Case Report

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Generalised recurrent hydatidosis with unusual subcutaneous polycystic localisation

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ABSTRACT

Introduction: Echinococcosis, also known as Hydatidosis, is a parasitic infection caused by Echinococcus species. Although the infection has a worldwide distribution with higher prevalence in certain regions, the incidence in western countries has been declining. As a result, both diagnosis and treatment have become a great challenge for health systems and medical practitioners.

Case Presentation: Such an example is the rare case of an 82-year-old woman first diagnosed with echinococcosis 50 years ago. Even though she had multiple recurrences and multiple surgeries in the past, she presented with multiple pelvic and intra abdominal cysts, one of which had ruptured and fistulised to the skin of the anterior abdominal wall. Due to the complexity of this case, the multidisciplinary team of our hospital suggested a four- week therapy with albendazole before surgical treatment in an outpatient setting.

Conclusion: Subcutaneous cysts of the anterior abdominal wall are rare findings, even in countries where the hydatid parasite is endemic. In complex cases of echinococcosis, the preoperative administration of albendazole aims to reduce the risk of recurrence and decrease the size of the cyst, facilitating surgical excision. Medical practitioners around the world must be fully aware of the unusual presentations of echinococcosis and the options that they have regarding the management of complicated patients.

KEY WORDS: *Echinococcosis, hydatidosis, albendazole, parasitic disease, echinococcus cysts, abdominal mass*

INTRODUCTION

Echinococcosis is a parasitic infection affecting both humans and animals, resulting from the infiltration of larval forms of various tapeworm species belonging to the Echinococcus subspecies. In Greece the infection is caused by *Echinococcus granulosus* most of the times [1].The disease is also called Hydatidosis because of the growth of hydatid cysts (metacestode) in internal organs (liver, lungs) of intermediate hosts, including humans [2]. The definitive hosts of the cestode are carnivores such as dogs. The infection is caused by humans and other hosts ingesting eggs or gravid proglottids that are excreted in the definitive host's feces. There is a worldwide distribution of the infection with higher prevalence in the Mediterranean, Russia, China, North and East Africa, Bulgaria, Australia, and South America [1]. In Greece the annual incidence is 0,13/100.000 (total number of cases 2004-2022: n=255), with yearly fluctuation and a steady decline since 2008,

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until 2022 when only four cases were reported [3]. Hydatid cysts can be found anywhere in the body either primarily or by secondary spread. The organ that is affected in over two-thirds of patients is the liver and the second most common localization are the lungs, followed by the brain, muscle, kidneys, bone, pancreas and heart [4]. In 85 % of the patients with E. granulosus, only one organ is involved and in most of these cases a single cyst is found. The objective of this case presentation is to highlight a rare instance of subcutaneous hydatidosis, an uncommon manifestation in an already infrequent condition.

CASE PRESENTATION

We report the case of an 82- year-old woman presented in the outpatient department with a palpable abdominal mass. The physical examination revealed effluence of pus and translucent sac-like cysts from the anterior abdominal wall, macroscopically resembling hydatid cysts (Figure 1). The patient was febrile but hemodynamically stable and had a known background medical history of atrial fibrillation on medication.

The patient was initially diagnosed with echinococcosis at the age of 32 years due to a palpable mass in the right hypochondrium. The patient was born and raised in an urban suburb of Athens working in non-agricultural posts and had no regular contact with animals. No medical history of hydatidosis in other family members was recorded.

Four years later, she underwent an urgent operation due to acute abdominal pain after a fall. A midline exploratory laparotomy was performed and multiple ruptured echinococcus cysts were resected. Seven years later, the patient was reoperated due to a subhepatic palpable abdominal recurrence. A segmental hepatic resection, cholecystectomy and excision of hydatid cysts were performed. Likewise 15 and 33 years following the index operation, the subhepatic echinococcosis relapsed and the patient underwent exploratory laparotomies with rooftop excision of hydatid cysts. After the last surgery in 2007, she had a three month course of post-operative treatment with albendazole although follow-up was regular thereafter.

During her presentation in our department, ultrasonographic examination showed a calcified lesion in the right hepatic lobe, multiple pelvic and intra abdominal cysts, one of which had ruptured and fistulised to the skin.

The patient was admitted to the surgical department of our hospital for further medical investigation. Immediate blood and wound cultures were taken while a sample of cysts was sent for parasitological examination – echinococcus granulosus was identified while she also had elevated titer of echinococcus antibodies.

The patient underwent a full body computed- tomography (CT) scan without intravenous contrast due to a severe allergic reaction she experienced in the past. The Brain and Chest CT showed no hard evidence of disease while the Abdominal CT showed multiple locations of echinococcus cysts in the anterior abdominal wall, intra abdominally and in the pelvis (Figures 2-4).

The multidisciplinary team, consisting of surgeons, radiologists and infectious disease specialists, suggested a four- week therapy with albendazole before surgical treatment in an outpatient setting. The patient was discharged seven days later, afebrile in a good general condition. After the completion of one month therapy with albendazole in the outpatient setting, the patient was readmitted for

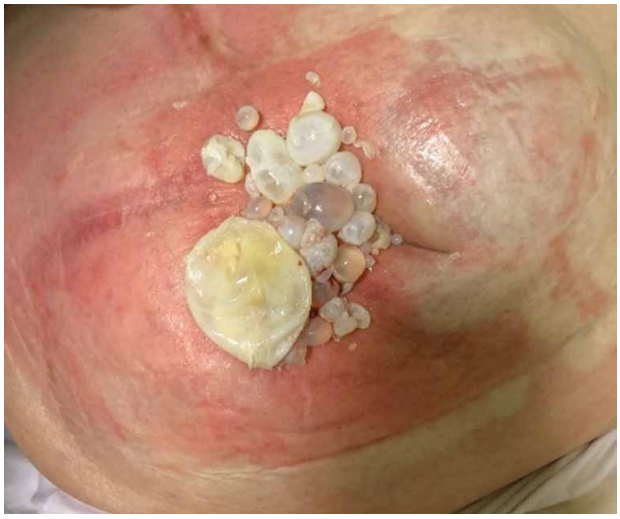

FIGURE 1. The patients presented with a palpable abdominal mass and macroscopic effluence of hydatid cysts.

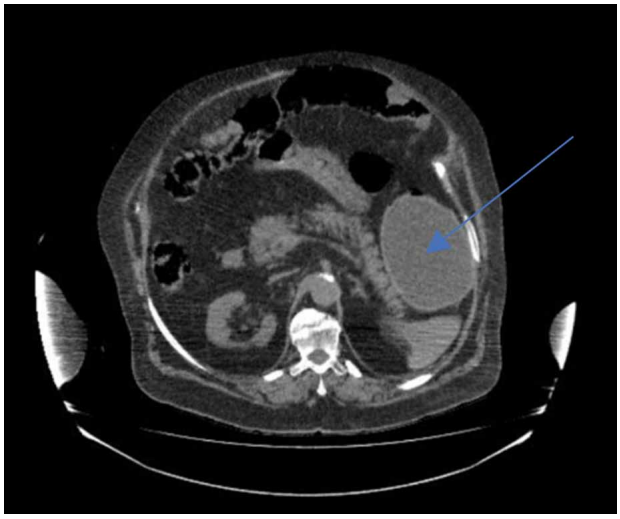

FIGURE 2. Computed Tomography, left subdiaphragmatic echinococcus cyst (blue arrow).

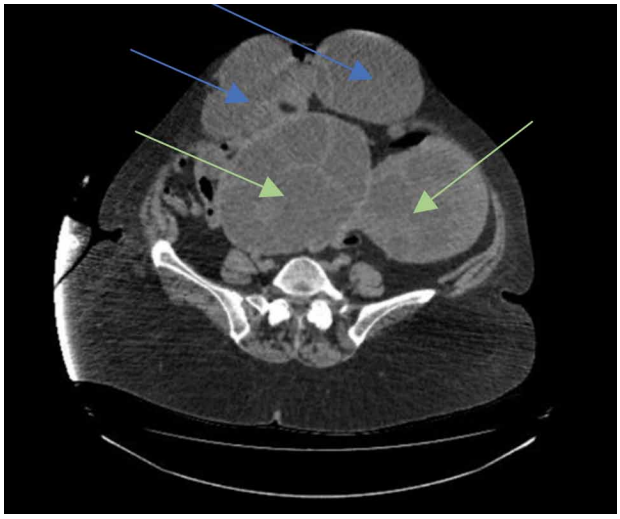

FIGURE 3. Computed Tomography, multiple fistulised subcutaneous and intrapelvic echinococcus cysts, green arrows: intrapelvic cysts, blue arrows: subcutaneous cysts.

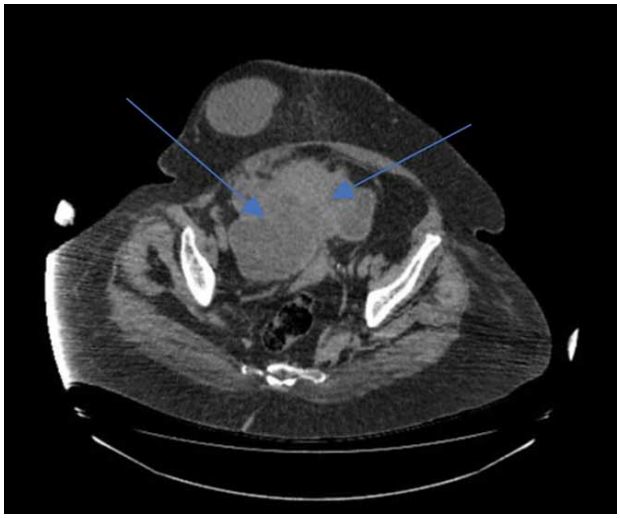

FIGURE 4. Computed Tomography, multiple intrapelvic echinococcus cysts (blue arrows).

further surgical intervention. A midline laparotomy was performed with extensive resection of the cysts from the subcutaneous space of the anterior abdominal wall. Multiple echinococcus cysts were identified within the pelvis in direct contact to the internal genitalia, whereas the presence of a solitary subdiaphragmatic cyst was also confirmed. Resection of three pelvic cysts, total hysterectomy and sigmoidectomy with primary anastomosis were performed, while unroofing and rinsing with hypertonic solution was chosen for the remaining cysts.

After surgery, the patient was admitted to ICU for four days before returning to the surgical ward where she had a stable postoperative course. She was finally discharged from the hospital on the 17th postoperative day.

Regarding the postoperative complications, we used the Clavien-Dindo classification in which the patient ranked 3b. The patient received a course of antibiotic treatment for positive blood cultures to Klebsiella pneumoniae carbapenemase (KPC) and Enterococcus faecalis. In addition, she developed an extensive skin necrosis which was treated with vacuum-assisted closure after wound debridement. For the moment, the patients have regular radiological and serological screenings with no warning signs of recurrence.

DISCUSSION

The patient presented with multiple hydatid cysts in the subcutaneous tissue of the anterior abdominal wall, in close proximity to intra-abdominal organs and the pelvis. Subcutaneous cysts of the anterior abdominal wall are extremely rare findings, even in countries where the hydatid parasite is endemic [2,5,6]. Primarily, subcutaneous cysts have been described mainly through lymphatic spread. However, in this particular case, the subcutaneous cysts are likely secondary to local dissemination from the previous four surgical interventions the patient underwent to address the hydatid disease.

Hydatid cysts are rarely localised in the abdominal wall, posing diagnostic challenges due to their atypical location and nonspecific symptoms. Radiological imaging and serological findings are essential in diagnosing hydatidosis, as they help differentiate hydatid cysts from other potential conditions. Prousalidis et al. reported that the frequency of extrahepatic and extrapulmonary hydatid cysts was 9%, based on a large series from Greece [5]. However, other studies report the frequency of subcutaneous tissue involvement to be approximately 2% [2,6].

The patient received preoperative albendazole for one month. Preoperative administration of albendazole aims to reduce the risk of recurrence and decrease the size of the cyst, facilitating surgical excision. The optimal duration of treatment is not well established. The World Health Organization (WHO) recommends that patients take albendazole or mebendazole for 4 to 30 days before surgery [7]. In our case, albendazole therapy for one month was chosen due to the patient's complicated medical history.

The management of echinococcal disease, according to both the Centers for Disease Control and Prevention (CDC) [8] and the Greek National Public Health Organization [3], includes a plethora of options, among which are the watch-and-wait strategy, medical therapy, surgical treatment, and the PAIR (percutaneous aspiration, injection of

chemicals, and reaspiration) procedure [3,8]. Watch-and-wait is recommended for uncomplicated echinococcal cysts in systemically well patients, which are routinely monitored with ultrasound. Surgical intervention should be reserved for cases with complicated cysts, such as superficial cysts at risk of traumatic or spontaneous rupture, infected cysts, those compressing adjacent organs, and cysts communicating with the biliary tree.

Medical therapy is the treatment of choice for small cysts (<5 cm), multiple cysts, and patients deemed to be poor surgical candidates. Furthermore, postoperatively, for the prevention of recurrences, patients are advised to undergo additional systemic treatment. Albendazole is the medication of choice at a dose of 400 mg/day orally for one-six months according to the CDC, and three-six months according to the Greek National Public Health Organisation, while mebendazole is an alternative. The PAIR procedure is a minimally invasive approach preferred in cases of multiple recurrences or failure of medical therapy.

After appropriate treatment, the prognosis for the patient is generally good, though it decreases in cases of cysts in difficult-to-reach locations such as the heart and spinal column. However, cardiac cystic echinococcosis is not associated with a higher mortality rate when there is timely diagnosis and appropriate surgical and non-surgical management [9]. Several factors raise the risk of recurrence. Two major factors are when the cysts are larger than 7 cm and when the primary location of the cysts is neither the liver nor the lungs. The surgical technique is also important since leaving viable material behind during conservative operative interventions, spillage during surgical removal, and the surgeon's experience and practice are associated with a higher risk of recurrence [10]. Castillo et al. reported that early diagnosis is important since it reduces high postoperative morbidity (POM) and mortality, which have been estimated at 24.4% and 2.0%, respectively [11]. Following treatment, measuring white blood cell and aminotransferase levels every 3-6 months initially and then annually can help identify recurrence. Surveillance with routine imaging is strongly advised. Serological methods do not allow for optimal follow-up because antibody titers may persist for years after the removal of the cyst.

CONCLUSION

Clinicians and medical researchers, even in areas where echinococcosis is not epidemic, should be aware of the public health importance of this disease. Subcutaneous hydatid cyst should be considered in the differential diagnosis even though this is a rare localization. Close precautions should be taken preoperatively and intraoperatively in order to prevent spillage and minimise the possibility of recurrence. In conclusion, despite the fact that the impact of this infection is waning across Greece and generally across Europe, when a subcutaneous mass is found in a patient, especially on a background of hydatidosis, hydatid cysts should be considered and treated within a multidisciplinary team.

REFERENCES

1. Tamarozzi F, Legnardi M, Fittipaldo A, Drigo M, Cassini R. Epidemiological distribution of Echinococcus granulosus s.l. infection in human and domestic animal hosts in European Mediterranean and Balkan countries: A systematic review. PLoS Negl Trop Dis [Internet]. 2020 Aug;14(8):e0008519. Available from: https://pubmed.ncbi.nlm.nih.gov/32776936/ Doi: 10.1371/journal.pntd.0008519.
2. Chevalier X, Rhamouni A, Bretagne S, Martigny J, Larget-Piet B. Hydatid cyst of the subcutaneous tissue without other involvement: MR imaging features. AJR Am J Roentgenol. 1994 Sep;163(3):645-6.
3. Εθνικός Οργανισμός Δημόσιας Υγείας. Εχινοκοκκίαση - Εθνικός Οργανισμός Δημόσιας υγείας. [Internet]. Updated 2023 Aug 18. Available from: https://eody.gov.gr/disease/echinokokkiasi/
4. Sayek I, Tirnaksiz MB, Dogan R. Cystic hydatid disease: current trends in diagnosis and management. Surg Today. 2004;34(12):987-96.
5. Prousalidis J, Tzardinoglou K, Sgouradis L, Katsohis C, Aletras H. Uncommon sites of hydatid disease. World J Surg. 1998 Jan;22(1):17-22. Doi: 10.1007/s002689900343.
6. Orhan Z, Kara H, Tuzuner T, Sencan I, Alper M. Primary subcutaneous cyst hydatic disease in proximal thigh: An unusual localisation: A case report. BMC Musculoskelet Disord [Internet]. 2003 Nov;4:(25). Available from: https://doi.org/10.1186/1471-2474-4-25
7. World Health Organization. Echinococcosis. [Internet]. Updated 2019 Dec 2. Available from: https://www.who.int/health-topics/echinococcosis#tab=tab_1
8. Centers for Disease Control and Prevention. Echinococcosis - Resources for Health Professionals. [Internet].
9. Banisefid E, Baghernezhad K, Beheshti R, Hamzehzadeh S, Nemati S, Samadifar Z, et al. Cardiac hydatid disease; A systematic review. BMC Infect Dis. 2023 Sep;23(1):600. Doi:10.1186/s12879-023-08576-3
10. Velasco-Tirado V, Romero-Alegría Á, Belhassen-García M, Alonso-Sardón M, Esteban-Velasco C, López-Bernús A, et al. Recurrence of cystic echinococcosis in an endemic area: A retrospective study. BMC Infect Dis. 2017;17:455. Doi:10.1186/s12879-017-2556-9
11. Castillo S, Manterola C, Grande L, Rojas C. Infected hepatic echinococcosis. Clinical, therapeutic, and prognostic aspects. A systematic review. Ann Hepatol. 2021 May-Jun;22:100237. Doi:10.1016/j.aohep.2020.07.009