University of Toronto Neurosurgical Rounds No. 4

Global Arteriovenous Malformation of the Cervical Region

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INTRODUCTION

This case was presented as an interesting example of a global arteriovenous malformation (AVM) involving muscle, bone, extradural and intradural spaces of the cervical region.

CASE HISTORY

A twenty-eight year old male in October, 1976, fell at work sustaining a blow to his lower back. The following day he complained of cervical pain. Physical examination was negative and he had a full range of movement of his cervical spine.

Plain films and tomograms of the cervical spine showed a lytic honeycomb lesion of C2 and C3, and a 4 mm. subluxation of C3 on C4 (Fig. 1). The bone scan showed increased flow and decreased static activity in the upper cervical region. The radiological diagnosis was vertebral hemangioma; tumor, infection and histiocytosis were unlikely but important differential considerations.

The lesion was further evaluated by myelography which showed a circumferential narrowing of the subarachnoid space without actual cord compression from C4 to the foramen magnum (Fig. 2). Angiography revealed an extensive angiomatous mass in the vertebral bodies and posterior elements of C2 and C3 with a large extradural collection of abnormal vessels in the spinal canal. An intradural component was not detected and the anterior spinal artery was not identified. The lesion received blood supply from the vertebral arteries, ascending and deep cervical arteries, and the ascending pharyngeal artery bilaterally. Muscular branches of these vessels, as well as the occipital arteries, were linked by direct arterial anastomosis to each other providing alternate routes of supply (Figs. 3, 4, 5 and 6).

Because of the complexity of the malformation it was decided to treat the patient conservatively. He was fitted with a neck brace and discharged.

Between his first admission in 1976 and his second admission in March, 1978, the patient continued to suffer from intermittent low back pain and sciatica. The physical findings were marked nuchal spasm, limitation of neck movements, lumbar muscle spasm, and straight leg raising on the right limited to 45 degrees. Treatment consisted of bed rest and lumbar traction. Improvement occurred and he was discharged with his neck brace.

Eight months later a third admission was necessary because of a sudden onset of headache and vomiting. There were no focal neurologic signs, but his neck was stiff. A lumbar puncture revealed bloody spinal fluid. A CT scan showed slight ventricular enlargement. Cerebral angiograms were normal.

After much deliberation it was concluded that in spite of the risks involved an attempt at embolization should be made. Via a right transfemoral catheter the left ascending cervical artery was embolized with a mixture of gelfoam and pantopaque (Fig. 7). This reduced considerably the abnormal vasculature. The right occipital and ascending pharangeal arteries were catheterized. There was a large aneurysm arising off the right ascending pharangeal artery. After embolization this aneurysm was occluded and the vascular shunt much reduced (Figs. 8 and 9).

After the procedure the patient complained of retrobulbar pain and that he could not see properly. Examination proved that with effort he could read and that his vision was 20/30 in both eyes. He stated that the print was blurred. Fundi were normal and visual fields, although difficult to assess, were probably normal. He showed a loss of convergence and paralysis (UMN) of the left arm and leg with incoordination. Eight days later the arm and leg weakness had improved. There was nystagmus in all direction of gaze, upward gaze was limited and pupillary reaction only moderately brisk to light. He had suffered a mid brain lesion resulting in a Parinaud's syndrome. By the time of discharge all findings had cleared. From the above complication the emboli must have reached the vertebral system from the muscular branches.

On February 26, 1980, he was carrying his child upstairs on his shoulders when he

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Figure 1 — Lateral view of the cervical spine. There is a minor forward subluxation of C2 and C3. The bone texture of C2 and C3 is abnormal showing the typical honeycomb appearance of a vertebral hemangioma.

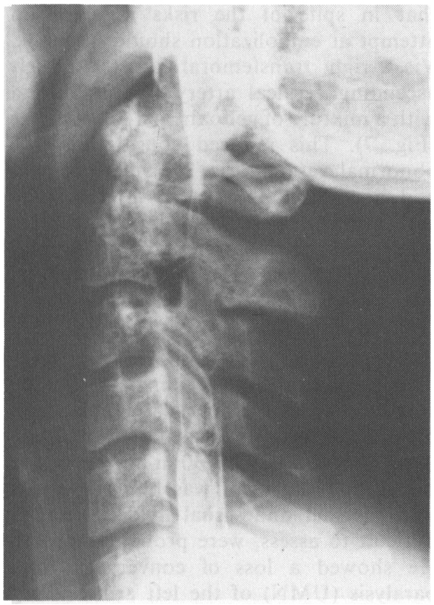

Figure 2 — Lateral view of the cervical myelogram. There is a circumferential narrowing of the dural sac (seen best anteriorly) which extends from C4 to the foramen magnum.

sneezed. This resulted in a sudden severe pain in his neck. The patient had noted in the last few months that every time he sneezed, coughed or strained he had to hold his head between his hands to prevent severe neck pain.

Figure 3 — A and B — Right and left vertebral angiogram. Subtracted views of the vertebral artery injections show extensive collections of abnormal vessels in the upper four vertebrae (right more than left). Lateral views confirmed the intraosseous position.

On admission, examination was normal except for limitation of neck movements. While in hospital the patient stated he could no longer hear the bruit. We had never heard a bruit, which had been complained of on admission, and there was no bruit audible on this examination. The patient was again discharged having improved.

DISCUSSION

Dr. W.M. Lougheed

The vascular supply to the nuchal muscles and vertebral bodies, extradural space and spinal cord is outlined in Figure 11 (Pia and Djindjian, 1978).

The ascending cervical and deep cervical arteries anastomose with both the spinal cord circulation and vertebral arteries. In turn, the vertebral artery contributed to the spinal cord circulation and with the external carotid circulation via the occipital artery. The occipital artery also anastomoses with the deep cervical. Due to the extensive network of communicating arteries each of these systems can therefore take part in the supply to the AVM making total irradication a complex problem.

Figure 12 (Pia and Djindjian, 1978), nicely illustrates the complex arrangements which may occur, especially when the AVM is global and is located in muscle, bone, extradural, intradural and intramedullary sites.

The extradural AVM's form about 15-20% of all vascular spinal abnormalities according to Bischof (1967). Feeders to the extradural malformations usually do not have any communications to the arteries feeding the spinal cord. However, Kendall and Logue (1977) reported one case draining to the intrathecal veins. The extradural AVM's are much less common in the cervical region, the most common site being mid thoracic.

According to Pia (1978), 27% show an apoplectiform course, 42% a remitting course, and 31% a progressive course.

Extradural hemorrhage or mass effect produces spinal cord compression with serious consequences. When the vertebral body is involved pathological fracture may occur.

Because of the partial block in the myelogram it would appear that in the future this patient will suffer from spinal cord compression. Pia (1978), stated that the timing for extradural AVM's to cause compression was 2-4 years following diagnosis. He made a plea for early treatment before a fixed neurologic deficit. Decompression would require laminectomy and extradural cauterization of the feeding vessels to the extradural component. The difficulty in staging the procedures becomes apparent as the muscular part of the malformation feeds the extradural component and hemostasis may be extremely difficult if the muscular component is not occluded first. Unfortunately, we cannot surgically remove the muscular component due to the extent and the anatomy, and embolization is not possible without first separating the extradural and intradural components. The only certain way of accomplishing this would be to occlude all the radicular arteries coming from the vertebral and ascending cervical branches. This, however, might compromise the normal blood supply to the spinal cord. The vertebral component might be eliminated by ligation of both vertebrals intradurally. This would have to be staged, necessitating two exposures. The proximal portion of the vertebral arteries could be embolized with large emboli. If the emboli were small there

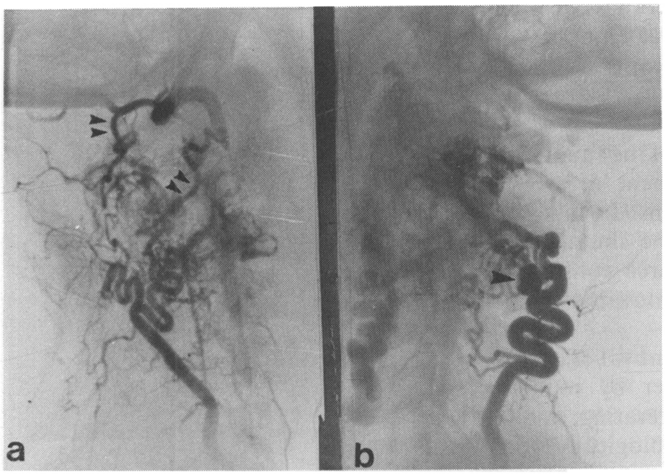

Figure 4 — A and B — Deep cervical artery. Selective injections into the deep cervical arteries show enlargement and tortuosity of the parent vessels and numerous abnormal vessels in the posterior elements and soft tissues. Of interest, an aneurysm can be identified on the left (B) (arrowhead), and direct anastomoses to the vertebral artery (arrowheads) can be seen on the right (A).

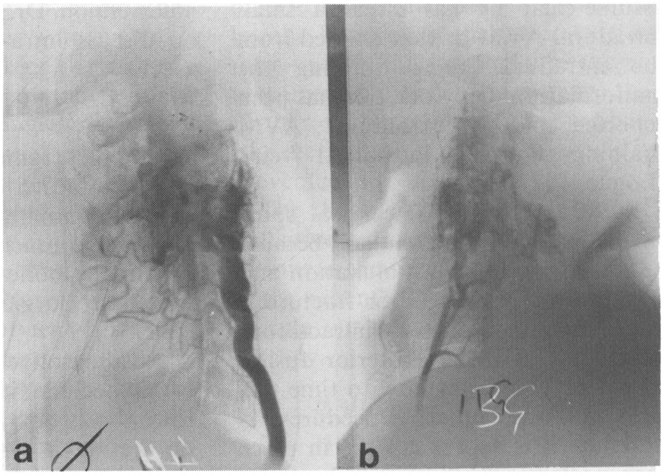

Figure 5 — A and B — Selective ascending cervical angiogram. The ascending cervical arteries also show hypertrophy (more marked on the right) A.) and supply of abnormal vessels to both the vertebral bodies and surrounding soft tissues.

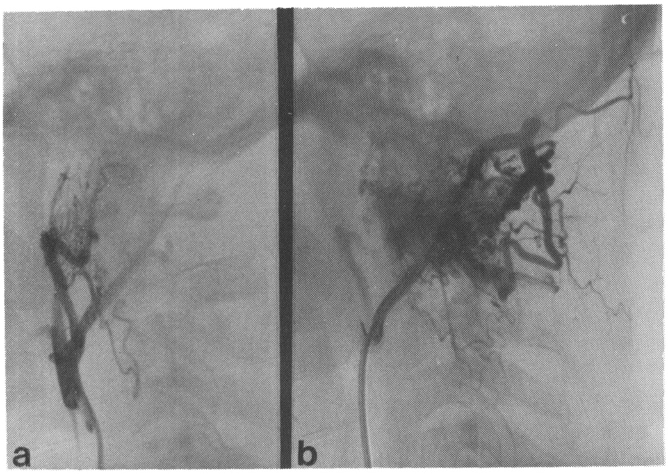

Figure 6 — Left external carotid injections. The left ascending pharyngeal artery (A) and the left occipital artery (B) also give direct and anastomotic supply to the hemangioma. Similar pictures were obtained on the right.

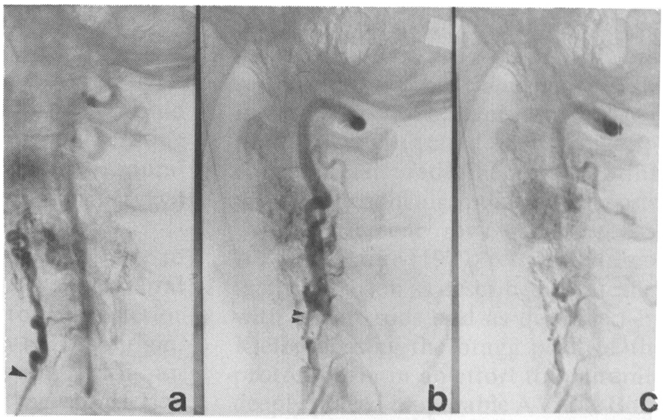

Figure 7 — Embolization, left ascending cervical artery. The catheter is well seated in the ascending cervical artery (A-arrowhead). Following embolization (A-B) there is obliteration of the parent vessel and a reduction in the flow to the malformation (compare with Fig. 5B). Despite good placement of the catheter a complication occurred as outlined in the text, presumably via the direct arterial-arterial anastomoses to the vertebral artery (B-arrowheads).

would be danger of embolizing normal cord circulation. Perhaps safer, but of more complexity would be to use a vein graft from the subclavian to the intradural segment of the vertebral artery and tie off the other vertebral intradurally. This would be carried out at the time of laminectomy. A third solution might be cannulation of the vertebral artery with a catheter balloon to obstruct the vertebral intradurally and then follow with proximal embolization. The sides could then be staged.

I am still perplexed as to how to deal with the ascending cervical branches because they give many branches to the spinal cord and embolization would probably result in a spinal cord infarct this time instead of a brain stem infarct as he had previously. Perhaps, if the emboli were large enough this could be avoided.

We did not visualize any intradural component by selective cord angiography. However, in the post embolization angiogram the anterior spinal artery became visible and no angioma was seen. Since the patient clearly had a subarachnoid hemorrhage we must assume that he has either a small intradural AVM or that he bled from the intradural veins draining the malformation. Only one case has been reported of an extradural AVM draining via the intradural veins (Logue, 1977).

Stabilization of the cervical spine should be considered eventually because he already has some subluxation and vertebral body collapse or fracture is one of the complications of intraosseous extradural AVM's. Posterior fusion would be better but how to time this with all the other procedures is difficult. If a bypass graft is in place then posterior fusion would be undesirable for fear of graft failure. Anterior fusion might be difficult unless the extraosseous component can be dealt with first.

I have decided to do nothing until his clinical state clearly demands intervention. Dr. Holgate has consented to discuss intra-arterial embolization (IAE).

Dr. R.C. Holgate

Complete removal of the shunt is the most definitive treatment of arteriovenous communications. In this case, because so much of the shunt occurs within the bone of three consecutive vertebrae, surgical removal is impossible.

An alternative to removal of a shunt is to occlude it either by inducing thrombosis or by obliterating, and/or obturating it with biologically compatible materials or devices — intra-arterial embolization (IAE).

Materials, devices, and methodology exist by which any vessel in the body can be successfully occluded. The hazard of IAE rests not with its ability to successfully occlude the target vessel,

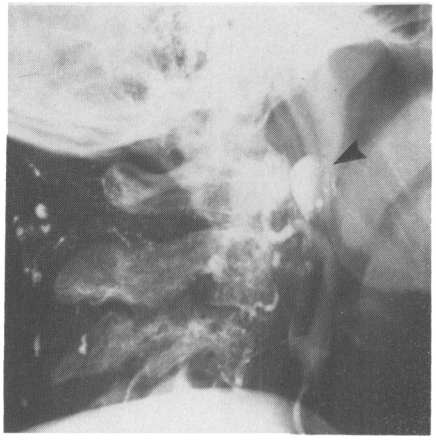

Figure 9 — Post embolization plain film. The plain film following embolization shows the distribution of the pantopaque labelled Gelfoam. Note the large collection in the ascending pharyngeal aneurysm (arrowhead).

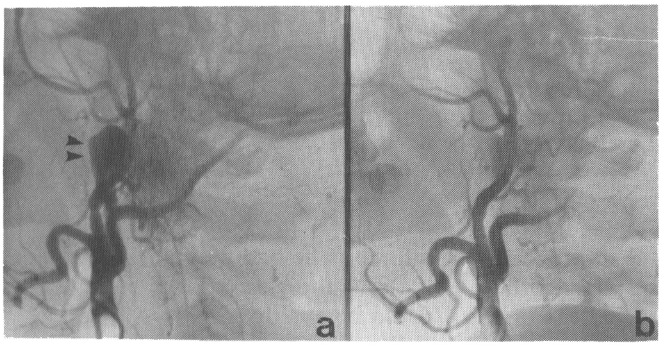

Figure 8 — A and B — Right external carotid embolization. A. Before embolization a large aneurysm (arrowheads) of the ascending pharyngeal artery and intraosseous supply to the upper cervical vertebrae is seen. B. After embolization the aneurysm is obliterated and the intraosseous vessels no longer opacify.

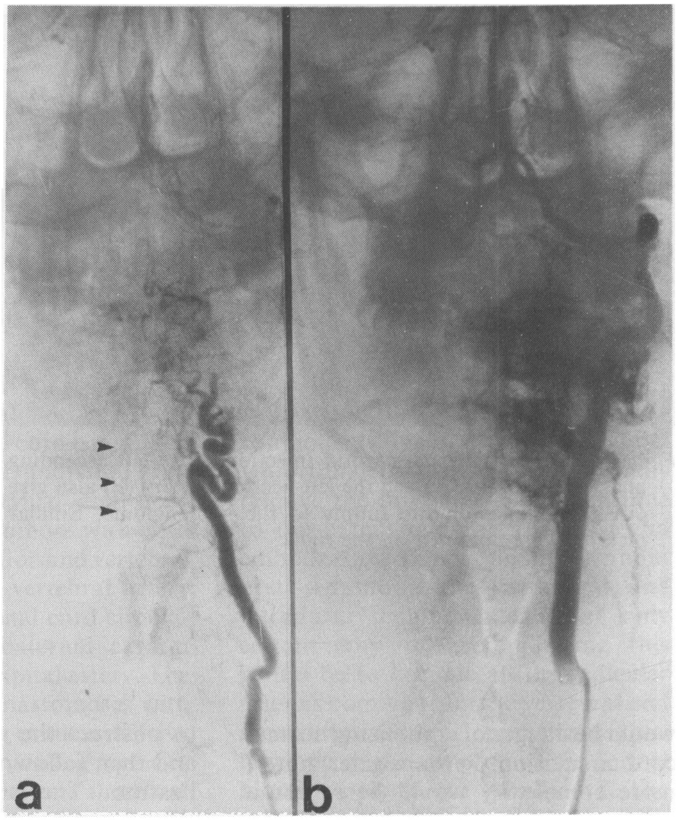

Figure 10 — A and B — Follow-up angiography. During his third admission a late post embolization angiogram was done. In Fig. 10A the previously obscured anterior spinal artery (arrowhead) can now be identified against the background of reduced left ascending cervical supply. The supply from the unembolized left vertebral artery has increased its contribution.

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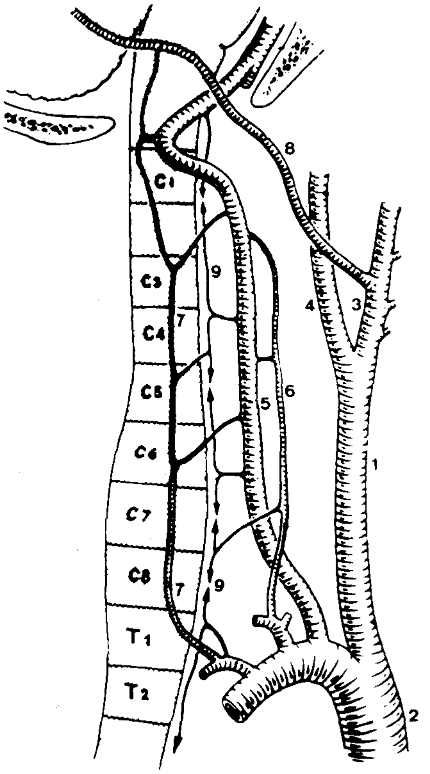

Figure 11 — The vascular supply of the cervical region from Pia and Djindjian (1978).

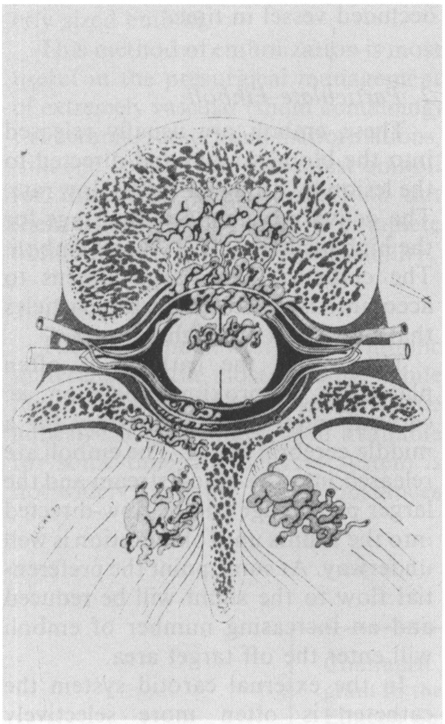

Figure 12 — Global arteriovenous malformation from Pia and Djindjian (1978).

but with its inability to avoid the occlusion of other vessels not part of the target. Assessment of the risk of IAE is an estimation of the relative chance and the degree of dysfunction which might attend the inadvertent occlusion of the off target vessels.

In this case there is global contribution and perhaps more importantly intercommunication between all of the vessels in the geographic area (vertebral, thyrocervical, costocervical and external carotid). As a consequence, although the majority of emboli released into any of these vessels will be drawn by sump effect into the shunt an irreducible minimum will be directed hemodynamically into the spinal cord, brainstem and higher centers.

The problem then reduces itself into how to isolate the neural tissue from the hemangioma (accepting the fact that one cannot be certain that none of the lesion is intra-axial).

There is probably no role for IAE in this case unless definitive therapy is anticipated. Definitive therapy would be aimed at: a) removing/obliterating the shunt; b) removing the extradural mass; c) restabilizing the cervical spine.

A plan of approach would have to include ligation of both vertebral arteries just proximal to the posterior inferior cerebellar artery (PICA) origin, probably in conjunction with an attempt to establish a new posterior fossa blood supply. Following this procedure all the feeding vessels except the left ascending cervical (which gives rise to the anterior spinal artery) would be embolized (Fig. 10). If one were to do IAE one would have to accept the risk and less devastating disabilities attendant upon dorsolateral spinal artery occlusion. The avascular angiomatous vertebral bodies would have to be removed and the cervical spine fused. The surgery would be done in the face of an unembolized left ascending cervical artery.

The attempt at definitive therapy would take place in a young otherwise healthy man with no neurological deficit. The risks of dorsolateral spinal artery occlusion(s) and bleeding from the unembolized vessels would have to be accepted. Furthermore, the treatment would occur without a clear picture of the natural history of the lesion, knowledge that a co-existing intra-axial component may be present and untreated and, no assurance that further subarachnoid haemorrhage (the mechanism is uncertain) would be prevented.

The therapeutic alternatives are: 1) palliative management of the complications as they occur, and 2) evaluation of the patient's fibrinolytic mechanisms to rule out the possibility of medical therapy.

Dr. A.R. Hudson

Dr. Hoffman would you consider radiation therapy in this case?

Dr. H. Hoffman

X-ray therapy for arteriovenous malformation has been advocated for many years. Although there have been sporadic reports of obliteration of an AVM by conventional radiotherapy, most reports using this therapy have not been studied angiographically and so the presumed beneficial effect of the radiation has not been proven. Furthermore, there is a very definite danger of conventional radiotherapy in this disorder producing radiation necrosis.

In his recent review of cerebral AVM's, Drake (1979) refers to stereotactic radiation as described by Steiner with cobalt rods and as described by Kjelberg using the bragg peak of the proton beam in an effort to obliterate deeply placed inoperable AVM's. Both these forms of radiotherapy resulted in complete obliteration of some AVM's, but these therapies were not completely free of the risk of radionecrosis.

Recently, Epstein (1980) reported on the beneficial effects of proton beam irradiation for deeply placed inoperable AVM's which were not amenable to embolotherapy. In his report, Epstein described cases in which deeply placed AVM's were completely obliterated by proton beam irradiation.

Consideration should therefore be given to the possible use of stereotactic radiation with the proton beam in this case if embolotherapy proves unsuccessful.

Dr. J.F.R. Fleming

Dr. Lougheed, have you considered using cardiac arrest with an injection of a fast setting glue to prevent it being swept into the neuraxis?

Dr. Lougheed

I would be concerned that our control of the direction and extent of spread might be inadequate. Also, I suspect that this malformation has more than one nidus so the glue might pass from one nidus to the other through normal intervening tissue.

Dr. Hudson

Dr. Humphreys you have just reviewed your experience at the Hospital for Sick Children, can we have your comments?

Dr. R.P. Humphreys

We have just finished reviewing our experience with spinal cord arterio-venous malformations at the Hospital for Sick Children (Humphreys, in print). These malformations are most commonly of the Type 111 (juvenile), with lush communications, multiple feeders and, in four instances, aneurysmal dilatations of component structures (Scarff, 1979). However, all of these lesions were intra-arachnoid, and in part or entirely intramedullary. There was none which had the extensive, multi-tissue involvement as Dr. Lougheed's patient.

In addition, we have had recent experience with two children who presented with extraosseous cervical vascular malformations. Both were obliterated by embolization techniques, a procedure successful no doubt because each lay within the territory of the appropriate external carotid artery only.

The operative approach to this patient would be overwhelming by virtue of the lush vascular supply beginning presumably within para-vertebral musculature, and extending through bone to the ventral dural surface. Even with radical operative techniques, such as total circulatory arrest (which we have not found to be useful for the vein of Galen malformations in the neonate), there is no guarantee that the anatomical exposure will allow the definition of the ventrally situated dural lesion.

It is tempting therefore, to recommend that nothing be done at this stage, particularly as the patient is asymptomatic. Presumably external spinal support has been provided, and in my view may be the only therapeutic aid required.

Dr. Hudson

Dr. Holgate could you review for us the present status of embolic techniques?

Dr. Holgate

The following section is meant to give the reader an overview of the technology available, the advantages and disadvantages of each class of embolus. Lastly, there is a questionnaire which reviews the mental hurdles encountered in assessing the advisability of embolizing a particular lesion. The case reviewed in this paper will serve as an example of the complexity of treatment planning in IAE.

CLASSIFICATION OF EMBOLI

1. Embolic Devices
   Guide wire fragments, coils, balloon catheters, detatchable balloons.
2. Particulate Emboli
   Blood clot (autologuos, amicar)
   Hemostatic agents: Gelfoam, Surgical, microfibrillary collagen
   Muscle
   Hemoclip — Gelfoam
   Polyvinyl alcohol sponge
   Molybdenum beads, lead pellets, iron filings
   Plastics: silicone spheres, polyurethane spheres
3. Liquid Emboli
   Pharmacologic agents ie: cytotoxic drugs, amicar
   Sclerosing solutions ie: 50% glucose
   Liquid plastics: acrylic tissues adhesives, liquid silicone, ferromagnetic silicone.

1. Embolic Devices

This class of embolus is delivered or placed in the vascular tree at the tip of an angiographic catheter. Because of their size (guide wire fragments, balloons) the complexity of their shape (coils) they remain in situ at the catheter tip and are hence, the least likely of all emboli to give rise to complications due to occlusion of off target vessels.

The devices enjoy effective use as intravascular ligatures in vascular tumors which are immediately removed surgically. Because of the short time interval between IAE and surgery there is little chance of recruitment by the tumor of collateral supply and the usual peroperative blood loss can be significantly reduced. These emboli are also of significant value in the management of arteriovenous fistulae where a balloon may be inflated and detached (or tethered proximally) to occlude the shunt. Guide wire lengths can similarly be placed so that both the afferent and efferent arterial loops of the fistula are occluded.

Despite their efficacy in the instances outlined above the devices have certain limitations. Because of their finite size they can only be used to occlude larger vessels and are not useful in the nonsurgical management of tumors. In the management of AVM's they can be used to isolate the lesions by pruning off some of the usual collaterals and, as a secondary benefit, increase the flow rate through the main feeding vessels for subsequent flow-directed emboli. Furthermore, the balloons may collapse and the metallic devices might erode through the occluded vessel in time.

2. Particulate Emboli

These emboli are usually released into the blood stream and directed to the lesion by its higher blood flow rate. The delivery system sets the stage for the high incidence of off target emboli. The catheters have large lumens to accommodate the size of the particles that must pass through.

The tip of the catheter is often placed in a proximal position ie: cervical internal carotid artery for a middle cerebral AVM. The emboli are released into the blood stream and the larger percentage will be flow-directed into the shunts until obliteration is well underway. At some point the preferential flow to the shunt will be reduced and an increasing number of emboli will enter the off target area.

In the external carotid system the catheter is often more selectively placed. Furthermore, occlusion of off target vessels has less devastating effects. Nevertheless, a point will be reached at which the shunt is partially occluded and the pressures used to inject the emboli will transiently exceed the pressure directing blood through the shunt. Once this imbalance obtains, flow can be reversed and emboli can be propelled backwards through the external carotid artery and thereby gain access to the internal carotid artery and the brain. Reflux of emboli can be prevented by wedging or ballooning the catheter tip, but at the cost of losing the preferential flow that directs the emboli to the lesion.

Apart from the increased hazards of stray emboli, particles have limitations by virtue of their composition. Blood clots, and the hemostatic agents though safe seldom give rise to permanent obliteration of the occluded vessels because they are rapidly removed by the fibrinolytic system. The plastic beads (silicone, polyurethane) are more permanent but only obturate the vessels and do not promote subsequent thrombosis. Polyvinyl alcohol sponge and muscle produce safe permanent occlusion, but are limited in usefulness by technical difficulties in delivering the emboli into the catheter and in the preprocedure production of regularly shaped, properly sized emboli.

This method of embolization is most useful in the presurgical management of extremely vascular (shunt containing) extracranial tumors and malformations. Silicone spheres are the safest emboli for intracerebral malformations but their efficacy is not high ie: complete obliteration is a virtual impossibility.

3. Liquid Emboli

These are the newest of the embolic materials and the most exciting. While liquid silicone rubber and the tissue adhesive acrylics have been available for some time, the delivery system is not widely available. These substances are delivered through balloon tipped microcatheters. These catheters are 1-2 Fr in size opposed to the 5-9 Fr size employed for devices and particulate emboli.

The balloon tip of the catheter allows for supraselective catheter placement ie: middle cerebral or ophthalmic artery. Once in place and inflated the balloon will also prevent reflux of the occluding liquid into off target vessels.

The drawbacks of this technique are inherent in the substances used. The acrylics are less viscous and can easily produce an intravascular cast which occludes both the arterial and venous side of a shunt, but the substance is too efficacious and enters not only the shunts but the normal capillary beds in surrounding tissues leading to a greater propensity to infarction of intervening normal tissue. There have been problems with intracerebral bleeding and catheters which have become "glued" inside the head.

Liquid silicone in its normal state is too viscous. Ingeniously treated, its viscosity has been greatly reduced, but at the cost of increasing its hardening time. Because of this slow setting time, isolation of the target vessels from the influx of untreated blood is a must, thus increasing the technical complexity of embolization and often requiring that multiple catheters be employed.

Liquid silicone is clearly the best material for the nonsurgical, stand alone, embolic treatment of such lesions as glomus jugulare tumors. It may well be the best material to use in the treatment of spinal cord AVM's.

CHOOSE YOUR WEAPONS

Pre-embolization Questionnaire

1. Is there an acceptable therapeutic alternative?
2. Is embolization to stand alone or used as an adjunct/assist to surgery? ie: if surgery is contemplated a more limited embolization may well suffice.
3. What tissues are exposed to potential off target emboli?
4. Can the damage (deficit) resulting from off target emboli be tolerated/accepted in the face of current knowledge of the natural history of the lesion untreated?
5. If the lesion is to be embolized what steps can be taken to avoid off target emboli?
6. Are the materials and expertise available to perform the type of embolization most efficacious for this lesion?

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