CASE REPORT

Arteriovenous Malformation of Pelvis Treated with Hypothermic Arrest and Hemipelvectomy

Steven C. Raithel, BS, CCP, Donna G. Vieth, CCP, Raymond M. Keltner, MD, Vallee L. Willman, MD

St. Louis University School of Medicine
St. Louis, Missouri

Key words: Arteriovenous malformation, cardiopulmonary bypass, hypothermia

Abstract

Arteriovenous malformations are lesions which may cause significant complications including congestive heart failure, venous insufficiency, limb ischemia, or difficulty with feeding and respiration. The treatment of arteriovenous malformations can be quite involved due to the complexity of the lesion.

The use of cardiopulmonary bypass equipment in noncardiac cases is not new as is evident in equipment use in liver transplantation, neurosurgery, and the delivery of hyperthermic anticancer therapy.

The authors report on a 30-year-old male with a large arteriovenous malformation involving the left thigh and pelvis treated with deep hypothermic cardiopulmonary arrest and hemipelvectomy.

Introduction

Arteriovenous malformations (AVM) are usually benign lesions which may cause significant complications including congestive heart failure (CHF), venous insufficiency, limb ischemia, or difficulty with feeding or respiration, depending upon the site affected (1-3). Congenital AVM's are most commonly found in the extremities with a higher frequency in the lower limb than the upper (4). The communications between the arteries and veins are always multiple and vary in diameter from several millimeters down to normal precapillary anastomosis (5). AVM occurring within the lower limb frequently involves the femoral vessels (4). The diagnosis of AVM requires a detailed history and extensive angiographic study. The treatment of congenital AVM occurring within the pelvis or upper thigh can be quite involved and includes elastic support, steroids, and radiation (3-8). Embolic therapy is also used, often followed by surgical excision (5,8,9). In extreme cases, as with certain aneurysms located within the brain, the use of hypothermic circulatory arrest is necessary to safely excise the malformation (10). This radical approach is sometimes used for angiomas involving the head, renal, or hepatic circulations with acceptable results (3,7, 11, 12).

Address correspondence to: Steven C. Raithel, BS, CCP, St. Louis University School of Medicine, Dept. of Surgery, 3635 Vista Ave. at Grand Blvd., P.O. Box 15250, St. Louis, MO 63110-0250.

History

The patient first presented in the mid-1960's with a mass in the left thigh along with elongation and increased circumference of the left leg. The thigh was first explored and some tissue excised in 1967, repeated in 1969 and 1972.

On all occasions, tissue, including small AVM, was removed but total excision was not possible. There was continued reoccurrence of the tumor with cardiomegaly and increasing CHF. Selective embolization of the femoral vessels was performed in 6/75, 12/76, 7/79 and 10/79. The last embolization was associated with extensive occlusion of the vessels and some clinical decrease in the size of the tumor mass; however, CHF symptoms increased and the patient could barely ambulate. In July 1982, a Goretex (a) graft was placed from the left proximal iliac artery to the left popliteal artery with ligation of the left iliac artery to decrease tumor size. There was graft closure in the immediate postoperative period with leg ischemia and gangrene. On 7/16/82, the patient underwent hip disarticulation with massive blood loss. A prosthesis was fitted, and the patient was able to ambulate for a time. On examination in 7/87, a large mass was noted in the left groin and stump region. The patient was studied extensively and underwent embolization of two collateral vessels. The distal end of the mass became gangrenous without relief of his heart failure. By 4/89, there was a huge mass at the amputation site extending into the left groin and scrotum (Figure 1). Arteriography and computerized axial tomography demonstrated extension of the process into the left psoas muscle. Hypertrophy of the lumbar vessels was evident, especially a very large artery feeding the angioma of the psoas muscle (Figure 2). The inferior mesenteric artery and the visceral branches of the right internal iliac artery were markedly hypertrophied. The patient had increasingly severe heart failure and difficulty ambulating. The decision was made to radically excise the mass in conjunction with left hemipelvectomy using deep hypothermic circulatory arrest.

a. Goretex, Flagstaff, AZ 86001

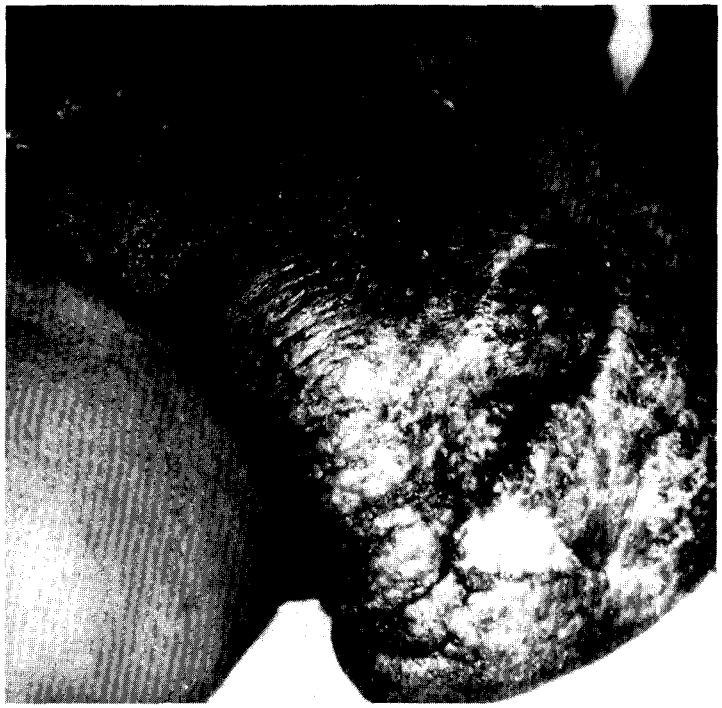

Figure 1: Preoperative view of tumor.

Figure 2: Anterior-posterior view of iliac vessels showing patent iliac and femoral vessels on the right. Large vessel in upper right is a lumbar artery feeding tumor of the left psoas muscle. Diffuse mass of radiopaque vessels is evident within pelvic circle.

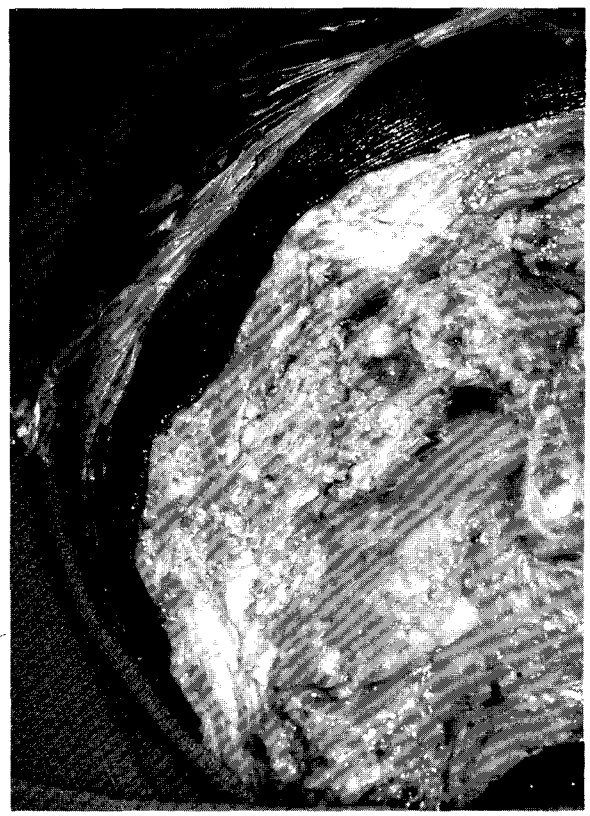

Figure 3: Cut surface of excised tumor.

Materials and Methods

The patient was anesthetized, intubated, and appropriate monitoring lines were placed. He was then positioned in a right semilateral position with the left hip elevated. A sternotomy was performed in preparation for cannulation and cardiopulmonary bypass. Simultaneously, an incision was made from the left midaxillary line at the crest of the ilium, carried medially along the crest of the ilium to the inguinal ligament, and extended to the pubic tubercle. The incision was deepened and the inguinal ligament removed. The peritoneum was identified and retracted to expose the retroperitoneum. The previously placed Gortex graft was identified and followed superiorly to its junction at the common iliac artery. The graft was occluded at both ends and heavily fibrosed. In the process of freeing the graft, the left ureter was transected. The hypertrophic blood vessels along the ureter were ligated and the ends of the ureter debrided for later anastomosis. The common iliac artery was identified proximally and followed to the aorta. The left side of the aorta was dissected to identify the hypertrophic lumbar vessels which were ligated and severed. The psoas muscle was transected at the upper border of the left kidney and dissected free from its origins. The left femoral nerve was transected and the psoas freed as far as the iliac crest.

On angiogram, the left external iliac vein was patent as were the visceral branches of the left internal iliac artery and vein, supplied by anastomoses from the right. Dissection was then begun to isolate the thrombosed and densely scarred left external iliac artery. In the course of this dissection, massive hemorrhage occurred from lacerations in the internal iliac artery. Local control was only partially possible by the placement of vascular clamps and it was evident that cardiopulmonary bypass would be necessary in order to proceed.

Heparin was administered (3.5 mg/kg), and the ascending aorta was cannulated with a 21 Fr. aortic cannula. The cavae were cannulated with a 30 Fr. venous cannula in the superior and a 40 Fr. in the inferior position. Cardiopulmonary bypass was initiated and an attempt was made to repair the iliac vessels. Severe bleeding continued with the operative field obscured by blood. Cooling was initiated in preparation for circulatory arrest. A left ventricular vent was placed in the apex and attached to low suction. The head was surrounded with ice and active cooling stopped at 13°C (nasopharyngeal) with the patient drifting down to 12.6°C. Shortly before the start of the circulatory arrest period, 1,550 milligrams of pentobarbital was given. Electroencephalographic activity became isoelectric at 15°C. The aortic cross-clamp was applied and the heart arrested with 1,000 cc of sanguinous cardioplegia (with added potassium and lidocaine) injected into the aortic root.

After adequate cooling, circulatory arrest was initiated leaving the operative field free of hemorrhage and the remainder of the hemipelvectomy was accomplished. The tumor weighed 10.7 kg and measured 30 x 30 x 18 cm. The bulk of the tumor consisted of fat and fibrous tissue studded with numerous tumor nodules. The nodules were reddish-brown in color, ovoid in shape, and averaging 1-2 cm in size. Sections of soft tissue involved by the nodules were noted for proliferation of variably sized capillaries, veins, and arteries. The vessels were haphazardly arranged and scattered in surrounding fat and subcutaneous tissue (Figure 3).

After 37 minutes of circulatory arrest, the aortic cross-clamp was moved distal to the left subclavian artery and flow reestablished at 1 l/min. This proceeded for 20 minutes with the perfusate warmed to 20°C. The aortic cross-clamp was then removed and the patient rewarmed to 37°C over a period of 45 minutes.

Following restoration of the circulation, a massive hemorrhage occurred requiring the addition of 16.6 liters of Plasmalyte A (b) 17 units of packed red blood cells (RBCs), one liter of 5% albumin, and two units of whole blood (WB) while on cardiopulmonary bypass to maintain flows of 3.1 to 4.8 l/min (cardiac index of 1.7 to 2.65 l/min/m^2). Additionally, 17 milligrams of phenylephrine were given in an attempt to maintain adequate arterial pressure. However, due to severe blood loss, the mean arterial pressure was generally less than 40 mmHg despite adequate flow rates. When the patient's temperature reached 37°C, cardiopulmonary bypass was terminated.

Protamine was given immediately in an attempt to control bleeding. Hemostasis was attempted with argon coagulator, ligature, clips, and cautery where appropriate. The most intense bleeding came from an opening in the spinal canal where the sacrum had been transected. Fibrin glue, thrombin soaked cellulose, and pressure were utilized; however, due to the immense vascularity of the site, severe bleeding continued. Approximately four hours were spent in an attempt to control bleeding, during which time, the patient received 37 units of packed RBCs, 16 units of fresh frozen plasma, 20 units of cryoprecipitate, and 30 units of platelets. He also received a total of 38 (225 cc) bowls of washed autologous RBC's while on bypass and immediately after bypass. Coagulation studies showed no diagnostic defect and the platelet count was 104,000 per cc^3=cubic centimeters.

During the time spent attending to hemostasis, his temperature dropped to 32°C, and his blood pressure ranged from 50 to 70 mmHg. It was decided that further attempts to attain hemostasis would be fruitless due to a coagulopathy, and it was elected to pack and close the wound. The lower abdomen and pelvis were wrapped tightly with an Esmark (c) bandage. The patient was then positioned in antishock trousers with the abdominal portion inflated to 15 mmHg and the right extremity inflated to 20 mmHg.

The patient was transported to the Intensive Care Unit. Over the next 12 hours, with tamponade of the wound and multiple infusions of blood products, the bleeding decreased. The wound was reexplored the following day and hemostasis achieved.

A protracted postoperative course followed, complicated by a stubborn wound infection requiring wound debridement. The

b. Plasmalyte A, Travenol Laboratories, Deerfield, IL 60015
c. Esmark, Zimmer, 84 Inverness Circle, Englewood, CO 80112

patient also had a suprapubic cystotomy placed as well as a colostomy performed.

One month later, the cystotomy and colostomy were closed, and two small areas on the stump received skin grafts. The patient was capable of spontaneously passing urine and stool the third postoperative day. Postoperative angiography showed marked improvement without recurrence of the lesion. The patient has some CHF which is compensated by digitalis. His exercise tolerance improved, and the patient was able to ambulate with crutches.

Discussion

While an arteriovenous fistula is defined as an "abnormal communication between an artery and vein bypassing the normal capillary bed," this term is best applied to traumatic communications, including those that are surgically placed. The term arteriovenous malformation should be used for the congenital lesion (4, 5).

AVM are sporadic, nonfamilial developmental errors which occur during the fourth through tenth week of intrauterine life. Not all malformations are obvious at birth; many manifest themselves years or decades later (1-3). Diffuse congenital AVM are most commonly found in the limbs. The connections are usually small, numerous, and may permeate the whole extremity (5).

Specific symptoms vary but may include pain, pulsation, heaviness, a palpable thrill, and functional impairment (1, 14-16). Pain is present in approximately 50% of patients (16). It may be ischemic in origin or the malformation may involve tissue causing pressure or distortion upon a specific nerve (8, 15). Thrombosis or deep hematoma may also be the reason for pain (15). The degree of pulsation or thrill depends upon the size of the malformation and blood flow (16). Heaviness is often position-dependent, decreasing with elevation. There may also be functional impairment of the limb or congestive heart failure (1, 8, 14-16).

In this patient, despite multiple selective embolization and several partial resections of the malformation, his condition continued to deteriorate. The decision to radically excise the malformation was made in an attempt to reduce his CHF and give some degree of mobility. The use of cardiopulmonary bypass and hypothermic circulatory arrest was utilized in an attempt to perform the resection under hemostatic conditions. However, once flow was reestablished, profuse bleeding occurred defying all attempts at control.

The primary problem encountered by our group and others (7, 14) was the massive blood loss which occurred due to the hypervascularity of the malformation. Despite the use of cardiopulmonary bypass and vasopressors, maintenance of an adequate blood pressure was almost impossible. The problem was not with infusing the blood into the patient but keeping enough volume in the cardiopulmonary bypass circuit. At one point, we had three people infusing volume into the pump and were barely able to maintain adequate flow. In retrospect, a second cardiotomy should have been filled with replacement fluids before reinstitution of blood flow and kept filled for immediate use. During liver transplantation at our institution, blood products in boxes of 10 are checked upon arrival in the room and can be hung without delay. That policy should have been applied for this case. Due to the profuse bleeding which may occur during surgical resection of an AVM, this procedure should only be attempted at facilities that can provide massive blood replacement.

References

1. Pitt DF: Congenital pelvic arteriovenous aneurysm. *CMAJ* 27:1200-1201, 1982.
2. Price AC, Coran AG, Mattern AL, Cochran RL: Hemangioendothelioma of the pelvis. *N Engl J Med* 286(12):647-649, 1972.
3. Little KET, Cywes S, Davies MRQ, Louw JH: Complicated giant hemangioma: Excision using cardiopulmonary bypass and deep hypothermia. *J Ped Surg* 11(4):533-536, 1976.
4. Borzatta MA, Klein SR: Congenital arteriovenous fistula. In: Wilson SE, Vieth FJ, Hobson RW, et al., McGrath RE, Kothman TH, Navrozov M, eds., Vascular Surgery Principles and Practice, New York: McGraw-Hill, Inc., 1987, pp. 873-885.
5. Young AE: Arteriovenous malformation. In: Mulligan JB, Young AE, McAlister L, eds., Vascular Birthmarks Hemangiomas and Malformations, Philadelphia: W.B. Saunders, 1988, pp. 228-245.
6. Mulliken JB: Treatment of hemangiomas, In: Mulligan JB, Young AE, McAlister L, eds., Vascular Birthmarks. Hemangiomas and Malformations, Philadelphia: W.B. Saunders, 1988, pp. 77-103.
7. Mulliken JB, Murray JE, Castaneda AL, et al: Management of a vascular malformation of the face using total circulatory arrest. *Surg , Gynecol, and Obstet* 146:168-172, 1978.
8. Flye MW, Jordan BP, Schwartz MZ: Management of congenital arteriovenous malformations. *Surg* 94(5):740-747, 1983.
9. Stanley RJ, Cubillo E: Nonsurgical treatment of arteriovenous malformations of the trunk and limb by transcatheter arterial embolization. *Diagn Radiology* 115:609-612, 1975.
10. Olcott IVC, Newton TH, Stoney RJ, et al: Intra-arterial embolization in the management of arteriovenous malformations. *Soc Vasc Surg* 79(1):3-12, 1976.
11. Spetzler RF, Hadley MN, Ragamonte D, et al: Aneurysms of the basilar artery treated with circulatory arrest, hypothermia, and barbiturate cerebral protection. *J Neurosurg* 68:868-879, 1988.
12. Ranne RD, Ashcraft KW, Holder TM, et al: Hepatic hemangioma: Resection using hypothermic circulatory arrest in the newborn. *J Ped Surg* 23(10):924-926, 1988.
13. Wickey GS, Martin DE, Larach DR, et al: Combined carotid endarterectomy, coronary revascularization, and hypernephroma excision with hypothermic circulatory arrest. *Anesth Analg* 67:473-476, 1988.
14. Young AE: Pathogenesis of vascular malformations. In: Mulligan JB, Young AE, McAlister L, eds., Vascular Birthmarks. Hemangiomas and Malformations, Philadelphia: W.B. Saunders, 1988, pp. 107-113.
15. Neifeld JP, Doppman JL, Chretien PB: Congenital pelvic arteriovenous fistulas: Report of a case and review of the literature. *J Urol* 114:648-652, 1975.
16. Young AE: Clinical assessment of vascular malformations. In: Mulligan JB, Young AE, McAlister L, eds., Vascular Birthmarks. Hemangiomas and Malformations, Philadelphia: W.B. Saunders, 1988, pp. 114-127.
17. Dry LR, Conn JH, Chavez CM, et al: Arteriovenous fistula: An analysis of 58 cases. *Am Surg* 38(3):154-160, 1972.