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Case Report
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Recurrent and Large Arteriovenous Malformation with Failed Interventions Managed Surgically in Deep Hypothermic Circulatory Arrest - A Case Report

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Introduction

The first recorded case of an arteriovenous malformation was in the late 16th century. In 1757, William Hunter described an arteriovenous malformation as an abnormal communication between an artery and a vein [1]. Arteriovenous malformations can occur anywhere in the body. They frequently occur as isolated, stable anomalies requiring no specific treatment. Some AVMs may be extensive, multiple,recurrent and progressive causing disfigurement with the potential for life-threatening hemorrhage, thrombosis, painful ischemia or high-output congestive heart failure. These occurrences usually are indications for intervention, which is often technically difficult and unrewarding [6].Surgery in deep hypothermic circulatory arrest has a role in this failed intervention.

Case Report

A 19 year old boy was referred to us with a diagnosis of recurrent and large arteriovenous malformation over the right side of the chest below the clavicle. He underwent embolisation of the feeding vessels in 2002, 2003 & 2005 with absolute alcohol, gelfoam and metallic coils respectively. He also underwent radiotherapy with cobalt fractions in 2006. Inspite of multiple embolisations and radiotherapy the lesion was progressively increasing along with symptoms of severe pain in the hand and increase in the severity of breathlessness associated with moderate cardiomegaly. On examination swelling measured 8x10 cm on the right subclavicular area which is compressible and associated with a thrill. He had small arteriovenous malformation in the right palm and in the back near the right scapula for which excision of the swellings was done in 2003. Biopsy specimen showed multiple arterio venous malformation. Recurrence of the arteriovenous malformation, severe pain in the hand and failed interventions made us to proceed to surgery.

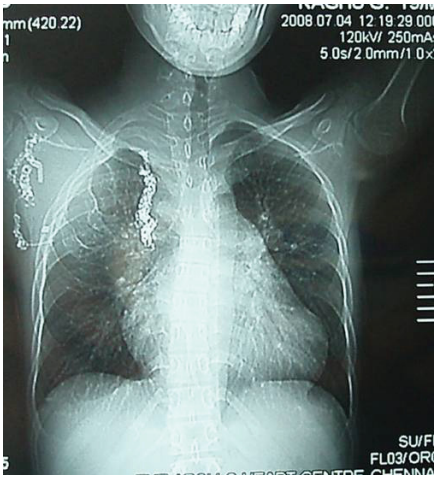

Figure 1:

Chest x-ray showed moderate cardiomegaly, scoliosis with embolisation coils [Figure 1].Computed tomographic angiogram showed large arteriovenous malformation with arterial feeders from the right subclavian artery, branches of both internal mammary artery ,upper and middle right intercostal arteries, both external carotid artery through the superior thyroid artery [Figure 2]. The venous drainage of the sac was through the right subclavian, right brachiocephalic and right internal jugular vein [Figure 3]. The brachiocephalic and internal jugular vein was stenosed at their origin from the sac. Echocardiogram showed good ventricular function with enlarged right atrium and ventricle. Electrocardiogram was normal sinus rhythm, right axis deviation with right ventricular hypertrophy. The blood investigations were found to be normal. After a thorough evaluation and discussion with the radiologist and the plastic surgeons, the patient was planned for surgery under cardiopulmonary bypass.

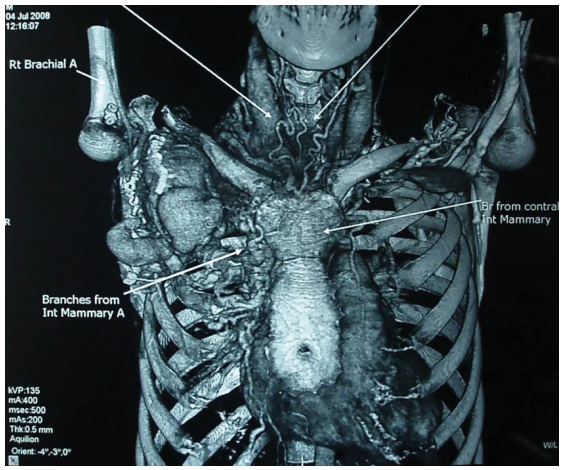

Figure 2:

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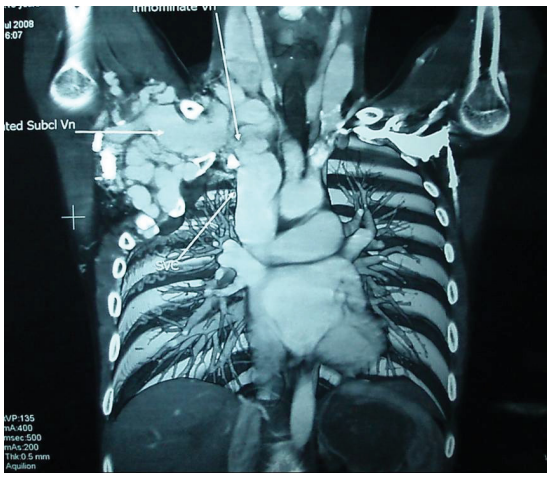

Figure 3:

Preinduction monitoring included electrocardiogram, pulse oximetry, invasive blood pressure and bispectral index. Two large bore intravenous cannulas were placed on the left hand. The patient was induced with injection midazolam, fentanyl, thiopentone and paralysed with vecuronium after checking for ventilation. Patient intubated and connected to the ventilator. Left internal jugular vein was cannulated as right internal jugular vein was stenos and draining the arteriovenous malformation. Right femoral vein was cannulated so that we had inferior vena caval access. Post induction monitors included central venous pressure, temperature, arterial blood gas and urine output. Maintanence was done with oxygen, nitrous oxide and sevoflurane. Midazolam, fentanyl and vecronium was given when required. Bispectral index was maintained around 40.

The sternotomy incision was extended to the neck, but dissection was not possible due to large number of collaterals. Further dissection was done on cardiopulmonary bypass. On cardiopulmonary bypass, the innominate vein, right subclavian vein, internal jugular vein and subclavian artery was dissected after excision of the middle two thirds of the clavicle. Internal mammary, thoracodorasal and cervicodorsal arteries were ligated. The aneurysmal sac was dissected on the anterior chest wall. Once it was opened there was continuous flow of blood, hence switched over to deep hypothermic circulatory arrest. The communications between the sac and the innominate vein was closed directly. Three other communications with systemic arteries were also closed. One large posterior communication could not be closed as it was against the ribs. Hence a pericardial patch was put, following which the sac was completely closed after disconnecting all the systemic connections.

Circulatory arrest was around 26 minutes, cross clamp time was around 45 minutes and cardiopulmonary bypass time was 90 minutes. The mean pressure in the cardiopulmonary bypass was around 40mm of Hg. Weaned off bypass with 10 mcg/kg/mt of dobutamine and 0.1mcg/kg/mt of adrenaline. Patient was extubated the next day. Postoperative period was uneventful except mild limitation of movements in the shoulder joint. Repeat computed tomographic angiogram showed no residual lesions. Post op chest x-ray showed mild reduction in cardiomegaly .The patient was discharged home after one week. Follow up after 3 months showed no residual lesions and patients was symptom free.

Discussion

The first recorded case of an arteriovenous malformation was in the late 16th century. In 1757, William Hunter described an arteriovenous malformation as an abnormal communication between an artery and a vein [1]. Arteriovenous malformations can occur anywhere in the body. However certain anatomical sites such as the pelvis, extremities, lungs and the intracerebral circulation seem to be more commonly affected [2]. In our patient the location was right subclavicular region although he had small arteriovenous malformation in the right palm and near the right scapular region.Many congenital arteriovenous malformation may regress spontaneously. Smaller arteriovenous malformations are usually asymptomatic. Larger arteriovenous malformations leads to decreased distal arterial pressures which might cause distal ischemia, increased peripheral venous pressures leading to swelling, visible veins [varicosities], and even ulcers in the limb. Extremity lesions may be associated with overgrowth of bone, but particularly of the soft tissues which may become massive and disfiguring. Finally it leads to cardiac decompensation and death if not treated [7].Our patient had severe pain in the right hand, right arm swelling, swelling below the right clavicle, tachycardia and moderate cardiomegaly.

Color flow doppler usually shows the direction and velocity of blood flow in the feeder vessels. Computed tomography is used to locate the abnormality, to evaluate for aneurysm formation, and to identify bony involvement [3]. Magnetic resonance imaging has become the new criterion standard in the preoperative evaluation of patients with arteriovenous malformations as it generates multiplanar views and can be used to accurately define tissue planes and extent of bone involvement [4].This patient was deferred of magnetic resonance imaging scan preoperatively as he was embolised with metal coils. The feeding vessels were accurately located with computed tomographic angiogram in our case. Angiographic evaluation is strongly recommended before considering any therapeutic procedures it allows precise evaluation of the number, location, and extent of the arteriovenous connections [5].

Indications for surgical intervention of arteriovenous malformations include hemorrhage, painful ischemia, congestive heart failure, nonhealing ulcers, functional impairment, failed interventions and limb-length inequality [6]. A lesion must be well localized for a chance at complete resection. Resectability depends on the degree of extension into adjacent structures. Our patient had painful distal ischemia, failed interventions and multiple feeders. Patients with disease that extends into the deep fascia or contiguous structures, such as muscle and bone, usually are not surgical candidate. The exact determination of the anatomical limits of the lesion may be very difficult, and successful surgical ligation and resection is frequently difficult and may not be possible [7].

The treatment options for large, high-flow arteriovenous malformations are multidisciplinary which includes surgical ligation, transcatheter embolization or combinations of surgical and embolization. Embolization has been used to reduce the vascularity of extensive arteriovenous malformations, either as a stand-alone procedure or before surgical ligation. Many endovascular embolic agents are currently available for treating arteriovenous malformations including autologous clot, gelfoam, polyvinyl alcohol particles, stainless steel, titanium coils, acrylic tissue adhesives, detachable balloons, and liquid sclerosing agents. Superselective angiographic catheters allow selective embolization of the arteriovenous malformation sources. Complications of embolization

include skin necrosis, nerve injury, distal emboli, inadvertent embolization of coils or glue in the pulmonary circulation [6]. In our patient although embolisation and radiotherapy did not lead to any complications he had recurrence. P.Clay Hass reported a similar case of huge subclavian arteriovenous malformation where surgical ligation and coil embolisation failed. Percutaneous wall graft endoprosthesis was done in that patient. However patient died due to fatal pulmonary embolis [6].

Even after apparently successful surgical ligation, arteriovenous malformations may eventually recur. High flow arteriovenous malformation and those with multiple large feeding vessels recurrence is common event. Either proximal feeding artery ligated stump stimulates the development of new channels or the inadequate initial ligation of all of the feeding arteries results in progressive dilatation of the residual arteriovenous communications. Our patient had recurrence of arteriovenous malformation after embolisation and radiotherapy. Amputation may be the best surgical option in some extreme cases. Upton in a review of two hundred sixty surgical resections came up with following indications for amputations: fast-flow type C arterial anomalies diffuse pulsating lesions with distal vascular steal and involvement of all tissues including bone [7].

The treatment options for large arteriovenous malformations are limited and often unsuccessful. Our patient had a large, high flow, recurrent and enlarging arteriovenous malformation in spite of interventions. The lesion was also involving the deep structures and multiple feeding arteries arising from the thorax. The surgical option was to repair the arteriovenous malformation in cardiopulmonary bypass under deep hypothermic circulatory arrest. Cardiopulmonary bypass with deep hypothermic circulatory arrest has been used in various major vascular procedures like aortic aneurysmal repair, gaint aneurysmal repair in neurosurgery and arteriovenous malformation in the brain [8]. Mc Creedy reported a patient with large recurrent pelvic arteriovenous malformation in whom transcatheter embolization was not feasible and the patient underwent surgical resection of arteriovenous malformation which was accomplished with deep hypothermic circulatory arrest. With deep hypothermic circulatory arrest the pump can be turned off enabling the resection to proceed in a bloodless field, multiple feeders can be ligated and enables the surgeon to identify adjacent structures thereby minimizing the risk for damage to those structures. The pump can be turned on briefly, as in our case, to identify any residual bleeding and enabling a more complete resection.

Some investigators have recommended maintaining low flow [500 ml/min] with profound hypothermia, because this may increase the duration of safe ischemia while not producing bleeding at the operative site. The risk for neurologic injury with deep hypothermic circulatory arrest is low, provided the period of circulatory arrest is less than 1 hour. There are no firm indications as to when deep hypothermic circulatory arrest should be used in the treatment of arteriovenous malformation. We agree with other authors who advocate transcatheter embolization as the primary treatment method. However there are cases in which experienced interventional radiologists may believe that embolization may not be feasible, or cases in which embolization has been associated with complications and further embolization is deemed hazardous. Patients with significant right-sided heart failure may benefit from surgical resection with deep hypothermic circulatory arrest, which would provide more prompt resolution of heart failure than multiple embolizations [8].

Conclusion

CPB with DHCA technique should be considered as one of the options in surgical repair of high flow arteriovenous malformation where usual surgical resection is not possible, multiple feeder involving deeper structures and in failed interventions with multiple embolisations. Complete resection, avoidance of damage to adjacent structures and bloodless field are some of the advantages of cardiopulmonary bypass with deep hypothermic circulatory arrest. Amputation can also be avoided in certain circumstances.

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