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Direttore Nicola Picardi

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A case report

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Giuseppe Salamone, Leo Licari, Brenda Randisi, Paola Di Carlo, Roberta Tutino, Nicolò Falco, Giuseppe Augello, Cristina Raspanti, Gianfranco Cocorullo, Gaspare Gulotta

Department of Surgical, Oncological and Oral Sciences University of Palermo
General Surgery and Emergency Policlinico Universitario "P. Giaccone", Palermo, Italy

A primary subcutaneous hydatid cyst in the thigh. A case report

AIM: *Hydatid disease is a parasitic disease considered endemic in many parts of the world such as South America, Middle East, Africa, Australia and the Mediterranean regions. Liver and lung hydatid disease accounts for 90% of all echinococcal cysts. Primary hydatid disease of subcutaneous sites is rare and the subcutaneous localization of a solitary hydatid cyst accounts for 1.6%. Not enough data exist for this localization, and only many heterogeneous data are described in order to define this rare condition.*
MATERIAL OF STUDY: *We present the case of a 68-year-old-woman affected by a mass in upper-medial side of her right thigh with a 12-year-growing history. Anamnestic data were accurately collected. Many different radiological and specific serum tests were performed in order to define the diagnosis. Surgical approach was decided in order to excide the mass, and a 6-months follow-up was performed.*
CONCLUSIONS: *Hydatid disease is common in endemic area but uncommon localizations, as in subcutaneous tissues, are a rare condition. Scientific Community lacks of complete and homogeneous data about the approach to this manifestation of the disease. Would be useful a complete review of the literature in order to plan guide-lines for the treatment of uncommon localization.*

KEY WORDS: Echinococcosis, Hydatid cyst, Subcutaneous localization

Introduction

Hydatid disease is a common cosmopolitan anthropozoonosis prevalent in endemic areas [1] caused by *Echinococcus* species. *E. granulosus* is the most common responsible of cystic formation; it is the least aggressive and the most treatable form [2-4]. Other three species could be responsible of infestation: *E. multilocularis*, uncommon and responsible for alveolar echinococcosis; *E. oligarthrus* and *E. vogeli*, very rare and responsible for multicystic echinococcosis [2-5].
Life cycle of the parasite does not include humans, even though they can occasionally ingest eggs becoming intermediate hosts [5]
The life cycle of *E. granulosus* is well known. Adult tapeworm lives in the small intestine of canids that are the definitive hosts. The eggs (gravid proglottids) are scattered through faeces. Herbivores (intermediate hosts) ingest the eggs during fodder and larvae (oncospheres) are released in the small intestine. Thanks to their con-

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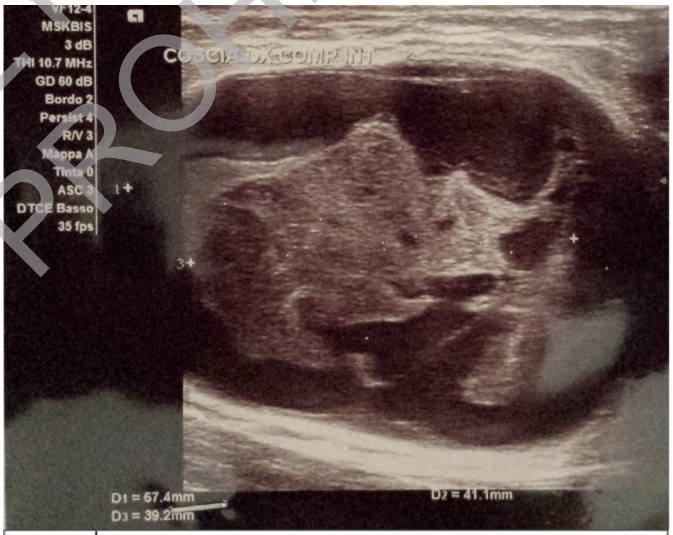
Correspondence to: Leo Licari, MD, Policlinico Universitario "P. Giaccone" Department of Surgical, Oncological and Science, University of Palermo, Via Liborio Giuffrè 5, 90127 Palermo, Italy (e-mail: lele.licari@gmail.com)

formation they penetrate the intestinal wall and once they reach the circulatory system they can potentially land in every part of the body, usually filtered and arrested in liver and lungs. At the moment of the filtering, when migration stops, larvae develop into a hydatid cyst (metacestode) that enlarge with time.
The capsule of the cyst consists of a pericyst, from the host tissue, which embodies the endocyst of metacestode origin. The outer wall of the endocyst is the chitinous layer; the inner wall is the germinative membrane, which proliferates. It produces a large number of small brood capsules filled with protoscolices (invaginated heads). If brood capsules leave the germinative layer (segmentation), they become daughter cysts which will float into the cyst. The life cycle perpetuate when dogs eat parasitized organs of herbivores [1-3,5].
Humans become accidentally intermediate hosts when ingest eggs; these remain viable for months and water contamination represents the major risk factor of infestation. They can also remain to the hand when a human pets an infested dog, or touches contaminated soil or vegetation. Uninfected pets may carry the eggs on their fur if they roll in parasitized faeces [2-5]. In order to justify uncommon localization other possible hypothesis of infestation were expressed such as airborne transmission and penetration of bronchial venules to enter heart and systemic circulation [6] or direct subcutaneous contamination through an injured skin.
Liver and lung are the most common organs involved in cystic formation and together account for 90% of all echinococcal cyst [1,7-9].
Subcutaneous localization is a rare event accounting for 1.6% of all locations [5]. The way of transmission and development in these cases is not clear even though many different hypotheses have been postulated [8,10,11].

Case Report

A 68-year-old-woman presented with a large soft tissue mass in the upper medial side of her right thigh. The mass existed since the last 12 years, gradually increased in size from few cm to approximately 6 x 4 cm altering the thigh profile, without signs of inflammation of overlying skin, anytime painless, neither fever nor related symptoms coexisted. On local examination it was possible to notice fixation of the mass at deep plans with mobility of the overlying skin, painless at palpation, definite and regular margins, tense-elastic thickness. Anamnestic investigation showed that the patient was living in endemic area for *Echinococcus spp* (Sicily), in rural house where lived dogs and where it was common use un-drinking-water; moreover she travelled in Italy, Russia, Germany and France during the last 12 years. She denied traumas or insects sting in the involved area; reported instead razor depilation. Initial diagnosis of lipoma was made but US revealed it to be a cystic mass of 6.5 x 4 cm in sub-cutaneous tissue, with a solid capsule and an inner structure with dishomogeneous echogenicity and iperechogen-septa; color-Doppler showed blood-flux in the capsule and in the septa; the suspect was of hydatid cyst (Fig. 1) In order to better define the lesion an MRI with contrast was performed three months later and it revealed an encapsulated cystic lesion, with multiple septa and a maximum diameter of at least 5.6 cm, lying in subcutaneous tissue over the gracilis and adductor magnus muscles without signs of infiltration. The capsule signal enhanced after gadolinium administration (Fig. 2).
The finding was suggestive of a hydatid cyst with inner daughter cysts. RBI fell within normal range. Parasitological serological test (Enzyme-Linked Immunosorbent Assay) was performed with border-line results (1/100). In order to investigate and to rule out lung and liver involvement, CT Chest-Abdomen with contrast were performed with negative results. Two-week neo-adjuvant anthelmintic chemotherapy was done with Albendazole 400 mg/die.



Gharbi 1981	WHO classification (cyst types)		
Type I	Univesicular anechoic cystic lesion with double line sign (CE1)	Active	CE 1
Type III	Multiseptated, "rosette-like"/ "honeycomb" cyst (CE2)	Active	CE 2
Type II	Cyst with detached membranes "water-lilly-sign" (CE3a) / Cyst with daughter vesicles in solid matrix (CE3b)	Transition	CE 3a, CE 3b
Type IV	Cyst with heterogenous content (hypoechoic/hyperechoic). No daughter vesicles (CE4)	Inactive	CE 4
Type V	= CE4 plus calcified wall (CE5)	Inactive	CE 5

Fig. 1: WHO US-classification of hydatid cyst is only for liver localization. It could be suggested to enlarge the use of US-classification also for uncommon localization. This could be CE3b.

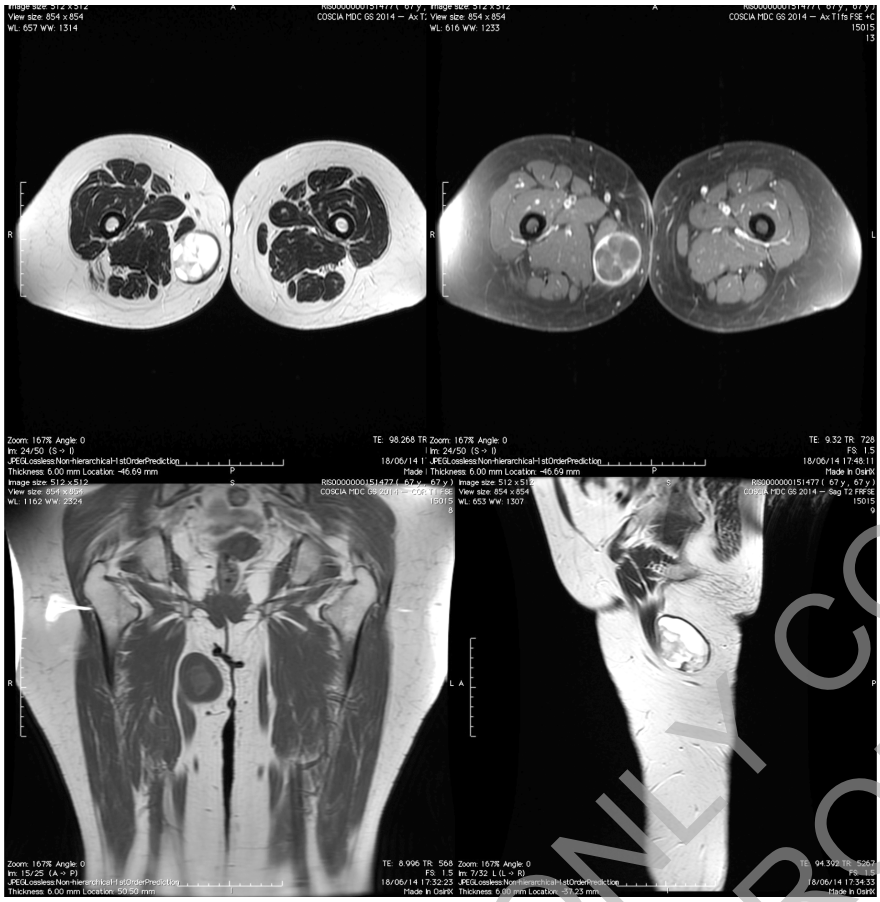


Fig. 2: MRI with contrast: the images show the encapsulated cystic lesion, with multiple septa and a maximum diameter of at least 5.6 cm, lying in subcutaneous tissue over the gracilis and adductor magnus muscles without signs of infiltration. The capsule signal enhanced after gadolinium administration.

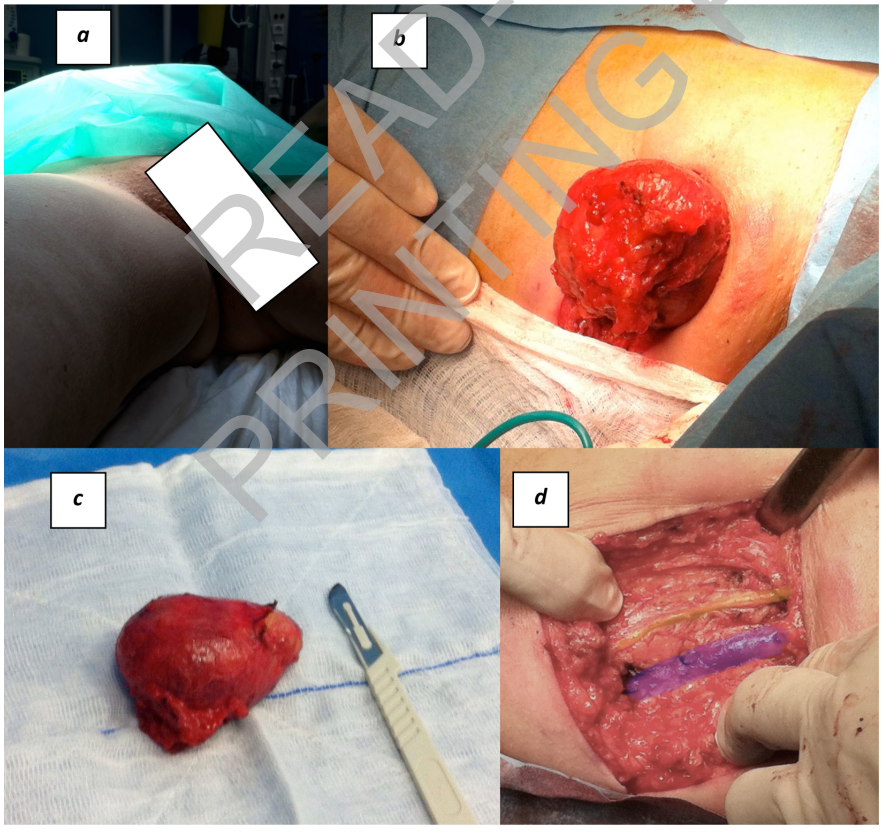


Fig. 3: Images of surgical operation performed: a) profile alteration of the thigh; b) c) isolation ed excision of the entire mass without spillage; d) cyst residual pouch after excision with digital reconstruction of upper thigh elements.

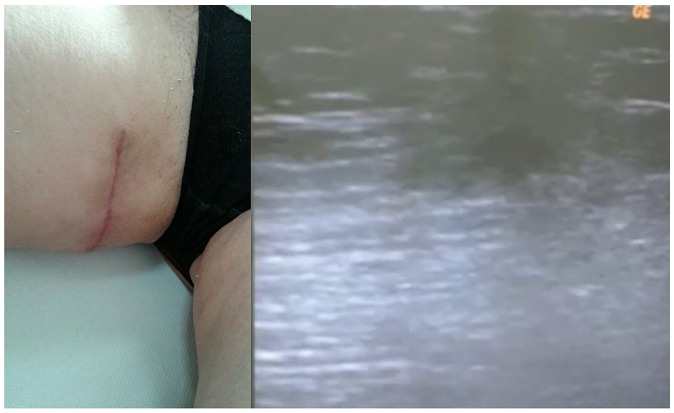


Fig. 4: Follow-up image and US Scan of the thigh at 6th month.

At the end a surgical operation was decided and done in local anaesthesia. During surgery, the wall of the cyst appeared to be delicate and soft, not very easy to dissect from adjacent structures, even though with accurate manoeuvre it was isolated and excised from the muscles plane without spillage. After the extraction, the cyst residual pouch was irrigated with hypertonic saline solution (Fig. 3).
The patient was dismissed from hospital the same day after surgical operation, with no related complications. A two weeks adjuvant therapy with Albendazole 400 mg/die was performed.
Histopathology described a chitinous layer surrounding a multi-loculated lesion and confirmed the diagnosis of cystic echinococcosis due to *E. granulosis.*
Follow-up was performed weekly in 1st month and monthly for 6 months; no recurrences have been observed nether clinically nor radiologically; six month after surgery an US scan of the thigh was performed documenting the absence of recurrent lesions. RBI fell within normal range (Fig. 4).

Discussion and conclusions

Primary hydatid disease of subcutaneous region is a rare condition as reported in literature [5,7], with no specific symptoms. Differential diagnosis includes lipoma, hematoma, abscess, soft tissue tumour, and hydatid cyst have to be suspected especially in endemic areas.
Echinococcus spp. have poor tropism for subcutaneous region, and it not represents a favourable environment for larval growth such as muscle and other tissues [12-14]. Many theories were advanced to explain this phenomenon. Kayaalp et al. [8] exclude the theory of direct contact for different reasons like the statistic evidence that uncommon localizations of pathology do not coincide with more exposed areas to traumas for example, nor for the fact that gastro-intestinal environment is required to transform eggs into larvae. It is possible instead that the parasite flee the portal-liver tough filter by using lymphatic or venous shunts directly to systemic circulation, as postulated by Abhishek et al [2,10].
A systematic review of the literature could give the possibility to obtain guide-lines on treatment of a rare pathology that could have dangerous consequences if not well defined. No homogeneous data exist in literature in order to have a clear pre-operative diagnosis (radiologic and serum tests), on surgical technique, neoadjuvant/adjuvant therapy, and follow-up time.

Riassunto

L'*Echinococcus spp.* rappresenta il movente eziopatogenetico della malattia idatidea, patologia a carattere endemico in diverse parti del mondo, con tipica localizzazione della cisti da echinococco, prodotto del ciclo vitale del parassita, in sede polmonare od epatica. Ciononostante le localizzazioni atipiche della malattia esistono e rappresentano circa il 1,6% dei casi, la più comune delle quali certamente quella sottocutanea. I dati a disposizione circa la diagnosi ed il trattamento delle rare forme a localizzazione atipica risultano disomogenei e pochi dirimenti circa uno standard diagnostico-terapeutico da adottare. Si è voluto presentare dunque il caso di una donna di 68 anni che lamentava la comparsa di una tumefazione al terzo prossimale della coscia dx che tendeva ad incrementarsi volumetricamente nel tempo. Dopo diverse indagini cliniche, oltre che una accurata anamnesi, il sospetto di cisti idatidea è stato confermato. La patologia è stata trattata secondo uno schema terapeutico chirurgico e farmacologico ben dettagliato. La raccolta di questi dati si prefigge lo scopo di pianificare, in accordo con la letteratura scientifica e la review dei case-report esistenti, linee guida nel percorso assistenziale di questa rara manifestazione di una comune malattia.

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