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HYDATID CYST MIMICKING PULMONARY HEMATOMA IN A PATIENT WITH HEMOPHILIA A*

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SUMMARY: Olcay L, Besim A, Şenocak ME, Göçmen A, Büyükpamukçu M, Kale G, Çetin M, Gürgey A. (Departments of Pediatrics, Radiology and Pediatric Surgery, Hacettepe University Faculty of Medicine, Ankara, Turkey). Hydatid cyst mimicking pulmonary hematoma in a patient with hemophilia A. Turk J Pediatr 1998; 40: 425-429.

We present a patient of 2.5 years of age with hemophilia A and a pulmonary hydatid cyst. A chest x-ray taken by chance showed a paracardiac opacity resembling an intrapulmonary hematoma which did not reduce in size after infusions of fresh frozen plasma and factor VIII but rather enlarged. Transabdominal ultrasound, colored echocardiography, thoracic computed tomography and magnetic resonance imaging findings were consistent with a cyst that was firmly attached on the border of the right atrium and also indented it; the wall was remarkably thick with no internal echoes. Hydatid cyst was dagnosed after thoracotomy. *Key words: haydatid cyst, hemophilia A.*

In countries with endemic regions, the diagnosis of "hydatid cyst" is easily made and treated[1-3]. Nevertheless, in a patient with hemorrhagic diathesis like hemophilia, presence of a pulmonary opacity suggests hemorrhage rather than a hydatid cyst and thus there may be some problems in the differential diagnosis.

Here in we present a hemophiliac patient with a pulmonary opacity which was suggestive of a hemorrhage at first but diagnosed as hydatid cyst postoperatively.

Case Report

A 2.5-year-old boy was admitted to our hospital because of bleeding of a cut on his foot. His past history revealed the diagnosis of hemophilia A at eight months of age, which had been followed in our hospital. He was hospitalized

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as the bleeding did not stop despite regular fresh frozen plasma infusions. A routine chest x-ray revealed a right paracardiac opacity. At that time, the borders of the opacity were not clear. Therefore, and in view of the patient's history, this opacity was thought to most probably represent an intrapulmonary hematoma. The patient was infused with fresh frozen plasma and factor VIII for seven days. Chest x-ray taken at the end of this therapy revealed that this opacity showed no reduction in size. On the contrary it had enlarged and this time with clear borders (Fig. 1). Transabdominal ultrasound and colored echocardiography

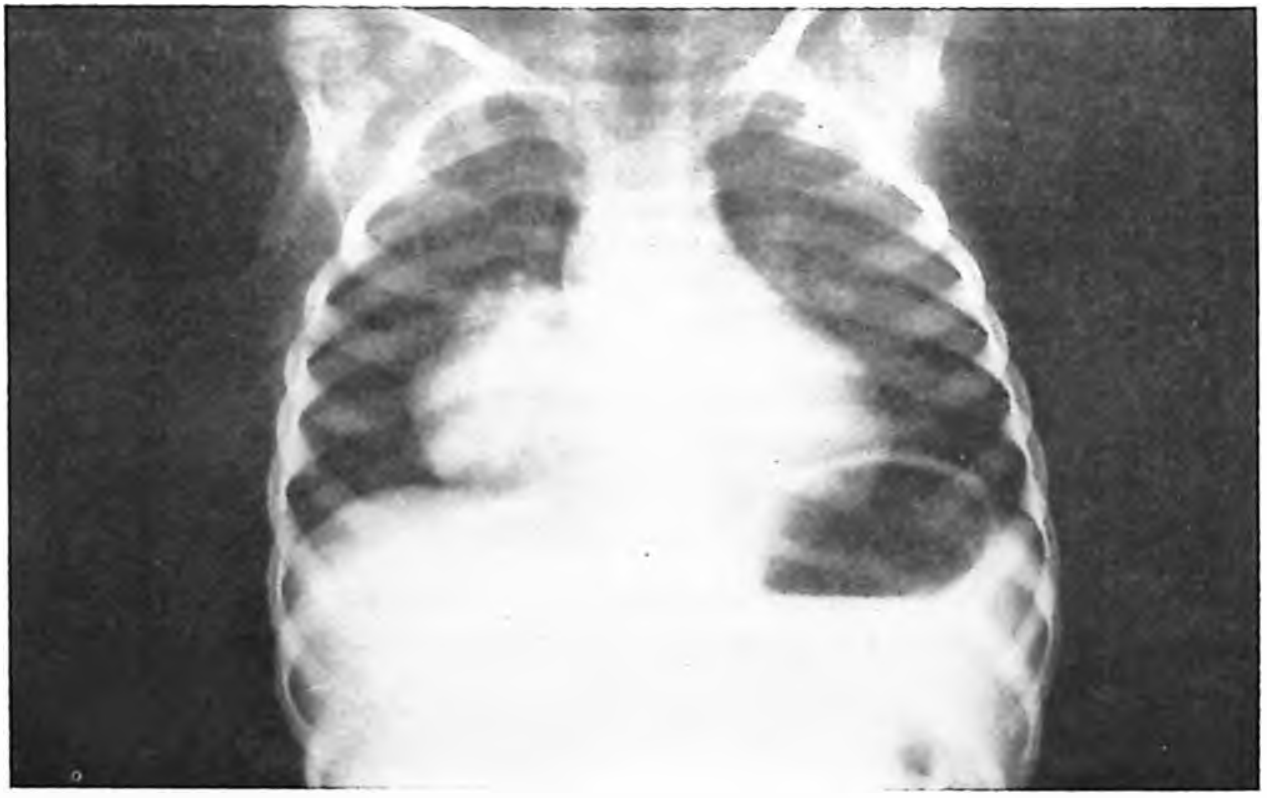

Fig. 1: The chest x-ray done after factor VIII and fresh frozen plasma therapy. Enlargement of the opacity, this time with clear borders, is seen.

revealed a pure cystic structure without internal echoes and with a thick wall. Thoracic computed tomograpy (CT) and magnetic resonance imaging (MRI) findings were also consistent with a cyst (Figs. 2 and 3) which was firmly attached on the border of the right atrium and also indented it. The wall of the cyst was remarkably thick. The patient's peripheral blood smear revealed eosinophils 27 percent anti-echinococcal antibody IgM and IgG 1/100, and Echinococcus specific IgE negative. In differential diagnosis, Echinococcus, pericardial cyst, thymic cyst bronchogenic cyst were considered.

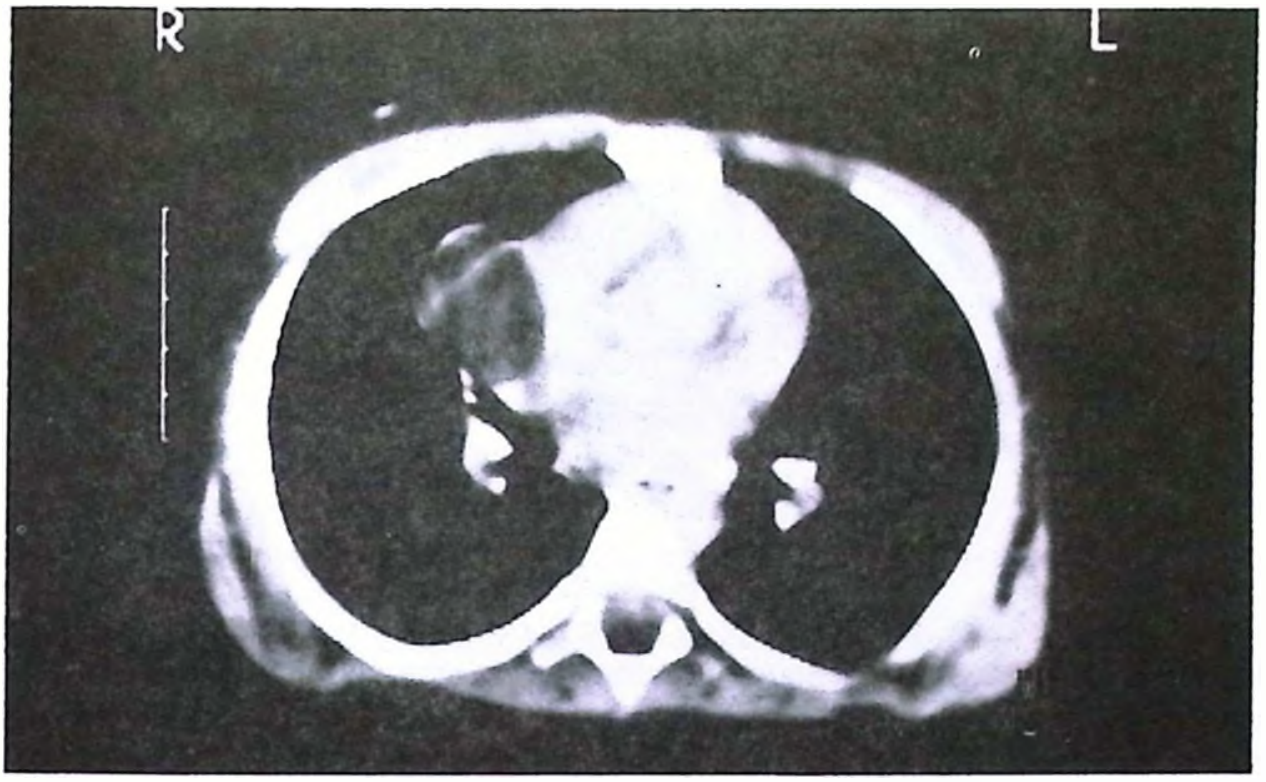

Fig. 2: Thoracal CT revealing presence of a cyst.

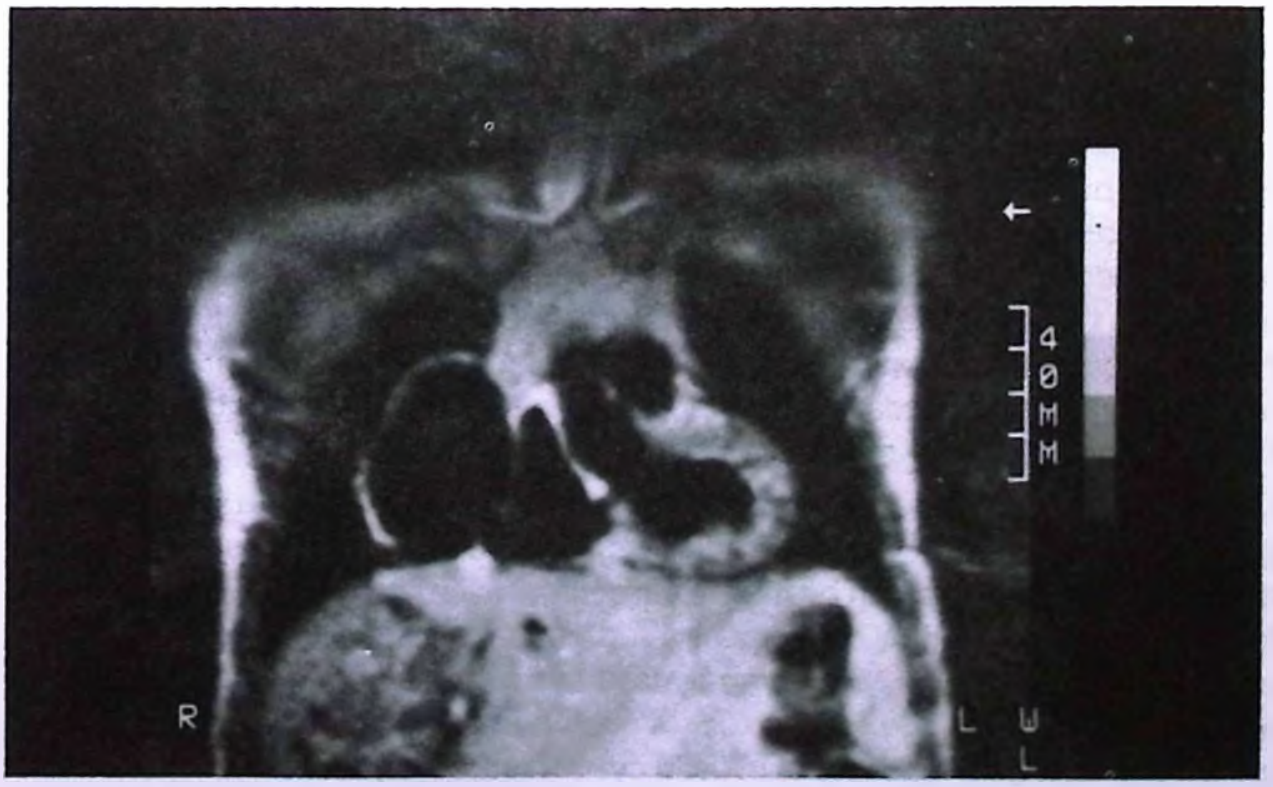

Fig. 3: Thoracal MRI showing a cyst firmly attached on the border of right atrium and indenting it. The wall of the cyst is remarkably thick.

Exploratory thoracotomy was performed after raising the factor VIII level over 60 percent. A hydatid cyst of 5 X 5 X 5 cm, with live scoleces on the right middle lobe, was found and the germinative membrane was explored. The inside of the cyst was washed with saline solution (10%), and the anterior wall of the cyst was then removed completely and the excision border was oversewn with an absorbable suture. Histological examination of the specimen also confirmed the diagnosis as a hydatid cyst. Albendazole treatment was then started at a dose of 10 mg/kg/day in two divided doses. On the 75th day of treatment, serum transaminases levels elevated mildly. The dose was tapered down by half and the drug was stopped at the third month when serum transaminases levels were normal. Chest x-ray taken 10 months after the operation was normal. The patient is fine and has been followed up at six month intervals.

Discussion

In the present case the most likely presumptive diagnosis was pulmonary hematoma. However, enlargement of the lesion instead of a reduction after appropriate fresh frozen plasma and factor VIII replacement suggested that the diagnosis of hematoma was unlikely. The symptom-free progress of the patient with such a large lesion, which was discovered by chance, is inconsistent with an abscess, or a primary or secondary malignant tumor. The radiographic appearance was not consistent with pulmonary tuberculosis or hamartoma. Although bronchogenic cysts and congenital arterio-venous fistulae could not be eliminated, as the symptoms may be delayed until adulthood[4], we did not stress them because of their rarity and focused instead on hydatid cyst.

The hydatid cyst disease is suspected with an history of residence in an endemic area. The patients are asymptomatic unless there is pressure of the enlarging cyst on the bronchial tree or pleura, secondary infection or cyst rupture. Our patient had no history of contact with possibly infected dogs; however, such history has been reported in only 29-48 percent of cases. Laboratory tests, other than radiographic examinations, are generally unreliable. The radiographic study, which is the main diagnostic tool, is accurate in 98-100 percent of cases, unless the cysts are ruptured or infected[5].

Ultrasonography distinguishes cystic lesions from solid tumors. If a desquamated germinative membrane, hydatid sand (debris) or daughter cyst (s) is seen, the diagnosis of hydatid cyst can be made confidently. These findings are pathognomonic for hydatid cyst and can be demonstrated by CT and MRI. It is striking that in ultrasonography, CT and MRI, some hydatid cysts, like in our case, might be seen as just simple cysts and nothing else. In such cases, the thickness of the wall, as in our case, can be suggestive of hydatid cyst. Thoracotomy was performed for both exploration and to prevent an anaphylactoid reaction and pneumothorax which could be fatal in the case of rupture of a possible hydatid cyst[6].

Mebendazole or albendazole is used in medical treatment of hydatid cysts[5]. In our patient, albendazole was started, as one-third of surgically treated patients reportedly had recurrences[7].

In conclusion, in hemophiliac patient such as ours, if a pulmonary lesion enlarges rapidly instead of getting smaller in spite of appropriate factor VIII replacement therapy, the diagnosis of hydatid cyst should be suspected and the appropriate therapy should be rapidly started. The diagnosis of hydatid cyst should especially be considered in areas with non-hygienic environmetal conditions.

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