PRIMARY ARTERITIS OF THE PULMONARY VESSELS AND THE AORTA

By Dhanwant Singh and Lenny Tan

SYNOPSIS

A 25 year old male Chinese presented with recurrent attacks of haemoptysis since the age of 14 years. Clinically he had a continuous murmur over the right upper chest. Pulmonary angiography showed multiple stenoses and occlusions of the pulmonary vessels. Aortogram showed calcification of the aorta with irregularity of it's outline and occlusion of the coeliac axis. The literature on pulmonary vessel stenosis and arteritis is briefly reviewed.

INTRODUCTION

Occlusive disease of the pulmonary arteries is not common. Some isolated case reports (William G. Smith 1958, Barbara Guller 1972) and several series have been reported (Eldridge *et al*, 1957; Oram *et al*, 1964; D'Cruz *et al*, 1964). Most of the cases described are in the paediatric age group (Oram *et al*, 1964; D'Cruz *et al*, 1964). This is understandable since the majority are of congenital aetiology. Pulmonary vessel narrowing with occlusion due to arteritis has been described in the aortic arch syndrome or aortitis (Judge, *et al*, 1962; Schrire, V. 1967), but for a patient with aortitis to present initially with symptoms and signs of pulmonary arteritis as our patient did is unusual and rare.

We describe a case of multiple, bilateral stenoses and occlusions involving both the main and peripheral pulmonary vessels with irregularity and calcification of the aorta and occlusion of the coeliac axis due to arterities in a young man.

CASE REPORT

Patient K.S.B., male Chinese, aged 25 years was first seen at Tan Tock Seng Hospital in February, 1973 with complaints of haemoptysis for 3 days prior to admission. The haemoptysis varied from slight staining to about a cup-full. There was no history of fever, chest pain, weight loss, dysnoea or trauma.

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Patient first had haemoptysis in 1961, when he was 14 years old. His second attack was in 1967 when he was diagnosed as having pulmonary tuberculosis and was given a course of anti-tuberculous drugs. In between the attacks he stayed well and led a normal active life playing badminton, swimming and even lifting weights. He had no other illnesses of any significance. There was no family history of tuberculosis or cardio-vascular disease. The mother could not recall having rubella during any of her pregnancies. Patient was the 4th child in a family of 9 children. The parents and the rest of the children were apparently well.

Clinically patient was of normal build and looked well. He had finger clubbing but no cyanosis. The significant findings were in the cardio-vascular system. All the pulses were felt and were equal. The radial pulse was collapsing in nature. B.P. was 130/50 in the right arm and 140/60 in the left arm. Heart size was normal. No thrills could be felt. There was a left parastenal heave. The second sound at the base was split with a loud pulmonary component. A continuous murmur was best heard at the right upper chest. It was widely radiating. There were some crepitations in the right lower zone posteriorly. There was a doubtful abdominal bruit heard in the epigastrium. The rest of the systems were normal.

Investigations Done

Hb. 11·7 gm.% White cell count: 6,900/c.mm. P.C.V. 48%

Platelets 235,000/c.mm.

Sedimentation rate: 14 mm./hr.; 8 mm./hr.

Urine: microscopy was normal. B. urea 31 mgm.%

VDRL: non-reactive

Direct sputum smear for acid-fast bacilli: (x6)—ve
Mantoux Test 17 mm. (+ve)
L.E. cell: (x4) —ve
Anti-nuclear antibodies: —ve
Rheumatoid Factor (x2) —ve
Direct and Indirect Coomb's test: —ve
S. Cholesterol: 185 mgm.%
S. Uric acid: 5·7 mgm.%
S. Calcium 9·6 mgm.%
S. Phosphates 3·8 mgm.%
Serum proteins : Alb:Glo. : 3·4 :3·8 gm.%
Protein electrophoresis:

alb	: 3·7 gm.%
$alpha_1$	: 0·4 gm.%
$alpha_2$	: 0·8 gm.%
beta	: 0·9 gm.%
gamma	: 1·9 gm.%

S. Immunoglobulins : Ig G 2400 mgm./100 ml. (Normal range):
Ig A 44 mgm./100 ml. (Ig G : 800-1680 mgm./100ml.)
Ig M 46 mgm./100 ml. (Ig A : 140-420 mgm./100ml.)
(Ig M : 50-190 mgm./100ml.)
Laryngeal swab for acid-fast bacilli culture (× 3) : —ve
Sputum culture grew: Klebsiella Pneumoniae

Blood gases — Normal values within brackets:
p.H : 7·42 (7·4)
Std. bicarb mM/L: 24 (24)
pCO_2 : mmHg. 37·5 (40)
PO_2 : saturation % 94 (97)

E.C.G. : Depressed ST and inverted T in leads III, VI, V2, V3.
Tall R in VI, V2 with deep S in V5, V6 indicating severe right ventricular hypertrophy.

Lung Function Studies

The results were within normal limits except for a 30% reduction in the resting diffusing capacity. This is consistent with multiple stenoses of pulmonary vessels.

Bilateral Bronchogram

Left : normal Right : Crowding of branches of basal segments of right lower lobe suggesting collapse.

X-ray Chest

(Fig. 1 a). Aneurysmal dilatations of the proximal pulmonary arteries bilaterally with remarkable lack of peripheral pulmonary vessels. Large area of consolidation involving lower lobe.

(Fig. 1 b). Note calcification of thoracic aorta and pulmonary arteries.

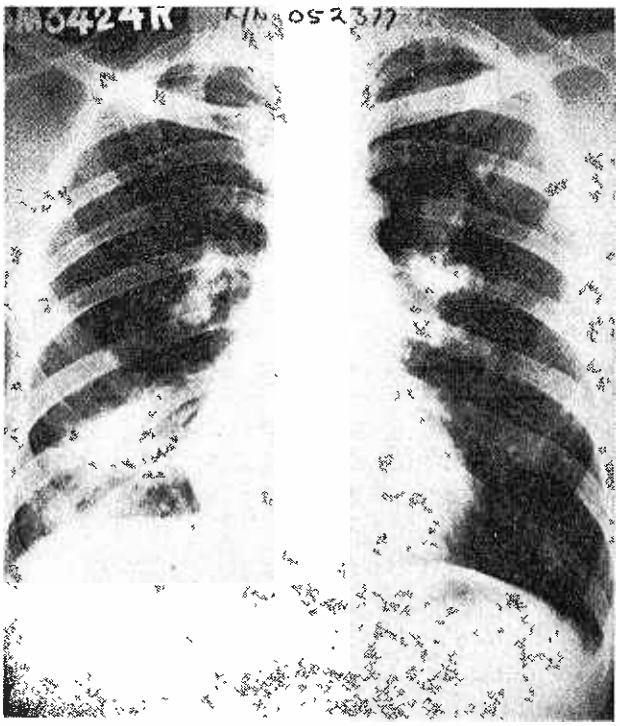

Fig. 1a.

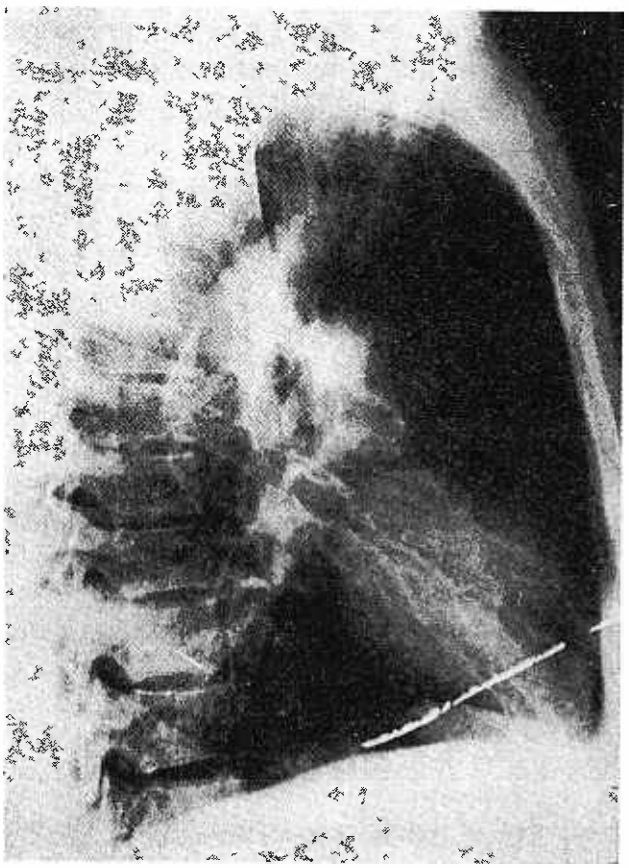

Fig. 1b.

Tomograms

(Fig. 2 a). Aneurysmal dilatation of left proximal pulmonary arteries with calcification. Note again relative absence of small vessels.

(Fig. 2 b). Aneurysmal dilatation of right proximal pulmonary arteries with relative absence of vessels in right lower zone.

Pulmonary Angiogram

(Fig. 3) Systolic pulmonary arterial pressure measured 90 mm. Hg. Complete occlusion of right lower lobe artery with aneurysmal dilatation of upper lobe vessels and loss of normal branching pattern. On the left side, remarkably few vessels are present with complete loss of normal branching pattern and severe stenosis of lower lobe artery.

Aortogram

(Fig. 4 a) Arch aortogram shows normal aortic valves and gross irregularity of the arch and thoracic aorta with areas of aneurysmal dilatations.

Abdominal Aortogram

(Fig. 4 b) shows irregularity of the aorta and a very large inferior phrenic artery and the coeliac axis filling retrogradely through collaterals via superior mesenteric artery. This was confirmed by a selective aeteriogram of the superior mesenteric artery. Renal arteries are normal.

Fig. 2b.

Progress

Patient has mild haemoptysis on and off but his general condition remains satisfactory and stable.

Fig. 2a.

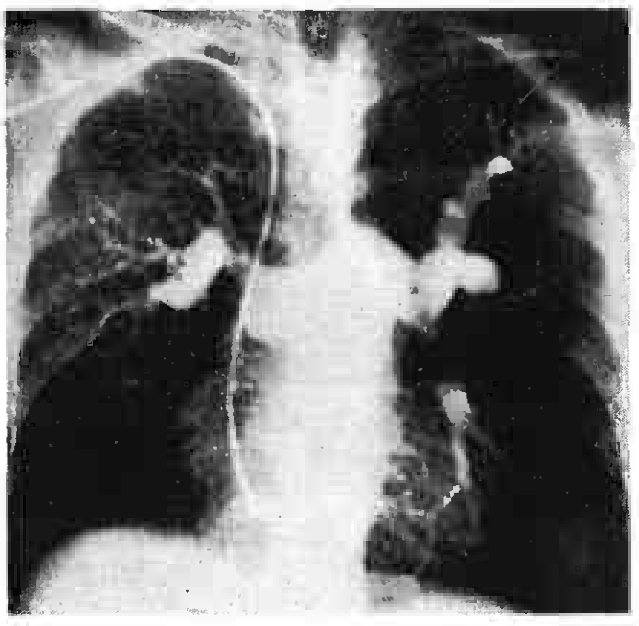

Fig. 3.

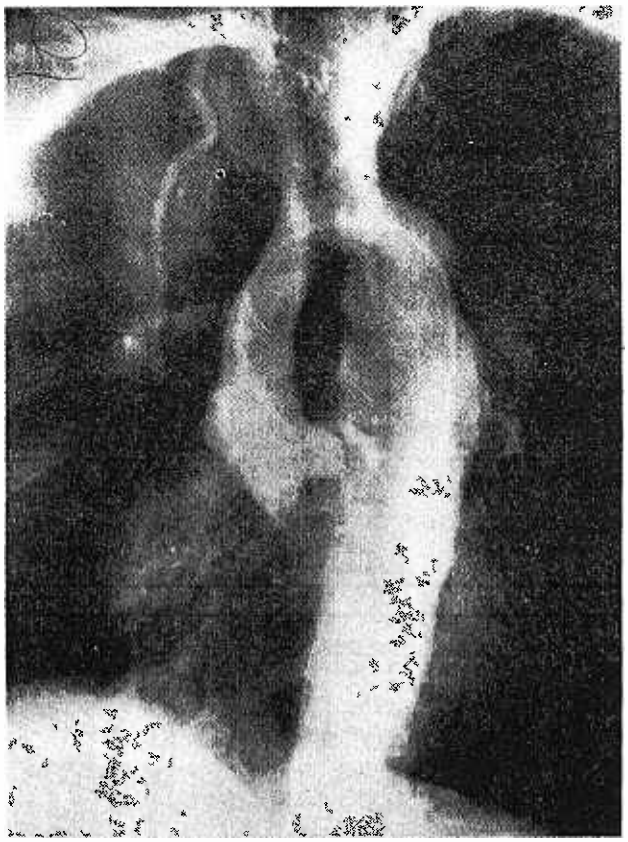

Fig. 4a.

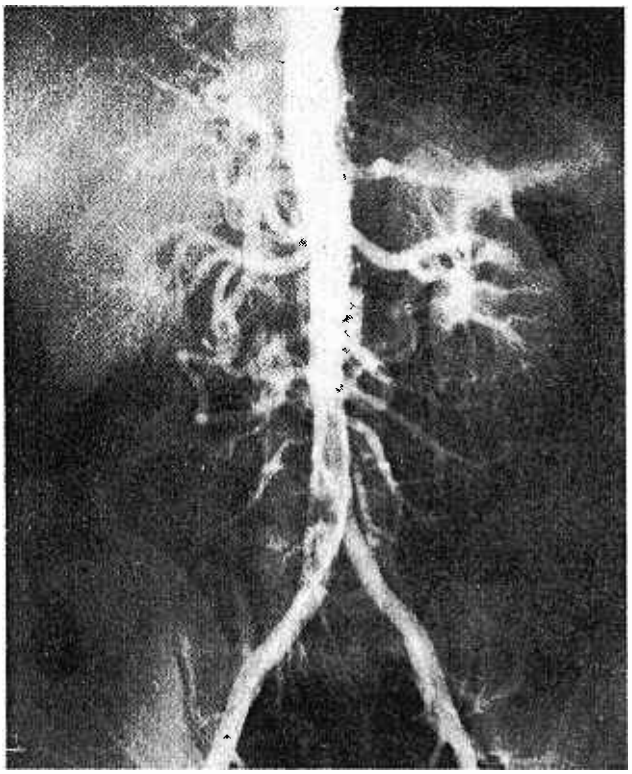

Fig. 4b.

DISCUSSION

The pathogenesis of stenosis of the pulmonary arteries is unknown in the majority of patients. Most lesions in which the aetiology is known are caused by maternal rubella infection during pregnancy (Liggins and Phillips, 1963), and in 60% of cases with bilateral stenoses the lesions are associated with some other cardiac or great vessel malformation (D'Cruz *et al*, 1964). Acquired causes have been due to pressure from tumour (Oram *et al*, 1964), recanalization of pulmonary emboli or thrombi (Saphir, 1932) and arteritis (Judge *et al*, 1962, Schrire, V. 1967).

For the reasons stated below, we believe that this is a case of arterities of the pulmonary vessels and the aorta. The age of onset, the absence of a history of maternal rubella during pregnancy and the absence of any other associated congenital cardiac or pulmonary vessel malformation would make the diagnosis of congenital pulmonary vessel stenosis unlikely. Moreover, the extent and nature of aortic involvement as demonstrated in our patient has not been documented before in other causes of occlusive disease of the pulmonary vessels especially congenital pulmonary vessel stenosis. There was no evidence to suggest the presence of any collogen disease, embolism or thromboses with recanalization. The angiographic abnormalities in the pulmonary vessels, the extent and nature of the aortic abnormalities with calcification and occlusion of the coeliac axis are consistent with the diagnosis of an arterities. Asherson, R.A. *et al* (1968) and Ghosh, M.B. *et al* (1971) found the IgG levels to be raised in cases of arteritis of the aorta. The high levels of IgG in our patient further strengthens the basis for the diagnosis of an arteritis. When the stenoses are multiple and severe as in our patient the pulmonary vascular bed is considerably reduced giving rise to pulmonary hypertension and right ventricular hypertrophy.

Though rare, dysnoea on exertion and haemoptysis may be the presenting complaints. Our patient had recurrent attacks of haemoptysis, which according to Van Epps (1957) is due to rupture of post-stenotic aneurysmal dilatations.

The auscultatory findings depend upon the severity of the stenosis. Commonly there is an ejection systolic murmur over the stenotic site (D'Cruz *et al*, 1964; Oram *et al*, 1964). A continuous or quasi-continuous murmur may be heard over the stenotic site or some distance away (Eldridge *et al*, 1958; William G.S. 1957). It is widely radiating. It is due to blood flow through a tight stenosis during systole and diastole. Our patient had a continous murmur over the right upper chest. It was widely radiating.

The 02 saturation of our patient was 94%. This excludes the presence of any significant arterio-venous communication. A fall of 30% in the diffusing capacity, which the patient had, is consistent with multiple pulmonary vessel stenoses and occlusions. The rest of the pulmonary function studies were within normal limits.

The aortitis that have been described under a variety of names such as Takayasu's arteritis, aortic arch syndrome, pulseless disease of the female, etc. by several authors (Danaraj, T.J. *et al*, 1963; Strachan, R.W. 1966, Schrire, V. 1967, Ghosh, M.B. *et al*, 1971, Ooi, B.S. *et al*, 1971) involve mainly as the name suggests the aorta and its branches, commonly the carotids, the subclavian, renal and the femorals. Pulmonary vessel involvement can occur (Judge *et al*, 1962; Schrire, V 1967) but is very rare. Judge *et al* (1962) described 4 cases of arteritis of the aorta. All had extensive involvement of the aorta and its branches. One of his patients had pulmonary involvement where the segmental branches to the right middle and lower lobes were irregular and of small calibre. Several of them were occluded. In Schrire's (1967) series of 34 patients with aortitis, all of whom had extensive involvement of the aorta and its branches, there was only one patient with pulmonary involvement. He had stenosis of the main vessel to the right lower lobe with post-stenotic dilatation. The right upper lobe vessels were large and dilated, resembling the angiographic findings in our patient though not as extensive as in our case. Most of the other series reported make no mention of pulmonary arteritis. One must bear in mind that pulmonary angiography is not routinely done in these cases: understandably too because clinically and otherwise there is little or nothing to suggest pulmonary involvement.

The management of these patients is usually unsatisfactory, and mainly symptomatic. The multiple peripheral type of stenoses are not amenable to surgery. Plastic reconstruction can be performed only if the lesions are few and localized (McGoon, D.C. *et al*, 1964). In progressive arteritis, immunosuppressive therapy may be tried (Strachan, 1966).

CONCLUSION

A case of multiple pulmonary vessel stenoses/occlusions with abnormalities of the aorta due to arteritis is described. The lesions were demonstrated by pulmonary angiogram and aortograms. The literature on pulmonary vessel stenosis and arteritis is briefly reviewed.

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REFERENCES

1. Asherson, R.A., Asherson, G.L. and Schrire, V.: 'Immunological studies in arteritis of the aorta and great vessels'. Brit. Med. J. 3: 589, 1968.
2. Barbara Guller: 'Stenosis of the pulmonary arteries: Evolution and haemodynamic effects'. Chest, 61: 70, 1972.
3. Danaraj, T.J., Wong, H.O. and Thomas, M.A.: 'Primary arteritis of the aorta causing renal artery stenosis and hypertension'. Brit. Heart J., 25: 153, 1963.
4. D'Cruz, I.A., Augustsson, M.H., Bicoff, J.P. *et al*: 'Stenotic lesions of the pulmonary arteries; Clinical and haemodynamic findings in 84 cases'. Am. J. Cardio, 13: 441, 1964.
5. Eldridge, F., Selzer, A. and Hultgren, H.: 'Stenosis of a branch of the pulmonary artery'. Circulation, 15: 865, 1957.
6. Ghosh, M.B., Chia, B.L.: 'Arteritis of the aorta and its major branches'. S'pore Med. J., 12: 217, 1971.
7. Judge, R.D., Currier, R.D., Gracie, W.A., Figley, M.M.: 'Takayasu's Arteritis and the Aortic Arch Syndrome'. Am. J. Med., 32: 379, 1962.
8. Liggens, G.G. and Phillips, L.I.: 'Rubella embryopathy'. Brit. Med. J., I: 711, 1963.
9. McGoon, D.C., Kincaid, O.W.: 'Stenosis of branches of the pulmonary artery. Surgery repair'. Med. Clin N. Am., 48: 1083, 1964.
10. Ooi, B.S., Chen, B.T.M., Yu, S.F., Toh, C.C.S. and Khoo, O.T.: 'Primary arteritis of the Aorta as a cause of Renovascular Hypertension in Singapore'. Aust. N.Z.J. Med. 2: 132, 1971.
11. Oram, S., Pattison, N. and Davis, P.: 'Post-valvular pulmonary stenosis'. Brit. Heart J., 26: 832, 1964.
12. Saphir, O.: 'Bands and ridges in the pulmonary artery. Their relation to Ayerza's disease.' Arch. Path., 14: 10, 1932.
13. Schrire, V.: 'Arteritis of the aorta and its major branches'. Aust. Ann. Med., 16: 33, 1967.
14. Strachan, R.W.: 'Pre-pulseless and pulseless Takayasu's arteritis'. Post. Grad. Med. J., 42: 464, 1966.
15. Van Epps, E.F.: 'Primary pulmonary hypertension in brothers'. Amer. J. Roentgenol., 78: 471, 1957.
16. William G. Smith: 'Pulmonary hypertension and continuous murmur due to multiple peripheral stenosis of the pulmonary arteries.' Thorax, 13: 194 1958.