THE TURKISH JOURNAL OF
PEDIATRICS
A HACETTEPE BULLETIN VOLUME 14 / NUMBER 2 / APRIL 1972

Schistosomal Pulmonary Hypertension: A Case Report

Muhsin Saraçlar, M. D.* / Behçet Tınaztepe, M. D. / Mustafa Öztürk, M. D.*** / Ali Ertuğrul, M. D.****

From the Department of Pediatrics, Faculty of Medicine, Hacettepe University.
* Associate Professor of Pediatrics and Pediatric Cardiologist.
** Professor of Pathology.
*** Resident in Pediatrics.
**** Professor of Pediatrics and Pediatric Cardiologist.

Clinically, there are two types of pulmonary schistosomiasis: Bronchopulmonary and cardiovascular.[1 2 3] In the former, the pathological lesions are in the bronchioles and alveoli. In the latter, those are primarily in the pulmonary arterioles, and secondarily in the pulmonary artery and its branches and in the right ventricle. Pulmonary hypertension develops in the advanced cases of cardiovascular forms of pulmonary schistosomiasis.

Shaw and Gharib reported that about one-third of patients with schistosomiasis showed involvement of pulmonary vessels at the autopsies.[1] However, the incidence of cor pulmonale caused by schistosomia is much less common. El Mofty[1] stated the incidence of schistosomal cor pulmonale as 2.1 % at autopsies, and 0.8 % to 1 % clinically. Among the 30 cases of schistosomal pulmonary hypertension, reported by Cavalcanti et. al., none was below the age of 10[4]. Schistosomiasis in Turkey is rarely diagnosed, since it is neither endemic, nor has it been seen in Turkish patients who have not travelled in countries where the disease is endemic. In Hacettepe Hospital, schistosomiasis has been diagnosed on a few occasions in patients who came from Arabian countries for some other problems of clinical diagnosis of this disease.

The purpose of this report is twofold. Firstly, to the best of our knowledge we could find no report of schistosomal cor pulmonale in Turkish medical literature. Secondly it is to bring to the attention of the physicians of our country that unless appropriate studies made,

schistosomal cor pulmonale may be diagnosed clinically as congenital heart disease, since it usually shows clinical features of congenital heart disease.

Case Report

History and Physical Examination: S. K. (282283), 8-year-old, Saudi Arabian male, who has had dispnea, dizziness and palpitation since one year prior to admission, was referred from his country to Hacettepe Children's Hospital for evaluation of congenital heart disease.

On physical examination he weighed 18.5 kg., and measured 118 cm. The blood pressure on the right arm was 100/60 mm/Hg, and right leg 120/80 mm/Hg. The pulse rate was 104/min, respiration was 24/min. He had no dispnea or cyanosis. The heart was hyperdynamic at the apex. The pulmonary component of the second heart sound was increased in intensity. An early diastolic decrescendo murmur, following P_2, was heard at the pulmonary area (Figure 1). The liver was 11 cm. below the right costal margin. The spleen was 12 cm. below the left costal margin.

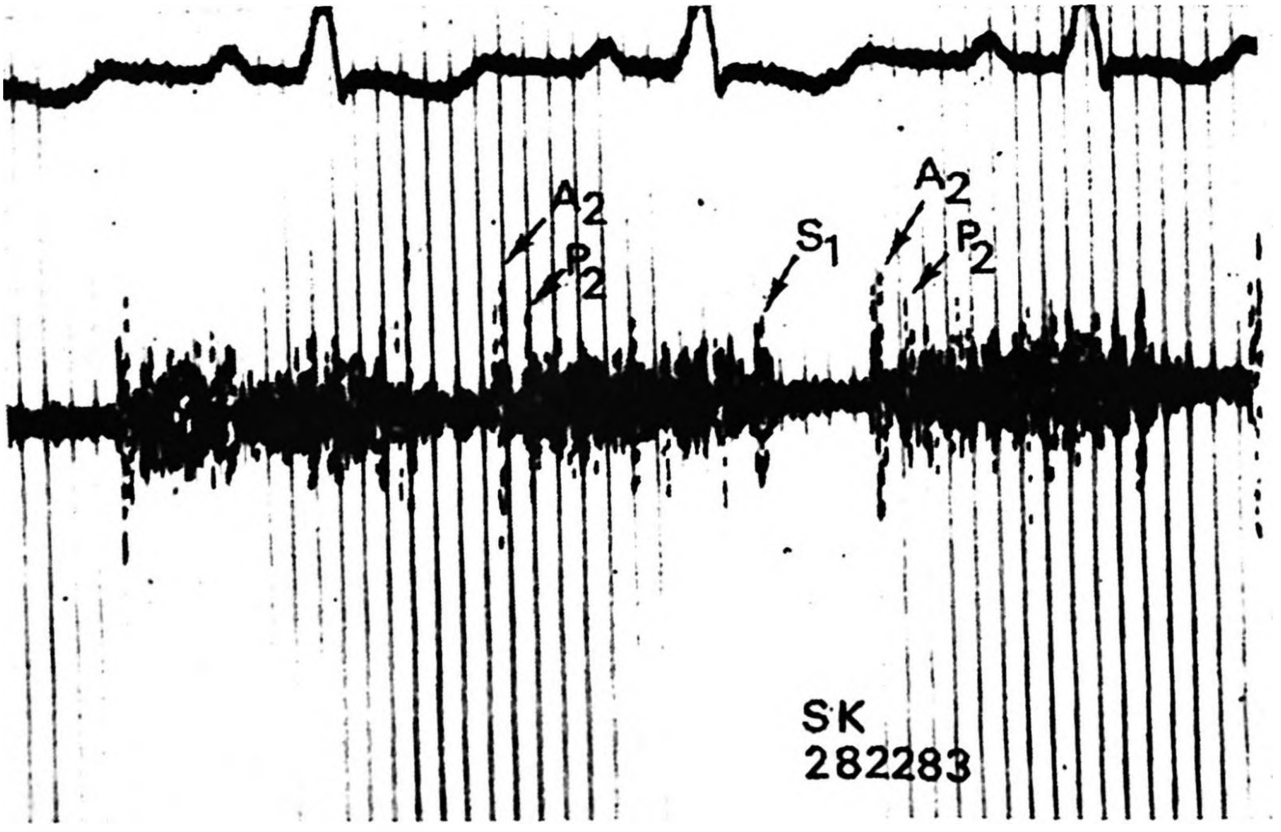


Figure 1

Phonocardiogram shows diastolic murmur which follows pulmonary component of the second heart sound.

Laboratory Findings: ECG showed + 12° right axis deviation, and right ventricular hypertrophy (Figure 2). Telecardiograms revealed right ventricular enlargement and a dilated main pulmonary artery (Figure 3). His haemoglobin level was 12 gm. per 100 ml., WBC 5.000 per cmm, total bilirubin 1 mg. per 100 ml., alkalin phosphatase 16 u, SGOT 78 u, SGPT 33 u, Tymol 6.6 u, total serum protein 8 gm per 100 ml. albumin 4.2 per 100 ml, globulin 3.8 gm per 100 ml, cholesterol 12 gm per 100 ml, CCF (++), prothrombin time 13 sec. EEG was normal.

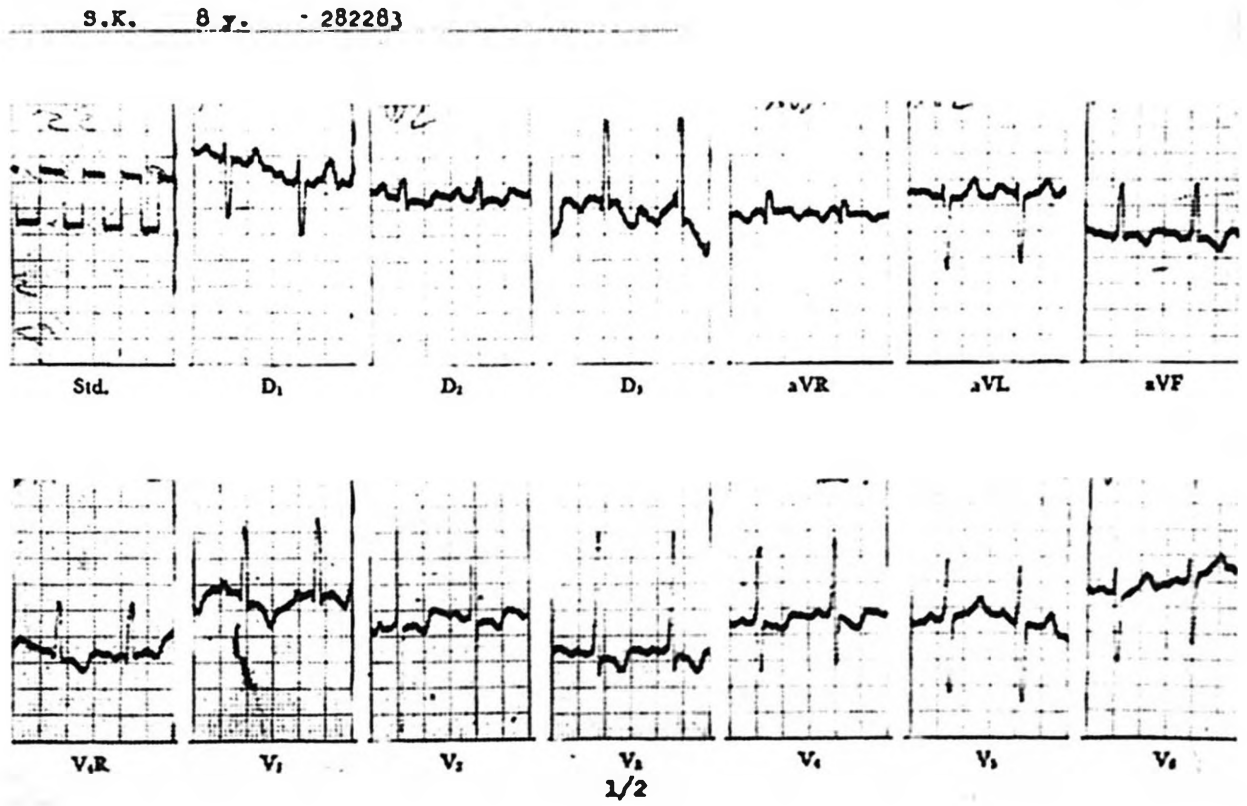


Figure 2

ECG of the patient shows + 120° right axis deviation, and right ventricular hypertrophy.

Heart catheterization was performed from the right arm. A Cournand catheter inserted in the right basilic vein entered the superior vena cava, right atrium, right ventricle, main pulmonary artery and left pulmonary artery. During the manipulation of the catheter in the right atrium, atrial tachycardia was seen several times. Blood samples for determination of oxygen saturation were obtained, and pressures were recorded from the chambers and vessels entered. After injecting radio-opaque material through a NIH catheter, placed into the right ventricle, angiocardiograms were obtained (Figure 4 a,b). Left ventricular angiography was not attempted because of the arrhythmias.

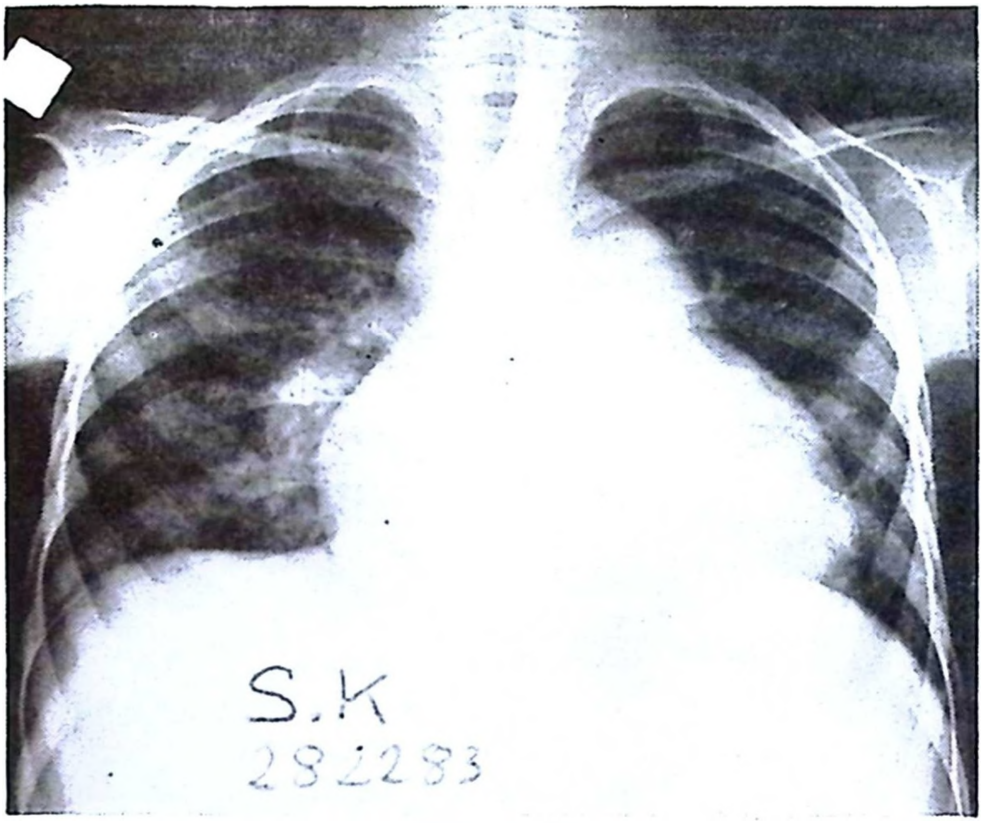


Figure 3
Posteroanterior telecardiogram of the case shows right ventricular enlargement. Main pulmonary artery is dilated.

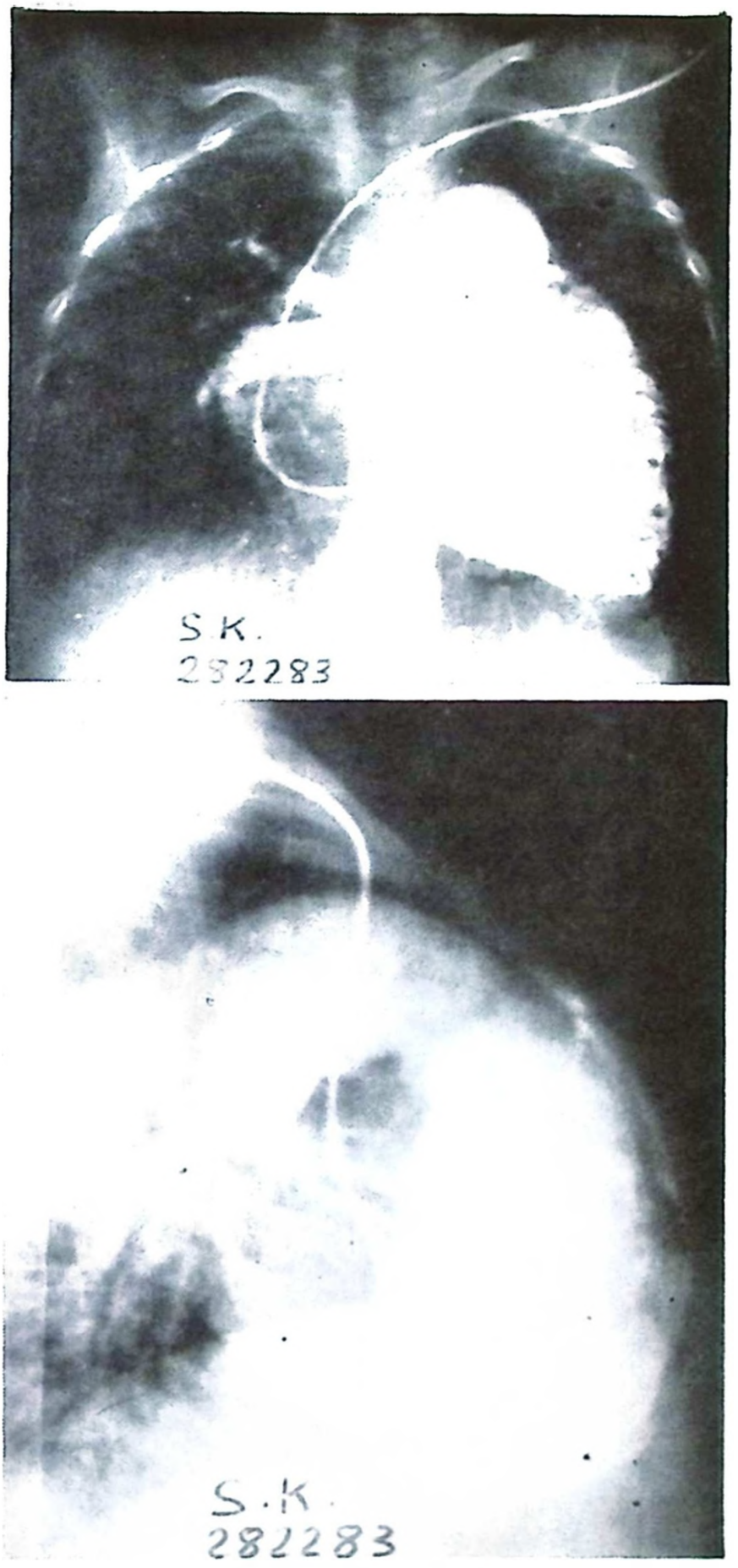
Analysis of oxygen saturations revealed no left-to-right shunt (Table I). There was pulmonary hypertension and no systolic pressure gradient at the pulmonary valve. Angiocardiograms revealed right ventricular and main pulmonary artery dilatation.

TABLE I
CATHETERIZATION DATA

	O_2 Saturations %	Pressures mm / Hg
Pulmonary artery	59	100 / 40
Right ventricle	57	100 / 5
Right atrium	58	m: 5
Superior vena cava	57	–
Brachial artery	95	120 / 70

Oxygen saturations reveal no left-to-right shunt. There is no arterial unsaturation. Pressures show pulmonary hypertension.

In spite of the lower systolic pressure at the pulmonary artery than the aorta, a left-to-right shunt could not be illustrated by oxygen studies. Because of the fact that the patient came from Saudi Arabia,



Figures 4 a and b

Angiocardiograms. Radio-opaque material, injected into the right ventricle, fills the right ventricle and pulmonary artery. Right ventricle and main pulmonary artery are dilated. a- Anteroposterior film, b- Lateral film.

where schistosomiasis is endemic, pulmonary schistosomiasis was suspected. For the definite diagnosis lung biopsy material was obtained.

Pathological Examination: Biopsy received consisted of 2.3x 0.8x0.3 cm pieces of lung tissue with focal areas of purplish-red, discoloration. It is fixed in 10 % formalin and embedded in paraffin wax. Multiple sections were stained by following methods: Haematoxylen-eosin, periodic-acid-schiff, Masson's trichrome, Haematoxylen and van Gieson'stain Gomori reticulin and Weigert's elastic tissue stains. The Main microscopic findings were of two types. One was the inflammatory cell reaction and the other was vascular changes. Inflammatory cell reactions were around the pulmonary arterioles and were mainly of granuloma type. These granuloma varied in their cellular components. Some lesions were composed of histiocytes and eosinophyls. The others contained multinucleated giant cells and still others consisted of a fibrous nodule (Figure 5). Fortunately in a focal area schistosoma was identified in one of these granuloma (Figure 6).

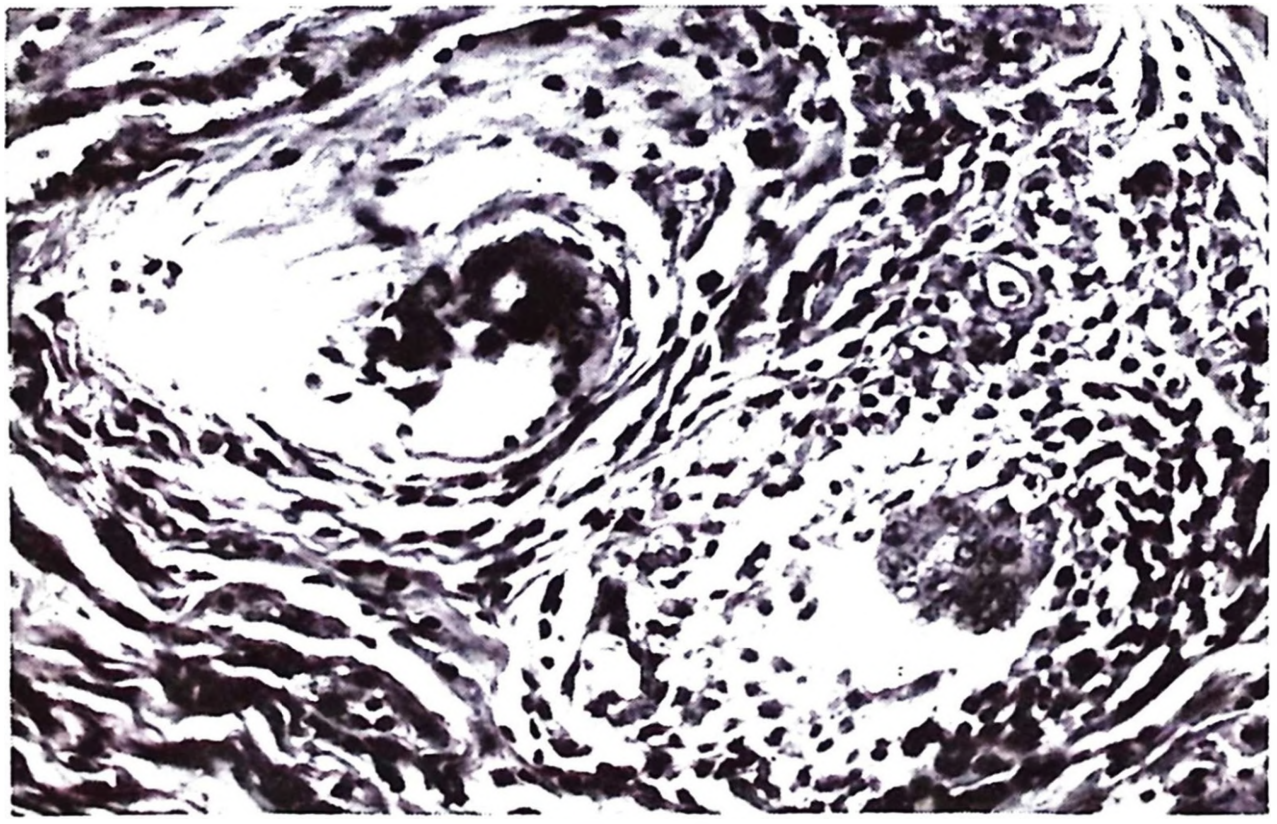


Figure 5
Granulomatous reaction with histiocytes, mononuclear cells and giant cells (Hemotoxylin-eosin x 250).

The vascular changes were of three types. Firstly there was necrotizing arteriolitis (Figure 7). Secondly there were narrowed lumina

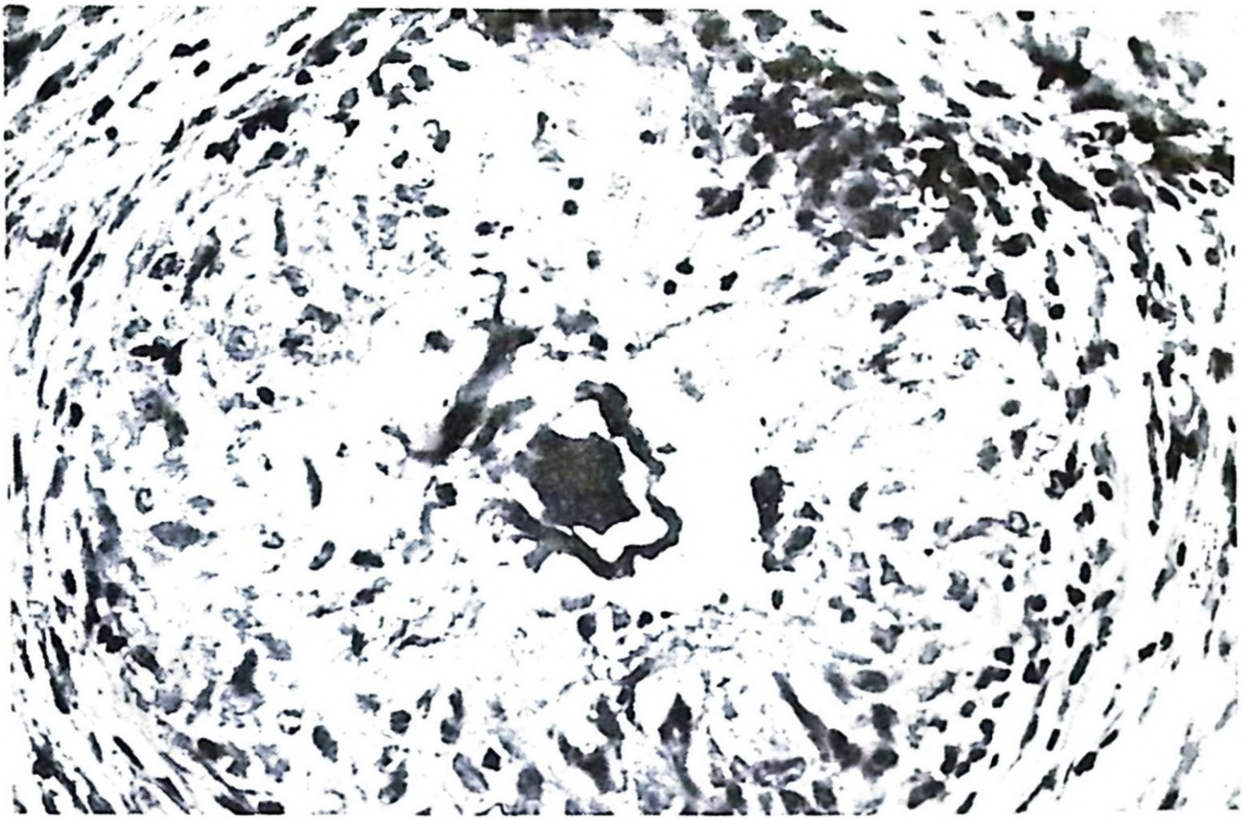


Figure 6
Egg of schistosoma can be seen readily in the center of the granuloma.

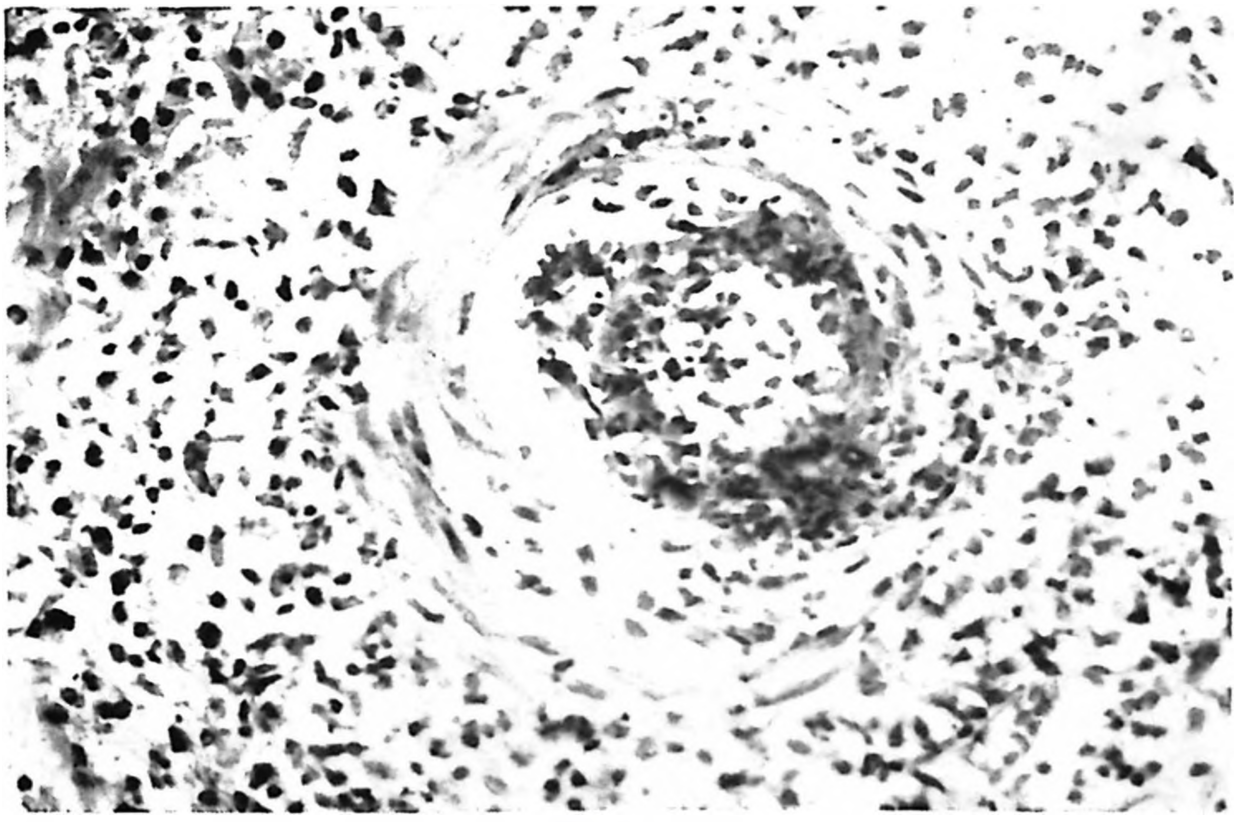


Figure 7
Acute necrotizing arteriolitis of a pulmonary arteriole (H+E x 250).

Figure 8
Advanced degree of intimal obliteration of the lumina of a pulmonary arteriole (H+E x 250).

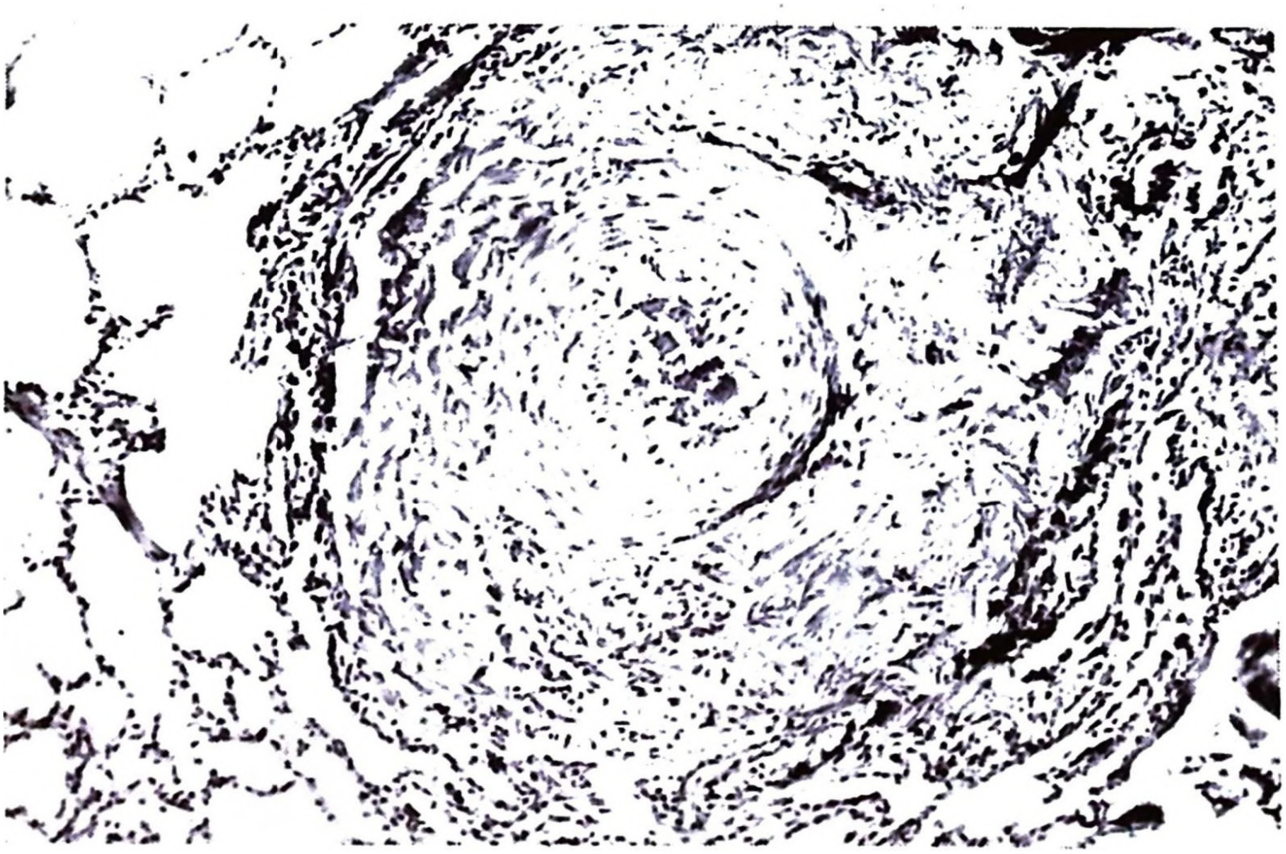


Figure 9
A pulmonary arteriole showing both intimal and medial thickening of the wall (considerable degree of periarterial fibrosis can also be seen) (H+E x 100).

of the vessels due to fibrous intimal proliferation (Figure 8). Thirdly there was conspicuous medial hypertrophy of pulmonary arterioles (Figure 9). Pathological diagnosis of pulmonary schistosomiasis and pulmonary arteriolar changes consistent with pulmonary hypertension was made.

Examination of a stool showed many ova of s. Mansoni type.

Discussion

Pathologically, Belleli[5] was first to record the presence of schistosome ova in the lungs in 1885. Symmers[6] reported the presence of schistosome worms in the blood vessels of the lungs in 1905. The ova and very rarely the adult worm reach the pulmonary circulation either from the veins of urinary bladder or from the intestines. Although the ova of s. hematobium can reach the lungs easily from the vesical plexus by way of the systemic veins, the route of those of s.Mansoni is more difficult to determine. Chaves[7] studied pulmonary vascular changes of 31 patients with pulmonary schistosomiasis due to s.Mansoni, and stated that this condition was almost always preceded by the hepatosplenic form of the disease. Most of the cases with pulmonary hypertension caused by s.Mansoni, also have vascular communications between the portal and systemic veins due to the cirrhosis of the liver, and ova can reach pulmonary circulation from the intestines by way of these communications. In those cases without cirrhosis and pulmonary hypertension s.Mansoni can be present in the venous plexuses of the bladder, as well as in those of the gut, so that the same route followed by the ova of s. hematobium can be used to reach the lungs[8].

Pathological findings in the lung biopsy, in our case, indicated chronic active schistosomal infection occurring in or around pulmonary arterioles. These parasites caused the obliterative arteriolitis, which increased the pulmonary vascular resistance, leading to pulmonary hypertension. The latter secondarily caused medial hypertrophy of the vessel walls, which is no way specific to the schistosomal infections. Since it is morphologically similar, if not identical, medial hypertrophy may occur in cases of pulmonary hypertension due to other causes.

Patients, with schistosomal pulmonary hypertension usually complain of palpitation, thoracic and precordial pain, weakness, and giddiness. Easy fatigue and exertional syncope may be present. These are related to the fixed cardiac output[9].

In a study made in 30 cases, 28 patients revealed dispnea on exertion, 23 palpitation, 5 anginal pain[4]. Syncope during exertion occurred

in 14 patients. Our case had weakness, palpitations and loss of consiousness on exertion, as most cases did.

The physical findings are similar to those seen in primary pulmonary hypertension. Cyanosis is not present. Blood pressure is usually normal. Arrhythmias are very rare. There is no clubbing of the fingers and toes. Hands and feet are usually cold. The right ventricular heave is palpated over xiphoid. The intensity of the pulmonary component of the second heart sound is increased. A pansystolic tricuspid insufficiency murmur may be heard over the left lower parasternal space. A systolic click and an ejection systolic murmur are usually heard. An early diastolic pulmonary insufficiency murmur may be present in some cases. This finding is due to the aneurysmal dilation of the pulmonary artery, secondary to pulmonary hypertension. A significant pulmonary insufficiency murmur was heard in our case.

Electrocardiographically, sinus rhythm was present in 30 cases reported by Cavalcanti et. al.[4]. Our case showed a sinus rhythm, however, atrial tachycardia developed during cardiac catheterization. Right axis deviation, and marked right ventricular hypertrophy are present in all cases. The P waves are usually normal, although occasional evidence of right atrial dilation is seen. Girgis[10] has found this finding in 6 of his 7 cases. Among 30 cases of schistosomal pulmonary hypertension reported by Cavalcanti et. al. 2 showed ECG evidence of right atrial dilation[4].

Radiologically, evidence of pulmonary arterial hypertension is seen. In most cases the heart size is increased. There is right ventricular enlargement. The main pulmonary artery is prominent, and the pulmonary vascular markings are normal in the periphery. Occasionally, miliary shadows of the schistosomal granulomata are seen.

Right heart catheterization reveals pulmonary hypertension, increased pulmonary vascular resistence and diminished pulmonary flow. The pulmonary arterial wedge pressure is normal. The right atrial pressure is always normal when congestive heart failure is absent.[4]

Selective right ventricular angiocardiography reveals a large right ventricle and a dilated pulmonary trunk. Radio-opaque material remains in the main pulmonary artery longer than normal.

The fact that the ova of s. hematobium or s.Mansoni are not present in the urine or stool does not eliminate this condition. In some cases it may be necessary to look for the eggs persistently. The ova seen in sputum are more helpfull. The best way for diagnosis is the

histological examination of the tissue material of the lung, obtained at necropsy or from biopsy.

Treatment with antimony compounds may prevent further embolisation; but it does not have any effect on the already obliterated pulmonary arteries.

Roux et. al.[11] stated that in the cases of pulmonary arteriovenous fistula, associated with schistosomal pulmonary hypertension, the resection of the fistula resulted in death.

Summary

A case of schistosomal cor pulmonale in an 8-year-old Saudi Arabian boy is described for the first time in Turkish medical literature.

The patient was referred to Hacettepe Children's Hospital for evaluation of congenital heart disease, and after appropriate studies in Hacettepe Children's Hospital the correct diagnosis was established.

Clinical and pathological features of the disease, pathogenesis of pulmonary hypertension and a brief review of the literature are presented.

REFERENCES

1. El-Mofty, A.: Clinical aspects of bilharziasis. Bilharziasis. Ed. G.E.W. Wolstenholme, and M. O'Conner J. and A. Churchill, London, England, 1962, p. 174.
2. Erfan, M.: Pulmonary schistosomiasis, Transec. Royal Sos. Trop. Med. Hyg. 42: 109, 1948.
3. Zaky H. A., El-Neneidy, A. R., Tawfick, I. M., Gemei, Y., and Khadr, A. A.: Broncho-Pulmonary shunts in schistosoma cor pulmonale, Dis. Chest. 36: 164, 1959.
4. Cavalcanti, I. L., Tompson, G., De Sonza, N., and Barbosa, F. S.: Pulmonary hypertension in schistosomiasis, Brit. Heart J. 24: 363, 1962.
5. Belleli, V.: Cited in reference 3.
6. Symmers, W. St. C.: Cited in reference 3.
7. Chaves, E.: The pathology of the arterial pulmonary vasculature in Manson's schistosomiasis, Dis. Chest. 50: 72, 1966.
8. Turner, P. P.: Schistosomal pulmonary arterial hypertension in East Africa. Brit. Heart J. 26: 821, 1964.
9. Badavi, H., Effat, H., Khalil, H., Nomeir, C. M., and Salam, M.: Cited in reference 1.
10. Girgis, B.: Pulmonary heart disease due to bilharzia, Amer. Heart J. 43: 606, 1952.
11. Roux, B. T., Gibb, B. H., and Wainwright, J.: Pulmonary arteriovenous fistula with bilharzial pulmonary hypertension, Brit. Heart J. 32: 571, 1970.