

PULMONARY ARTERIOVENOUS MALFORMATION: STILL A SURGICAL CONSIDERATION

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Abstract - Pulmonary arteriovenous malformations are rare clinical entities that are associated with right to left shunts, and are often clinically presented by the triad of dyspnea, cyanosis, and digital clubbing. Currently, transcatheter embolization of fistula is gaining increasing popularity as the superior therapeutic option. In certain situations, however, surgery is preferred. Two cases of direct pulmonary artery - left atrial fistulas are described who, were treated simply by surgery with no recurrence. *Acta Medica Iranica* 36 (2): 92 - 96; 1998

Key words: Pulmonary artery, arteriovenous fistula, pulmonary arteriovenous malformation, pulmonary arteriovenous

INTRODUCTION

In variance with their systemic counterparts pulmonary arteriovenous fistulas tend to trigger rarely a substantial hemodynamic instability. Instead, there have been a multitude of threatening events reported, including neurologic complications; massive pulmonary bleeding into the pleural space; pulmonary hypertension and endocarditis (1 - 7) which might adversely affect the life history of patients involved with these malformations. Accordingly some therapeutic intervention finds justification to treat these lesions. Few authors have claimed survival of variable durations on patients treated by nonsurgical methods and a selective approach has been proposed (8,9). However, others have not consented on their conclusions. The experience of the majority of authorities reported on this issue has shown lower mortality and morbidity with the invasive intervention versus the conservative treatment.

Since the first surgical excision by Shenstone (10) in 1940 until the recent years, surgery was thought to be the only therapeutic option to manage patients. Surgical outcomes, however, were at times not satisfactory, especially with multiple lesions affecting one or both lungs. Technologic advances and technical innovations developed in interventional catheterization

procedures have in recent years encouraging promises to the diagnosis as well as the treatment of cardiovascular diseases. Accordingly, transcatheter angioembolization of arteriovenous fistulas has gained increasing popularity so that it is viewed by a dominant number of practitioners as a superior alternative to surgery (11, 12). Despite the safety and relative efficacy pertaining to the embolization therapy, yet it is unwarranted to refute surgery entirely as an outdated traditional mode of treatment. There are likely specific occasions where by that might merit properly as the procedure of choice. In this paper we are presenting two cases of pulmonary arteriovenous malformation whom were treated simply and successfully by surgery. The rationale for their surgical indications will be discussed.

CASE REPORT

Two consecutive patients were referred to our institution because of unexplainable dyspnea and cyanosis in May 1993 and August 1997. Their clinical features were almost identical and they are summarized in Table 1. Chest X - ray findings were unremarkable. The arterial blood gas analysis revealed an obvious oxygen desaturation of 73% and 84%, at room air, in case 1 and 2 respectively. The color flow mapping echocardiography scanned a shunt flow from the pulmonary arterial system towards the left atrial chamber in both patients (Fig. 1). The pulmonary arterial cineangiogram was diagnostic by outlining a fistula between the right pulmonary artery and the left atrium in each individual. The cardiac catheterization data are brought in Table 2. The final diagnosis of pulmonary arteriovenous fistula was established in both cases by clinical findings and diagnostic work up studies. The patients were scheduled, as elective cases,

fistulas was carried out through standard midline sternotomy incisions while using extracorporeal circulatory support. The morphologic findings met at surgery, on case 1 included a large defect in the interatrial septum occupying the fossa ovalis membrane. The cusps of the pulmonary valve were somewhat thickened and there was mild fusion at commissures as well. Similarly case 2, presented with a moderate size patent foramen ovale in association with a dilated left atrial chamber. The common finding to both was a fistula track that made its path through the mass substance of the roof of the left atrium. The fistula extended between the right pulmonary artery and the left atrial chamber.

Total correction was accomplished, in both cases, by suture closure of entry and exit openings of the fistulas through an arteriotomy incision in the right pulmonary artery and the pre - existing defect in the interatrial septum. Subsequently, the atrial septal defect was repaired accordingly.

The post - operative course was uneventful in both cases. The pre - operative arterial oxygen saturation on case 1 and 2 improved from 72.5% and 84 %, at room air, to 94% and 97% respectively post - operatively. The post - operative echocardiogram successfully illustrated complete elimination of the shunts (Fig. 2). The patients were followed periodically at our outpatient clinic for 52 months and 3 months respectively. Repeated color mapping echocardiographies have shown complete extirpation of the fistulas and no recurrence. The patients have remained asymptomatic so far, leading normal lives and are in good health.

DISCUSSION

Pulmonary arteriovenous fistulas are rare lesions that might be large or small, single or multiple, and nearly all are congenital. Their association with hereditary hemorrhagic telangiectasia has been well recognized (13). The peculiarity of our two cases, is related to their unique morphologic features. The

Table 1. Patients' Clinical Features

Case No	Sex/Age	HHT	Dyspnea	Cyanosis	Clubbing	Bruit	Pre op.Hct/Hb	CNS complications
1	F/23	No	Yes	Yes	Yes	Yes	51/17	No
2	M/11	No	Yes	Yes	Yes	Yes	46/15	No

HHT=Hereditary hemorrhagic telangiectasia;

Table 2. Pre - operative catheterization data

Site	Pressure in mm Hg	
	Case 1	Case 2
Right Atrium	4	2
Right Ventricle	40/4	18/2
Pulmonary Artery	16/2	18/5
Capillary Wedge	6	Not measured
Left Ventricle	Not measured	105/8
Aorta	Not measured	105/7

anomalies, morphologically alike in both patients, are specifically characterized by an isolated aberrant vascular channel that runs within the roof of the left atrial chamber. The vessel bridges between the right pulmonary artery and the interior of the left atrium. It conveys the unsaturated venous blood to the left side, creating a right to left shunt. This malformation could be regarded as a specific subtype of pulmonary arteriovenous fistulas in accord to Szilagy's (14) classification.

The occurrence rate of pulmonary arteriovenous malformations, including all varieties collectively, has been recognized exceedingly low; not more than few hundred cases have been reported to date. Puskas and coworkers (15) from Massachusetts General Hospital were only able to document 21 cases antemortem, from 1964 to 1992. The mean age was 37.5 years. Diagnosis in the pediatric age group has been yet more unusual. In a series of 63 cases from Mayo Clinic, only 4 were found in children over a period of 20 years (8). Our case 2 represents a yet more scarce subgroup. In a review of the literature, Schumacker and associates (16) collected 26 reported patients treated surgically at the age of 16 or younger over the years 1947 to 1963. Amongst them there was only one instance who was similar to our case described here.

Pulmonary Arteriovenous Malformation

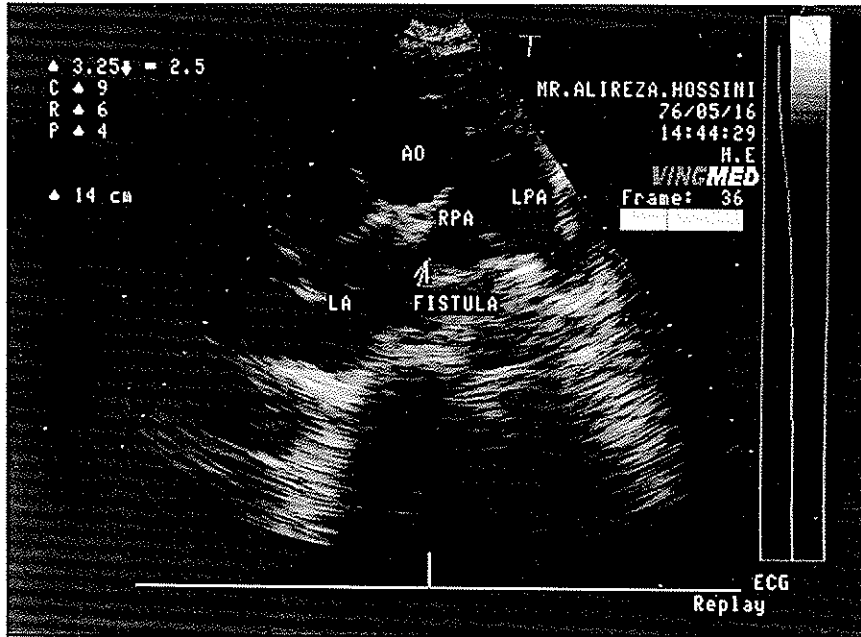


Fig. 1. Pre - operative echocardiogram showing a fistula between the right pulmonary artery and the left atrial chamber

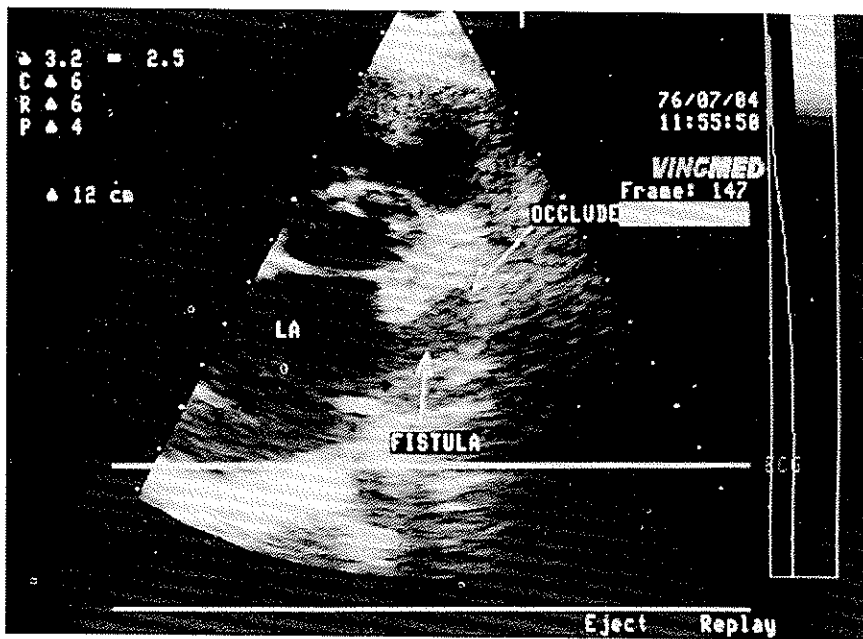


Fig. 2. Post - operative echocardiogram. The fistula is closed

The developmental basis of pulmonary arteriovenous malformations has been well elucidated by others (17). A developmental error during the growth process of the pulmonary arterial system, and the rete- form venous plexus formation arising from the lung bud, leads to the retention of primitive arteriovenous connections. Although the process of maldevelopment in our two cases is hard to explain, we speculate that a developmental error has probably resulted in the persistence of the early arteriovenous connections. The subsequent involution into the left atrial wall has created a fistula extending between the right pulmonary artery and the left atrium.

Unexplained clubbing of digits and effort cyanosis noted in our patients are, in essence, the usual presentations. Clearly, these findings reflect a major arterial desaturation due to a large pulmonary shunt. Repeated major or minor cerebral accidents complicate as high as 5% to 30% of cases according to several authors (1, 2, 13). We did not observe these complications in our patients. These events are more frequent in patients beyond the 3rd decade of life (2,5). Septic or aseptic emboli streaming through the abnormal route escape the natural filtering process in the lungs. Life threatening cerebral thromboses or abscesses are related to the existing polycythemia, cyanosis, or migrating paradoxical emboli (1).

We positively believe that cardiac catheterization and selective pulmonary angiography are still superior, as the gold standards, to other investigative measures. They should be implemented in all cases (18) to specify the site and size of fistula. Echocardiography, on the other hand, has proved valuable as a noninvasive screening means as well as a simple method of assessing the efficacy of therapeutic interventions (19).

Historically, since the first surgical excision in 1942 until 1978, surgery was the only therapeutic option recognized. Various procedures were exercised including total excision, segmentectomy, lobectomy and even pneumonectomy. Results however, were not consistently gratifying; as very often lesions happened to be multiple, situated in one or both lungs. The complete extirpation was often not feasible, or required

sacrifice of the healthy lung parenchyma along with the removal of the lesion. At times, staged thoracotomies were mandatory. In a series including 17 patients with multiple lesions treated surgically, 4 failed to improve (8). The introduction of coil embolotherapy by Taylor and coworkers in 1978, and subsequently balloon embolotherapy by Terry and coworkers (12) in 1980 were promising advances. These procedures soon became favored by many physicians as the preferred alternatives for surgery. Currently, procedures involving the transcatheter therapeutic embolization of fistulas are thought advantageous over surgery by a number of practitioners. The logics supporting this notion include: 1) they are less invasive, and as such are especially well suited for surgically high risk patients and elderly people; 2) there is no need for anesthesia; 3) they are as curative as surgery; 4) they might be repeated as many times as needed for multiple or bilateral lesions; 5) there is no need to sacrifice the normal lung tissue to remove the lesion; 6) hospital stay is short; and 7) the procedure is less costly. On the other hand, surgical approach can be equally accepted when malformation happens to present itself as a well-defined extra- parenchymal solitary lesion. Surgery remains the only option when embolic procedures have failed or are not technically feasible. Accordingly, we elected surgery as our preferred choice to manage our patients; as their morphologic features rendered them suitable for perfect surgical correction. On the other hand, we believe that transcatheter embolic methods, despite their relative simplicity and safety, require experience and expertise of the practicing physician. These procedures are not recommended in the absence of proper facilities or qualifications.

Following surgery the arterial oxygen saturation in our patients rose up to normal values and both remained noncyanotic with their usual activities. The post - operative color flow mapping echocardiography verified the elimination of the shunts in both cases. Although the follow up on the second case is too short to make any comment, the first case has remained free from symptoms and the shunt has been nonexistent after nearly four years. Both patients are, presently,

Pulmonary Arteriovenous Malformation

leading normal lives and enjoying good health.

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