Hydatid disease in a very unusual location: the adrenal gland. A case report

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Riassunto

Le cisti idatidee sono più frequentemente localizzate nel fegato e nel polmone (55-90%), la loro presenza nel surrene è considerata molto rara (0.5%). Abbiamo osservato un paziente affetto da cisti idatidea a localizzazione surrenalica, alla cui diagnosi siamo giunti in maniera casuale attraverso un'ecotomografia ed una tomografia assiale computerizzata. L'esperienza acquisita in Sicilia in virtù dell'endemia della malattia idatidea rende non necessaria l'esecuzione dei test seriologici, anche perchè spesso i loro risultati richiedono una certa attesa e sono costosi. Il paziente da noi trattato è stato molti anni prima operato per una cisti idatidea epatica. Quindi la localizzazione surrenalica potrebbe essere spiegata quale conseguenza di una disseminazione secondaria attraverso la circolazione sanguigna. La cisti si è sviluppata all'interno della ghiandola surrenalica ed ha determinato l'atrofia del tessuto ghiandolare. Il trattamento chirurgico ha richiesto la surrenalectomia dato che il surrene era interamente interessato dalla cisti.
Parole chiave: cisti idatidea surrenalica, surrene, surrenalectomia

Summary

Hydatid disease in a very unusual location: the adrenal gland. A case report. *A. Di Cataldo, G. Trombatore, R. Greco, R. Lanteri, G. Li Destri, A. Licata*

Hydatid cysts are most often located in the liver and lungs (55-90%), while their location in the adrenal gland is very rare (0.5%). We observed a patient with a hydatid cyst in the adrenal gland, the diagnosis of which was incidental during ultrasonography and computed tomography. The experience acquired in Sicily where hydatid disease is endemic makes serological tests unnecessary, also because they often require a lengthy waiting period and are expensive. The patient had undergone surgery for the treatment of hepatic hydatid cysts. The adrenal localization may be explained as a consequence of secondary dissemination via the blood stream. The cyst developed inside the gland and caused atrophy of the glandular tissue. The surgical treatment called for adrenalectomy as the adrenal gland was entirely occupied by the cyst.
Key words: hydatic cyst, adrenal gland, adrenalectomy
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Introduction

Hydatid disease is a parasitic disease which is endemic in Western Europe, South America, North Africa, Australia and the Middle East. In Italy it is endemic in the South, especially in Sicily and Sardinia.

Hydatid cysts are most often located in the liver (55-70%) and lungs (20%). If the parasite goes beyond the double hepatic and pulmonary filters, it spreads to other organs (5-10%). These locations have been named rare or peripheral locations.

In 1964 Grassi subdivided the peripheral locations of hydatid disease into 3 groups[1]: a) rare locations: kidney, spleen, bone, muscle, bone marrow; b) very rare locations: brain, thyroid, pancreas, diaphragm, salivary glands, subcutaneous tissue; c) extremely rare locations: thymus, lymphatic nodes, tonsils, hypophysis, adrenal gland.

Therefore, hydatid disease in the adrenal gland corresponds to one of the least frequent locations (about 0.5%)[2].

Case report

The patient was a 49-year-old male who, at the ages of 7 and 11 years underwent surgical operations to treat hepatic hydatid cysts. One month before hospitalization in our department he noticed dyspeptic symptoms characterized by nausea and abdominal pain. Physical examination was negative.

Ultrasonography of the liver was carried out in order to verify the presence of a relapsing hepatic hydatid cyst, as the patient had undergone surgery for the treatment of hydatid cysts of the liver.

A cystic lesion probably originating from the tail of the pancreas was discovered incidentally. Computed tomography confirmed the presence of the cyst, which measured about 10 cm in diameter, but established its location in the left adrenal gland with a clear boundary between the cyst and the upper pole of the kidney (Fig. 1).

The data obtained by ultrasound and CT and the presence in the patient's medical history of previous surgery for hydatid disease suggested a diagnosis of hydatid cyst of the left adrenal gland. Left adrenalectomy was performed as the adrenal gland was entirely occupied by the cyst. Histology confirmed the diagnosis of hydatid disease.

Discussion

Rare locations of hydatid disease prompt interesting reflections upon both the pathogenesis and diagnosis of the condition.

A peripheral location of this parasitic disease may be primary, secondary or tertiary. The mechanism of primary localization is unknown and several hypotheses have been expressed in order to justify the bypassing of the hepatic and pulmonary filters with consequent spread of the disease through the arterial circulation.

One theory supposes that the parasite has such a small diameter (25-35 microns) that it is able to pass through the hepatic sinusoids which have a diameter of between 10 and 100 microns. According to this theory the bypassing of the sinusoids is thought to depend on the biophysical and structural peculiarity of the parasite (variable dimensions in the different stages of its biological cycle, amoeboid movement) and a number functional factors such as pH, surface tension, or the presence of colloids which could modify the parasite for whatever vascular system[1]. However, as the size of the parasite probably does not allow it to bypass the hepatic capillary circulation, it may be possible for it to reach the peripheral organs, bypassing the hepatic sinusoids via arterio-venous anastomoses upstream of the sinusoidal circulation or through anastomoses between the portal and hepatic veins. The parasite could bypass the liver via a portocaval shunt, via Retzius' retroperitoneal system, the oesophagogastric system and/or haemorrhoidal veins.

According to these hypotheses the incidence of hydatid cysts in the lungs should be higher than in the liver. This is not the case, and therefore to justify the localization of hydatid cysts in peripheral organs without a pulmonary localization we would have to accept the possibility that the parasite may be able

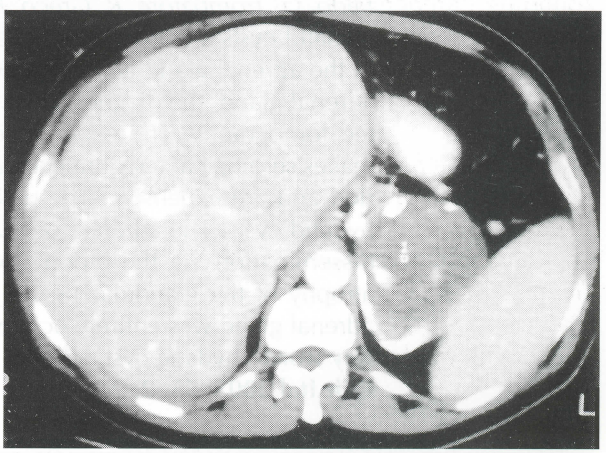

Fig. 1. Computed tomography. The left adrenal gland is entirely replaced by a cystic lesion.

to bypass the pulmonary capillary vessels as well as the hepatic capillary vessels.

Rare localizations of hydatid cysts could also appear as a result of secondary dissemination through the blood stream or by contiguity[3].

Heterotopic hydatid disease occurs as a consequence of a breach in the outer covering that releases the hydatid cyst which then continues to grow in the new location[1].

Spread via the lymphatic system bypasses the liver and carries the parasite directly to the right heart through the thoracic duct, subclavicular vein and superior vena cava. If there is communication between the auricles it can also bypass the pulmonary filter[1].

According to Aurousseau[4], after an abdominal trauma, even of only minor severity, very small fissures would allow some hydatid fluid to spread into the abdominal cavity or a blood vessel, thereby justifying the presence of peripheral cysts, which are multiple in 50-60% of cases.

Tertiary spread occurs when the hydatid fluid of secondary cysts spreads and the disease then appears in other organs.

In our opinion the most reliable hypothesis it is that which considers the larva as an "emulsoid" which adapts itself easily to the various calibres and bypasses all the filters, and even the pulmonary one. The final location, furthermore, might depend on the patient's immunological system, body temperature, peripheral oxygenation of the blood, perfusion of the organ and anatomical and functional tissue integrity.

In our patient the adrenal location was probably secondary. The hypothesis that some hydatid fluid spread during a previous surgical treatment of hepatic hydatid cysts is likely. In this case the cyst would have developed inside the gland, pressed the surrounding parenchyma and caused atrophy of the glandular tissue. Such cysts are usually asymptomatic as was the case in our patient. Lumbar pain, arterial hypertension and urinary disorders may be present.

Since only 6-7% of adrenal cysts are hydatid cysts, for the purposes of differential diagnosis we need to consider:

- adrenal pseudocysts (40%) which appear after haemorrhage in a normal or pathologic adrenal gland;
- cystic lymphangioma or angioma (30%);
- cystic adenoma (7%);
- congenital cysts (9%).

The differential diagnosis is based mainly on imaging studies in the form of ultrasound and CT which allow a clear-cut diagnosis of cystic lymphangioma, angioma and adenoma. It is probably less easy to differentiate between adrenal pseudocysts, congenital cysts and hydatid cysts.

In our opinion, however, when the differential diagnosis involves very rare pathological features or conditions, such as the above-mentioned adrenal diseases, another important factor which may serve as a guide to diagnosis is personal experience. In Sicily, hydatid disease is endemic and therefore the experience acquired by Sicilian surgeons and radiologists in the diagnosis and treatment of the disease is appreciable. In the past, when ultrasonography and CT were not available, serological tests were the only diagnostic tools available. The Casoni skin test (intradermoreaction of the hydatid antigen) and Ghedini's reaction (compliment fixation) were commonly performed, though the substantial number of false positive and false negative results made their use unsatisfactory. Later, better results were obtained with immunological tests, such as RAST (radio-allergosorbent-test) with assay of specific hydatid IgE, but on the basis of our personal experience, in a region like Sicily where hydatid disease is endemic and very frequent, the data provided by radiologists through ultrasonography and CT are sufficient for a correct diagnosis because they provide useful information about the nature of the cyst - presence of "septa", daughter cysts - and its location and connections with contiguous organs[2].

Nowadays we rarely perform serological tests which often require a long waiting period and are expensive. In our daily practice, unfortunately, the costs of our health-care activity are increasingly obliging us to curb public spending. Serological tests and especially RAST are useful in the follow-up of patients who have undergone surgery for echinococcosis. High levels of antibodies or antibody elevation months after surgery suggest a strong suspicion of an unrecognized cyst or a relapse[5].

The treatment of the adrenal hydatid cyst in our case required adrenalectomy as the cyst entirely occupied the gland so that conservative treatment was impossible. In our opinion a laparoscopic approach might be advisable but accidental tearing of the cyst could cause the disease to spread.

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