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SHORT COMMUNICATIONS & CASE REPORTS

Disseminated hydatid disease in a child: Albendazole treatment of orbital cyst

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PURPOSE. *Orbital hydatid cyst is rare and represents a small percentage of all echinococcal systemic locations. It is usually a primary echinococcosis. A new case of orbital hydatid cyst with asymptomatic coexistent lung and liver cysts is described in a child.*
METHODS AND RESULTS. *The patient underwent surgical excision of the lung and liver cysts and successful application of albendazole treatment for orbital echinococcal cyst without evidence of recurrence after a 4-year follow-up.*
CONCLUSIONS. *Hydatid cyst should be included in the differential diagnosis of unilateral exophthalmos in patients from countries where echinococcosis is endemic. (Eur J Ophthalmol 2008; 18: 1034-6)*

KEY WORDS. *Hydatid cyst, Orbit, Albendazole*

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INTRODUCTION

Hydatid disease is endemic in most sheep-raising countries in the Middle East, Mediterranean Basin, South America, New Zealand, and Australia. The main form is due to *Echinococcus granulosus*, a dog parasite tapeworm. Orbital hydatid cyst is rare and represents less than 2% of all echinococcal systemic locations (1). Surgery has been the traditional treatment. Response of abdominal and cerebral hydatid disease to treatment with albendazole has been previously reported (2, 3).
The present article describes a case of orbital hydatid cyst with asymptomatic coexistent lung and liver cysts in a child. The patient underwent surgical excision of the lung and liver cysts and successful application of albendazole treatment for orbital echinococcal cyst without evidence of recurrence after a 7-year follow-up.

Case report

A 7-year-old girl presented with a unilateral painless proptosis of the right eye of 2 months' duration. Visual acuity was 20/25 and downward and temporally displacement of the globe with impaired extraocular motility was present. A soft mass was palpable superonasally and fundus examination was normal. Hematologic and routine biochemical examinations were also normal. Erythrocyte sedimentation rate was 10 mm. Orbital ultrasonography revealed a well-demarcated uniocular cyst in the superior orbit with minimal internal echoes. CT scan showed an extraconal low-density well-circumscribed mass in the superior orbit, 3 x 5 cm (Fig. 1, left). MRI showed a low-intensity signal on T1- and a high-intensity signal on T2-weighted images (Fig.1, right). The cyst gave signal identical to that of the vitreous. Further clinical and radiologic evaluation including chest radiographs, abdominal ultrasonography, and CT scans of the brain, thorax, and abdomen revealed two cystic lesions in the lower lobe of the right lung and one more liver cyst near the liver hilus (Fig. 2). Radiologic characteristics and cysts' location were indicative of echinococcal disease. On serologic tests for echinococcosis, both the indirect hemagglutination (IHAT) and the enzyme-linked immunoabsorbent assay (ELISA) were negative.
The patient underwent surgical excision of the lung cysts and 1 month later of the liver cyst. Gross appearance and histopathologic examination of the cysts confirmed the diagnosis of hydatid disease. Postoperative period was uneventful. Chemotherapy started 2 weeks preoperatively and continued

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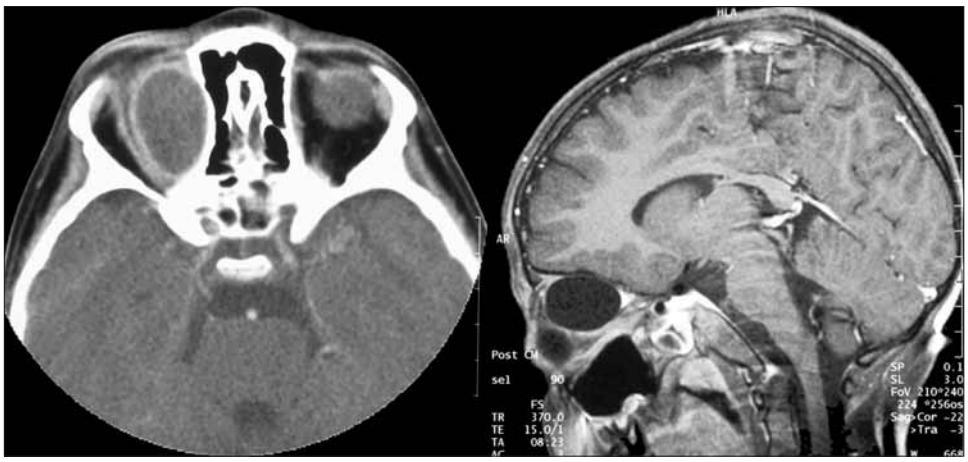

Fig. 1 - (Left) *CT scan shows a well-circumscribed cystic mass with hyperdense borders in the superomedial angle of the right orbit.* (Right) *Parasagittal magnetic resonance image (T1 weighted) shows a low-intensity cystic lesion with sharply defined borders causing downward displacement of the globe.*

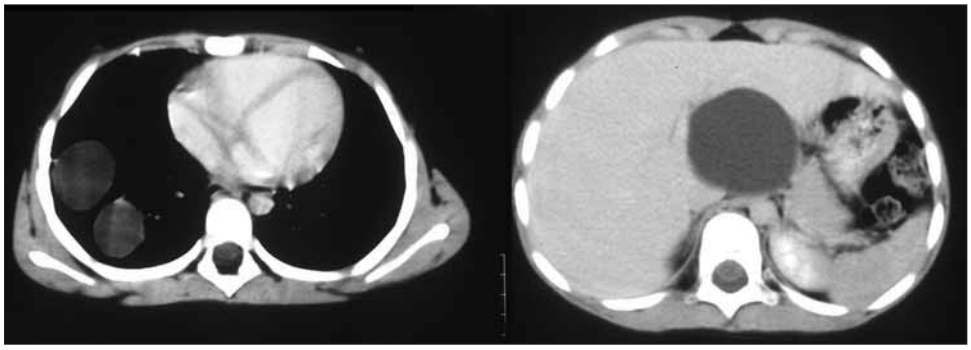

Fig. 2 - (Left) *CT scan shows two cystic lesions in the lower lobe of the right lung.* (Right) *CT scan shows cystic lesion in the liver located near the liver hilus.*

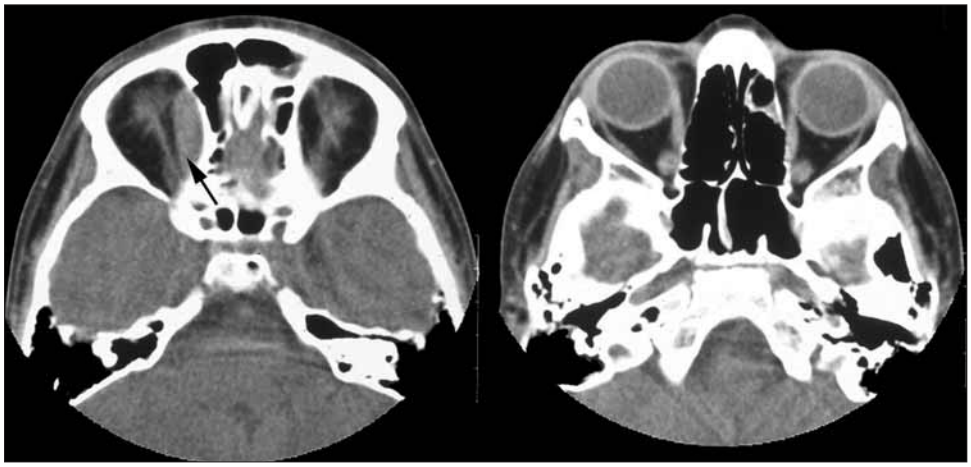

Fig. 3 - (Left) *CT scan after a 6-month treatment shows a residual solid shrunk cyst (black arrow) located in the superomedial angle of the orbit.* (Right) *Proptosis has resolved.*

postoperatively. The patient was administered albendazole, 200 mg twice daily with meals, for six courses of 4 weeks with 2-week drug-free intervals. Treatment was tolerated well, without developing side effects or laboratory test abnormalities.

One month after starting albendazole treatment, a notable decrease in size and volume of the orbital cyst was noticed. Proptosis decreased significantly and motility abnormalities improved. The patient's good response to chemotherapy postponed the surgical excision of the orbital cyst. Regular close follow-up was instituted. CT scan at the end of the sixth month of treatment demonstrated a 26 x 9 x 12 mm residual, solid, shrunk cyst (Fig. 3, left). The patient's clinical condition remains stable 7 years after treatment, without changes or evidence of recurrence of hydatid cysts in the imaging studies. Visual acuity

and optic nerve appearance remained normal during the entire follow-up period.

DISCUSSION

Hydatid cyst should be included in the differential diagnosis of unilateral exophthalmos in patients from countries where echinococcosis is endemic. Orbital hydatid cyst is usually a primary echinococcosis. It is uncommon to find other simultaneous hydatid cysts elsewhere in the body (1, 4, 5). In the present case, the patient had simultaneous orbit, lung, and liver hydatid disease.

Preoperative diagnosis is important in order to avoid cyst rupture during surgery and further dissemination of the disease. Serologic tests are used in diagnosis, but due to the integrity of orbital cysts the results are usually negative (1, 6). Therefore, imaging techniques, ultrasonography, CT scan, and MRI scan play an important role.

Surgical extirpation is still the most favored treatment for orbital cysts (1). Lateral osteoplastic orbitotomy, fronto-orbital approach, or inferior orbitotomy are the most frequent surgical approaches used. Surgical removal of orbital hydatid cysts is frequently complicated by cyst rupture, which may result in a severe postoperative anaphylactic reaction, incomplete removal, and secondary implantation due to spillage of the cyst contents.

The benzimidazole compounds, albendazole and mebendazole, are used in hydatidosis. Albendazole is more effective than mebendazole, with a higher degree of systemic absorption and penetration into the cysts (7). Albendazole acts as a parasiticidal agent by blocking glucose uptake by susceptible larvae and adult parasites. Their glycogen stores are depleted and ATP formation is blocked. The parasite is thus immobilized and killed. Chemotherapy is used preoperatively to reduce the size of a big cyst, in cases of intraoperative rupture of cysts, and in inoperable cases of echinococcosis, like cases of multiple cerebral hydatid cysts (3, 7). Smaller and younger cysts respond better. The penetration of the drug depends on the thickness of the cyst wall and the presence of calcification (2). The suggested dose of albendazole is 10 mg/kg/day for a minimum of four 1-month courses, separated by 15-day intervals. The drug is tolerated well and the most important side effect is hepatic toxicity. Pathologic elevation of liver enzymes prompts discontinuation of the drug. In cases of partial success or initial failure, retreatment is recommended. Some studies of combined chemotherapy (mebendazole or albendazole plus praziquantel) suggest that this is more effective than either agent when given alone (6). Treatment success is indicated by a parasiticidal effect characterized by radiologic findings of reduction in the size or disappearance of the cysts, increased density of the cyst contents, and a totally echogenic picture and/or calcification of the cyst walls (2).

In conclusion, orbital hydatid cysts seem to respond well to chemotherapy. Chemotherapy can be used preoperatively to reduce the size of the cyst and ease subsequent surgical excision and to prevent dissemination of the disease in case of accidental intraoperative rupture of the cyst. Long-term chemotherapy may also be effective as monotherapy provided there are not existing vision-threatening complications and a close long-term follow-up of the patient is instituted.

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