and minimal-change disease.\(^6\) (4) Antibody response to type-3 pneumococcal polysaccharide can be enhanced in patients with minimal-change disease by treatment with antithymocytic globulin, suggesting the presence of abnormal suppressor T-lymphocytes.\(^7\) Based on these clinical observations an association between minimal-change disease and thymoma was predicted by Shalhoub;\(^4\) however, to the best of our knowledge, this association has not been described in the literature.

In our case, additional circumstantial evidence supports the fact that minimal-change disease was a paraneoplastic process, rather than a coincidental event. Minimal-change disease in elderly patients is rare and generally occurs in relation to malignant disease other than thymoma.\(^8\) When minimal-change disease is the only disease in the elderly patient, it is usually responsive to steroid therapy.\(^10\) The minimal-change disease presented in this case was resistant to steroid treatment, suggesting a possible independence of this nephropathy with the malignant thymoma, which does not respond to steroid therapy.\(^11\) Remission of Hodgkin’s disease also results in a remission of minimal-change disease in these patients.\(^1\) Unfortunately, in our case, this was not achieved, since the thymoma was unresectable. The minimal-change disease developed late in the course of the thymic malignant disease. An explanation might be that the mutant clone of lymphocytes originated towards the end of the course of the disease.

In conclusion, this case provides further indirect evidence that minimal-change disease might be the result of an imbalance in cell-mediated immunity. Nevertheless, until an experimental model for minimal-change disease induced by disorders of cell-mediated immunity is created, our assumption concerning the mechanism underlying this clinical observation remains hypothetical.

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REFERENCES


Massive Hemothorax Secondary to Pulmonary Arteriovenous Fistula*

Treatment by a Catheterization Procedure


Massive pulmonary hemorrhage secondary to an acquired arteriovenous fistula is a rare event associated with high mortality. Cotton wads mounted on steel coils were inserted by percutaneous catheter and successfully occluded a pulmonary arteriovenous fistula in a patient who had massive hemothorax and contraindications to thoracotomy.

Massive pulmonary hemorrhage is life-threatening and associated with high mortality.\(^1,2\) Suffocation occurs in 50 to 100 percent of patients treated without operation. The most common causes of hemothorax are tuberculosis, arthropilosis, bronchial tumors, and bronchiectasis, but pulmonary arteriovenous fistulas, multiple or single, are well-documented causes of massive hemothorax.\(^1\)

Various nonoperative treatments of massive hemothorax have been attempted. None has been uniformly successful, and mortality rates approximate 85 percent.\(^3\) Operation is the treatment of choice, but contraindications to surgery include inadequate pulmonary function, inability to locate the site of bleeding, bronchial carcinoma with involvement of the mediastinum, multiple bleeding sites, or severe coagulation deficiencies.

The following case illustrates a new method of managing massive hemothorax from a pulmonary arteriovenous fistula without thoracotomy.

CASE REPORT

A diagnosis of single left ventricle, L transposition of the great arteries, and pulmonic stenosis was made at cardiac catheterization at age four years in a 15-year-old boy who had been cyanotic at birth. At age five, a Glenn anastomosis was performed for palliation of cyanosis and polycythemia. During the summer of 1977, he developed progressive dyspnea, cyanosis, and polycythemia. In October 1977, he had his first episode of hemothorax which terminated spontaneously.

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time, and platelet count were all within normal limits. Cough produced only minimal amount of blood-stained sputum until 9:30 AM. At this time, the patient coughed up 400 ml of bright red blood in a ten-minute period, but again bleeding stopped spontaneously. The pulmonary arteriovenous fistula was the most probable source of hemorrhage. Bronchoscopy was not done because of the rapidity and amount of hemorrhage.

The two episodes of massive hemoptysis demonstrated an urgent need for definitive therapy. There were several considerations, however, which weighed against resection. First, because of cyanotic heart disease and the previous Glenn operation, many tortuous thin-walled vessels were expected in the adhesions between the right lung and chest wall. Second, the patient needed all of his lung for oxygenation; it was uncertain whether or not he could tolerate lobectomy. Lastly, the open sternal wound increased the possibility of infection. These considerations favored an attempt to occlude the arteriovenous malformation with Gianturco stainless steel coils.

Under fluoroscopic control, a 6.5 French cobra-shaped catheter was introduced percutaneously through the left internal jugular vein into the right pulmonary artery branch supplying the fistula. Seven stainless steel coils with attached wads of cotton-wool were then selectively passed through a nontapered 7F Teflon catheter into the subsegmental arterial branches supplying the fistula until repeated injections of contrast material had confirmed that flow to the fistula had been completely obliterated (Fig 2).

The patient coughed up small amounts of blood over the next 48 hours and complained of right pleural pain. He also developed prominent neck veins, pitting edema of both upper extremities, and swelling of the soft tissues of the head. Chest x-ray film showed consolidation of the right lower lobe and a right pleural effusion. These findings were consistent with the superior vena caval syndrome and infarction of the right lower lobe and were direct consequences of acute occlusion of the pulmonary arteriovenous fistula with the Gianturco coils.

Over the following days, the patient made a progressive recovery, but occasionally produced blood tinged sputum.
His upper body edema had resolved by hospital discharge on the 29th postoperative day.

Seventeen weeks following his surgery, the patient presented with massive right heart failure. He was treated vigorously but died in intractable failure on June 23, 1978, having had no recurrent hemoptysis. Autopsy showed the arteriovenous fistula was totally obliterated and the lung distally demonstrated multiple areas of infarction with beginning organization (Fig 3).

DISCUSSION

Hemoptysis secondary to pulmonary arteriovenous fistula is uncommon, accounting for less than 2 percent of cases in most series. In those patients who develop a fistula, which is a recognized complication of the Glenn superior vena cava-right pulmonary arterial anastomosis, the incidence of hemorrhage complications is most common in the second and third decades, and is approximately 20 percent.

Silastic spheres, blood clot, autologous tissue, tissue adhesives, and gelfoam have all been used in the past in an attempt to stop localized hemorrhage. These materials, however, when injected into an arteriovenous fistula, can be life-threatening because of possible passage of the material into the pulmonary veins with resultant systemic embolization.

In 1976, Wallace et al. reported the use of a stainless steel occluding device in 24 patients, two of whom had renal arteriovenous fistulas. The coil successfully occluded the fistula in each case. The Gianturco coil was subsequently used in surgically created fistulas between the carotid artery and jugular vein in dogs where it produced a fixed obstruction at the point of placement and complete occlusion of the fistula within 5 minutes. Fibrous encasement eventually occurred and resulted in permanent closure. This same technique has been utilized, clinically, to correct hypoxemia resulting from arteriovenous fistulas.

In our patient, occlusion of the pulmonary arteriovenous fistula produced two complications: the superior vena cava syndrome and pulmonary infarction. After a Glenn anastomosis, resistance of vessels in ventilated portions of the right lung progressively increases. In this patient, as vascular resistance increased, more unoxygenated blood was shunted through the arteriovenous fistula into the pulmonary veins. This prevented systemic venous hypertension but increased arterial hypoxemia. Acute occlusion of the right-to-left shunt disclosed the increased vascular resistance of the ventilated portions of the right lung and produced the superior vena cava syndrome.

Occlusion of a pulmonary artery does not generally cause hemorrhage consolidation unless the affected lung also becomes atelectatic. If the affected lung remains inflated, infarction does not occur. In this patient, hemorrhagic atelectasis of the right lower lobe developed after occlusion of the fistula in those areas of the right lower lobe not supplied by bronchial arteries.

Gianturco steel coils may have wider application in patients with multiple pulmonary arteriovenous fistulas or in patients with other causes of hemoptysis in whom thoracotomy is contraindicated or unduly hazardous. In patients with arteriovenous fistulas and severe hypoxia secondary to a Glenn anastomosis, the technique may be preferable to ligation of the right pulmonary artery and reanastomosis of the superior vena cava to the right atrium.

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Echocardiographic Features of an Unruptured Aneurysm of the Right Sinus of Valsalva*

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A 2-cm aneurysm of the right sinus of Valsalva was documented in a patient with a prosthetic aortic valve. The M-mode findings differed from prior reports and mimicked those of aortic root dissection or a catheter placed in the right ventricular outflow tract. Two-dimensional echocardiograms readily distinguished the aneurysm of the right sinus of Valsalva from the alternative possibilities.

Aneurysms of the sinus of Valsalva are usually asymptomatic until rupture.1 They result from a defect in the attachment of the aortic root to the fibrous ring connecting it to the aortic valve and left ventricle. In the case of an aneurysm of the right sinus, growth of the sac and retraction from its initial position lead to its pointing toward the right ventricular outflow tract, anterior to the ascending aorta. Rupture is usually into the right ventricle just below the pulmonic valve.

We describe a case of a presumably acquired aneurysm of the right sinus of Valsalva secondary to bacterial endocarditis, a known cause of such a condition.2 Special attention to heretofore unreported echocardiographic features indicates that findings on M-mode echocardiograms can closely simulate those consistent with an ascending aortic dissecting aneurysm. Real-time two-dimensional echocardiograms can help make an accurate noninvasive distinction between the two conditions.

MATERIALS AND METHODS

Both M-mode and two-dimensional echocardiograms were obtained with the patient in a semirecumbent 30° left lateral decubitus position. M-mode recordings were made with a multichannel recorder (Irex Continumtrace 101) interfaced with a cardiac ultrasonic module (Irex 150-149) using a 2.25-MHz transducer (Aerotech) at a paper speed of 25 mm/sec. Two-dimensional study was performed with a phased-array sector scanner (Varian V3000).

CASE REPORT

In 1969, after intravenous abuse of drugs, a 23-year-old white man without known antecedent heart disease developed bacterial endocarditis. Medical therapy was successful in controlling the infection, but the patient was left with residual aortic regurgitation. He suffered repeated bouts of endocarditis in 1970 and 1972. Cardiac catheterization in June 1973 showed severe aortic regurgitation and an aneurysm of the right sinus of Valsalva. In September 1973, at another hospital, the patient underwent aortic valvular replacement with a No. 2 Kaye-Suzuki prosthesis. At surgery, a nonperforated aneurysm of the right sinus of Valsalva was described.

In March 1976, the patient again developed endocarditis, and four months later, after successful medical treatment, a soft murmur of aortic regurgitation was noted. Catheterization in July 1976 revealed 1+ aortic insufficiency with a paravalvular leak in the area of the aneurysm of the right sinus of Valsalva, without evidence for a left-to-right shunt. The right sinus of Valsalva was aneurysmal and calcified. Surgery was subsequently planned, due to fear of rupture of the aneurysm. Furthermore, the patient had suffered several episodes of cerebral embolization. The patient refused surgery but underwent repeat catheterization in 1978 when, after suffering repeated embolic episodes, he agreed to undergo surgery.

Preoperative M-mode echocardiograms demonstrated an abnormal thick echocardiographic density 21 mm anterior to the anterior aortic wall. Its motion was identical to that of the aorta (Fig 1, left). Scanning showed this structure to converge inferiorly with the upper interventricular septum.

On the longitudinal view, two-dimensional echocardiograms demonstrated an echo-dense saccular structure 2 cm in diameter at the base of the anterior aortic root (Fig 1, top right). When the two-dimensional cursor was used to derive an M-mode record, a pattern identical to that described previously herein was obtained. A representative frame from the patient's sortogram on his most recent catheterization is shown to compare with the two-dimensional image (Fig 1, bottom right).

In January 1979, the patient successfully underwent repeat aortic valvular replacement with a prosthesis (No. 21 Björk-Shiley), as well as aneurysmal repair by exclusion with a Dacron patch. Postoperative echocardiograms were essentially without change from the preoperative studies, with the exception of better definition of the (new) aortic valvular poppet.

DISCUSSION

The few echocardiographic reports of aneurysm of a sinus of Valsalva have indicated variable findings. Roth-