CHRONIC PULMONARY GRANULOMATOUS SCHISTOSOMIASIS WITH ATYPICAL PRESENTATION

Hala A. Ibrahim*, Azza Attallah**, Abdulaziz A. Al Mustafa***, M.H. Maghout***, Ayman F. Youssef***, Tarig S. A. Al-Khuwaitir* MRCP(UK)
and Arab BIM, FRCP Edin
Departments of *Medicine, **Histopathology, ***Cardio-Thoracic surgery
King Saud Medical City, Ministry of Health, Riyadh KSA

ABSTRACT:

Patients with chronic *Schistosomiasis* may feature a range of pulmonary symptoms and radiological findings. The pulmonary lesions of schistosomiasis manifest frequently as granulomas or tubers diffusely scattered around the bronchioles and alveoli and rarely occur as mass lesions. The finding of bilateral mass lesions in the chest leads one to entertain differential diagnoses of carcinoma or pulmonary tuberculosis. A 52-years-old patient who lived in a schistosomiasis endemic area with chronic productive chough and a pulmonary mass lesion with multiple pulmonary nodules suggestive of malignancy underwent exploratory thoracotomy. The lung-biopsy specimen showed several granulomas with *Schistosoma* eggs, and hyperplasic connective tissue with no sign of malignancy. The patient presented no clinical signs of pulmonary or portal hypertension; nor was either identified through diagnostic tests.

KEYWORDS:

Chronic schistosomiasis Parasitic infection.

INTRODUCTION:

Schistosomiasis (SSM) is a helminthic infection, endemic to tropical and subtropical regions. The infection is common in parts of Africa, South America, the Middle East, the Caribbean, and Asia and is a leading cause of morbidity and mortality in these areas.[1]

In individuals within these regions, chronic complications most often develop from progressive and recurrent infection.[2]

Pulmonary involvement in SSM is uncommon, but has been described in populations live in these endemic areas.[3]

In S. haematobium infection, this results from sustained and heavy embolisation of ova from the vesical plexus to the lungs 4. In S. mansoni and S. japonicum, ova from the adult worms living in the mesenteric veins are deposited in intrahepatic portal venules and cause granulomata, portal occlusion and ultimately, over many years, fibrosis leading to portal hypertension. Portosystemic shunting then allows ova to pass directly to the lungs and 26% of patients with advanced hepatosplenic disease go on to develop interstitial pulmonary schistosomiasis.[5]

Given the above, we report a case of man with the finding of granulomatous pulmonary nodules (eggs of Schistosoma) after many years coming from an endemic region without any hepato-intestinal manifest-tation.

Case Report

A Sudanese male patient, 52-years-old shepherd, non- smoker was seen in the out-patient department complaining of chronic productive cough with no loss of weight or appetite with occasional shortness of breath. At no time during his illness had he any fever, nor was there foul sputum. No past history of pulmonary tuberculosis or other chronic illness. On clinical examination: the patient was conscious alert and oriented, his general condition being fair. He was not dyspnoeic, hemodynamically stable. There was no pyrexia., no clubbing of the fingers. Examination of the abdomen was normal. On chest examination an impaired percussion note was detected over the right infra-scapular area, but the breath sounds were normal with diminished intensity and there were bilateral scattered mid-crackles. Other systems were normal. Laboratory evaluation showing blood picture of hemoglobin of 11.6 g/d; red blood cells 4.47 million/l; white blood cells 8,600 (polymorphs 69.9%, lymphocytes 17.5%, eosinophil count 3.2%, basophils 3.2%, monocytes 8.75%) Erythrocyte sedimentation rate 55mm/hour and arterial blood gases on room air were within normal range.

Sputum microscopy for acid fast bacilli 3 samples and direct DNA technique were negative. Purified Protein Derivative test was less than 5mm.Sputum Gram stain showed few pus cells, no organism seen and culture was negative. Urine analysis at this time showed no red blood cells, and no parasites were identified on urine or stool microscopy, urine cytology showed no evidence of malignancy. Biochemical indices of hepatic and renal function were normal.

Chest radiograph and high resolution computed tomography of the chest (HRCT) revealed bilateral old fibrotic changes, with non-homogeneous opacities occupying the upper lobes of both lung and showing internal calcification. In addition to multiple calcified mediastinal and hilar lymph nodes, wide spread, ill defined nodules in a predominantly peri-bronchovascular and sub-pleural distribution and a minimal right pleural effusion, fig. (1). CT Abdomen with contrast was normal. By abdominal ultrasound liver was average in size with mild coarse echotexture and no focal lesions or intrahepatic biliary radicular dilatation. Common bile duct and portal vein were not dilated.

Clinical, laboratory, and radiological findings did not reveal the nature of the lesion, also fiber optic bronchoscopy revealed no changes in the bronchial tree. Search for the tuberculosis bacillus and fungi and other bacteria in bronchoalveolar lavage were negative. Cytology was negative for malignant cells. Eventually right mini thoracotomy was done and revealed multiple nodules and masses involving most of the right lung, with no involvement of parietal pleura, and a wedge biopsy of lung was taken from the right upper lobe.

Macroscopic analysis showed two brownish soft tissues lumps together measuring 2×1×1cm. Cut surface shows multiple whitish areas. Microscopically it showed large granulomata with central suppuration and fibrin deposition, surrounded by fibroblasts, lymphocytes and histocytes. Few eosinophils, plasma cells and foreign body giant cell reaction were also present. Initially we tried to role out T.B and pulmonary mycosis. AFB and Grocott's stain revealed negative result. Deeper sections showed collection of multiple schistosomal ovi. Some of which were

viable and others were calcified. The rest of the lung tissue showed atelectatic and emphysematous changes. Several sections revealed no significant arterial lesion caused by Schistosoma infection nor other non-schistosomal lesions. Again there was no evidence of malignancy (figs. 2,3)

A diagnosis of schistosomiasis should prompt initiation of treatment, even if the patient is asymptomatic, since adult worms can live for years .The patient was treated with praziquantel (40 mg/kg, single oral dose) without complications. One month after treatment, a subjective improvement occurred as regard to his general condition, there was a decrease in the amount of sputum which became whitish, a chest CT scan showed a no changes on previous findings.

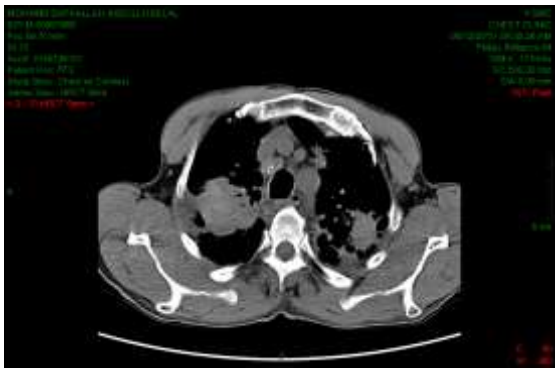

Figure 1:Bilateral mass lesions and fibrotic changes with calcified paratracheal lymph node (mediastinal window)

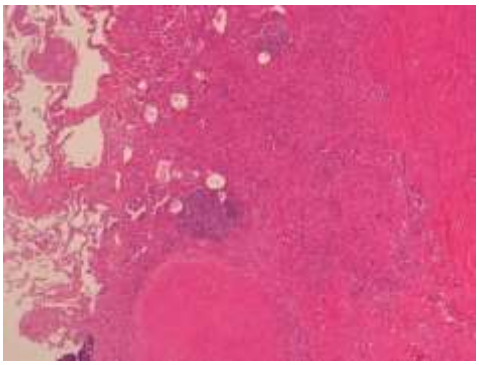

Fig. 2: Lung tissue with large granulomata with central suppuration surrounded by fibrosis and chronic inflammation (H and E stain 20× objective)

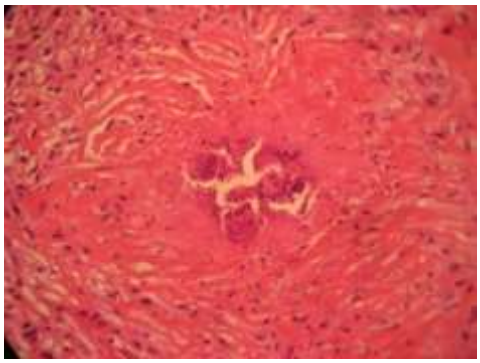

Fig. 3: Schistosomal ova . The spines are not clear for sub-classification (H and E stain 40× objective).

DISCUSSION:

Chronic schistosomiasis results from granuloma formation in response to the schistosoma eggs; however, a complex relationship exists between the severity of clinical disease, the intensity of infection, and the infecting species.[6]

Two types of chronic pulmonary schistosomiasis are now recognizable pathologically:

1. Cardiovascular type which is characterized by a necrotizing arteriolitis with endarteritis obliterans and perivascular tubercles. If these lesions are widespread, dilatation and thickening of arteries in the lungs and of the pulmonary artery and its primary branches may occur, the right ventricle hypertrophies, dilates and finally fails.[7]

2. Parenchymatous or broncho-pulmonary type. This is the one which occurred in our patient. Its pathologic incidence is more common than the former type and it is less serious clinically as in the present case. While the cardiovascular manifestations of schistosomiasis are well established, the parenchymatous type's are still the subject of ongoing debate due to the fact that there is no definite correlation between the pathologic findings and the clinical features of broncho-pulmonary schistosomiasis.[7]

Patients with chronic schistosomiasis may have pulmonary involvement featuring a variable range of symptoms and radiological findings.[8] The most common clinical manifestation of chronic lung disease is dyspnea and reduced exercise tolerance, but severe hypoxemia, chest pain and digital clubbing may also be found. Cor pulmonale develops in 5% of patients.[9] This case shows some features that fit into the established categories of chronic pulmonary schistosemiasis.

On clinical grounds, Mainzer et al.,[10] and Erfan et al.,[11] described a chronic type presenting as chronic bronchitis, bronchiectasis, emphysema and fibrosis or presenting as bronchial asthma.

Laboratory findings showed bilharzial ova are present in the urine or stools unless the bilharzial infection has died out as in this case or is the rare visceral form described by Mainzer et al.,[10] 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. 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 cirrhotic liver may be taken as almost sure evidence of bilharziasis, even in the absence of ova from the urine and stools like our case.[12]

Radiographic appearances including HRCT are of interstitial infiltrates, typically nodular or micronodular, and there may be frank fibrosis with calcification. This particular pattern of lung infiltration, however, is more usually described in longstanding interstitial pulmonary disease[13,14] and may simulate tuberculosis silicosis, sarcoidosis, atypical pneumonia or polyarteritis nodosa. These radiological appearances could be due to widespread granulomatous pulmonary schistosomiasis. .In advanced cases marked affection of the heart and pulmonary artery occur.[12]

Pulmonary nodes secondary to schistosomiasis as in our case are rare, and pose a differential diagnosis with lung neoplasia, are frequently defined only after exploratory thoracotomy.[15] The term 'pseudotumoral schistose-

miasis presentation' was first used in 1975 describing a pulmonary nodule in a patient who died, who had autopsy findings of granulomatous reaction, Schistosoma mansoni eggs, and pulmonary arteriolitis obliteration.[16,17] The presence of a large mass associated to *Schistosoma* infection (bilharziasis) was described in 1953 in Cairo.[11] The authors pointed out that, as first hypothesis, the lung mass producing these pathology findings was secondary to schistosomiasis. In 2003 Marcelo et al reported a case of man with finding of granulomatous lung nodule (adult worm of Schisto-somamansoni) after 25 years of having been treated.[18] Another case report by Claudio et al., 2009 was related to a 25-years-old patient who lived in a schistosomiasis endemic area with a pulmonary mass showed several granulomas with *Schistosoma mansoni* eggs.[19]

The granulomas have no preferential location, and may be found in all lung segments and pleura. Inter-alveolar thickening and connective tissue fibrosis were also reported.[20] The tubercle is formed when the egg provokes a granuloma surrounded by histiocytes, few eosinophils and fibroblast within the lung parenchyma and the inflammatory reaction becomes chronic, with the granuloma resolving to a focus of fibrosis and scarring as in our case. These small granulomas are always related to the bronchioles or alveoli. In some patients the alveoli fill with a serofibrinous exudate which may progress to become patchy consolidation characteristic of bronchopneumonia,[17]

Lesions from dead worms are rarer, and characterized by necrotic areas surrounded by intensive exud-ation, usually reabsorbed and involved by cicatricle tissue.[21]

CONCLUSION:

The interest elicited by this case is twofold. First, it is an example to stress the well known fact of the necessity for having bacteriologic proof before accepting the clinical diagnosis of tuberculosis. Second, it is a rare clinical manifestation of a pulmonary pathologic process caused by schistosomal infestation. The difficulty is the absence of sure diagnostic criteria, apart from the histopathologic picture of the lesion. From the review of the observations of previous workers, as well as the present case, it is seen that pulmonary manifestations in cases of bilharzias are numerous and variable. It is proposed that, in schistosomiasis endemic areas, pulmonary schisto-somiasis is considered a differential diagnosis for complex structures, as pulmonary masses. Pulmonary bilharziasis may be arrested at any stage, and the patient may live his normal span of life.

REFERENCES:

1. World Health Organization. Prevention and control of schistose-miasis and soil-transmitted 1991; 4: 1023-1026.

2. Lam-Quynh Nguyen1 Jeannelyn Estrella1 Elizabeth A. Jett1 Eduardo L. Grunvald2 Laura Nicholson2 David L. Levin1. Acute Schistosomiasis in Nonimmune Travellers: Chest CT Findings in 10 Patients a*AJR* 2006; 186: 1300–1303

3. Schaberg T, Rahn W, Racz P, Lode H.: Pulmonary schistosomiasis resembling acute pulmonary tuberculosis. European Respiratory Journal 1991; 4: 1023-1026 .

4.. Damluji SF, Al-Khouri M, Johnstone RM. Cor pulmonale due to chronic infestation with S.haema-tobium. J Trop Med Hyg 1964;67:220.

5. Andrade ZA, Andrade SG. Pathogenesis of schistosomal pulmo-

nary arteritis. Am J Trop Med Hyg 1970;19:305–10.

6. Morris W, Knauer CM. Cardiopulmonary manifestations of schistosomiasis. Semin Respir Infect 1997; 12:159–170

7. Abdel-Hakim M. and Anwar M. Elwi: Bronchopulmonary Schistosomiasis: Case Report, Dis Chest 1963; 43;317-319

8. Schwartz E. Pulmonary schistosemiasis. Clin Chest Med. 2002; 23(2):433-43.

9. Bethlem EP, Schettino GDPP, Carvalho CRR. Pulmonary schistosomiasis. Curr Opin Pulm Med1997; 3:361–5.

10. Mainzer, F: "On a Latent Pulmonary Disease Revealed by X-ray in Intestinal Bilharziasis (Schistosome Mansoni)," Puerto Rico I. Pub. Health and Trop. Med., 1939;1:121.

11. Erfan, M., Moussa, A. H., Garem, A. and Hazerian, N.: "Clinical Aspects of Pulmonary Bilharziasis," Gaz. Kasr ci Amy Fac. Mcd,, 1957; 13:2,.

12. Abdel-Aziz Sami: Pulmonary Manifestations of Schistosomiasis. Dis Chest 1951;19;698-705

13. 1A D B Waldman, 2J H Day, 3P Shaw, and 2A D M Bryceson: Case report: Sub acute pulmonary granulomatous schistosomiasis: high resolution CT appearances—another cause of the halo sign The British Journal of Radiology, 2001; 74:1052–1055 .

14. Rocha RL, Pedroso ERP, Rocha MOC, Greco DB, Ferreira CE. Forma pulmonar crônica da esquistossomose *mansoni*: avaliação clínico-radiológica. Rev Soc Bras Med Trop. 1990;23(2):83-9.

15. Fatureto MC, Correia D, Silva MBO, Barra MFC, Silva AV, Tarquinio DC, et al. Nódulo pulmonar esquistossomótico simulando neoplasia: relato de caso. Rev Soc Bras Med Trop. 2003;36(6):735-7.

16. Akoun G, Huchon G, Rolland J, Barrière L, Marsac J, Ronco P.: Pseudotumoral pulmonary schistosomiasis due to Schistosomamansoni Nouv Presse Med. 1975; 4(33):2408. French

17. Santiago Martı´nez, C. Santiago Restrepo, Jorge A. Carrillo, Sonia L. Betancourt, Toma´s Franquet, Claudia Varo´n,: Education Exhibit **135** Thoracic Manifestations of Tropical Parasitic Infections: A Pictorial Review1 Radio Graphics 2005; 25: 135–155

18. Marcelo Cunha Fatureto, Dalmo Correia, M rcia Benedita de Oliveira Silva, Maria, Pulmonary schistosomotic nodule simulating neoplasia: case report: Revista da Sociedade Brasileira de Medicina Tropical 2003; 36(6):735-737, nov-dez,

19. Cláudio Dornas de Oliveira, Glaucio de Oliveira Nangino, Paulo César Correia, Carlos Vechio Salomão, Marcelo Alencar Resende, Leonardo da Cruz Peixoto, Tumoral pulmonary mass secondary to *Schistosoma mansoni* infection resembling neoplasia: case report Rev Bras Ter Intensiva. 2009; 21(4):461-464.

20. Bogliolo L.: Schistosomiasis mansoni. Pathology. Rev Bras Malariol Doencas Trop. 1959; 11:359-424. Portuguese.

21. Barbato EC. :Schistosomal pneumopathy and chronic cor pulmonale. Arq Bras Cardiol. 1953; 6(3): 195-305.