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This 45-year-old farmer has a two-year history of fatigue and exertional dyspnea. There is a past history of urinary schistosomiasis. Physical examination reveals a noncyanotic patient with raised jugular venous pressure. There is a diastolic thrill palpable along the left sternal border. The first sound at the apex and the second sound over the pulmonary area are accentuated. There is a long holodiastolic murmur and an ejection systolic murmur over the pulmonary area. Hepatosplenomegaly is present. The electrocardiogram shows right ventricular hypertrophy.

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Diagnosis:

**Cor Pulmonale Secondary to Schistosomiasis**

The roentgenogram reveals moderate cardiomegaly with marked dilatation of the pulmonary arc segment. The right main pulmonary artery is huge, with abrupt tapering of its branches. The peripheral pulmonary vascularity is greatly diminished. At fluoroscopy, abnormal intrinsic exansile pulsations of the pulmonary arteries were seen.

Cardiac catheterization revealed a mean pulmonary artery pressure of 45 mm Hg, a mean right ventricular pressure of 30 mm Hg, an arterial oxygen saturation of 91 per cent, and no evidence of a shunt. Schistosoma ova were found in the urine, but not in the sputum.

Schistosomiasis is caused by one of four species of Schistosoma which inhabit the blood stream in man: *S. mansoni*, *S. haematobium*, *S. japonicum* and *S. intercalatum*. Man, the definitive host, is infected by the fresh water larval form which penetrates the skin or buccal mucosa. The larvae reach the right atrium from the systemic venous circulation or lymphatics. They pass through the lung capillaries and become disseminated throughout the body. Only those which reach the portal venous system attain maturity. The female worm leaves the male, swims upstream against the direction of the blood flow, and reaches the venules and capillaries of either the bladder, in the case of *S. haematobium*, or the intestine, in the case of *S. mansoni*. Here the ova are deposited. The ova rupture the surrounding vessels and leave the host in urine or feces. The ova hatch in water. The snail is the intermediate host.

Pulmonary lesions are found in about one-third of the cases of schistosomiasis. These lesions are caused by ova which reach the lungs as emboli.

Three types of pulmonary lesions occur, depending on the number of ova deposited. The most common type is the parenchymatous lesion, in which the ova obstruct small arterioles, pass through their walls and lie immediately outside. Here a tissue reaction occurs, resulting ultimately in a fibrous scar.

A second type of lesion is the focal arterial lesion, in which necrosis results after the ova pass through the vascular wall. With healing, there is intimal thickening which compromises the lumen of the vessel. Some recanalization occurs, but there are many vessels that are narrowed or occluded. Medial and adventitial thickening may occur. The lesions, though irreversible, are focal and consequently do not cause right ventricular hypertrophy. The third type of pulmonary lesion, seen in about 2 per cent of all cases of schistosomiasis, is widespread arterial occlusion which ultimately produces cor pulmonale, as in the present case.

With widespread pulmonary artery disease, pulmonary hypertension develops, the main pulmonary arteries enlarge, and right ventricular hypertrophy and dilatation occur. The pulmonary arteries may even become aneurysmal, as in the present case. Abnormal intrinsic hilar pulsations may be present if pulmonary insufficiency supervenes. In long standing cases, pulmonary artery atheroma usually develops.

References


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