PULMONARY HYDATID CYST IN SINGAPORE — A CASE REPORT

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SYNOPSIS

We report a rare case of a solitary pulmonary hydatid cyst in a 37-year-old Indian woman who presented with allergic manifestations due to a small rupture of the cyst and leakage of hydatid fluid. The chest x-rays showed the typical features of pulmonary hydatid cyst. Both the Casoni test and the fluorescent antibody test were positive. The patient was operated on successfully. Although the patient had been a resident in Singapore for 20 years, her infection was probably acquired during one of her visits to India, as hydatid disease is very rare in Malaysia and Singapore.

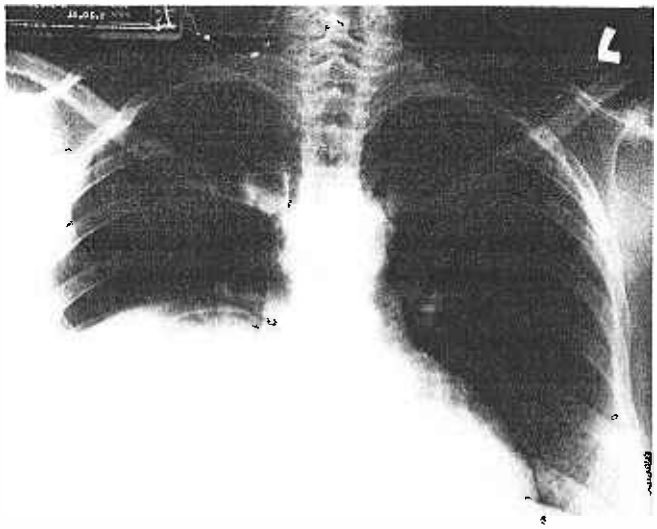
INTRODUCTION

Hydatid disease is endemic in sheep raising countries. It is rarely seen in Malaysia and Singapore. Indeed, only 3 cases have been reported in this region, all of them from Kuala Lumpur, West Malaysia. The first case was reported in 1955 by Khaira(1); the patient was a 34-year-old Indian man who had hydatid cyst of the liver. Duguid at al(2) reported the second case in 1968 in a 6-year-old Chinese boy who had hydatid cyst of the lung. The third case was reported in 1970 by Kutty et al(3). The patient was a 6-year-old Indian boy who had hydatid cyst of the lung. Of the 3 cases, only the patient reported by Duguid had never been out of the country and his infection was presumably acquired locally. This is a report of a case of pulmonary hydatid cyst diagnosed pre-operatively in a patient living in Singapore.

CASE REPORT

A 37-year-old Indian woman was referred by her general practitioner to the Emergency Unit of Tan Tock Seng Hospital in September 1981 for sudden onset of giddiness, vomiting, pruritus and rash of 1 day's duration. She was given an injection of Phenergan and was then admitted to the medical unit by the doctor on duty. The patient did not have a history of allergic diseases or self medication. Apart from menorrhagia, there was nothing of significance in the past history. She did not seek treatment for her menorrhagia which had been present for 3 years.

A



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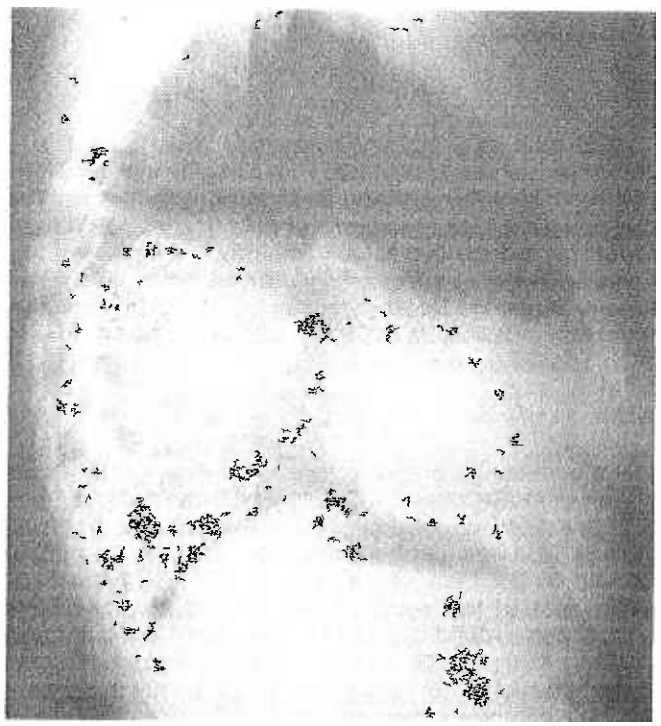

Fig 1 PA and lateral chest x-rays (A&B) showing a large homogeneous opacity with a well defined convex upper border situated in the lower right hemithorax.

Our patient was born in Malacca in 1945. She lived near a fruit estate. When she was a child, she used to play with 2 dogs kept by her family. At the age of 17 years, she married and left her hometown to settle in Singapore with her husband. She is now a permanent resident. She visited her inlaws twice in Madras, India in 1966 and 1971 and stayed for about 3 months on each visit. There she came in contact with sheep and dogs.

On examination, she was afebrile, pale and drowsy. Her pulse was 80 per minute and her BP was 100/60 mm Hg. Her rash cleared up after admission and was not detected subsequently. There was impairment of percussion note, decreased vocal resonance and diminished air entry over the right lower chest; liver and and spleen were not enlarged.

Investigations

Haemoglobin 6.6g/dl; white cell count 12,000 c mm; neutrophils 64%, lymphocytes 12%, monocytes 2%, eosinophils 22%, reticulocyte count 3%. Peripheral blood film showed moderate hypochromia, microcytes, poikilocytes and polychromatic cells. Serum iron 35ug/dl, total iron binding capacity 280ug/dl. Serum folate 1.0ug/l, serum vitamin B_{12} = 330ng/l. Urea and electrolytes were normal.

Chest x-ray (fig 1) showed a large homogeneous opacity in the lower right hemithorax and pleural effusion.

Clinical Progress

On the next day after admission, she developed a fever. A pleural aspiration was done. 300 ml of cloudy yellowish fluid was removed. Analysis showed specific gravity of 1.034, total protein 5.5g/dl, lymphocytes + + +, polymorphs +. Pleural biopsy showed an area of necrosis with polymorph leucocytes and mononuclear cells. No granulomas or caseation were seen and no acid fast bacilli were detected. Sputum cultures were also negative for acid fast bacilli.

She was given 3 units of blood and received a course of Ampicillin for 1 week after which she discharged herself on 8.10.81 against medical advice.

Second Admission

She was re-admitted on 25.3.82 for giddiness and vomiting for 1 day. She did not have a rash or itchiness. On examination, her general condition was satisfactory. Her temperature was 39.5°C, pulse 120 per minute and BP 110/60 mm Hg. The physical signs on the right side of the chest were similar to the findings in the first admission.

Investigations

Haemoglobin 13.4g/dl, white cell count 18,000 c mm, neutrophils 91%, lymphocytes 5%, monocytes 2%, eosinophils 2%.

Chest x-ray showed a huge cavity with a fluid level in the lower lobe of the right lung (fig 2).

Blood cultures were negative for pyogenic organisms. Serology for E histolytica and Pseudomonas pseudomallei were also negative.

Clinical Progress

Our patient was treated initially as for anaerobic lung abscess with intravenous crystalline penicillin 2 mega units every 6 hours and flagyl 400 mg tds. A bronchoscopy was done which showed pus in the basal segmental bronchi but no other abnormalities were detected. Examination of the bronchial aspirate did not reveal any scolices and cultures were negative for pyogenic organisms or fungi. At this stage, the possibility of hydatid disease was considered when a chest x-ray (fig 2) showed a curvilinear shadow looking like a folded membrance inside the cavity. Casoni test was positive with both early and late reaction. Fluorescent antibody test was positive in a titre of 1 in 256. Liver function test showed only a raised alkaline phosphatase (224U/l). Liver scan showed a moderately enlarged liver but no space occupying lesion to suggest a

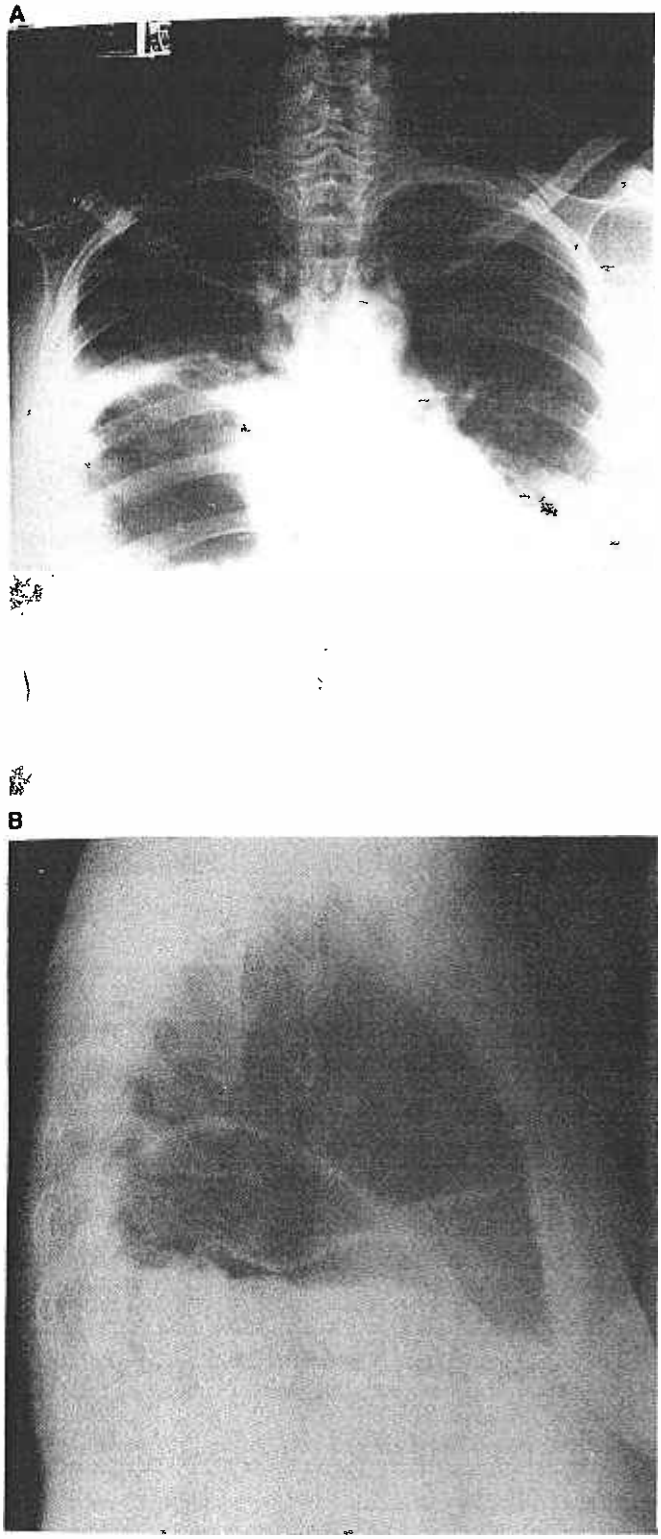

Fig 2 PA and lateral chest-rays (A&B) showing a large thin walled cavity with fluid level after rupture of cyst. Note presence of collapsed membrance within the cyst and irregularity of the fluid level (water lily sign).

cyst or abscess.

During her stay in the ward, she coughed out copious amounts of yellowish sputum which became progressively less in quantity with antibiotic treatment. Her fever also subsided gradually. A chest x-ray (fig 3) taken pre-operatively showed a marked reduction in the size of the cyst.

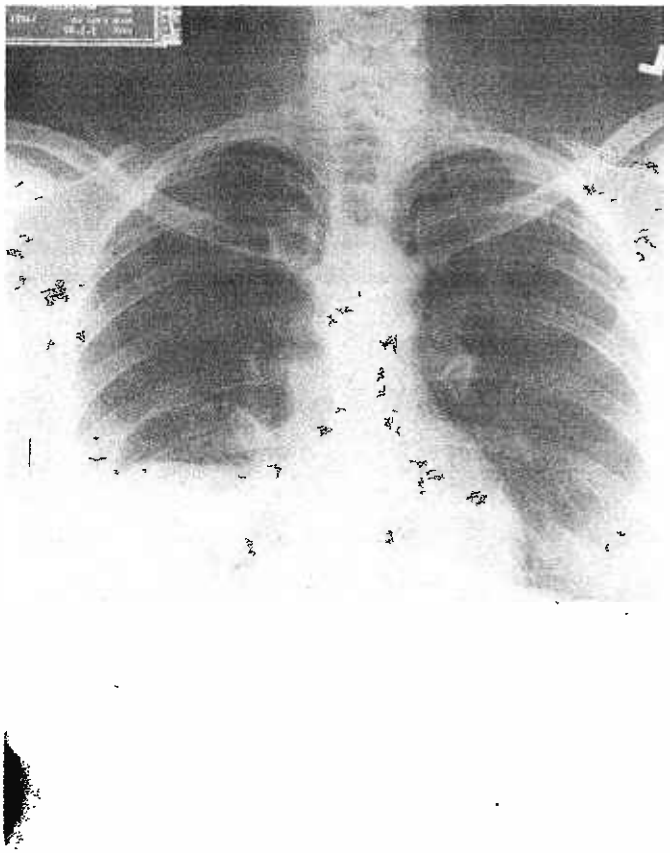

Fig 3 Pre-operative chest x-ray showing reduction in size of cyst.

Operation

A thoracotomy was done on 12 May 82. At operation, the whole of the right lower lobe was noted to be stuck to the diaphragm and pericardium. The cyst was found to be intact and situated in the anterior and medial segments of the lower lobe (fig 4). To prevent spilling and contamination of the surrounding structures, packs soaked in 3% hypertonic saline were placed around the cyst which was then enucleated after making an incision through the pericyst. The pericyst cavity was obliterated by purse string sutures and a drain was left behind. Her post-operative course was uneventful and she was discharged on 28.5.82. When seen as an out-patient on 16.7.82, she was noted to be well.

Pathology

Gross Examination

The specimen was a collapsed unilocular cyst with a transparent gelatinous like wall (fig 5). It measured 11 cm in maximum diameter and averaged 1.5 mm in maximum thickness of the wall. There were focal areas of translucency and the inner surface was smooth. Free lying daughter cysts and scolices were not identified.

Microscopic Examination

Histological examination showed the cyst wall to be composed of a thick laminated membrane which was devoid of nuclei (fig 6). Apposed to this and comprising the inner lining of the cyst was a darkly stained membrane; this was

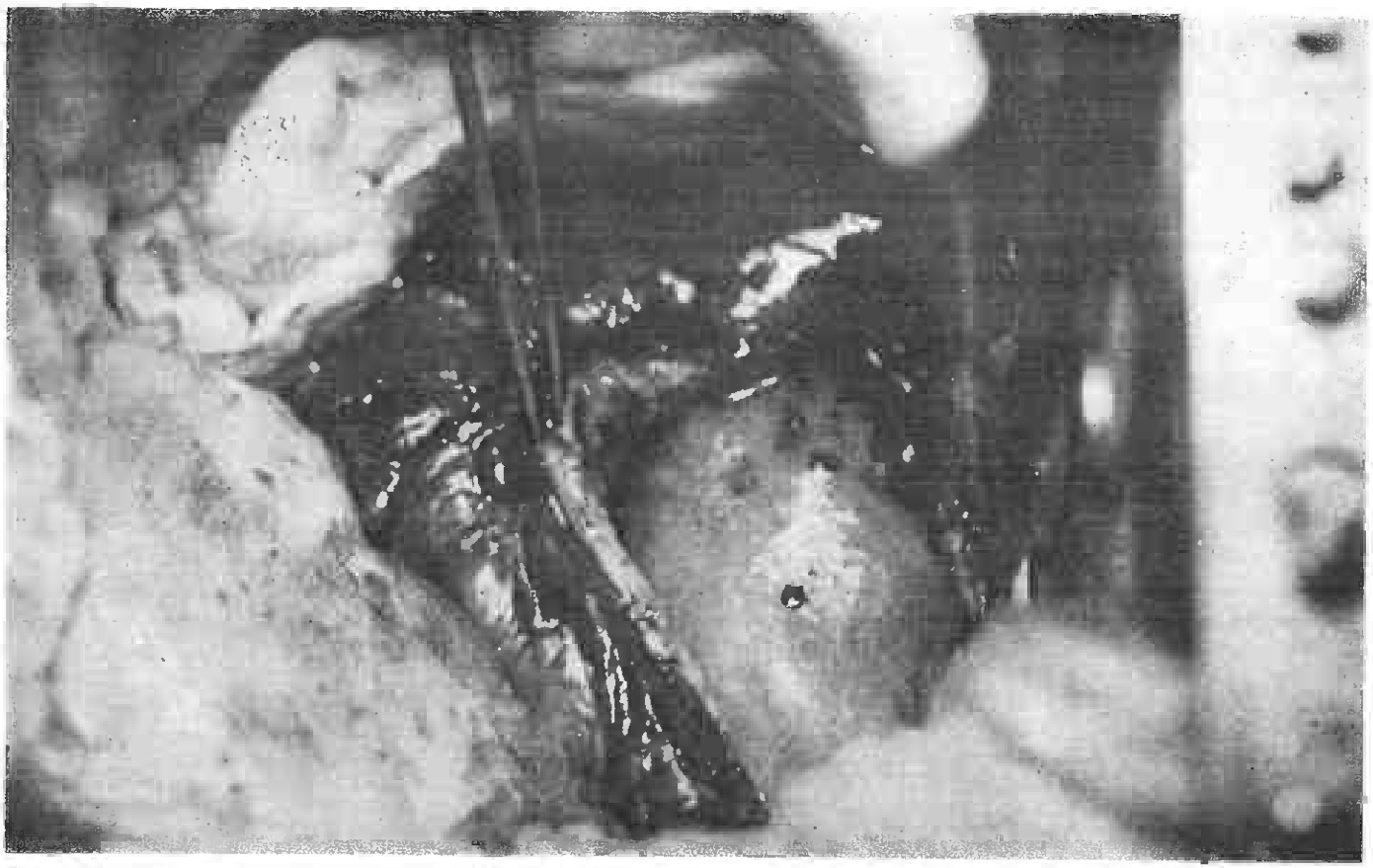

Fig 4 View of cyst at operation.

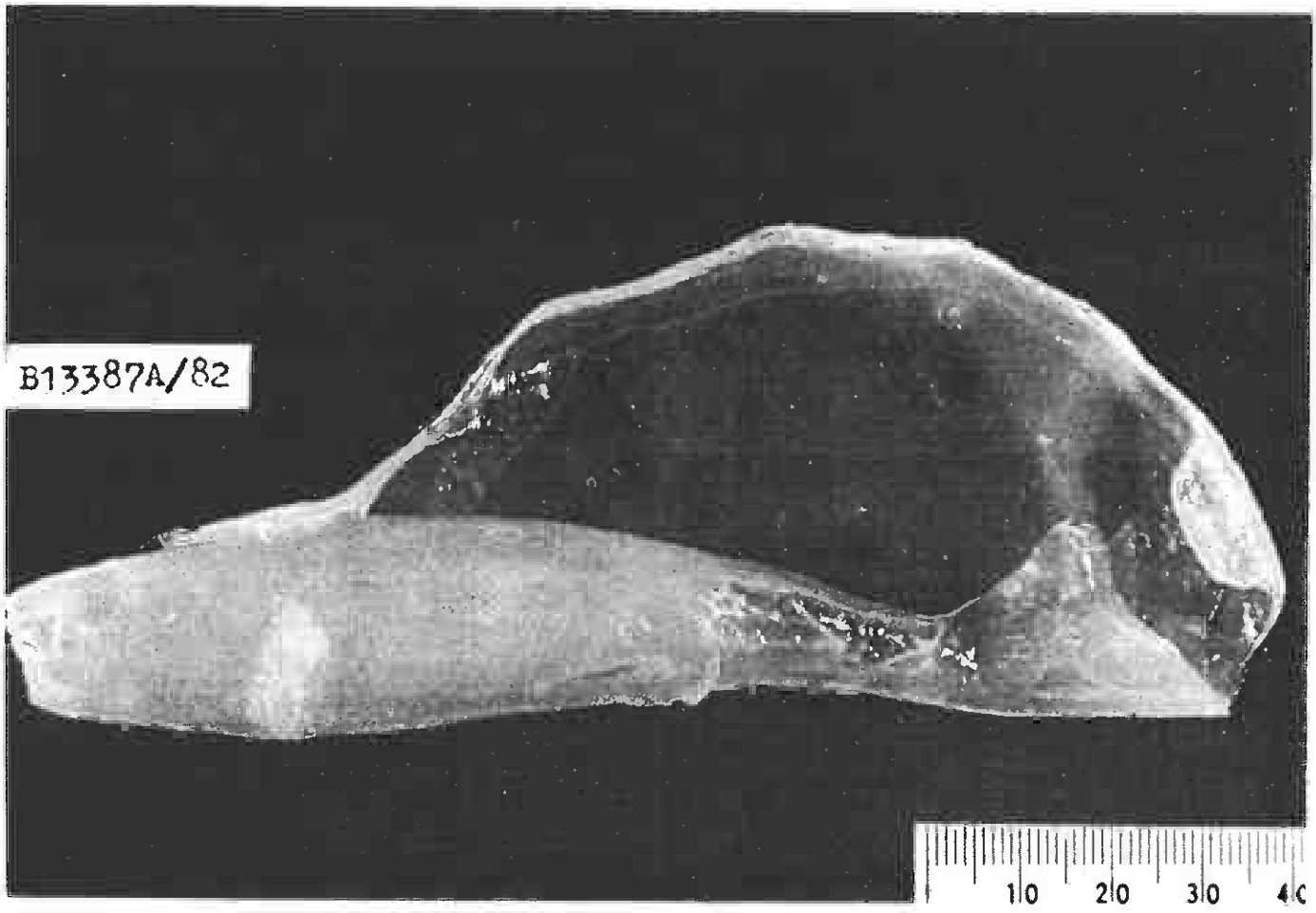

Fig 5 Part of hydatid cyst showing the glistening transparent wall.

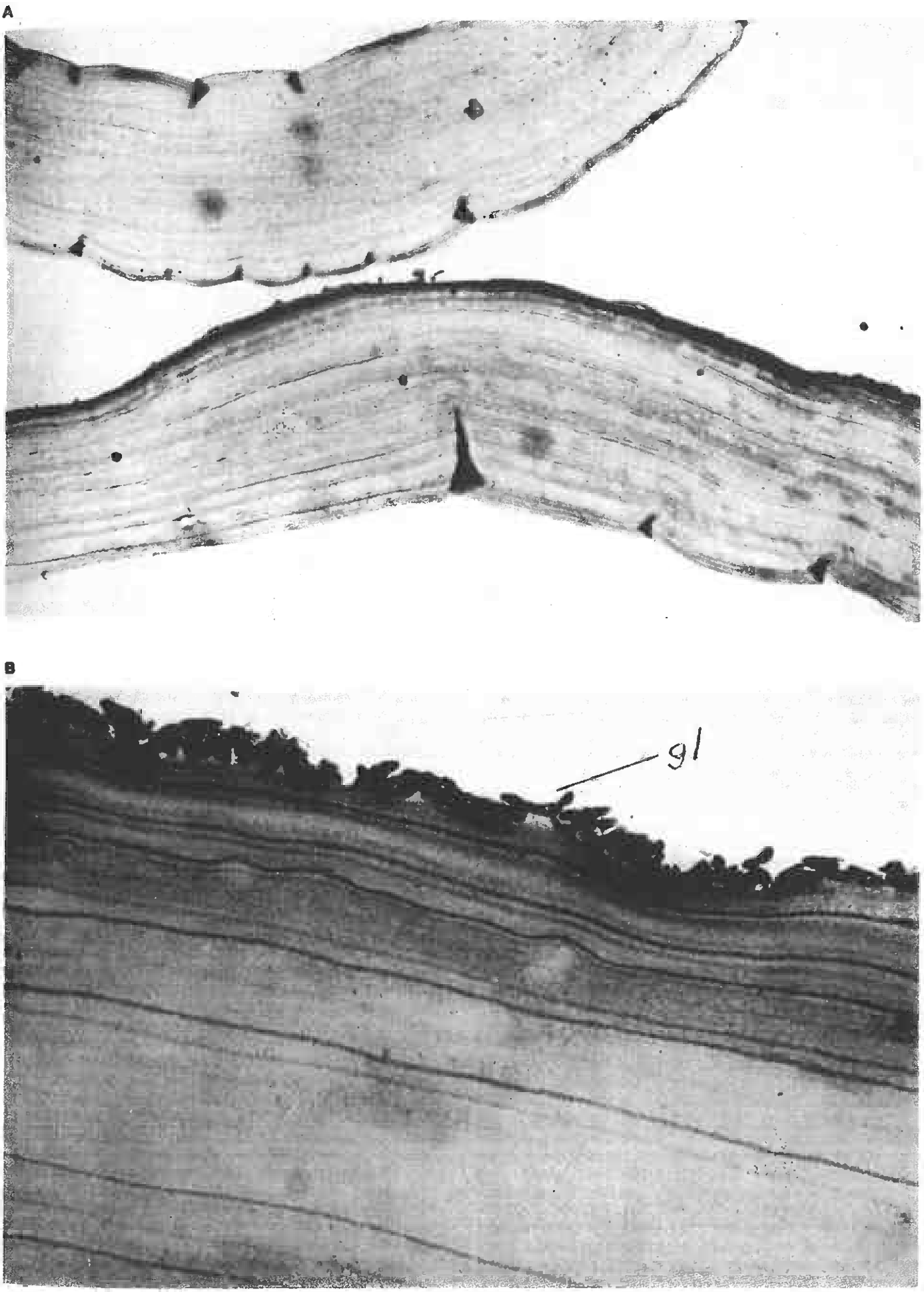

Fig 6 Wall of hydatid cyst x 40 (A) and larger magnification of wall x 250 (B) showing laminated membrane and nucleated germinal layer (gl).

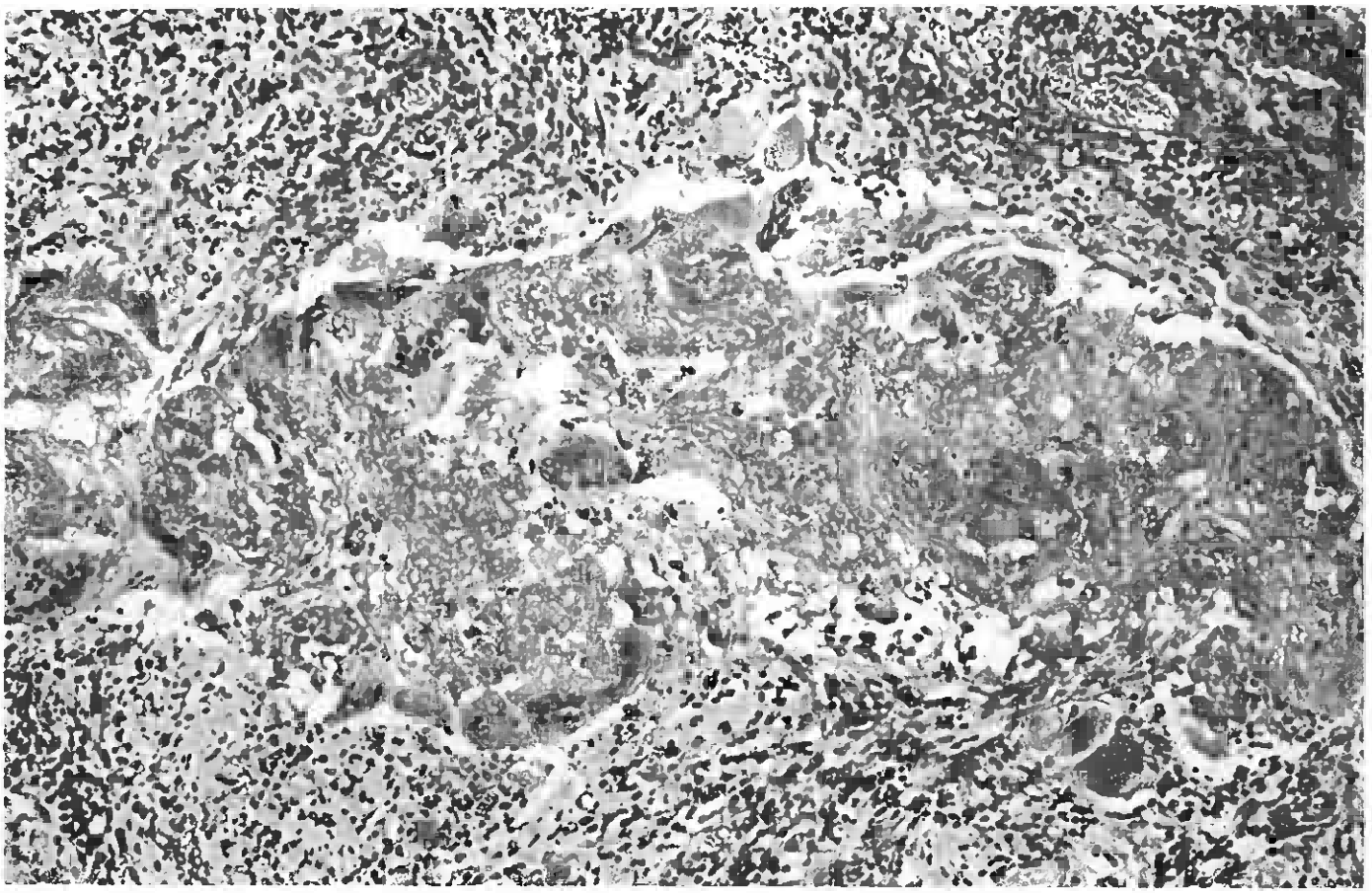

Fig 7 Section of pericyst showing thickened fibrous tissue with a heavy lymphocytic infiltrate and a focus of granulomatous inflammation. Multinucleate giant cells surround amorphous fibrin-like material x 100.

unilayered and although the individual cells forming this were not well identified in this specimen, the appearance was consistent with that of the germinal layer of a hydatid cyst. Focal stippled calcification was present. Although no free brood capsules or scolices of the Echinococcus were identified, the appearances were that of a hydatid cyst, the larval stage of the tapeworm genus Echinococcus.

A small piece of the pericyst wall (fig 7) showed thickened fibrous connective tissue with increased vascularity, congestion and lymphocytic infiltrate together with scattered polymorphs. Foci of multinucleate giant cell reaction and histiocytes indicative of a granulomatous response were present. In areas these encircled amorphous flibrin-like material. The features were indicative of a host reaction to the cyst. No brood capsules or scolices were identified in the lung tissue.

DISCUSSION

Hydatid disease is caused by the larval stage of the tapeworm belonging to the genus Echinococcus. There are 2 forms of the disease; the most common is the unilocular hydatid disease caused by E granulosus, while the multilocular or alveolar hydatid disease which is less common but more serious infection, is caused by E multilocularis. In this article, only unilocular hydatid cyst of the lung will be discussed as this was the infection found in our patient.

Unilocular hydaited cyst is endemic in sheep-rearing countries such as Australia, New Zealand, Middle East, Mediteranean countries, and South America especially Argentina, Chile, Uruguay. In Echinoccoccus granulosus infection itself, there are 2 distinct biological types. In the pastoral type, the dog is the primary host and the intermediate hosts include sheep (the most common intermediate host), cattle, pigs and man. In the less common sylvatic type which is found in Canada and Alaska, the primary host is the wolf and the intermediate hosts are reindeer and moose.

The dog harbours the adult worm in its small intestine. The ova produced by the gravid tapeworm are passed out in the faeces and contaminate the environment. The ova are then ingested by sheep when they graze in pastures which have been contaminated. After passing into the stomach of the sheep, the chitinous coat of the ova is dissolved by gastric juice and the embryo worm then migrates through the intestinal wall and is carried by the portal circulation to the liver where most of the embryo worm are arrested. Some pass on to settle in the lung, while a small number may lodge in virtually any part of the body eg spleen, kidney, bone, brain, skin and muscles. In patients with hydatid cyst of the lung without concomitant liver involvement, it is postulated that the embryo worm after migrating through the stomach wall enters the lymphatics and passes directly to the lung by way of the thoracic duct and mediastinal lymphatics thus avoiding the liver(4). In general, the liver is involved in 60-70% of cases, the lung in 20-30% and other organs or tissues 5%. However the lung may be more frequently involved than the liver in some countries(5). Once lodged in an organ, the larva grows slowly and may take 5 to 20 years before producing symptoms. The life cycle of the tapeworm is completed when the infected viscera of an infected sheep are fed to dogs as offal. Once ingested, the larva develops into an adult worm in the intestine of the dog.

Man is usually infected during childhood when an infected dog is fondled or handled with the result that the ova are transferred to the fingers of a child and then in-

gested during a meal. In our patient, the large size of the cyst suggests that she could have been infected many years ago, probably during one of her visits to India, where there were sheep and goats at the home of her in-laws. Infection acquired in here hometown of Malacca or in Singapore is possible but less likely in view of the extreme rarity of this disease locally. Inquiries with local veterinary officers revealed the absence of Echinococcus granulosus infestation in dogs in Singapore.

The hydatid cyst has 2 layers: an outer, thick, non-nucleated, laminated layer (ectocyst) and an inner, single cell, germinal layer (endocyst), from which brood capsules and daughter cysts develop. There is also an adventitial layer or pericyst which is formed by the reaction of the host to the pressence of the larva. The cyst contains clear fluid which is antigenic to the host and can cause an anaphylactic reaction when it is released into a serosal cavity. In fertile cysts, the deposit of brood capsules and scolices at the bottom of the cyst is known as hydatid sand. A cyst may be sterile ab initio(6) or with age(4) thus explaining why there is no dissemination of cysts in some patients after spilling of hydatid fluid either spontaneously or accidentally during surgery. Secondary infection of the cyst can lead to death of the scolices and daughter cysts. In our patient, brood capsules and scolices were not seen in the cyst or surrounding lung tissue suggesting that the cyst was sterile (acephalocyst). The possibility that the scolices and daughter cyst were killed by infection cannot be excluded.

During the stage of growth and expansion, symptoms are due to irritation and compression of lung tissue. The main respiratory symptoms are cough, chest pain, and haemoptysis. Fever and dyspnoea due to lung collapse may also be present. Many patients are symptomless as was the case in our patient before she was admitted to hospital. It is interesting to note that our patient presented with allergic manifestations which were probably due to leakage of hydatif fluid following a spontaneous tear or rupture of the cyst wall. The loss of fluid must have been slight as our patient did not develop an anaphylactic reaction. Rupture of cyst is also confirmed by the finding of a cavity containing a fluid level on the chest x-ray taken subsequently and the reduction in the size of the cyst. A cyst which has ruptured or become infected is referred to as a complicated cyst. Patients with ruptured cyst may cough out fragments of laminated membrane or daughter cysts which look like grape skin. An interesting feature noted by Barrett(7) is that for some unknown reason, leakage of hydatid fluid into the bronchial tree does not or very seldom lead to bronchial dissemination of parasites.

Radiology

In most patients, pulmonary hydatid cyst is diagnosed on the appearances of the chest x-ray. The radiological features are as follows:

(i) Round or oval pulmonary opacities (fig 1)
A round or oval opacity is usually seen in the lower lobe of the right lung attributed to greater perfusion of blood to that region. The opacity may be solitary or multiple, occurring in one or both lungs. In our patient, there was a huge opacity in the lower lobe of the right lung. Sometimes the cyst assumes a lobulated or bizarre appearance due to impingement against a rigid structure resulting in indentation of the cyst.

(ii) Crescent sign
This sign is due to air occupying the space between the pericyst and ectocyst and is usually due to a tear in the pericyst.

(iii) Cavity with fluid level (fig 2)
This feature is seen when there is rupture of the ectocyst into the bronchial tree allowing fluid to escape and air to enter the cyst producing an air fluid level. This feature was seen in our patient's chest x-ray.

(iv) Water lily sign (fig 2)
When a cyst ruptures, the membrane or lining of the cyst may collapse inwards and become visible e.g. as a folded membrane on the chest x-ray of our patient. The fragmented membrane may also float on the surface of the hydatid fluid imparting an irregularity to the fluid level which has been likened to water lilies floating on a pond.

(v) Calcification
Calcification of pulmonary hydatid cyst is rare(8). There is a tendency for the cyst to rupture before any calcification can occur, as resistance to the growth of the cyst is less in the lung than in the liver.

Diagnosis

Although the diagnosis of hydatid cyst is not difficult in endemic countries, it can be a problem in non endemic countries where the differential diagnoses include among others, primary or secondary lung tumour, pyogenic lung abscess, amoebic lung abscess, granuloma, neurogenic tumour and aneurysm. That hydatid disease can be acquired locally is shown by the case reported by Duguid(2).

In our diagnostic approach, it is important to question any patient who has an unexplained pulmonary opacity, his place of birth, residential and occupational history. Eosinophilia is not a very helpful sign as it is present in 20-25% of cases and may also be due to other helminthic infestation in patients living in developing countries. In recent years, ultrasonography and computerised tomography have added precision to the diagnosis as they are able to differentiate solid from cystic lesions eg. in the liver. Unfortunately, ultrasonography is not helpful for pulmonary hydatid cyst.

Suspicion of hydatid cyst is strengthened when there is a positive Casoni skin test. It is a sensitive but not a very specific test and is possitive in 60-80% of cases. Serological tests include complement fixation(CF), haemagglutination (HA) using tanned red cells, latex agglutination, fluorescent antibody and arc 5 immunoelectrophoresis or IEP test. The latex agglutination and IEP are reported to be sensitive and highly specific(9). Although the Casoni test can remain positive for as long as 5 years after removal of pulmonary hydatid cyst(10), the complement fixation and haemagglutination tests become negative within 1 year after its removal(10). The CF and HA tests are thus useful in assessing the success of surgical treatment.

Treatment

Surgical removal of the cyst is the treatment of choice. The intact pulmonary hydatid cyst is enucleated after an incision through the pericyst. The pericyst cavity left behind is either obliterated by suturing or left open as advocated by Sarsam(11). Resection of the lung is done for heavily infected or ruptured cysts. Some surgeons aspirate the cyst first after which they introduce a scolicidal agent such as 10% formalin or 10% hypertonic saline into the cyst before removing it. However there is a risk of spilling the hydated fluid(7). During operation, packs soaked in 2.5% formalin(12) are placed around the cyst to prevent contamination and dissemination of scolices should the cyst

rupture. Solutions of 0.5% formalin and 5% sodium chloride have also been found to be effective scolicidal agents(13).

Medical therapy has not been successful until recently. Mebendazole, a drug related to thiabendazole has been found to be effective in the treatment of hepatic cysts given in dosages of 400-600 mg tds for 3 weeks(14). Serious toxicity has not been a problem. The drug limits the uptake of glucose by the germinal membrane leading to glycogen depletion and death of the cyst. Mebendazole would appear to be indicated for elderly patients with hgh surgical risks and those patients with underlying medical disorders who are unfit for surgery. Although the drug was originally used for patients who had a recurrence of inflection after surgery, it has been suggested that Mebendazole should also be given to patients who had rupture of cyst and spilling of hydatid fluid during operation. Further research is necessary to establish the role of Mebendazole in treatment.

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