MULTIPLE PULMONARY HYDATID CYSTS

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As with other diseases, the diagnosis of hydatid infestation has come to be made more frequently and with greater confidence in recent years, owing to developments in both diagnostic and surgical techniques. In the past, some cases of hydatid disease of the chest may have been misdiagnosed under any of the following headings:[1] acute pneumonia or pleurisy, spontaneous pneumothorax, empyema, lung abscess, lung tumour, tuberculosis with haemoptysis, or bronchiectasis. Improved radiology and earlier surgery have now led to a more accurate diagnosis in some of these cases.

Thus the reported incidence of hydatid disease in the lung, relative to other organs, has undergone modification, and multiple hydatid cysts are found more often than is commonly thought.

This paper discusses the mode of infestation in multiple pulmonary hydatid cysts and their treatment.

ROUTE OF INFECTION

With the concept that the liver forms the primary filter of ova that are absorbed *via* the portal system from the bowel, it has been held that perhaps 75% of cysts are to be found in the liver, and that when other organs are affected the liver is involved as well in 50% of such cases.[2] The lung, as the second filter, was said to be the site of cyst development in 10 - 20% of cases. Jenkins,[3] in 1949, put the figures at 60 - 70% for the liver and 23% for the lung. However, some recent figures would indicate a much higher percentage for the lung. Jidejian,[4] in the Lebanon, found that 52% of his patients had hepatic cysts (178 cases) and the lung was affected in 30% (104). In New Zealand,[5] between 1946 and 1955, 946 patients were admitted to hospital for hydatid disease. Of these, 374 had hepatic cysts, i.e. approximately 40%, while during the same period 320 patients had thoracic cysts, a figure not far short of the incidence for the liver; this would seem to be more in keeping with experience in South Africa. It would appear that the more recent the series, the greater the incidence of pulmonary infestation. However, in Iceland lung cysts have apparently been very uncommon[6] for no very definite reason.

Experimentally it has been found that in animals the introduction of live ova into the bowel usually results in a predominance of pulmonary cysts, and in the squirrel as many as 97% of the cysts form in that site.[7] It has been suggested that the hepatic capillaries are particularly large in such animals, and that the ova can therefore pass through quite readily.

Most authors have favoured the portal system as the route of entry, since the emphasis has been so largely upon the liver in the past. However, if there is indeed a much higher incidence of pulmonary cysts than was thought, attention may be given to alternate routes which have been postulated. Thus, the lymphatic route has been suggested, whereby the ova travel up the thoracic duct to reach the right side of the heart, and thus bypass the liver. Occasional cysts have been reported in the posterior mediastinum, but it has been suggested that they should be found more frequently if this is a common route. However, the embryo is about 25 microns in size and thus could easily be carried further on by the lymph stream.

In most series there is a predominance of cysts in the right lung, and the lower lobe is particularly affected. This has led to the suggestion that the embryo may actively migrate through the diaphragm. The embryo is indeed capable of some motility, but if it did behave in this way one would expect to find many more cysts in other organs adjacent to the liver, without such a predominance in the lung. Also, the difference in incidence in the two lungs is of the order of 65 : 35[19] — one would expect a far greater incidence on the right if this route were common.

It was suggested as far back as 1877, by Bird,[8] that the ova may reach the lungs by inhalation. These small ova could be carried readily in the dust which is so common in sheep-rearing districts. Barrett,[7] Dew, and others have discounted this possibility on somewhat flimsy grounds. Experimentally, Dévé[9] insufflated live ova into the tracheas of rabbits and was able to obtain lung cysts by this method in many cases. Live ova can survive for long periods on the coats of animals and, while most inhaled particles are probably removed by ciliary action,

it does not seem inconceivable that ova could survive in the bronchial mucus. Barrett[7] stated that this is unlikely, but does not substantiate his statement.

Proved cases of bronchogenic spread of hydatid disease are extremely rare. Dévé in 1931 could only find records of 5 cases of intrabronchial rupture of a hydatid cyst which had resulted in further pulmonary cysts. In 1952 Susman,[25] in Sydney, reported the case of a 7-year-old child in whom the rupture of a single primary lung cyst was followed, 6 months later, by 8 widespread cysts, which were almost certainly the result of bronchogenic dissemination. In such instances one is dealing with the contents of a cyst, i.e. scolices and possibly small daughter cysts, which for one reason or another may be expectorated more readily than ova, or rendered harmless in other ways. However, this evidence would weigh against inhaled ova producing cysts.

The development of multiple cysts in the lung could be explained on a basis of multiple infestation by any of the four routes mentioned, and it is perhaps surprising that in so many cases there is only a single cyst in the lung. Moreover, in contrast to what has sometimes been stated,[2] the liver seems to be affected uncommonly when the lung shows the presence of cysts. I have rarely seen patients with cysts in both liver and lung. Barrett[7] has even gone to the extent of opening the diaphragm in the hope of finding cysts which could not be detected clinically, but it is also his experience that if the lung is involved there is rarely a cyst in the liver. Some degree of host resistance may be responsible for this.

The incidence of multiple cysts in the lung is illustrated by the various series shown in Table I. These figures suggest that in some 20 - 30% of cases of pulmonary

TABLE I. MULTIPLE HYDATID CYSTS IN THE LUNG

Author	*Date*	*Bilateral cysts*	*Patients with more than one cyst*	*Total number of patients*
Waddle[10]	1950	(*a*) 6·6%	?	246
		(*b*) 9·3%	?	232
Jidejian[4]	1957	?	34	104
Taiana[11]	1957	16	41	200
Howden[12]	1955	4	11	31
Brown[13]	1958	9	30	152
Majano[14]	1957	20	31	103
Present series	1962	3	7	24

echinococcosis one can expect to find multiple cysts, and that in about one-third of these both lungs will be affected. Authors of small series have varied in their opinion on the incidence, as may be expected, some stating that multiple cysts are uncommon,[15] while others consider them common.[16] It is of interest that in one of the few reports on hydatid cysts in the South African literature, Schrire[17] recorded one case of multiple bilateral cysts among 4 patients.

DEVELOPMENT OF MULTIPLE CYSTS

There are three methods by which multiple cysts may develop:

1. As mentioned above, multiple ova may be ingested and reach the lung. This would appear to be a likely explanation in those cases where the cysts are of different size.

It has been suggested that cysts grow at variable rates. There is radiological evidence of cysts increasing by 2 cm. in 2 months, or 3 cm. in nearly 2 years;[18] others have noted similar differences. However, one would anticipate that 2 cysts in the same lung would grow at the same rate, although it is possible that the environment may be different in various areas within the lung.

2. A second method is by the rupture of a cyst into a vein in some other organ. This has apparently been recorded in only 3 organs, viz. the heart, liver and iliac bones.[7] Dévé analysed 31 cases and found that the primary cyst was in the heart in 20, in the liver in 8, and in the iliac bones in 3. Only 250 cases of primary cardiac cysts had been recorded up to 1956,[7] and few of these had produced pulmonary daughter cysts; so it seems this method of development is rare. Experimentally, however, it is relatively easy to obtain multiple daughter cysts by intravenous injection of scolices; it is of interest to note that these cysts are relatively uniform in size and tend to settle peripherally in the lung substance.

3. Thirdly, multiple cysts may arise following partial rupture of a primary cyst in the lung. As has been mentioned above, bronchogenic spread is rare, but multiple cysts can form within the adventitial sac of the primary cyst. This is also uncommon, but Barrett[7] recorded such a case.

Partial rupture which threatens the life of the cyst often leads to the formation of daughter cysts within the primary cyst, and even if this is fully expectorated later, one or more of the daughter cysts may grow in the remaining space. Daughter cysts can survive in pus and other secretions, though Dew was doubtful whether scolices can.[20]

Rupture into the pleural space may lead to multiple cysts, and this is particularly so after spontaneous rupture. The so-called 'hydatid pneumothorax'[23] and empyema may well be followed by the growth of numerous cysts within the pleural cavity. In these cases one can easily visualize the formation of daughter cysts within the primary cyst before rupture, owing to the threat of disintegration from its peripheral situation. However, from 104 primary lung cysts which ruptured into the pleural space, Dévé[24] found that secondary cysts developed in only 8%.

Though surgical rupture has traditionally been fraught with danger, there are not very many cases on record of recurrence after spillage of hydatid material in the chest during an operation. Brown[13] recorded 3 instances of recurrent pleural hydatids following operation for simple cysts in 85 patients. However, here in Cape Town I have not seen or heard of such recurrence, although a number of cysts have ruptured over the years during attempted intact removal. This has made one wonder whether scolices can indeed form new cysts, but this seems to be beyond doubt from experimental evidence.

Dew,[20] on the basis of work by d'Alexinsky and Dévé in Europe, by Lagos Garcia in South America, and by himself in Australia, was able to state categorically: 'If washed living scolices are injected into rabbits by the subcutaneous, intraperitoneal or intravascular routes, positive results in the production of secondary cysts are

obtained in about 75% of the experiments'. Dévé referred to this as 'retrograde metamorphosis', since the scolex can rapidly form a vacuole and develop into a true replica of the primary parasite.[21] This has recently been confirmed in a different way by Abello,[22] using radioactive materials; he claimed that scolices can produce ova or oncospheres at a certain stage of development and that these can grow into cysts. He suggested that it is a form of parthenogenesis, occurring only at a certain stage, which allows recurrences in some patients and not in others.

TREATMENT

Before the present era of adequate results from thoracic surgery, expectant treatment was the norm, and there are various reports of apparently complete cure of pulmonary hydatids by expectoration. Thus Waddle,[10] in reviewing some 30 years' experience with the Australasian Hydatid Registry, was able to record, in 1949, the recovery in this manner of 36 patients out of 91 who did not undergo some form of operative procedure. Fitzpatrick,[1] in 1950, reported the case of a patient who was found to have 5 cysts scattered in both lungs at the age of 42 years. These cysts were expectorated soon afterwards, and in a 28-year follow-up the patient showed no sign of residual damage.

Berry and Francis[5] have recently reported an extraordinary case of multiple cysts, largely cured by spontaneous expectoration. This young adult was first seen in 1947 when he had between 20 and 30 lung cysts — the exact number could not be determined since they were too numerous. He collapsed under anaesthesia when removal was to be attempted and, for the next 6 years, proceeded to cough up the cysts at varying intervals. The final cyst became too large under this expectorant regime so that it was removed by thoracotomy, since when the patient has been fit.

This, however, is only one side of the picture. The same authors[5] noted 27 deaths over a 10-year period due to pulmonary cysts. The morbidity, too, must be high, as can be realized when the natural history of the disease, with its complications of chronic lung sepsis, haemoptysis, etc., is considered. Thus, all authorities agree today that surgical treatment is indicated when a lung cyst is diagnosed.

Surgical removal of hydatid cysts is a regular procedure today, with very satisfactory results. Various methods are adopted, depending on the extent and nature of the disease.

The effects of a hydatid cyst on the surrounding lung have been investigated fully[26,27] and, in many cases, these effects are reversible. However, particularly in the case of infected cysts, there may be permanent lung damage which requires treatment, apart from the hydatid. The related compression-atelectasis and emphysema is usually reversible, and even the marked bronchial changes may not be permanent. The bronchi are compressed to form an oval-shaped lumen and there may be cartilage degeneration with gradual erosion of the wall until bronchial openings are made into the adventitial sac.

With infection, the lesion becomes more destructive, with surrounding pneumonitis and possible bronchiectasis. An infected cyst is virtually a chronic lung abscess, with contained sequestra in many instances. Since these features affect the ultimate outcome of treatment, it is important to try to assess the position as adequately as possible before operation. For this reason, bronchography is certainly essential in patients with infected cysts, and it has been recommended that it should always be done, even when uninfected cysts are present.[16,28]

Surgical treatment can be divided into: (1) the removal of the cyst; and (2) the procedure used for the surrounding lung.

Removal of the Cyst

The cyst may be removed in a number of ways. The majority of authors seem to favour simple suction without the introduction of any antiseptic into the cyst. A useful method is to remove a good deal of the fluid through a needle and then, after making a small incision, to insert a large cannula, attached to the suction apparatus, which will remove the actual wall of the cyst. It has been shown by Fitzpatrick[29] that so-called 'hydatid sand' is heavier than the fluid and therefore sinks within the cyst. If the cyst is not shaken, scolices will settle at the base within a minute; thus this method of aspiration is unlikely to produce the spillage which might lead to recurrence.

Another method of removal, which has been advocated by Barrett[7] and others, is intact extrusion of the cyst through an incision in the adventitial wall. Gaseous pressure by the anaesthetist gradually opens wide an initial rent in the adventitia (pericyst), and the whole cyst is extruded into an awaiting dish. When this occurs as planned, it is a very satisfying procedure, and Fig. 1 shows 2 large cysts which were removed from the right

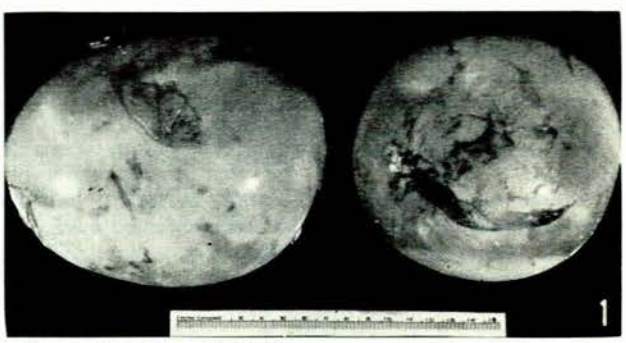

Fig. 1. Two large hydatid cysts removed intact from the right lung of an adult male (scale in cm.).

lung of an adult male by this method. However, it can be more dangerous in respect of spillage, since the cyst not uncommonly ruptures; this can occur with such suddenness that the entire contents of the cyst are scattered over the wound. Usually the surgeon has packed swabs around the area of removal so that the contamination is not gross, but the basal material is nevertheless thrown over this part. Although it was popularized by Barrett, Brown[13] pointed out that Quick in Australia used this method in 1933, and Ugon in Uruguay in 1947.

Extracapsular dissection has been recommended by Fontana and by others.[30] This involves excision of the pericyst, but is not as haemorrhagic as might be expected. Bronchi which have become involved are dealt with as encountered, and there is little residual air leak. The cyst is delivered

intact and the procedure is a satisfactory one except that it entails a more laborious dissection than the other methods.

Treatment of the Lung

Once the cyst has been removed, attention must be directed to the surrounding lung. If the pericyst has already been removed as suggested above, little more need be done apart from adequate pleural drainage, as after any thoracotomy. If, however, the pericyst is still *in situ*, there will almost invariably be some bronchial openings which must be closed with sutures. On the other hand, some have suggested that the pericyst be excised at this stage, removing the layer of compressed lung. Although cysts are usually not restricted to a single pulmonary segment,[27] it is unusual to perform lung resection, e.g. lobectomy, for simple or uninfected cysts. Brown[13] has suggested that resection may be indicated if the cysts are very large, e.g. more than 6 inches in diameter, or occasionally if there are large cysts in the middle lobe. Generally, however, even with large cysts, the lung expands readily once the compressing factor is removed. This is naturally of importance in dealing with multiple cysts in the same lung; a number may be removed without the excision of potentially functioning lung tissue.

Infected cysts pose rather a difficult problem, and require resection of permanently damaged lung tissue more frequently. The lesions may be in the form of bronchiectasis or chronic interstitial infection. It is often possible to deal with these in a similar way to the uninfected cysts, namely, by removal of the sequestrum-like cyst contents and then closure of the bronchial leaks. However, with infected cysts of the lower lobes it is usually wisest to resort to pulmonary resection, since the lack of adequate drainage does not allow of proper recovery once the cyst has been evacuated.

It is usually possible to decide pre-operatively on the question of resection, with the help of bronchography. Complete pneumonectomy is rarely indicated, but may occasionally be necessary in unilateral infected cysts.

In patients with cysts in both lungs, the same principles of treatment apply, with the proviso that as much lung tissue as possible be preserved. It is a feasible procedure to deal with both lungs simultaneously, and this has been advocated for infected and also uninfected cysts.[31]

SUMMARY

In hydatid disease, the lungs appear to be affected more commonly than was supposed, and infestation with more than one cyst is not uncommon. Some figures of published cases are quoted to support this statement.

The routes of entry to the lungs, and, in particular, the methods by which multiple cysts may arise, are discussed.

The treatment of pulmonary hydatid cysts is reviewed, with brief reference to various reported methods. Of a recent Cape Town series of 24 patients with hydatid disease 7 had multiple pulmonary cysts, and their case histories illustrated many of the points made in this article.

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