

## PROPTOSIS CAUSED BY HYDATID DISEASE\*

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HYDATID disease is a very rare cause of proptosis even in countries where it is endemic. It is therefore worth recording an analysis of the main features of three recent cases.

**MODE OF DEVELOPMENT.**—This disease is also termed echinococcosis, or hydatid cyst formation; it results from the ingestion of food contaminated with the eggs of the tape-worm *Echinococcus granulosus*. The embryo is released in the intestines, attaches itself to the mucosa, and makes its way into the portal circulation, by which it is carried to a capillary barrier, where it settles down, and, if not overcome by the reaction of the surrounding tissue, develops into a "hydatid cyst". This hydatid cyst is the larval form of the tape-worm, man being the intermediate host. The adult worm is usually harboured by the dog, which is one of the main definitive hosts.

The commonest site for hydatid cyst in man is the liver. Statistics in endemic areas show that in man hydatid cysts of the liver constitute 75 per cent. of cases. This is because, on account of their size, most of the embryos cannot pass through the capillary connection between the portal and hepatic veins and therefore settle in the liver. The smaller embryos are carried by the blood to the hepatic vein, reach the pulmonary circulation, and meet with a second barrier in the pulmonary capillaries. Here they are filtered and some have to settle and develop into hydatid cysts there. In man these form about 5 to 10 per cent. of the cases.

Embryos that escape the two barriers of the liver and lung capillaries are carried to the left side of the heart and distributed to various parts of the body. Statistics show that the kidneys and brain are the two main organs infected in this way. The orbit is one of the rarest sites to be involved; according to Khalil (1939), the incidence of hydatid disease of the face, orbit, and mouth together, ranges between 0.8 and 2.3 per cent. in endemic areas.

**TYPES OF CYST.**—Under optimum conditions an echinococcus embryo normally develops into a unilocular cyst, containing hydatid fluid, scolices (taenia heads), and in some cases brood capsules. As a result of the normal reactions of the tissues of the body, this cyst is enveloped by a thick fibrous capsule termed adventitia or pericyst. The wall of the

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hydatid cyst is whitish, glistening, and firm; it is made up of a laminated layer, essentially free of nuclei, lined by a protoplasmic nucleated germinal layer. From this lining layer, scolices and brood capsules develop. Therefore, in the course of exploration a dense fibrous layer is found surrounding the true hydatid cyst which is whitish and shining.

Occasionally the echinococcus embryo does not develop into a typical cyst with circumscribed boundaries, and the germinal layer is thus capable of sending out processes into the neighbouring tissues and giving rise to metastasis along the lymphatics and blood vessels in distant organs. This is termed the alveolar type. According to Faust (1939) this type of infection is more common in older patients.

An embryo that is caught in a closely confined bony canaliculus seems incapable of developing into an ordinary unilocular cyst, but grows and permeates all the available bony spaces causing serious destruction of the bone involved; this is termed the osseous type of cyst.

The symptoms produced by the unilocular hydatid cyst depend mainly on its size and site, but hydatid toxin may leak out and be absorbed into the system of the host, thereby producing allergic manifestations. Eosinophilia occurs in only 20 to 25 per cent. of diagnosed cases (Faust, 1939), and the specific precipitin complement-fixation tests, and intra-dermal reactions are not constantly present. In view of this and because of the rarity of the disease in the orbit, hydatid disease in this region is likely to be missed, which may lead to such serious consequences as rupture of an unsuspected fertile cyst, resulting in a spread of infection to the neighbouring tissues.

#### CASE REPORTS

**Case 1. L. H., female, aged 10 years.**—When seen in 1936 she complained of gradually increasing right-sided proptosis of 4 years' duration (Fig. 1). The condition was painless and associated with no constitutional disturbances. The general condition of the patient was quite good, there had been no allergic manifestations and diplopia was not complained of. The right eyeball was proptosed mainly forwards with no limitation of movements; there was severe congestion of the conjunctiva with chemosis of its lower part; the lower eyelid and the inner part of the upper, were also chemosed. The lids could not close over the proptosed eyeball, but fortunately the cornea was free of ulceration.

On palpation a very tense swelling (? cyst) was felt below and to the inner side of the eyeball; it was not tender. The margins of the orbit were intact, but the right inferior orbital margin was felt to be lower than the left one.

The right fundus showed papilloedema.

Intra-nasally no abnormality was detected.

Radiologically the cyst gave a very dense shadow overlapping the right side of the nose, the lower half of the right orbit, and the right maxillary antrum. It was of uniform density (Fig. 2).

General medical examination revealed no abnormality.

Systematic laboratory investigations revealed 11 per cent. eosinophilia; the precipitin and complement-fixation tests were negative.

During exploration a fibrous mass was found adherent to the floor of the orbit and

to all the neighbouring soft tissues; and a firm, whitish, shining sac bulged out separating it from the bone. This was shelled out and the fibrous mass removed. The sac was found to be a hydatid cyst full of opalescent fluid, and containing three grape-like daughter cysts. The fibrous mass surrounding it was recognized as the adventitia.

Microscopic examination proved that the cysts were all sterile and contained only degenerate scolices.

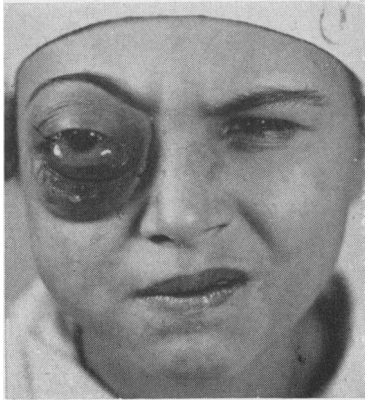


FIG. 1.—Case 1, showing right proptosis produced by development of intra-orbital hydatid cyst.

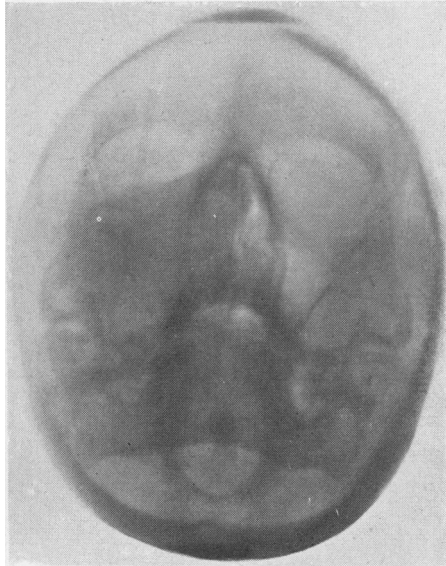


FIG. 2.—Case 1, radiograph demonstrating dense opaque shadow of hydatid cyst.

**Case 2.** N. S. R., female, aged 12 years. When seen in 1946 she complained of right proptosis of 3 months' duration which was ascribed to trauma (Fig. 3, overleaf).

As a result of panophthalmia in infancy the patient had developed a nebula on each cornea. She had no history of any allergic manifestations and diplopia was not complained of. The proptosis was mainly forwards and outwards, associated with slight inward and downward limitation of eyeball movements. The right conjunctiva was congested.

On palpation a cystic swelling was felt to be stretching the inner part of the right upper eyelid, and extending deeply into the right orbital cavity; its extent and attachments could not be ascertained. It was not tender and appeared to follow the movements of the eyeball. The skin of the lid was not adherent to it. The orbital margins were felt to be intact.

Nasal examination revealed no abnormality.

Radiological examination showed that the right orbit was dilated mainly at the expense of its inner wall (Fig. 4, overleaf), but the cyst was not visible.

The right ethmoid and maxillary sinuses were opaque and the right frontal sinus absent.

Systematic clinical and laboratory tests revealed anaemia (Haemoglobin 58 per cent.) and eosinophilia 15 per cent.

Precipitin and complement-fixation tests were negative. On exploration a unilocular hydatid cyst was found mainly inside the muscle cone, lying above and on the sides of the optic nerve, following it back to the optic foramen; the inner and anterior part

of the cyst was protruding out into the orbital tissues outside the cone, and this was the part that was presenting under the lid. The adventitia was thinner in this case than in Case 1.

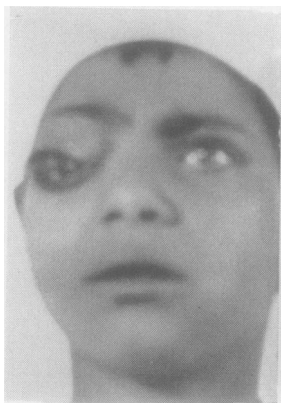


FIG. 3.—Case 2, right proptosis produced by hydatid cyst lying mainly inside the muscle cone.

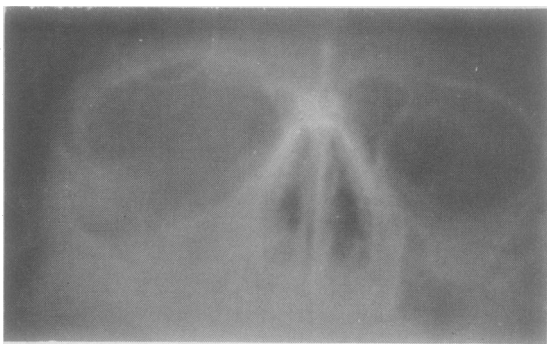


FIG. 4.—Case 2, radiograph showing primary dilatation of the orbit resulting from development of hydatid cyst.

This cyst was full of hydatid fluid and scraping of the germinal layer proved it to be fertile; Fig. 5 demonstrates the cyst wall with a scolex inside.

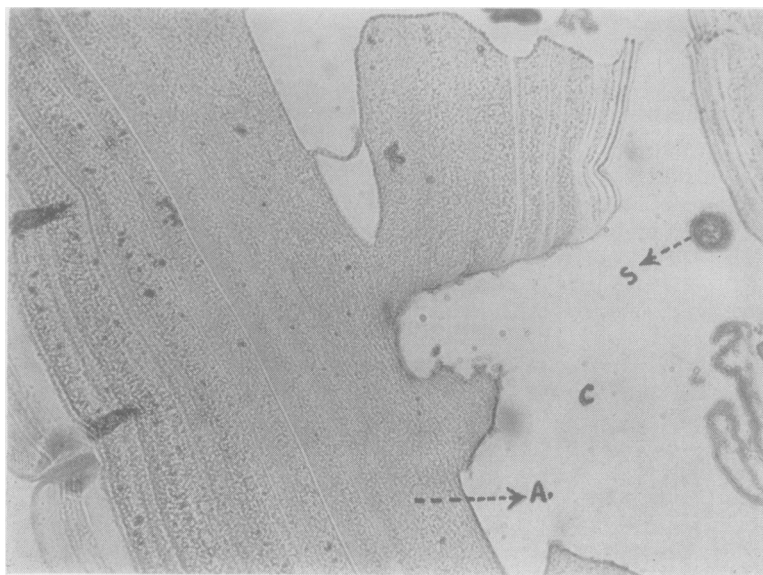


FIG. 5.—Case 2, photomicrograph of portion of hydatid cyst.

A—Germinal layer. C—Cyst cavity. S.—Scolex in section.

**Case 3, male, aged 9 years.** When seen in 1950, he complained of left-sided, gradually increasing proptosis, associated with failing vision, of 5 months' duration.

The condition was not associated with fever or any constitutional disturbances. It was painless. There was no history suggestive of allergic phenomena, and diplopia was not complained of. The proptosis was directed forwards, upwards, and inwards with limitation of eyeball movements downwards and outwards. The cornea was intact. The left pupil was slightly dilated compared with the right, but reacted well to light. A cystic swelling thought to be the cause of the proptosis was felt in the lower and outer part of the left orbital cavity. The bony margins of the left orbit were felt to be intact. Vision in the left eye was 6/24, and the left fundus appeared to be normal.

Nasal examination revealed double maxillary sinusitis and an adenoid mass. The general medical condition was good.

Radiological examination did not reveal the cyst, but the left orbital cavity was seen to be bigger than the right (primary orbital dilatation).

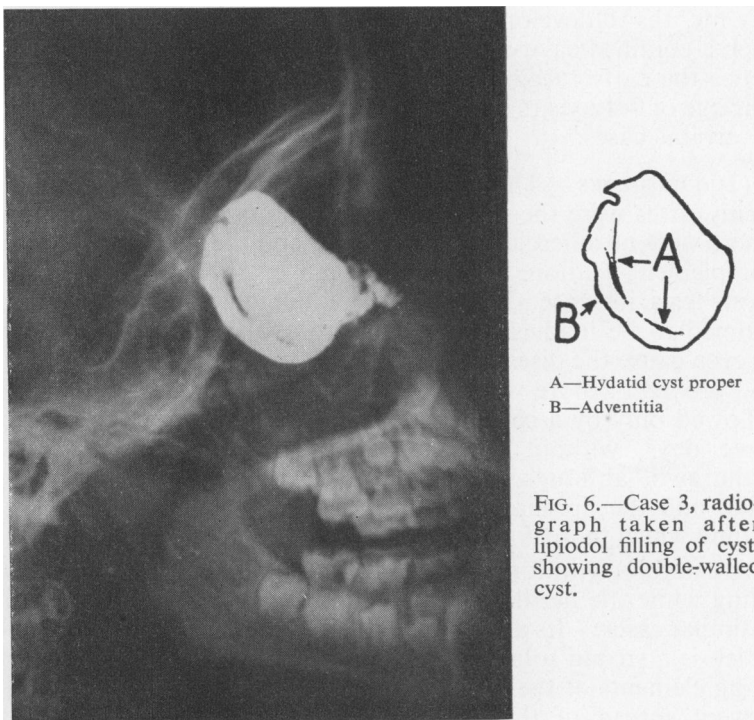


FIG. 6.—Case 3, radiograph taken after lipiodol filling of cyst, showing double-walled cyst.

A—Hydatid cyst proper  
B—Adventitia

Laboratory tests showed eosinophilia 7 per cent. The precipitin and complement-fixation tests were negative.

In view of the position of the cyst and in the light of previous experience, a hydatid cyst was suspected; aspiration, lipiodol filling, and re-x-raying were attempted. The cyst fluid showed no cellular elements or scolices, and was bacteriologically sterile.

Chemically it contained:

Protein ...	...	250 mg./100 ml.
Sugar ...	...	63 mg./100 ml.
Chloride ...	...	710 mg./100 ml.

X ray demonstrated a double-walled cyst (Fig. 6). The radio-opaque lipiodol filled the cyst cavity and the spaces between the two walls. This favoured the diagnosis of hydatid cyst.

On exploration, a unilocular cyst was found lying in the orbital cavity outside the muscle cone; embedded (as in Case 1) in thick fibrous tissue (adventitia) which was firmly adherent to the surrounding tissues. There were no daughter cysts. Microscopically scolices and echinococcus hooks were demonstrated, proving the cyst to be fertile.

Caseni's intradermal test was carried out 2 weeks after operation and was found to be negative, denoting the absence of any other hidden focus.

#### COMMENT

(1) AGE INCIDENCE.—According to Faust (1939), echinococcus infection occurs as a rule in childhood and an embryo takes only five months to develop into a cyst 1 cm. in diameter. Such a cyst in such a confined cavity as the orbit is sure to produce symptoms, and hence the early recognition of this disease when it affects the orbit. The age of 9–12 years in my cases represents the average age incidence in orbital cases.

(2) DIAGNOSIS.—The positive findings in favour of hydatid disease in my cases were the presence of a cystic swelling and eosinophilia. There were no allergic manifestations and the specific precipitin and complement-fixation tests were negative. The negative results of these tests increase the difficulty of diagnosis. Cyst puncture is claimed to be inadvisable as liable to evoke allergic manifestations or even cause the disease to spread. It was, however, tried in Case 3 and proved of more value than the specific tests usually done, being followed only by a certain amount of oedema which disappeared in three days, without much discomfort to the patient. The fluid withdrawn, although negative for *scolices*, was typical of hydatid fluid, and the lipiodol filling demonstrated a double-walled cyst, a finding which to my knowledge could be given only by a hydatid cyst.

A careful aspiration of the cyst, followed immediately by lipiodol filling while the needle is still inserted is, in my opinion, worth trial in similar cases. Its diagnostic value is seen in Case 3. The lipiodol, which is a strong solution of iodine, may also be enough to kill the living elements of the parasite before surgical intervention, and thus prevent spread of the disease if the cyst is accidentally ruptured during operation.

Simple radiography demonstrated two important points in my three cases:

(a) The cyst may throw a demonstrable shadow and its boundaries may be fully defined as in Case 1. This is of value in guiding the operator.

(b) The orbit could be dilated by the development of a cyst in its cavity. This was seen in Cases 2 and 3, but in Case 1, although the orbital cavity was dilated, the dense opacity of the cyst overshadowed a large part of the orbital cavity so that its size could not be judged from the radiograph.

This point was discussed in a previous communication (Handousa Bey, 1947), and it was shown that the same appearance could also be caused by other intra-orbital tumours.

Exploration has proved to be the most reliable method of diagnosis, and microscopic examination of scrapings from the lining of the cyst has been the best way of finding out whether the cyst is fertile or sterile.

(3) POST-OPERATIVE COURSE.—Recovery after operation was uninterrupted. There was no sign of spread or recurrence of the disease, but the first two cases developed enophthalmos. This was avoided in Case 3 by preserving part of the adventitia, which is due to a reaction of the tissues of the host and is not a part of the parasite. A portion of the adventitia of appropriate size was left behind and was enough to prevent sinking in of the globe.

#### SUMMARY

A study of three cases of intra-orbital hydatid cysts shows that:

(1) X-ray examination after lipiodol filling of the cyst can be of diagnostic value and is worth trial in similar cases.

(2) The enophthalmos which is liable to follow removal of the cyst can be avoided by leaving *in situ* a portion of the adventitia sufficiently large to prevent sinking in of the globe.

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