Brachial artery rupture complicating a pregnancy with neurofibromatosis: A case report

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A 30-year-old black woman was first seen at 34 weeks’ gestation with pain in the right arm, which was already afflicted with neurofibroma. She was in hypovolemic shock on presentation, and after she was resuscitated, a live infant was delivered. The patient was diagnosed with a pseudoaneurysm of the right brachial artery with extravasation of blood in the interstitium of the arm. Although there have been other reported cases of arterial rupture in pregnancies with neurofibromatosis, the report of brachial artery rupture in this situation is believed to be the first documented case. (Am J Obstet Gynecol 1998;179:832-4.)

Key words: Neurofibromatosis, brachial artery rupture, hemorrhage

Neurofibromatosis is one of the most frequent and complex genetic disorders in humans. It was first described by Robert Smith in 1843. In 1882 Von Recklinghausen correctly identified the relationship between the nerves, tumors, and cutaneous lesions. It is transmitted in an autosomal dominant fashion with an incidence of 1 in 3000 births. The incidence in pregnancy is less frequent and has been reported as 1/5000 to 1/18,500 deliveries. There have been 2 reported cases of arterial rupture in pregnancy with neurofibromatosis, neither of which involved the brachial artery. Recently we cared for a patient with a pseudoaneurysm of the brachial artery that spontaneously ruptured, leading to hypovolemic shock.

Case report

A 30-year-old primigravid woman with an intrauterine pregnancy at 34 weeks’ gestation and an uncomplicated prenatal history, except for neurofibromatosis, was seen with a chief complaint of right arm pain. She was evaluated and found to be in hypovolemic shock with a blood pressure of 70/50 mm Hg and an enlarging area that encompassed the patient’s entire elbow and forearm (Fig 1). An orthopedic tourniquet was placed above the patient’s elbow, and multiple intravenous lines were secured. The fetus was examined by ultrasonography, which revealed a fetal heart rate of 60/min. The initial hematocrit level was 20%, and after fluid resuscitation an emergency cesarean section was performed. A viable male infant weighing 6 lb was delivered with Apgar scores of 2 and 3 at 1 and 5 minutes, respectively. The only evidence of neurofibromatosis in the infant were 4 café au lait spots. An arteriogram of the patient’s right upper extremity revealed massive extravasation and leaking about the brachial artery above the bifurcation (Fig 2). The patient was then taken to the operating room, where an attempt at a saphenous vein harvest with graft was unsuccessful. A total of 6 units of blood and 6 units of fresh-frozen plasma was required to correct the anemia and coagulopathy. The patient recovered with decreased function of the affected arm. On the day of scheduled discharge the patient again experienced pain in her arm. After another hemorrhage was identified, it was then agreed on between the patient and the surgeon that a transhumeral amputation would be performed. After an uneventful course in the nursery, the infant was discharged home 30 days after the delivery. The pathology examination revealed that the vascular rupture was potentiated by impingement of the brachial artery by large neurofibromas and invasion of a neurofibroma into the vasculature wall.

Comment

Neurofibromatosis is rare in pregnancy with an incidence of 1/5000 to 1/18,500 deliveries. Reports of neurofibromatosis occurring in pregnancies date back as early as 1906, when Brickner coined the term fibroma mollicum gravidarum, for neurofibromas that enlarged during pregnancy. Most current information on pregnancy and neurofibromatosis is derived from case reports, which may allow for speculation rather than fact in the management of these cases.

Pregnancy can have several effects on the clinical course of patients with neurofibromatosis. They include worsening of the skin lesions with partial remission in...
the postpartum period, increased growth of neurofibromas leading to pain, compromised blood supply, which may lead to uteroplacental insufficiency, dystocia from pelvic tumors, and vascular rupture. Vascular changes may predispose the patient to hypertension. Growth of central nervous system tumors can occur that require intervention. An increased incidence of pheochromocytoma is reported, as well as malignant degeneration of neurofibromas.

The case presented is believed to be the first report of brachial artery rupture in a pregnancy with neurofibromatosis. Review of the literature has revealed several cases that did involve other vascular ruptures but not of the brachial artery. There are a couple of hypotheses regarding the cause of vascular rupture in neurofibromatosis. The first theory involves the poor integrity of vessels in a patient with neurofibromatosis, leading to aneurysms that are potentiated by the increased blood volume of pregnancy. The second theory is explained by trauma to the vessel from the mass effect of a plexiform neurofibroma.

Arterial vasculopathy in patients with neurofibromatosis was first described by Reubi. He postulated that all patients with this disorder might have varying degrees of some type of vascular dysplasia. Reubi described the different types of arterial lesions that one may see, and Sayler confirmed these same lesions in a study of 18 patients. The 4 main types of lesions that are commonly seen in small arterioles and arteries are the pure intimal, advanced intimal, intimal aneurysmal, and nodular. In all of these forms proliferation of spindle cells occurs within arterial walls and may be responsible for the associated arterial wall disruption. The exact origin of proliferating spindle cells is unknown, but Schwann cells or smooth muscle cells have been implicated as precursors. Several cases of arterial rupture with neurofibromatosis have been associated with renal arteries. This is alluded to because of the abundance of Schwann cells that are located in these vessels.

Fatal or near-fatal hemorrhages have been reported in neurofibromatosis in association with vasculature rupture in several different sites including pleural or abdominal cavities, the retroperitoneum, soft tissues of the trunk, and the extremities. All of these cases were thought to stem from the previously mentioned hypothesis including rupture as a secondary effect of impingement or arterial dysplasia. A large majority of the morbidity and mortality associated with the above-mentioned cases were caused by severe hemorrhages during or just after surgery from difficulties in achieving hemostasis.
The vessels have been described as friable and necrotic and not supporting arterial ligatures or responding to electrocautery in the control of hemorrhage. This is in support of other authors who have reported on neurofibromatosis and arterial dysplasia and on vascular invasion. This may explain why the surgeons at our hospital had trouble with the saphenous graft.

When care is being given to a pregnant woman with neurofibromatosis, a thorough history focusing on cognitive and motor neurologic functioning and a detailed physical examination with particular attention to hypertension, scoliosis, focal neurologic deficits, skin lesions, and neurofibromas should be obtained. Counseling should be provided for all patients and their families, with explanations of risks of progression through puberty and pregnancy. Genetic counseling should note a 50% inheritance rate but with varying expressivity within the family. Psychologic and social adjustments may be needed regarding the disease and the possible deformities that it may present.

The patient should be placed in a high-risk clinic for close observation of blood pressure and evaluation of pain and growth in the neurofibromas. Antenatal testing should be performed once the fetus is viable, including ultrasonography for fetal growth and nonstress testing for fetal well-being. Unless there is an obvious malformation of the pelvis from neurofibromas, a trial of labor is appropriate.

We believe our case to be the first report of brachial artery rupture in a pregnancy with neurofibromatosis. Review of the literature has revealed several cases that involved other incidences of vascular ruptures but none that occurred spontaneously in the brachial artery. Possible explanations of vascular rupture involving neurofibromatosis have been reviewed, and specific patient care recommendations have been given. Although we believe the cause of our patient’s vascular rupture was both impingement and invasion by the neurofibroma, the literature review suggests additional etiologies of vascular rupture in pregnancy.

REFERENCES

A complete list of references is available from the authors on request.