Pulmonary Manifestations of Schistosomiasis*

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Trematodes of the genus Schistosoma, which occur as parasites in some parts of the venous circulatory system in human beings, are very frequent in Egypt. Infection is almost the rule among Egyptian peasants, and it is estimated that 60 to 70 per cent of the inhabitants are infected. It used to be considered that the affection was limited to the lower urinary and intestinal tracts, with visceral complications limited to the abdominal organs, particularly the liver, spleen, and kidneys. It is only relatively recent that affection of the lungs has attracted attention.

Two species of the parasite occur in Egypt. Sch. mansoni inhabits the tributaries of the portal vein, while Sch. haematobium thrives in the vesical, prostatic and ureteric veins. They may, however, occasionally exchange localities, and double infection is frequent.

Infection occurs by contact with water which has been contaminated with ova passed by an infected person. The ova hatch out a "miracidium" which then infects a special kind of snail. In this it undergoes further development, and emerges as a "cercaria" which swims about in the water. If a human being comes in contact, the cercaria penetrates the skin, and is carried by the circulation through the lungs to its final habitat where it develops into the adult worm. This lays its eggs in the venules of the bladder or rectum, and during muscular contraction of these organs the ova are expelled into the lumen, from which they escape with the urine or feces.

The chief clinical manifestations are the passage of blood in the urine or stools. The most frequent complications are cirrhosis of the liver, urinary tract infection, pyonephrosis and urinary calculi.

Historical: Belleli1 reported Bilharzia ova in the lungs in 1885. Symmers2 described Schistosoma worms in the vessels of the lungs in 1905. In 1928 Sorour3 described the pathology of Bilharzial lesions in the lungs, and further pathological studies were made by Shaw and Abu Ghareeb in 1938.4

*More usually called "Bilharzia" after Bilharz who first described the parasite in 1851. In the present article the two terms will be used synonymously.

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Azmy and Effat\textsuperscript{5} reported two cases of pulmonary arteriosclerosis due to bilharzia in 1932. Mainzer\textsuperscript{6,7} reported on bilharzial asthma and on latent x-ray findings in 1939.

Other studies were published by Kenawy and Guirgis,\textsuperscript{8} by Bedford, Aidorous and Girgis,\textsuperscript{9} by A. H. Moussa,\textsuperscript{10} M. Erfan,\textsuperscript{11} and Erfan et al.\textsuperscript{12}

\textbf{Incidence:} It is difficult to estimate the true incidence of pulmonary affection in endemic regions. The figures that are available are taken from the material of general hospitals, or from post-mortem material, both of which represent cases with the heavier grades of infection.

Bilharzial cor pulmonale, which is a manifestation of advanced pulmonary affection, was recorded in 0.8 per cent of 520 cases of visceral bilharziasis admitted to Kaer-el-Ainy hospital. It was found in 2.1 per cent of 282 consecutive autopsies of cases with schistosomiasis. Bilharzial lesions in the lungs were found in 33 per cent of post-mortem cases with bilharzial infection (Shaw and Abou Ghareeb\textsuperscript{4}).

\textbf{Pathogenesis:} The bilharzia parasite can affect the lungs in various ways and at various stages of infection. (1) During the passage of the invading cercaria through the pulmonary circulation, symptoms and signs of pulmonary irritation may occur which have long been known as “verminous pneumonia.” (2) All along the duration of infestation, but particularly during the earliest stages, and especially so in members of European races, allergic reactions of the type of Loeffler’s syndrome may appear. (3) Asthma may also occur, as especially stressed by Mainzer.\textsuperscript{6} Our experience with Egyptian patients however, shows these allergic reactions to be distinctly rare, except possibly during the stage of actual invasion, when they usually escape detection.\textsuperscript{4} The most striking and most frequently observed manifestation, however, is due to embolisation into the pulmonary circulation of ova, and occasionally of the worms themselves. This embolisation becomes more frequent the more chronic the infection, and possibly occurs at one time or another in almost all bilharzial cases.

With Sch. haematobium, which inhabits systemic veins, the embolisation is direct. With Sch. mansoni, which lives in the portal system, pulmonary embolisation can only occur when hepatic cirrhosis, which is a frequent complication, has led to the development of anastomosis between the portal and systemic veins.

Mainzer\textsuperscript{7} believes that there is a type of bilharzial infection which is purely visceral, all or most of the ova being embolised to the liver or lungs instead of following their normal course through the bladder or rectum.

\textbf{Pathology:} Only the embolic lesions have rendered themselves
to pathological study. Allergic lesions have not been observed post-mortem, except as an accompaniment to the former.

Sorour described bilharzial tubercles, and in addition bronchial lesions leading to a condition of "endobronchitis obliterans." The thorough studies of Shaw and Abou Ghareeb, however, showed the main effection to be vascular and perivascular. The other pulmonary structures are free of lesions except in the case of embolisation by worms, when lesions of a pneumonis type may occur. The rare occurrence of ova in the sputum, however (Erfan), shows that direct involvement of the bronchial tree may occur.

Shaw and Abou Ghareeb describe the essential lesion as a bilharzial tubercle occurring round the dead ovum (Figure 1). In slight cases only isolated parenchymatous tubercles are present. With massive infection, healing of these multiple lesions causes an obliterative arteritis (Figure 2), often followed by canalisation of the occluding tissue. The newly formed capillaries hypertrophy, producing a structure they call "angiomatoid," which they consider characteristic of the bilharzial lesion (Figure 3). With massive and repeated infection the arterial changes are widespread, with dilatation of the main pulmonary arteries, and right ventricular hypertrophy. The dilatation of the pulmonary artery and its main branches may reach aneurysmal proportions, as in the case described by Bedford et al.

Embolised worms cause no lesions unless they die, when there occurs an acute focal necrotising pneumonia. This is later cicatrised, and the worm may become calcified (Figure 4).

Clinical Picture: These may be described under: (1) Allergic manifestations, (2) Non-allergic parenchymatous manifestations, and (3) vascular manifestations.

(1) Allergic manifestations: These have been particularly described by Mainzer. He reported four cases of asthma which responded to treatment with antimony, which is the specific
treatment for bilharzias, and in one of which bilharzia ova were found in the sputum. We note however, that in some of the cases the asthma recurred after antimony treatment, and repeated courses had to be given. My own experience of asthma in bilharzial subjects suggests that the two are associated rather than casually related, though the latter possibility cannot be excluded in some of the cases.

Lesions of the Loeffler type have been observed in bilharzial infection. They are especially likely to occur at the time of invasion, while later they have been observed during antimony treatment, presumably from the liberation of toxins from the dead worms. As these allergic lesions are asymptomatic, they are rarely seen in hospital practice.

(2) Non-allergic parenchymatous manifestations: These are hitherto ill-defined. Chronic bronchitis, emphysema, bronchiectasis and pulmonary fibrosis have been reported. Erfan et al., describe among the symptoms noted in their series, cough, thoracic pain, and slight haemoptysis; rales were sometimes present over the bases of the lungs. They noted that four cases of chronic bron-
chitis improved on antimony treatment. In addition Erfan reported a case of bronchitis and emphysema in which bilharzia ova were found in the sputum.

While there is no proof that bilharzial lesions are directly responsible for these conditions, it may be assumed that they at least play a predisposing role.

(3) Vascular manifestations: These are the most important, and the best studied, of the manifestations of pulmonary bilharziasis. They constitute the clear-cut picture of pulmonary vascular hypertension resulting eventually in cor pulmonale.

We owe the first description to Azmy and Effat, who described two cases in 1932, one of which was confirmed post-mortem. Later reports followed by Kenawy and Girgis, Moussa, Bedford et al., Erfan, Erfan et al., etc.

Bilharzial hepatosplenomegaly is usually present. The main complaint is of general weakness, and some dyspnoea on effort may be present. There may be some pallor from associated anemia, but cyanosis is conspicuous by its absence. This striking difference from Ayerza's disease, which is similar in many other respects, has been explained by the localisation of the lesion in the pulmonary arterioles, leaving the pulmonary capillaries and alveoli free. There is thus no cause for the development of cyanosis as long as the right ventricle is able to overcome the obstruction.

Physical examination reveals nothing characteristic in slight cases, but with sufficiently developed cases there is an accentuation of the second pulmonary cardiac sound, often with a palpable shock. When the condition is further developed, there is dullness over the dilated pulmonary trunk, in the third left interspace. The pulsation of the artery may be felt, and a soft diastolic murmur of pulmonary regurgitation often develops. The cardiac dullness is extended to the right owing to enlargement of the right
ventricle. In the final stages, the picture of systemic venous congestion may be found.

The disease occurs chiefly in young adults, especially between 20 and 35. It has however, been observed as early as the age of 12.

Radiological Appearances: Mainzer\(^7\) described opacities either accompanying asthma, or occurring without symptoms in bilharziasis. These may be miliary, may consist of coarser foci, or may form a honey-comb or "birch-broom" appearance.

In the more recent studies of Erfan et al.,\(^1\) three grades are described: (1) an intensification of the shadows of one or more of the second or third degree arteries, mostly the basal. The arteries appear beaded with nodules 0.5 to 1.0 mm. in diameter, which have a more or less hazy outline (Figure 5). (2) Here the lesions are more widely distributed. Clusters of mottling in relation to the arteries give the lung fields a granular background. The cardiac outline shows beginning changes in the form of a slight prominence of the pulmonary conus, with slight enlargement of the transverse diameter (Figure 6).

If the disease is arrested at this stage, we get fewer nodules with only moderately granular fields, while the cardiac changes are the same (Figure 7).

(3) In the third grade the lesions are more severe and patches of localized opacity may be present. The pulmonary conus and trunk are ballooned and may reach aneurysmal size. The heart shows the typical configuration of cor pulmonale with enlargement of the right ventricle and right auricle (Figure 8).
If healing occurs at this stage, the lung fields become more or less clear, but the arterial and cardiac changes persist or increase (Figure 9).

Laboratory Findings: Bilharzia ova are present in the urine or stools unless the bilharzial infection has died out or in the rare visceral form described by Mainzer. The sputum may contain eosinophils, or rarely actual ova. Eosinophilia in the blood is present with active infection and may reach as high as 70 per cent. The cutaneous reaction of Fairley has not been used routinely in the Egyptian material, but in the cases described by Mainzer it was regularly positive.

Diagnosis: This rests on the presence of the above-mentioned clinical and radiological pictures, together with evidence of bilharzial infection. In endemic areas the presence of a cirrhosed liver may be taken as almost sure evidence of bilharziasis, even in the absence of ova from the urine and stools.

The pulmonary shadows may simulate tuberculosis, silicosis, sarcoidosis, atypical pneumonia or parieteritis nodosa.

In advanced cases with marked affection of the heart and pulmonary artery, the condition has to be distinguished mainly from congenital atrial septal defect. The differentiation rests mainly on the presence of evidence of present or past bilharzial infection, and the almost constant association with cirrhosis of the liver.

Course and Prognosis: Pulmonary bilharziasis may be arrested at any stage, and the patient may live his normal span of life. More often he succumbs to the associated hepatic cirrhosis. In advanced cases, failure of the right side of the heart may terminate the picture. This failure, as pointed out by Erfan, is late in appearance, and its cause is mechanical, as there is no direct myocardial damage. Sudden death from pulmonary infarction is not uncommon.

Treatment: In cases which still present evidence of active bilharzial infection, antimony (tartar emetic, Fouadin) should be given. The course may have to be repeated. In advanced cases with enlargement of the right ventricle, however, it must be given with caution. Focal reactions in the lung are very frequent during antimony treatment and may be observed radiologically as a transient increase in the already existing shadows. Heart failure should be treated along ordinary lines.

SUMMARY

An account of pulmonary schistosomiasis has been given. The lungs are frequently affected in endemic areas. Asthma, pneumonic episodes or lesions of the Loeffler type may occur. But the most frequent condition is a progressive obstruction of the pulmonary
ar arterioles due to embolisation by ova, eventually leading to pulmonary hypertension, dilatation of the main pulmonary arteries and enlargement of the right side of the heart.

The condition should be constantly borne in mind in regions in which bilharzia is endemic.

RESUMEN

Se describe la esquistosomiasis pulmonar. Los pulmones son frecuentemente afectados en las áreas endémicas. Asma o episodios neumónicos del tipo de los infiltrados de Loeffler pueden ocurrir. Pero lo más frecuente es la obstrucción progresiva de las arteriolas pulmonares debida a la embolización por huevos, lo que puede llevar a la hipertensión pulmonar, dilatación de las arterias principales pulmonares y crecimiento del corazón del lado derecho.

Estas condiciones deben tenerse presentes en las regiones donde la bilharzia es endémica.

RESUME

Dans les pays où la schistosomiasis est endémique, les poumons sont souvent atteints. Ainsi peuvent apparaître des crises asthmatiformes, des épisodes pneumoniques, ou des lésions du type de la maladie de Loffler. Mais ce qu'on constate le plus fréquemment, c'est l'obstruction progressive de l'artère pulmonaire, qui peut mener à une hypertension de la circulation pulmonaire, à une dilatation des artères pulmonaires, et à une augmentation des cavités droites du cœur.

Il faut toujours avoir présente à l'esprit la possibilité d'une telle affection dans les régions où sévit la bilharziose.

REFERENCES