Embolization of a hemorrhaging abdominal plexiform neurofibroma

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ABSTRACT

In rare instances, neurofibromas may spontaneously bleed. In this case, a 21-year-old woman with a known cutaneous neurofibroma presented with a rapidly enlarging right abdominal wall mass. After embolization of a focal pseudoaneurysm and the right internal mammary artery, her hemoglobin continued to decline. The right internal mammary artery and inferior epigastric artery were then embolized. The patient eventually underwent mass resection, hematoma evacuation, and flap reconstruction. Final surgical pathology confirmed the expected diagnosis of plexiform neurofibroma. The Schwann cells of plexiform neurofibromas, present in approximately 50% of patients with neurofibromatosis-1, have invasive and angiogenic properties, often resulting in hemorrhage.

KEYWORDS Embolization; hemorrhage; interventional radiology; neurofibromatosis; plexiform neurofibroma; pseudoaneurysm

Neurofibromatosis type 1 (NF-1) is a neurocutaneous autosomal dominant disorder that affects approximately 1 in 3500 people.¹ NF-1 has pathologic manifestations in the integumentary, nervous, and orthopedic systems. Characteristic findings include café au lait macules, axillary or inguinal freckling, Lisch nodules, long bone deformities, soft tissue sarcomas, and neurofibromas.² A neurofibroma is a tumor of the peripheral nerve myelin sheath and is composed of a mixture of Schwann cells, fibroblasts, mast cells, and perineural cells, with Schwann cells as the primary cell type.³ NF-1 can rarely be associated with spontaneous hemorrhage. When this occurs, it is usually a result of friable vasculature secondary to arterial dysplasia or vascular invasion by the neurofibroma.⁴ Most bleeding is due to plexiform-type neurofibromas.^{4,5}

CASE DESCRIPTION

A 21-year-old woman with known sickle cell anemia and NF-1 presented to the emergency department with a rapidly enlarging right-sided abdominal mass. The patient had a

history of chronic right abdominal wall discoloration and loose skin. Over the course of 2 days, a mass began to form in that region with associated mild pain. The patient attempted to continue daily activities and go to work; however, the mass was rapidly enlarging and she had generalized weakness, which prompted her presentation to the emergency department. Hypotensive and anemic (hemoglobin of 9.4 g/dL), she was resuscitated with fluids and 2 units of packed red blood cells. On physical examination, there was a large, hyperpigmented right abdominal mass with central ulceration (*Figure 1*). Computed tomography of the abdomen/pelvis demonstrated a large right-sided abdominal wall soft tissue mass/hematoma with active arterial extravasation (*Figure 2a*), corresponding to a complex, right abdominal wall neurofibroma noted on magnetic resonance imaging in 2005 (*Figure 2b*).

On angiogram, there was a hypertrophied right inferior epigastric artery with numerous vessels feeding the inferior aspect of the mass. This was subsequently embolized with Gelfoam (absorbable gelatin sponge). Follow-up angiogram demonstrated stasis of the distal branches of the right inferior epigastric artery. Angiography of the right internal mammary

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artery demonstrated a hypertrophic artery with a focal pseudoaneurysm (*Figure 3a*) at the site of active extravasation within the superior aspect of the mass. Using a microcatheter, the distal inferior mammary artery and pseudoaneurysm were catheterized and embolized using Gelfoam and microcoils. Completion angiogram demonstrated complete stasis without evidence of extravasation.

Following the procedure, the patient was taken to the surgical intensive care unit for continued monitoring, and her hemoglobin continued to decrease to 6.0 g/dL. The following morning she was taken back to the angiography suite, where angiography demonstrated an enlarged right inferior epigastric artery and persistent enhancement of the inferior aspect of the

mass (*Figure 3b*). Selective angiography of the right inferior epigastric artery demonstrated recanalization of the previously embolized artery. A microcatheter was then utilized to catheterize the right epigastric and right superficial circumflex iliac arteries with no evidence of active extravasation. These were again embolized utilizing Gelfoam and microcoils with no residual filling. Selective angiogram of the right internal

Figure 1. The abdominal wall mass at the time of excision.

Figure 2. (a) Computed tomography of the abdomen and pelvis on admission with active extravasation within a large right abdominal wall mass. (b) Magnetic resonance imaging of the abdomen and pelvis in 2005 demonstrating a heterogeneous mass within the right abdominal wall, compatible with a plexiform neurofibroma.

Figure 3. Angiography showing (a) a right internal mammary artery pseudoaneurysm; (b) persistent flow within the inferior epigastric artery prior to repeat embolization; and (c) persistent flow within the right internal mammary artery prior to repeat embolization.

mammary artery demonstrated persistent enhancement of the superior portion of the tumor without active extravasation (Figure 3c). The treated pseudoaneurysm demonstrated no residual filling. Subsequently, third-order branches arising from the right internal mammary artery were embolized. Completion angiogram demonstrated no residual enhancement of the entire right abdominal wall tumor.

The patient's postprocedural course included persistent fevers, probably due to superimposed infection. She eventually underwent mass resection, hematoma evacuation, and flap reconstruction. The postsurgical course was uneventful, and the patient was discharged home in stable condition.

The gross and microscopic description of the mass confirmed the diagnosis of a neurofibroma. Centrally located within the specimen was an irregular area of transmural defect measuring 6.6 × 4.2 cm. This defect was surrounded by irregular areas of tan-pink ulceration and skin slippage. Sections through the specimen showed numerous foci of fleshy and somewhat myxoid tissue, along with areas of hemorrhage and fat necrosis. Sections of skin and subcutaneous tissue showed a large plexiform neurofibroma with a prominent associated diffuse component. There was focal tumor infarction and hemorrhage, secondary to prior embolization.

DISCUSSION

Neurofibromas are composed mainly of Schwann cells. In plexiform neurofibromas, the Schwann cells may have invasive and angiogenic properties.⁶ An estimated 50% of NF-1 patients have a plexiform neurofibroma discovered on whole body imaging.⁷ Plexiform neurofibromas carry a greater risk of uncontrolled bleeding and hemorrhage as opposed to other types, such as cutaneous or nodular

neurofibromas. Neurofibromas can be slow growing and eventually spontaneously bleed. These patients should be educated on the risk, albeit small, of spontaneous hemorrhage of a neurofibroma and provided anticipatory guidance on the steps to take if they suspect a hemorrhage. Patients should watch for signs including a rapidly enlarging preexisting mass, weakness, and tachycardia.

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