

An Uncommon Cause of Soft Tissue Mass of the Extremities, Report of 2 Cases and Review of Literature of Cystic Echinococcosis

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Abstract

Cystic echinococcosis or hydatid disease is a rare zoonosis caused by *Echinococcus granulosus*. The most frequently affected organs are the lung and liver, whereas solitary involvement of muscles and skin (2,2%) or bone (0,6%) is very uncommon. We report two cases of solitary hydatid disease of the extremities mimicking soft tissue sarcoma of the gluteal region in case 1 and infection of the lateral thigh involving the greater trochanter in case 2. Both patients were treated successfully by surgical intervention and perioperative anthelmintic drugs. Solitary hydatid disease adds to the broad spectrum of differential diagnoses in examining soft tissue masses.

Keywords: Solitary hydatid disease soft tissue; Osseous solitary hydatid disease; Cystic echinococcosis

Case Presentation

History and physical examination

Case 1: A 56 year old woman presented with a slow growing mass of the left gluteal region. The lesion had been noticed 12 months ago but caused pain only recently. She had no history of trauma, infection or previous surgery. The patient had been living in Austria for 25 years but originated from rural Turkey. Physical Examination revealed a dense lesion adherent to the pelvis not involving the skin.

Laboratory tests were without pathological findings.

Case 2: A 44 year old man presented with a painful soft tissue lump of the left lateral thigh present for months. The overlying skin had signs of inflammation (erythema, fluctuation, and induration) imminent to spontaneous perforation (Figure 1). Clinically the

process didn't involve the proximal femur region except for tenderness on palpation of the greater trochanter. The patient was afebrile and his laboratory workup was unremarkable. Medical history of the patient who had immigrated to Austria from Eastern Turkey 12 years ago was uneventful except for pharyngitis 3 months after onset of symptoms.

Imaging

Case 1

The AP radiograph of the pelvis showed an unspecific soft tissue mass in the dorsal aspect of buttocks without signs of calcification or osteodestruction. MRI revealed a lobulated mass measuring 21 × 12 × 7 cm affecting the gluteal muscles and the subcutaneous fat with low intensity on T1 weighted images, high intensity on T2 and peripheral enhancement of contrast agent (Figure 2). Combined with the clinical presentation soft tissue sarcoma was expected.

Case 2

The AP and lateral radiograph of the left hip joint revealed well defined osteolysis of the greater trochanter partially with a sclerotic rim. The cortical bone was thinned out but intact and no periosteal reaction was present (Figure 3). MRI showed a large subfascial soft tissue mass of the proximal lateral thigh with extension into the greater trochanter. The diameter measured 20 cm; intensity on T1 weighted images was low with high intensity on T2 and marginal contrast medium enhancement (Figure 4). Linked to the physical examination a bacterial infection seemed to be likely.

Histology

Case 1

The patient underwent an open incisional biopsy (Figure 5).



Figure 1: Clinical presentation of case 2. Around the trochanter region exists a not well defined inflamed area.

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Figure 2: The fat-suppressed MRI showed a multi-cystic formation in the gluteus maximus muscle (PD-weighted SPIR).



Figure 3: AP plain radiograph showing an osteolysis involving the greater trochanter.

Hematoxylin and eosin staining of the specimen showed components of the cyst's wall as well as scolices, the heads of tapeworms (Figure 6).

By means of DNA analysis the parasite could be identified as *Echinococcus granulosus* sheep strain genotype G1. Immunoglobulin E was elevated up to 455 IU/ml (reference <100 IU/ml). Because the

ELISA was positive for two types of Echinococcus – *Echinococcus granulosus* and *E. multilocularis* – a Western blot was accompanied to confirm the diagnosis.

Case 2

An aspirate culture of the abscess-like formation was negative. However a chronic bacterial infection was suspected and therefore



Figure 4: Contrast enhanced MRI visualized the subcutaneous abscess-like formation in the thigh (fat-suppressed T1 weighted TSE).



Figure 5: Intraoperative situs of the incisional biopsy. Direct under the skin a cyst formation was present.



Figure 6: Hematoxylin and eosin staining of scolices (arrow) and the membrane of the cyst (asterisk).

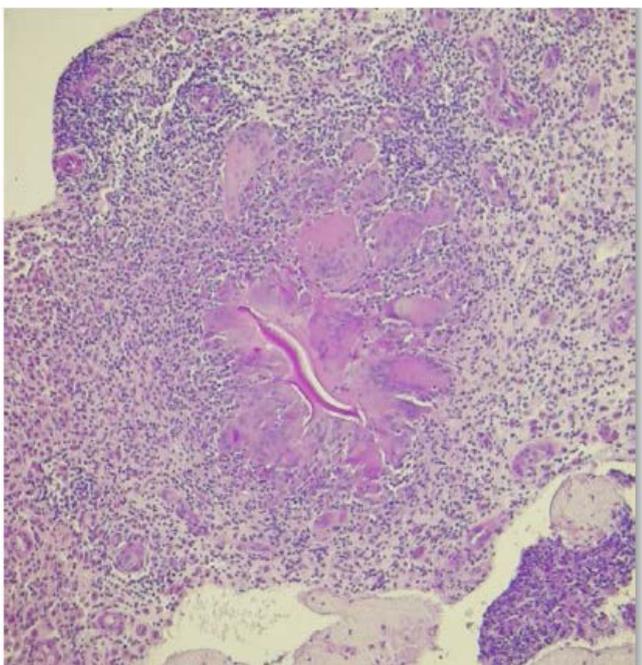


Figure 7: Hematoxylin and eosin staining of the inflammatory reaction caused by the parasite's membrane.

resection of the lesion and high speed burring of the affected bone were performed. The PCR of the specimen revealed *Echinococcus granulosus* with the sheep strain genotype G1. The staining with hematoxylin and eosin found a foreign body reaction surrounding the parasite's membrane (Figure 7). Immunodiagnostic assays consisting of ELISA and Western blot were positive for *Echinococcus granulosus*.

Treatment

Case 1

Albendazole was started immediately after receiving the histological

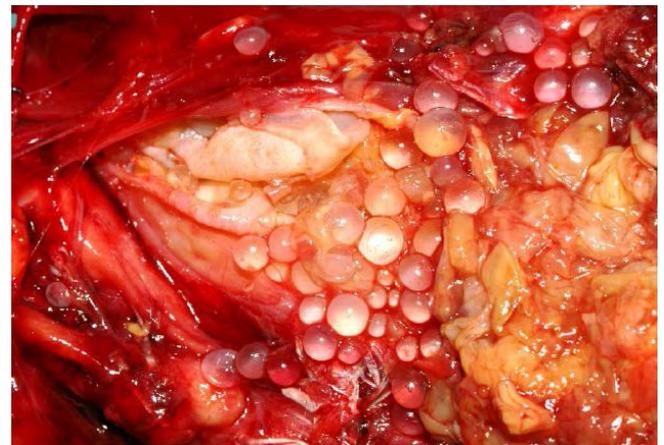


Figure 8: Resected and sliced hydatid cyst formation. Notice the innumerable daughter cysts.

results. Two weeks after the biopsy the marginal resection of the cyst was performed (Figure 8). The oral anthelmintic medication was continued for duration of three months.

Case 2

The patient received albendazole postoperatively, because of distinct side effects medication was switched to mebendazole. In both cases staging with CT scan of chest and abdomen revealed no further lesions. The last follow up (case 1: 7 years and 10 months; case 2: 17 months after surgery) including MRI scan and serological tests showed no evidence of recurrence.

Background, Discussion and Conclusion

Echinococcus granulosus is globally distributed, high parasite prevalence are found in Eurasia, Africa, Australia and South America (prevalence of 2,1% to >3%) [1]. This specimen contributes to more than 95% of the estimated 2-3 million human cases of hydatid disease worldwide. Its importance in public health and economic is evident [2]. Solitary hydatid disease initially was not considered in the differential diagnosis primarily because of its sporadic occurrence in Austria (2009-2014: 7,2 reported cases per year) [3]. Most infections are diagnosed in immigrants from countries where this zoonosis is endemic such as the Mediterranean region [4,5].

Cystic echinococcosis or hydatid disease is by *Echinococcus granulosus*. Its natural cycle is as a cyst in sheep and as a tapeworm in dogs [1,6]. Humans become infected by ingesting food or water contaminated with dog's faeces containing the eggs of the parasite or when they handle infected dogs [1]. So far 9 genotypes are described, most human cases of cystic echinococcosis are caused by the sheep strain (G1) [1,7,8]. Usually the infection is asymptomatic for a long period of time. After ingesting the parasite's eggs, they are spread via circulation of the blood. By passing the portal vein the specimen predominantly settles down in liver (65%) and lung (25%), other anatomic sites are rare. In the majority (40-80%) a single-organ involvement is described [1,9]. At the site of infection the development of a fluid-filled cyst called hydatid can be observed. It consists of three layers: the fibrocollagenous pericyst, the laminated acellular eosinophilic ectocyst and the inner germinal layer called endocyst [10]. Typically a univesicular cyst is formed, a multivesicular mass results from a partial rupture of the mother cyst causing smaller daughter cysts [1,5].

Both of our cases grew up in Turkey and migrated to Austria in adult age. Since the incubation period is months to years and the cyst's growth are slow and indolent, we suppose that both patients got infected in their country of origin [11].

The two cases are special because of the isolated musculoskeletal affection. In literature a solitary occurrence in bone is reported in 0,5-4% and in soft tissue only in 0,2-2,2% of cases [1,12-14]. Osseous hydatid disease mostly occurs in the spine (35%-60%) and the pelvis (14%-21%) followed by the femur (16%-19%). Tibia, humerus and ribs are rarely affected [5,7,15]. Vecchio performed a review of literature concerning the distribution of solitary subcutaneous hydatid cysts. The analysis of 23 patients showed that 35% of the lesions are located in the thigh, the lower extremities accounting for 61% of all cases [16].

Due to the latent infection and the slow growth of the hydatid cyst, the disease often remains undetected for years. In general symptoms arise from the expansive effect of the lesion: number, size and developmental status of the cyst (active or inactive) and anatomical site being the critical factors [1]. Musculoskeletal affection usually comes along with a sometimes painful mass, osseous involvement can result in a pathological fracture or -in the case of a spinal lesion - paraplegia and scoliosis [6,14]. The clinical presentation of our 2 cases was quite unspecific. Both patients reported a slow growing mass initially causing no pain. In case 1 a malignant tumour could not be ruled out by preoperative imaging, for which reason a biopsy was performed. In case 2 the clinical presentation favoured a chronic bacterial infection despite a negative aspirate culture.

Especially in countries with a low incidence of hydatid disease solitary musculoskeletal echinococcosis seldom is a first line differential diagnosis of a soft tissue mass. Therefore – although a variety of diagnostic procedures are available – such cases of cystic echinococcosis most frequently are diagnosed in the wake of surgery [13]. Plain radiographs usually show no pathognomonic calcifications as in cysticercosis [7,14]. Ultrasound and MRI allow visualizing the cystic nature of the disease and are valuable tools for planning surgical resection [15].

Eosinophilia may be the finding on haematological investigation but it is seen in only one fourth of the cases with musculoskeletal echinococcosis [12,15]. Immunodiagnostic tests like the ELISA isolate the patient's antibodies against *Echinococcus granulosus* from the serum [17]. The sensitivity of this test is about 95% but the specificity is low [1]. Other available tests are the Western Blot and the IHA (indirect hemagglutination test) [1,15,17,18]. However, limitations of these methods are sometimes based on the low induction of antibody production in the affected bone [1]. Most cases of cystic echinococcosis are diagnosed intraoperatively by detecting the cyst itself or after histological analysis [13,15,19]. The staining with hematoxylin and eosin visualizes parts of the cyst or the scolices [7]. Another technique to obtain material for the histological analysis is a needle aspiration of the lesion [1]. The risk of rupture of the cyst with spreading of the parasite and a potential anaphylactic shock reaction has to be kept in mind [12]. Several therapeutic strategies are known. In musculoskeletal solitary hydatid disease total excision is the treatment of choice [1,12,16]. Meticulous surgery in accordance to the principles of musculoskeletal tumor surgery is mandatory to avoid residual infectious tissue and spreading of the parasite [4]. Surgery should be accompanied with the oral administration of an anthelmintic drug aiming at the reduction of the recurrence rate [6,11,14]. Chemotherapy with benzimidazoles (albendazole, mebendazole) is recommended. They disrupt the reproduction of the parasite by inhibiting the maturing of fertile eggs.

Furthermore starving the parasite is achieved by interfering its glucose metabolism. The major side effect is an elevation of the liver enzymes. In 10-30% a complete remission and in 50-70% at least a reduction in size or degeneration of the cyst is observed. In 20-30% no effect is described. Nevertheless the effect of these substances is limited by the difficulty to achieve therapeutic tissue levels in bone [1]. If the complete removal of the unsevered cyst is unlikely, albendazole should be administered already before surgery. A nonsurgical monotherapy with albendazole can be considered if the patient is symptomatic, refuses operation or in the case of a multilocular, surgically incurable disease [12]. If the patient is asymptomatic and the cyst is inactive, a watch-and-wait strategy is arguable [1]. Another available option usually practiced in the liver is the PAIR-technique where puncture, aspiration, injection of 95% alcohol and re-aspiration devastate the cyst [1,12].

The follow up should include physical examination, serologic testing and imaging. There is no need for testing relatives because a transmission in humans is impossible. Persons with a single exposure should be tested serologically 1, 6, 12 and 24 months after the suspected contact and in case of recurrent exposure twice a year [1]. Solitary hydatid disease adds to the broad spectrum of differential diagnoses in examining soft tissue masses.

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