Aneurysm of the Pulmonary Artery
Due to Schistosomiasis*

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Schistosomiasis is an ancient and endemic disease in Egypt. It exists under two varieties: a haematobium and a mansoni. The worms of the former produce ova with a terminal spine which they deposit in the small veins of the urinary system, while in the latter the worms favor the intestinal tract and their ova have a lateral spine. S. mansoni is prevalent in the lower Nile Delta and is commonly attended with visceral complications in the liver, spleen and lungs, while S. haematobium is more frequent in the upper Nile Valley and its noted complications are mainly urinary.

The earliest clinical recognition of schistosomiasis of the cardio-pulmonary system was first reported by Azmy, Effat and Sorour in 1932, which disease they labelled Bilharzial Ayerza; later an extensive pathological study from the same school was conducted by Shaw and Gharieb in 1938.

Though Schistosoma worms may reach the pulmonary artery or its branches and upon their death (accidentally or from antimony treatment) may follow a verminous dull congested lobular pneumonic allergic infiltrations, yet the usual picture is not due to the migrated worms but is due to the deposition of living ova in an around the terminal pulmonary arteriole. Schistosoma mansoni ova are particularly injurious, in that respect, especially to the vessels, though the incidence of schistosoma haematobium deposited ova may be higher in the lung.

There are two main forms of pulmonary schistosomiasis which are by no means individually separate; a cardiovascular and a parenchymatous, the one often merging into the other. The ova reach the lesser circulation either from the vesical veins in the schistosoma haematobium or across a porto-caval anastomosis in S. mansoni, the residence of which is confined to the portal tract. Only living ova are capable of exciting a histiocytic and a fibroblastic reaction in and around the arteriole, which they are capable of penetrating. They lie in its immediate vicinity causing an end-arteritis obliterans and the so-called bilharzial tubercle which is 0.5 to 1 mm. in diameter, greyish in color, firm in consistency

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194
and well fixed to its bed (Sorour). Microscopically, it is at first composed of histiocytes and esinophiles and later lymphocytes and giant cells. Gradually the tubercle is fibrosed and replaced by a nodular scar. Parenchymatous lesions are less frequent than the periarterial.

The deposition of the ova and the consequent train of events are patchy at first but the process gets more generalized as more and more ova reach the lung from the prolific schistosoma worms. The deposition favors the perihilar areas and peripherally the lower lobe than the upper, while the apices are noticeably free.

The bilharzial tubercle may end by calcification and this cause must be added to other known causes of calcified nodules in the lung. Histologically, the impacted ova in and around the arteriole show a necrotising arteriolitis with destruction of the media, that heals with oblitative endarteritis. The distinctive histological feature of the schistosomal pulmonary arteritis is the formation of angelomatoids. The occluded vessel becomes canalized by new capillaries some of which dilate forming blood spaces lined by endothelium and in the absence of an intact medial coat this vascularized tissue expands beyond the normal boundaries of the vessel and may reach cavernous dimensions (Shaw and Ghareeb). There is also a process of oblitative endarteritis in the vasa vasorum of the big vessels (Sorour). These pathological processes lead to weakening of the arterial coat of the pulmonary vessels which begin to enlarge even in the absence of a constant rise of pressure either in the right ventricle or in the pulmonary artery.

**FIGURE 1**
*Figure 1:* Impacted ova in a pulmonary vessel with obliteration of the lumen and marked histocytic reaction and fibrosis.

**FIGURE 2**
*Figure 2:* Obliteration of the lumen and angelomatoid formation.
Of 10 cases studied by Nasr Soliman in Cairo by means of cardiac catheterization there were only two that showed a distinct rise of pressure in the pulmonary artery, 74 and 47 mms. Hg. with the circulating pulmonary blood volume 1.6 and 0.8 litres respectively. Both cases showed aneurysmal dilatation of the main trunk and its branches. It seems probable that gross dilatation of the pulmonary artery is a late event partly due to local weakening of the vessel wall and partly to a rise of pressure as the process becomes increasingly generalized from repeated implantation of schistosoma ova. In ordinary emphysema the pressure in the pulmonary circuit may rise comparatively much higher yet the aneurysmal dilatation is lacking in spite of the secondary atheroma.

FIGURE 3
Ovum in vasovasoum.

FIGURE 4
Bronchitis obliterans.

FIGURE 5a: Postero-anterior.
in the pulmonary artery. In atrial septal defect, in patent ductus arteriosus and in Roger's disease the gross enlargement encountered is due, besides the rise in pressure, to the increase of the circulatory blood volume in the pulmonary circuit.

**FIGURE 5b:** Right ventricular enlargement and strain.

**FIGURE 5c**
*Figure 5c: Two seconds after Diodrast.*

**FIGURE 5d**
*Figure 5d: Four seconds after Diodrast.*
Figures 6a and 6b: Four seconds after Diodrast (22/5/50). Large pulmonary artery and its branches—a diastolic pulmonary murmur and hilar dance. Diodrast.—Figure 6c: Digitaalis and mercurials for 17 days (10/6/50).
Though Schistosoma ova have been reported in the myocardium, yet this rare event is in no way responsible for the schistosoma cor pulmonale. The hypertrophy and enlargement of the right ventricle are due to an increased strain on that chamber probably of various factors: (a) arteriovascular, (b) associated emphysema from schistosomal bronchiolitis obliterans (Sorour), (c) and the need to fill a widely dilated pulmonary artery and its branches.

We have had under our care in the past five years six instances of schistosomal aneurysm of the pulmonary artery. All of them were from the lower delta and had associated hepatomegaly and splenomegaly. The youngest was 14 and the oldest 55 years. Dys-

**FIGURE 6d:** Right ventricular enlargement and strain. Patient died suddenly after apparent improvement.
pnoea on slight exertion and cyanosis were present in all. Cyanosis is not an early feature in ordinary cases of vascular pulmonary schistosomiasis, but it is present when right cardiac strain develops. The presence of pulmonary aneurysm is in itself an expression of an end result in a long series of events, in other words the last few strokes in a picture that took years to draw.

The neck veins were congested in all but the pulsations were vigorous, seen and felt in two, so much as to be mistaken for arterial at the first glance. The waves were throbbing synchronously with the ventricles. This means that in addition to an incompetent tricuspid valve there was an uninterrupted column of blood with a sufficiently tense venomotor tone as to be able to conduct a haemodynamic wave from the right ventricle across a distended right auricle without appreciable loss.

**FIGURE 7**

*Figure 7: Four seconds after Diodrast.*

**FIGURE 9**

*Figure 9: More parenchymatous lesion than vascular.*

**FIGURE 8a**

*Two seconds after Diodrast.*

**FIGURE 8b**

*Five seconds after Diodrast.*

Diodrast and pericardial effusion associated; a boy of 14 years with pyelonephritis secondary to Schistosomiasis and B. coli.
Generalized oedema and ascites were present in all, probably due to a combination of: (a) hypoproteinaemia; from a poor protein diet, associated parasites as ankylostoma and ascaris, deficient albumin formation from a cirrhotic liver and excessive loss from bilharzial dysentery, haematuria or intestinal bleeding; (b) cardiac failure with venous congestion; (c) in one case there was ascending pyelonephritis secondary to bilharzial ureters.

Clubbing of the fingers was present in one case.

The chest showed the usual configuration seen in cases of schistosomal hepatosplenomegaly: conical, emphysematous, short in the vertical axis and with a wide everted subcostal angle. The heart was pushed upwards and placed horizontally, the apex frequently in the fourth interspace. The big vessels were likewise higher than normal. There was bulging and systolic pulsation in the second interspace, which in one case reached the midclavicular line laterally. The right border was one half to one inch outside the right sternal border. Auscultation showed besides haemodynamic systolic murmurs over the orifices, an accentuated second sound over the pulmonary in all, reduplication in one and diastolic murmur in two.

The electrocardiogram showed right ventricular preponderance and strain.

Radiography and angiocardiography disclosed a widened right ventricular cavity and conus, a diffuse aneurysmal enlargement and ballooning of the pulmonary artery and its branches; the hilum standing out rigidly away from the mediastinum. Hilar dance was present along with a diastolic pulmonary murmur. There was no enlargement in the left auricle against the visualized esophagus but the pulmonary artery marked an indentation. A hypoplastic aorta suggests an atrial septal defect. The left ventricle showed moderate or slight enlargement. The cardiac surface area as well as the throbbing of the neck veins became appreciably smaller in two cases after mercurials and digitals but the pulmonary vessels remained almost unchanged in size. The rest of the lung showed tortuous beaded vessels, pseudohoneycombing and scattered hard nodular mottling more marked in the middle and lower zones; the whole engrafted on top of areas of increased translucency due to localized or diffuse emphysema.

**SUMMARY**

Schistosomiasis exists in Egypt as: (a) Haematobium, mainly urinary and (b) Mansoni, mainly intestinal.

The lungs may be affected as follows:

1) Migration and death of the worms in the lungs leading to a verminous pneumonia (uncommon).
2) The deposition of carried living ova in terminal arteriole, Mansoni causing the most mischief. This leads to parenchymatous, bronchiolar and vascular changes. Endarteritis obliterans with angiomatoid formation and the development of the “bilharzial tubercle” are characteristic.

3) Aneurysmal dilatation of the pulmonary artery and its branches is a late manifestation probably due to a local weakening of the arterial coat from schistosoma ova deposit as well as associated malnutrition aided by increased pulmonary circulating blood volume and a gradual rise of blood pressure in the pulmonary circuit.

4) Dyspnoea is a constant feature in cardiopulmonary schistosomiasis, cyanosis signifies right ventricular strain and/or bronchiolar narrowing. The congested veins may conduct right ventricular pulsations. A pulmonary diastolic murmur may be heard.

5) The radiographic appearance shows enlargement of the right ventricle and the pulmonary artery, and its branches, beading and tortuous appearance of the small vessels. Pseudohoneycoming of the lung parenchyma and exaggerated bronchial shadows make the background.

6) The E.C.G. shows right ventricular hypertrophy with strain when failure sets in.

RESUMEN

La esquistosomiasis existe en Egipto como: (a) Hematobia, principalmente urinaria y (b) Mansoni, principalmente intestinal.

Los pulmons pueden afectarse como sigue:

1) Emigración y muerte de los gusanos en los pulmones conduciendo a una neumonía tóxica (poco común).

2) La detención de huevos vivos arrastrados por la corriente sanguínea hasta una arteriola pulmonar, este tipo Mansoni causando el daño mayor. Esto conduce a cambios parenquimatosos, bronquiolares y vasculares. Son características la endarteritis obliterante con formación angiomatoid el desarrollo de “tubérculos bilharzianos.”

3) Dilatación aneurysmal de la arteria pulmonar y de sus ramas como una manifestación tardía, probablemente debida a un debilitamiento local de la túnica arterial por los depósitos de huevos de esquistosoma asociada a desnutrición y a un aumento del volumen de sangre circulante y aumento gradual de la presión arterial en el circuito pulmonar.

4) La disnea es un síntoma constante en esquistosomiasis cardiopulmonar; la clanosis significa esfuerzo del centriculo derecho y/o estrechamiento bronquiolar. Las venas congestionadas pueden
conducir pulsaciones del ventrículo derecho. Un murmullo diastólico pulmonar puede auscultarse.

5) La aparicién radiológica muestra aumento del ventrículo derecho y de la arteria pulmonar y de sus ramas con aparicién de rosario o tortuosa de los vasos pequeños. Un aspecto falso de panel del parénquima pulmonar y sombras bronquiales exageradas pueden hallarse en el fondo.

6) El electrocardiograma muestra hipertrofia ventricular con esfuerzo cuando se establece la insuficiencia.

RESUME

La schistosomiase existe en Egipto sous forme: (a) de “Haematobium,” le plus souvent à localisations urinaires, (b) sous forme de Mansoni, le plus souvent à localisations intestinales.

Les poumons peuvent être atteints de la façon suivante:

1) Migration et mort des vers dans les poumons menant à la constitution d’une pneumonie vermíneuse (cas peu commun).


3) La dilatation anévrismale de l’artère pulmonaire et de ses branches réalise une manifestation tardive. Il est très probable que cet accident est dû à un affaissement local de la paroi artérielle à la suite du dépôt d’oeufs de schistosomes. Il s’y ajoute les troubles de la nutrition, favorisés par l’augmentation du volume du sang circulant, et l’élévation progressive de la pression sanguine dans la circulation pulmonaire.

4) La dyspnée est un facteur constant dans la schistosomiase cardíopulmonaire. Lorsqu’il y a cyanose, cela doit faire penser à l’existence d’une atteinte du ventricule droit ou d’un rétrécissement bronchiolaire. Les veines congestionnées peuvent être secouées par les pulsations du ventricule droit. On peut entendre un souffle diastolique à l’orifice pulmonaire.

5) La radiographie montre l’augmentation de volume du ventricule droit et de l’artère pulmonaire, ainsi que de ses branches. Les petits vaisseaux prennent un aspect moniliforme et sinueux. L’ensemble est caractérisé par un aspect du parenchyme pulmonaire simulant de multiples petites cavités et par une exagération des ombres bronchiques.

6) L’électrocardiogramme montre une hypertrophie du ventricule droit.
REFERENCES