

Vertebral Hydatid Disease: White Cancer

Vertebral Kist Hidatik: Beyaz Kanser

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Abstract

Hydatid disease (HD), or echinococcosis, is a complex, chronic, neglected zoonotic disease caused by cestodes of the genus *Echinococcus*. The infection is found worldwide affects an estimated 1.2 million people. Vertebral involvement is rare, affecting less than 1% of all HD patients, even in endemic regions. Diagnosis and treatment of osseous HD is quite challenging; it is also called "white cancer." Despite surgical and medical treatment, recurrence ratio for HD is 40% to 100% within average of 30 months. Challenges arising from anatomical and physiological features in cycle of treatment and recurrence can be frustrating for clinicians. New methods, particularly medications that can better penetrate the central nervous system, are needed to prevent recurrence. Presently described is a case of vertebral HD penetrating the spinal canal.

Key words: Olağandışı lokalizasyonu, vertebra, omurga, kemik, hidatik kist.

Özet

Kist hidatik (KH), dünya çapında tahminen 1,2 milyon insanı etkileyen sestod *Echinococcus granulosus*'un neden olduğu kronik, kompleks ve ihmal edilen zoonotik bir enfeksiyondür. Vertebral tutulum endemik bölgelerde bile tüm KH hastaların %1'den daha azını etkileyen nadir bir durumdür. Tanı ve tedavisi oldukça zordür. Bu nedenle "Beyaz kanser" olarak adlandırılmıştır. Bütün cerrahi ve medikal tedavilere rağmen rekürrens oranı %40-100 arasında ve ortalama süresinde 30 aydır. Bölgenin anatomik ve fizyolojik özelliklerinden kaynaklanan güçlüklerde eklenince hastaların tedavileri daha da zorlaşmaktadır. Klinisyen tedavi ve hastalık arasında kısır bir döngünün içine girmektedir. Hem cerrahi hem de medikal açıdan rekürrensi engelleyecek; kısır döngüyü kırarak yeni yöntemler ve özellikle santral sinir sistemine daha iyi penetre olan ilaçlara ihtiyaç duyulmaktadır. Burada spinal kanala uzanım gösteren Vertebral KH olgusunu sunduk.

Anahtar Sözcükler: Unusual localization, vertebra, spine, bone, hydatid cyst.

Hydatid disease (HD) is a complex, chronic zoonotic infection. Geographically very widespread, it affects an estimated 1.2 million people, especially in rural communities due to inadequate environmental health, preventive medicine, and veterinary services (1). There are 4 forms of the disease caused by different species of parasite. The most frequently seen in humans is *E. granulosus*, which is found in domestic animals, and causes cystic echinococcosis. More rarely, infection with *E. multilocularis*, which is found in wildlife, causes alveolar echinococcosis. *E. vogeli* and *E. oligarthrus* are

underlying causes of polycystic and unicystic diseases, respectively, and are rarely seen in humans. The most frequent localizations are the liver and the lungs; however, HD may be primary or secondary anywhere in the body (2). Vertebral involvement is rare, affecting less than 1% of all HD patients, even in endemic regions (3). Diagnosis and treatment of osseous HD is quite challenging; severity and behavior led to it also being called "white cancer" (4). We hereby present a case of vertebral HD in the spinal canal.

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Submitted (Başvuru tarihi): 06.06.2016 Accepted (Kabul tarihi): 21.10.2016

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CASE

A 64-year-old female patient was admitted to our hospital 3 years ago with complaints of back pain and weakness and numbness in her hands and feet. No outstanding circumstances were specified in physical examination or lab tests at admission. Thorax computed tomography (CT) scan of the left lung upper lobe apicoposterior segment revealed smooth, contoured mass lesion 45 x 35 mm in size penetrating the spinal canal and causing damage at lateral fourth thoracic vertebra (T4) (Figure 1). Suspecting neurogenic tumor, histopathological confirmation and surgery were suggested; however, the patient refused any treatment or operation. The patient was admitted to emergency room of our hospital after complaints had increased over 3 years. It was reported that she was hardly walking or using her hands. At admission, she was generally in good condition, conscious, cooperative, and oriented. There was only minimal bilateral lower extremity weakness observed in physical examination. All lab test results were normal and indirect hemagglutination assay was positive for hydatid cyst. In thorax CT of the left lung upper lobe apicoposterior segment, a smooth contoured 45x35mm mass lesion was observed, penetrating into spinal canal, causing destruction at lateral T4 vertebra (Figure 2). Thoracic magnetic resonance screening revealed 40 x 37 x 35 mm, smooth, marginated, and macrolobulated mass lesion. It explicitly restricted the neural foramen at T4, extending to arcus aorta and neighboring the left lung hypointense at lateral T1 and hyperintense at T2, and having involvement of minimal homogenous contrast after intravenous gadolinium injection. The specified lesion was observed extending into the neural foramen, causing damage in vertebral body and posterior elements (Figure 3). Posterolateral thoracotomy was performed through fourth intercostal space. After accessing the thorax, adhesions were separated with obtuse and sharp dissections. Exploration yielded 4 x 3.5 x 3.5 cm, smooth, marginated, lobulated lesion, neighboring T4 lateral to the arcus aorta and the left lung. Lesion extending into spinal canal was separated from surrounding tissue and multiple germinative membranes were removed. Medulla spinalis was intact. No additional surgery was performed since the location was vertebral body. Thorax tissues were closed. No perioperative complications developed. Diagnosis was pathologically verified. The patient was discharged on ninth postoperative day without any complaint and treated with albendazole (10 mg/kg/d) for 6 months. Following operation, her complaints regressed and then were completely alleviated.

At 1-year follow-up, no recurrence or complications were observed.

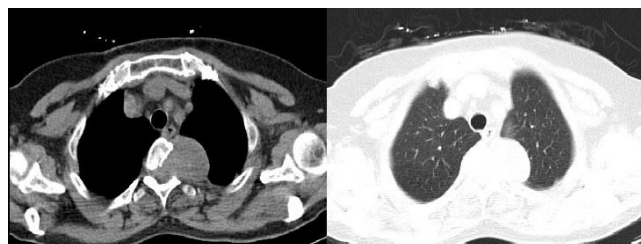


Figure 1: Thoracic computed tomography scans of the patient (at T4 level)

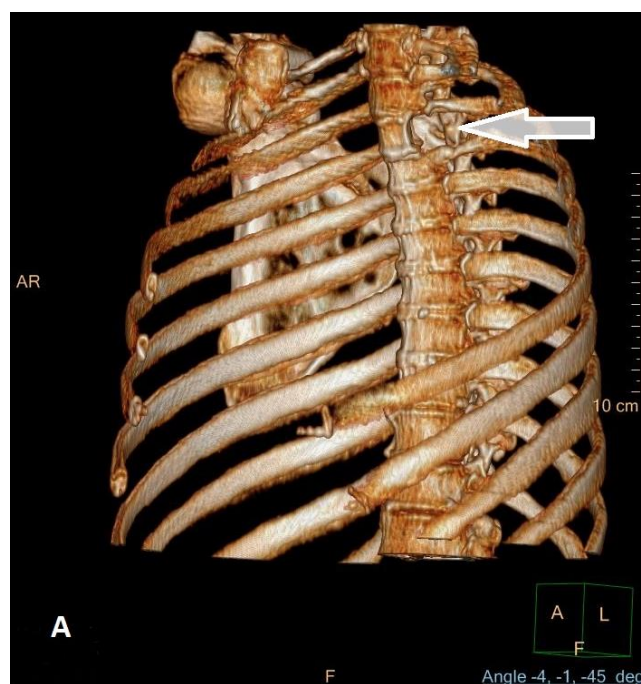


Figure 2: Three-dimensional computed tomography scan showing expanding cystic mass (white arrows) at T4 level and destruction of vertebral body

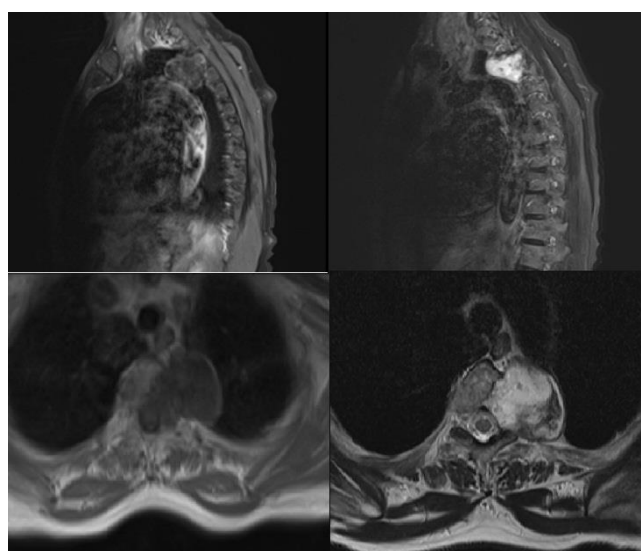


Figure 3: Magnetic resonance images of the patient (at T4 level)

DISCUSSION

Understanding of the physiopathology and lifecycle of cyst has developed over the last 2 centuries, although mankind has been aware of HD for 2000 years (5). New explorations continue to reveal more about HD. In addition to the 4 well-known types, recently, *Echinococcus shiquicus* and *Echinococcus felidis* species were identified in small mammals in the Tibetan plateau and in lions in Africa, respectively. The zoonotic potential and spread have been investigated (6). Although it is primeval, HD is still an important human health problem, particularly in endemic regions of the Mediterranean basin, Eastern Europe, Middle East, Far East, Central and South America, as well as former Soviet republics. It seems it will continue to exist into the future. Humans accidentally become host outside the normal life cycle of the parasite by swallowing cystic form. Cysts in the duodenum may spread anywhere in the body hematogenously or lymphogenously via the vena porta (2,6). As natural filters, the liver and lungs are the most frequent locations for HD to develop, with 90% of cases found in these organs. The remaining 10% localize in soft tissue organs, so-called "rare locations," such as the spleen, pancreas, gall bladder, adrenal gland, pelvis, seminal vesicle, heart, bone, breast, kidney, thyroid gland, and muscles. Many hypotheses have been put forward, but pathogenesis has not yet been revealed. We agree with the most widely accepted of them, the shunt-escape theory (2).

Vertebral localization of HD is extremely rare. Only 0.5% to 2% of HD cases involve the bones, and half are in the vertebrae (3). In order of frequency, most cases are localized in thoracic, lumbar and cervical vertebrae. Bone, and particularly spinal involvement, when compared with other rare locations, is quite interesting. It is usually localized in compact bone, which contains calcium, although it is porous, in contrast with soft tissue. It was demonstrated by Dew that HD embryos localize in the corpus vertebrae due to high vascularity and invade other parts of vertebrae, surrounding tissues, and even spinal canal by forming microvesicular polycystic lesions. These grow 1 to 5 mm a year (7). In our case, consistent with the literature, there was involvement in the corpus of T4 vertebra; however, it grew only 2 mm, less than the rates found in the literature.

Diagnosis may be achieved through holistic evaluation of clinical, radiological, and lab data, along with anamnesis. HD findings and symptoms depend on the involved organ, localization of the organ and the tissue, effect on neighboring tissue, rupture or complications, secondary infec-

tions and immunological reactions (2). Symptoms range from the most frequent, localized back pain, to complete quadriplegia (7,8). Similarly, numbness in the hands and feet, weakness, and back pain were the preliminary complaints of our patient. Radiological screening methods are the first and most important options for diagnosis. Lab tests have limited effect. As also seen in our case, appearance of HD in rare locations may lead to diagnostic difficulty, as hydatid cysts may be confused with benign or malign tumors, abscess or other cysts (2). Preoperative diagnosis in our case was neurogenic tumor destroying or extending into spinal canal.

Most of the endemic regions for HD are underdeveloped or developing countries. In these regions, especially the Mediterranean basin and the Middle East, animals have always been depended upon as sources of food, transportation, labor, and security. Ideal conditions for HD are constituted when appropriate climate and ecological conditions, nomadism, low level of hygienic conditions and education, local customs and beliefs, joint feeding grounds, uncontrolled slaughtering that may take place in poor conditions and even on open ground, and presence of carnivorous animals, particularly stray or pet dogs accessing the remnants of slaughtered animals are added (6). Migration and travel from these regions spreads HD worldwide. As might be expected, the largest published case series, with 36 cases, is from our neighbor, Iraq, where HD is endemic (7). HD has serious negative effects on national revenues due to increasing health costs, as well as losses in livestock industry.

In theory, HD is a controllable, even eradicable, disease. It has been controlled in small island countries like Iceland, New Zealand, Tasmania, Cyprus, and the Falkland Islands, as well as limited areas of Argentina and Chile (6). Nevertheless, factors that cause HD to spread, such as environmental conditions, hard-to-change cultural traditions and human behaviors are still in place, and it seems it will continue that way. Therefore, easiest and least expensive solution is to prevent human HD infection. Although vertebral HD is rare zoonotic infection, treatment and eradication of the cyst is challenging due to its effects on vertebral body, intervertebral disk, and epidural space. Despite surgical and medical treatment, recurrence ratio is 40% to 100%, within average of 30 months (3). Together with the challenges arising from anatomical and physiological features, treatment is very challenging. Clinicians go round in circles pursuing treatment and disease. New methods, particularly medications that can

better penetrate the central nervous system, are needed to prevent recurrence, and to break the vicious cycle.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - M.Ç., S.G.Ç., H.E.; Planning and Design - M.Ç., S.G.Ç., H.E.; Supervision - M.Ç., S.G.Ç., H.E.; Funding - M.Ç., S.G.Ç., H.E.; Materials - M.Ç., S.G.Ç., H.E.; Data Collection and/or Processing - M.Ç., S.G.Ç.; Analysis and/or Interpretation - M.Ç., S.G.Ç.; Literature Review - M.Ç., S.G.Ç.; Writing - M.Ç., S.G.Ç.; Critical Review - M.Ç., S.G.Ç.

YAZAR KATKILARI

Fikir - M.Ç., S.G.Ç., H.E.; Tasarım ve Dizayn - M.Ç., S.G.Ç., H.E.; Denetleme - M.Ç., S.G.Ç., H.E.; Kaynaklar - M.Ç., S.G.Ç., H.E.; Malzemeler - M.Ç., S.G.Ç., H.E.; Veri Toplama ve/veya İşleme - M.Ç., S.G.Ç.; Analiz ve/veya Yorum - M.Ç., S.G.Ç.; Literatür Taraması - M.Ç., S.G.Ç.; Yazıyı Yazan - M.Ç., S.G.Ç.; Eleştirel İnceleme - M.Ç., S.G.Ç.

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