.[5,13,15] In all of the cases, the time to death was less than 12 months.

Our patient was a 79-year-old woman with angiosarcoma arisen from thrombotic fistulas. She was on CAPD without any use of her AVF or history of kidney transplantation. She did not have any comorbid conditions either, such as diabetes mellitus, human immunodeficiency virus infection, or other immunocompromised states. Six months after surgical excision and radiotherapy our patient was still alive.

Arteriovenous fistula is the best form of vascular access for hemodialysis patients. Pain, swelling, and any soft tissue growth at or near the AVF site is the usual presentation and have some differential diagnosis such as thrombosis, infection, or aneurysmal dilatation. Physicians and surgeons should have a high index of suspicion about angiosarcoma in patients who present with unexplained pain and growth at or near a fistula site.

CONFICT OF INTEREST

None declared.

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