Case Report

A Case of Pulmonary Artery Bypass Surgery for a Patient with Isolated Takayasu Pulmonary Arteritis and a Review of the Literature

Nobuyuki Nakajima, MD,[1] Masahisa Masuda, MD,[1] Mizuho Imamaki, MD,[1] Atsushi Ishida, MD,[1] Nobuhiro Tanabe, MD,[2] and Takayasu Kuriyama, MD[2]

A 45-year-old female was presented with progressive dyspnea and bilateral leg edema. Pulmonary angiography revealed total occlusion of the right pulmonary artery and significant stenosis of the left pulmonary artery. The inferior lobar artery as well as the segmental arteries were well patent. No pathology was detected elsewhere at the aorta and its branches. The diagnosis of chronic pulmonary arterial occlusion by isolated Takayasu arteritis was made because of the characteristic pattern of angiographic findings and the presence of unusual shunt formation from the coronary artery to the peripheral portion of the pulmonary artery, as well as a characteristic presentation of HLA typing in blood analysis, which strongly suggested the diagnosis of Takayasu arteritis. To restore the pulmonary blood flow, we employed reconstructive surgery by means of bypass procedure, using PTFE graft. Postoperatively there was marked improvement in cardiopulmonary function and the quality of life of the patient. The graft was proved to be patent at long-term follow-up study. An extremely rare case of chronic occlusive pulmonary arteritis, which was surgically treated by means of bypass procedure, is reported herein, and a brief review of previous reports on this subject was attempted. (Ann Thorac Cardiovasc Surg 2007; 13: 267–271)

Key words: Takayasu arteritis, isolated pulmonary arteritis, pulmonary artery, pulmonary artery bypass surgery

Introduction

Most cases of chronic steno/occlusive lesion of pulmonary artery are attributed to thromboembolism; therefore pulmonary arteritis as the primary cause is extremely rare. We experienced a middle-aged woman who presented with dyspnea and right heart failure. Pulmonary angiography revealed total occlusion of the right pulmonary artery and significant stenosis of the left pulmonary artery. Bypass surgery was performed and satisfactory results were obtained. The etiology of this rare case, selection of surgical procedure, and a brief review of literature are presented herein.

From [1]Cardiovascular Surgical Service, Department of Surgery, and [2]Department of Respirology, Chiba Graduate School of Medicine, Chiba, Japan

Received September 13, 2006; accepted for publication December 15, 2006

Address reprint requests to Nobuyuki Nakajima, MD: Kashima Rosai Hospital, Japan Labor Health and Welfare Organization, 1–9108–2 Doaihoncho, Kamisu, Ibaraki 314–0343, Japan.

Case Report

A 45-year-old female patient developed dyspnea on exertion and bilateral leg edema, which progressed to facial and upper extremities at the later stage. She was diagnosed as chronic pulmonary thromboembolism and referred to us for surgical treatment. Various studies were undertaken to clarify the nature and the extent of this disorder, and the following results were obtained. Immunochemistry showed IgG: 2,180 mg/dL, CRP<0.3 mg/dL; ERS: 30 mm/H; ANA: (–); antiDNA antibody: (–); C3: 79 mg/dL; C4: 28 mg/dL; CH50: 40 U/mL; C-ANCA: (–); P-ANCA: (–); HLA typing: A24 (+) and B52 (+). Hemodynamic-respiratory functions are illustrated in

Table 1. Hemodynamic-respiratory functions before and after surgery in room air and 10 min after inhalation of 100% oxygen

	Before surgery	After surgery	Oxygen inhalation response (100%, 10 min after)	
			Before surgery	After surgery
VC	3.23	2.36		
%VC	118.7	87.0		
FEV1.0	2.69	1.91		
%FEV1.0	85.4	83.7		
PaO2	63.9	67.0		
PaCO2	39.1	40.8		
SaO2	92.8	93.4		
(Room air)				
HR	92	108	72	92
Psa	126/64 (86)	116/64 (84)	114/50 (74)	118/56 (86)
Ppa	98/18 (46)	84/18 (39)	90/16 (74)	62/14 (30)
CO	4.68	5.36	4.53	5.18
CI	3.10	3.53	3.00	3.41
SVR	1,435.3	1,223.4	1,271.0	1,296.8
PAR	734.7	432.7	617.9	308.8

Table 1. A ventilation-perfusion scan showed there was no defect on the bilateral lung in a ventilation scan, but a perfusion scan demonstrated as a near total defect on the right lung and a partial defect (S1+S2a+S3b) on the left lung. The pulmonary angiography was taken, which showed a complete occlusion of the right pulmonary artery. The left pulmonary artery was dilated at the proximal portion, and 70% of irregular, circular stenosis was observed from the upper lobe artery to the beginning of the A6 segment (Fig. 1). From this point to distal areas, no pathology was found. The pressure measurement at A9 was 22/10(14) mmHg, compared to 98/18(46) mmHg of proximal portion. The pulsatory index was (92–14)/43=1.81.

Aortobronchial arteriography was taken to clarify whether the associated pathology was presented at the aorta and its branches, but it was found to be normal (Fig. 2). The pulmonary artery at peripheral segments were visualized by retrograde fashion through bronchial artery circulation. The coronary angiography demonstrated the presence of collateral flow to the right pulmonary artery from #13 via sinus artery (Fig. 3). The gallium scintigraphy showed no accumulation of gallium in the body portion, which indicated the absence of active inflammatory reaction.

The surgical treatment was undertaken with the use of total cardiopulmonary bypass. The initial plan was patch reconstruction of the stenotic segment; however, this attempt was abandoned because of the presence of a dense adhesion between pulmonary tissues. As an alternative procedure, the bypass procedure using a 14-mm ring-supported PTFE graft was placed from the left pulmonary artery to the basal artery.

The postoperative course was smooth, and the follow-up study on cardiorespiratory function is illustrated in Table 1. In comparison to the preoperative values, there was marked improvement in hemodynamic-respiratory functions. The pulmonary arterial resistance was greatly reduced in response to the inhalation of 100% oxygen. After discharge from the hospital, the patient has been living a normal life. A follow-up pulmonary arteriography at 5 years demonstrated well patent bypass graft and normal appearance of the left peripheral segment of pulmonary arteries (Fig. 4).

Discussion

The cases with chronic steno-occlusive lesion of pulmonary artery are rare. Among them, thromboembolism is most popular; on the other hand, pulmonary arteritis-vasculitis is rarely encountered. Most pulmonary arteritis and vasculitis are attributed to Takayasu arteritis. Although the association of pulmonary pathology in Takayasu arteritis appears to be not so high, approximately 20% of involvement was reported in the literature.[1)] Contrary to this, isolated Takayasu pulmonary arteritis was very rarely

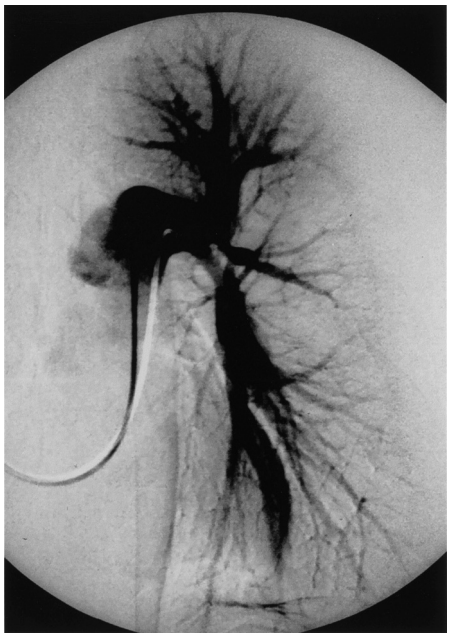

Fig. 1. Pulmonary angiography before surgery. Complete obstruction of right and significant dilatation and stenosis of the left pulmonary artery are visualized.

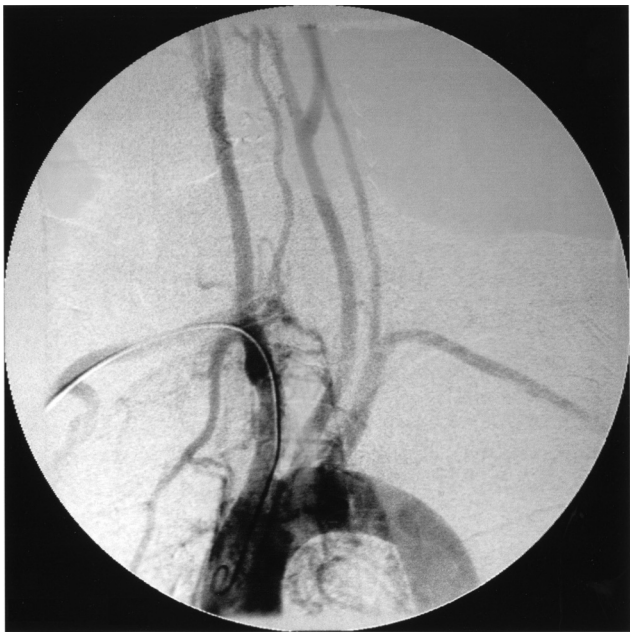

Fig. 2. Aortography shows there is no pathology at the aorta and its branches.

Table 2. Summary of the details of the status of occlusion of the pulmonary artery and the method of surgical correction reported in the literature

Reference	Occlusion (total/subtotal)	Laterality	Surgical Procedure	Material applied
Nakamura et al. (1984)[13]	Subtotal	Left	RA-RP bypass	16 mm Dacron
Moore et al. (1985)[14]	Subtotal	Right	Patch + bypass	14 mm Dacron
Chauvaud et al. (1987)[15]	(–) Severe stenosis	Bilateral	Patch	Pericard*
Okubo et al. (1988)[16]	Total	Left	Patch	Pericard#
Nakajima et al. (1992)[17]	Total	Left	Patch	Pericard#
Sundt (2001)[18]	Total	Right	Bilateral bypass	PTFE

*, glutaraldehyde-preserved autologous pericardium; #, fresh autologous pericardium.

reported.[2–6] The new classification of Takayasu arteritis, which was lately proposed by Hata and Numano, firstly mentioned pulmonary involvement as a type; however, the isolated involvement of the pulmonary artery was not described in their classification.[7] As a consequence, isolated or single involvement of the pulmonary artery in Takayasu arteritis is not classified as an independent type.

We have made a diagnosis of isolated Takayasu pulmonary arteritis in this case by the following findings; (i) young female, (ii) HLA type, (iii) angiographic findings (Figs. 1 and 2), (iv) presence of unusual communication from coronary to pulmonary artery (Fig. 3).

It is well known that most patients with Takayasu arteritis are relatively young females (about 80%)[8] and that there is a strong association with the particular site of HLA types, B52 and B39.[9,10] B52 was proved to be positive in our patient. The angiographic study showed that there was no pathology at the aorta or its branches (Fig. 2). The characteristic finding was presented only at the pulmonary artery. Through the review of literature, we would like to emphasize that there were certain characteristic presentations of steno-occlusive lesion demonstrated by pulmonary angiography in pulmonary arteritis patients. In 10 reported cases of isolated pulmonary arteritis, including Takayasu arteritis, reviewed in the literature, it was found that 80% of patients presented with total or near total occlusion of either side (right in 5 cases and left in 3) of the main pulmonary artery in addition to

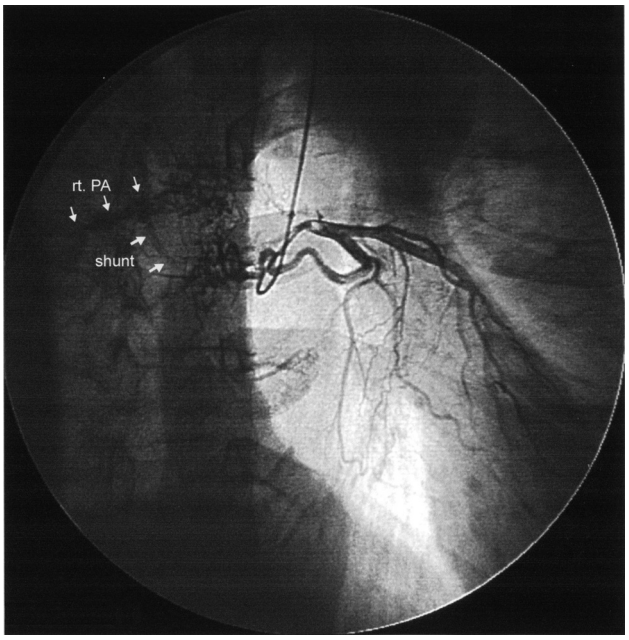

Fig. 3. Coronary angiography.
The presence of unusual communication/shunt from the left coronary branch to the peripheral segment of the right pulmonary artery.

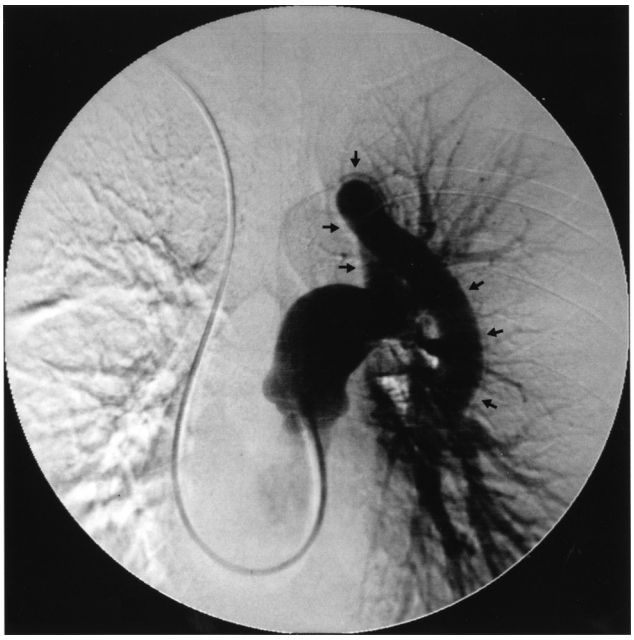

Fig. 4. Follow-up pulmonary angiography at 5 years.
Well patent bypass graft and a good perfusion of blood flow distally to the stenotic segment.

the significant stenotic lesion at the remains of the arteries. These findings would suggest that the chronic inflammatory process is insidiously proceed without a clear manifestation of clinical symptoms. Once major changes such as significant stenosis/obstruction in the pulmonary artery are developed, a significant reduction of pulmonary blood flow occurs, and the signs and symptoms of cardiorespiratory failures are initiated for the first time.

The presence of an unusual shunt formation or communication between the coronary or brachial artery to the peripheral portion of pulmonary arterial trees is also a characteristic finding associated with the significant reduction of pulmonary blood flow, and it is interpreted as a compensatory mechanism.[2,11,12)]

The surgical treatment is indicated when cardiorespiratory failure is presented. Until now, only 6 cases of detailed description on surgical treatment have been reported in the literature, including 2 of my (first author, N. Nakajima) own experience.[16,17)] Since stenosis/occlusion occur at the major portion of the pulmonary artery and the distal/peripheral portion of pulmonary arteries are patent, as is demonstrated by the presence of the coronary or bronchial artery to pulmonary arterial shunt, the basic concept of surgical repair is either an enlargement of the stenotic segment by patch technique or an application of the bypass graft distal to the steno/occlusive segment. This concept is completely different from that of the surgery for chronic thromboembolism where endoarterectomy is the essential procedure for surgical intervention.

In this reported case, our initial plan of reconstructive procedure was an enlargement of the stenotic segment by patch using her own pericardium; however, this procedure could not be accomplished because of the presence of a dense and firm adhesion of interlobular space. As an alternative, a bypass was created from the main pulmonary artery to the basal artery distal to the stenotic segment using a 14-mm ring-supported PTFE graft. The presence of dense fibrous adhesion at the visceral pleural space was a sign of chronic inflammatory reaction associated with pulmonary arteritis/vasculitis and has been documented by our own previous case,[17)] as well as by reports from others.[18)] In our previous case,[17)] we had to transect the ascending aorta to approach and to facilitate the exposure of the stenotic portion of the right main pulmonary artery; then firstly could be able to undertake patch reconstruction.

The follow-up pulmonary angiography at 5 years was documented by the presence of patent of the bypass graft as well as by clearly visualized blood flow to the periph-

eral segment of the pulmonary arterial trees (Fig. 4). From my own experience of 3 cases of surgical intervention for isolated pulmonary arteritis, it will be said that once the pulmonary blood flow is significantly restored, the prognosis of the patient appears to be convalescent.

References

1. Sheikhzaseh A, Tettenborn I, Eftekharzadeh M, et al. Occlusive thromboaortopathy (Takayasu Disease): Clinical and angiographic features and a brief review of literature. *Angiology* 2002; **53**: 29–40.
2. Hayashi K, Nagasaki M, Matsunaga N, et al. Initial pulmonary artery involvement in Takayasu arteritis. *Radiology* 1986; **159**: 401–3.
3. Nakabayashi K, Kurata N, Nagasawa T, et al. Pulmonary artery involvement as first manifestation in three cases of Takayasu arteritis. *Int J Cardiol* 1996; **54** (Suppl): 177–83.
4. Lie JT. Isolated pulmonary Takayasu arteritis: clinicopathological characteristics. *Mod Pathol* 1996; **9**: 469–74.
5. Hara M, Sobue R, Ohba S, et al. Diffuse pulmonary lesions in early phase of Takayasu arteritis predominantly involving pulmonary artery. *J Computer Assisted Tomogr* 1998; **22**: 801–3.
6. Elsasser S, Soler M, Bolliger C, et al. Takayasu disease with predominant pulmonary involvement. *Respiration* 2000; **67**: 213–5.
7. Hata A, Noda M, Numano F, et al. Angiographic findings of Takayasu arteritis: new classification. *Int J Cardiol* 1996; **54** (Suppl): 155–63.
8. Tanabe S, Kuriyama T. Takayasu arteritis (in Japanese). *Kokyukika* 2003; **4**: 493–500.
9. Numano F, Ohta N, Sasazuki T. HLA and clinical manifestation in Takayasu disease. *Jpn Circ J* 1982; **46**: 184–9.
10. Kitamura H, Kobayashi Y, Numano F, et al. Association of clinical manifestation with HLA-B alleles in Takayasu arteritis. *Int J Cardiol* 1998; **66** (Suppl 1): 121–6.
11. Ishikawa T. Systemic artery—pulmonary artery communication in Takayasu arteritis. *Am J Roentgenol* 1977; **128**: 389–93.
12. Matsunaga N, Hayashi K, Sakamoto I, et al. Coronary-to-pulmonary artery shunts via the bronchial artery: analysis of cineangiographic studies. *Radiology* 1993; **186**: 877–82.
13. Nakamura C, Watanabe T, Isoda N, et al. A case of RA–PA bypass operation for acquired pulmonary artery stenosis due to non-specific arteritis (in Japanese). *Shinzou* 1984; **16**: 368–73.
14. Moore JW, Reardon MJ, Cooley DA, et al. Severe Takayasu's arteritis of pulmonary arteries: report of a case with successful surgical treatment. *J Am Coll Cardiol* 1985; **5**: 369–73.
15. Chauvaud S, Mace L, Carpentier A, et al. Takayasu arteritis with bilateral pulmonary artery stenosis—successful surgical correction. *J Thorac Cardiovasc Surg* 1987; **94**: 246–50.
16. Okubo S, Kunieda T, Nakajima N, et al. Idiopathic isolated pulmonary arteritis with chronic. *Cor Pulmonale Chest* 1988; **94**: 665–6.
17. Nakajima N, Masuda S, Tanabe S, et al. A case of surgical treatment for pulmonary arteritis with stenosis of main pulmonary artery (in Japanese). Report of Research Committee on Intractable Vasculitis sponsored by Health Sciences Research 1992; 78–81.
18. Sundt TM. Clinical–pathological conferences in thoracic surgery: proximal pulmonary artery obstruction secondary to Takayasu arteritis. *J Thorac Cardiovasc Surg* 2001; **121**: 163–75.