

A Case of Spinal Intradural Extramedullary Hydatid Cyst Mimicking a Spinal Tumor

Spinal Tümörü Taklit Eden Spinal İntradural Ekstramedüller Hidatik Kist Olgusu

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ABSTRACT A patient with multiple cystic involvements in rare locations, who is immunosuppressed due to co-existing conditions, has been presented in this case report. A 48-year-old male with numbness in the right leg and a medical history of Behçet's disease, hypertension and ankylosing spondylitis, using prednisolone, methotrexate, sulphasalazine and colchicine admitted to our hospital. He had left hemihypoesthesia below the level of thoracic vertebra 3 and bilateral Babinsky positivity. Cervical magnetic resonance imaging showed an extramedullary mass lesion in the spinal canal, at the level of first thoracic vertebra, which caused compression. Diffuse edema was noted at the spinal cord segments between cervical 5 and thoracic 2 levels. Cervical 7- thoracic 1 laminectomy was performed. The mass was totally excised. Histopathological examination confirmed the diagnosis of a hydatid cyst. Indirect hemagglutination test for echinococcosis was positive. Radiological examination showed additional cysts in lung, liver and spleen. Two cysts in liver and spleen were drained under computerized tomographic guidance. The patient is still on sulphasalazine, methotrexate and albendazole therapy and is under regular follow-up.

Key Words: Echinococcosis; immunocompromised host; spine; spinal cord compression

ÖZET Bu olgu sunumunda nadir yerleşimli çoklu kistik tutulumları olan ve aynı zamanda eşlik eden durumlar nedeniyle immün sistemi baskılanmış olan bir hasta sunulmaktadır. Sağ bacakta uyuşma ve Behçet hastalığı, hipertansiyon ve ankilozan spondilit öyküsü olan, prednizolon, metotreksat, sulfasalazin ve kolşisin kullanan 48 yaşındaki erkek hasta hastanemize başvurdu. Üçüncü torakal vertebranın altındaki seviyede sol hemihipoestezi ve iki taraflı Babinski pozitifliği vardı. Servikal manyetik rezonans görüntülemesinde spinal kanalda birinci torakal vertebra seviyesinde kompresyona neden olan ekstramedüller kitle lezyonu görüldü. Servikal 5 ve torakal 2 seviyeleri arasındaki spinal kord segmentlerinde yaygın ödem mevcuttu. Servikal 7-torasik 1 laminektomi yapıldı. Kitle tamamen çıkarıldı. Histopatolojik muayene hidatik kist tanısını doğruladı. Ekinokok için indirekt hemaglutinasyon testi pozitif sonuçlandı. Radyolojik muayenede akciğer, karaciğer ve dalakta da kistler görüldü. Karaciğer ve dalaktaki iki kist bilgisayarlı tomografi rehberliğinde drene edildi. Hasta halen sulfasalazin, metotreksat ve albendazol almaya devam etmekte olup düzenli kontrolleri sürmektedir.

Anahtar Kelimeler: Ekinokokkozis; immün yetmez konak; omurga; omurilik sıkışması

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Echinococcosis (cystic hydatid disease) is a zoonotic disease which is not rare in Turkey. A patient with cystic involvement in a very rare location, who is immunosuppressed due to co-existing conditions, is presented in this case report.

CASE REPORT

A 48-year-old male admitted to the Department of Neurosurgery with numbness in the right leg present for two months without any pain or weakness. His medical history revealed presence of Behçet's disease for 20 years and hypertension and ankylosing spondylitis for 2 years. He was on prednisolone, methotrexate, sulphasalazine and colchicine treatment.

He had left hemihypoesthesia below the level of thoracic vertebra (T) 3 and bilateral Babinski positivity on neurological examination.

On cervical magnetic resonance imaging, located at the level of T1 vertebra, an oval shaped extramedullary mass lesion with a size of 22x10x14 mm, necrotic/cystic regions at its center, smooth iso/mild hyperintense margins on T2 images and hypointense margins on T1 images was noted within the spinal canal. Intensive contrast enhancement was observed in the solid margins of the lesion. The mass caused significant narrowing of the spinal canal, right lateral displacement and thinning of the spinal cord due to compression at this level. Diffuse edema was noted at the spinal cord segments between cervical (C) 5 and T2 levels. Mild expansion of the central canal was observed at C5 and C6 levels due to cord compression (Figures 1, 2).

Cervical 7-thoracic 1 laminectomy was performed by neurosurgeons. It was noted that the mass was located intradurally and extramedullary,

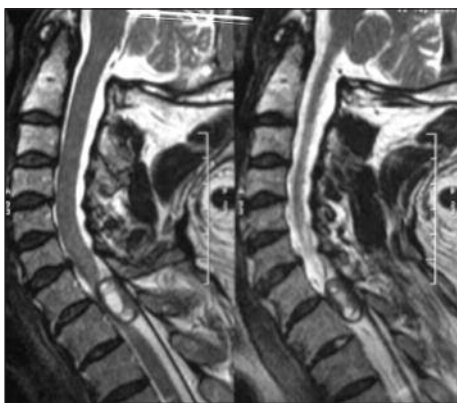


FIGURE 1: Cervical magnetic resonance imaging of our case (vertical).

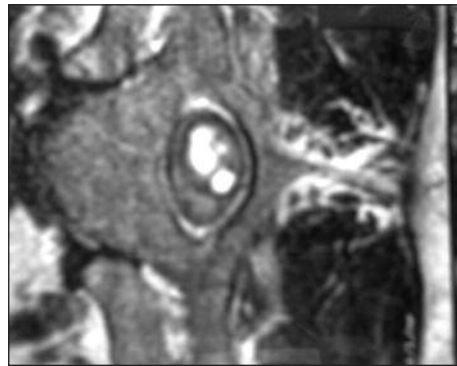


FIGURE 2: Cervical magnetic resonance imaging of our case (transverse).

and adhered strongly to the dura and the spinal cord. Pus was drained out of the mass with occasional calcified regions and a thick capsule. The mass was totally excised macroscopically. After postoperative consultation of the Department of Infectious Diseases, treatment with albendazole 400 mg twice daily was initiated for suspected hydatid disease. Indirect hemagglutination test for echinococcosis resulted as positive at 1/16,384 titer, and radiological examinations were performed to investigate possible cysts in other locations. The patient was discharged on postoperative day 1 and followed-up at the outpatient clinic of the Department of Infectious Diseases thereafter. Histopathological examination confirmed the diagnosis as hydatid cyst.

A round shaped nodule with regular margins and a diameter of 2 cm was noted in the middle zone of the right lung on chest X-ray. Abdominal ultrasound revealed cystic lesions with regular borders and homogenous content, one with a diameter of 5 cm located in the posterior of the right lobe of the liver, and one with a diameter of 7 cm located within the spleen parenchyma. High resolution computerized tomography revealed a 20x18 mm, round, cystic nodule in the apical segment of the lower lobe of the right lung, a 10x6 mm nodule in the apical segment of the upper lobe of the right lung with surrounding millimetric satellite appearances, as well as round hypodense lesions including a 4x4 cm cyst located in the liver subsegment 6, and a 4.5x4.5 cm cyst located in the middle-lower pole of the spleen.

The lesions in the liver and the spleen were drained under computerized tomographic guidance by interventional radiology. The patient continues to take sulphasalazine (1 g twice daily, oral), methotrexate (10 mg, once a week, oral) and albendazole (as cycles of 400 mg twice a day, oral, for 28 days, followed by a two-week-rest, yet completed his 4th cycle), and is under regular follow-up.

DISCUSSION

Cystic hydatid disease is a zoonotic disease frequently involving the liver and the lung, and can be seen worldwide. The most frequent microorganism is *Echinococcus granulosus* leading to unilocular cystic hydatid disease, and is frequent in Turkey.^{1,2}

Echinococcus granulosus is mostly located in the liver (50 to 75%) and lungs (10 to 30%). Other organ involvements are approximately 10-20%. While hydatid cyst is often primarily located in the liver and lung, it may appear in the other organs, both primarily and secondarily. Spinal location is rare and constitutes about 1% of all cases.³⁻²² Spinal involvement has been reported as 3.8% in Turkey.^{16,17} It may be located at intramedullary, intradural extramedullary, extradural intraspinal, vertebral or paravertebral locations.¹⁸ Spinal hydatid cysts may be located in thoracic (49-82%), lumbar (18-39%), sacral (2-12.5%) and very rarely, in cervical segments.^{16,19,20} Symptoms are related to spinal cord compression and may include back pain, radicular pain, weakness in extremities, sensory disturbances, bladder and bowel dysfunction, and even paraplegia.²¹⁻²⁵

An important feature of our patient was his immunosuppression. which might have played a role in multiple organ involvement. Any similar case has not been published before. There was only a report of a 6-year-old child with AIDS, who had disseminated alveolar echinococcosis in her liver.²⁶

Surgical removal of the cyst is the most effective treatment. Percutaneous drainage can be performed as an alternative. Preoperative and postoperative chemotherapy is routinely recommended, since it reduces the risk of recurrence and facilitates the removal by reducing intracystic pressure. Adjuvant chemotherapy should be initiated four days before surgery, and treatment should be continued at least for one month with albendazole (or as an alternative agent, mebendazole). Medical treatment is indicated when the general medical condition of the patient is not suitable for surgery, when the cyst is uncomplicated, when surgical access is not possible due to its location, when there is bilateral and multiple involvements, when dissemination or recurrence develops following surgery, or when the patient does not provide consent for surgery. Either albendazole (10-15 mg/kg/day in two or four doses) or mebendazole (40-50 mg/kg/day) should be administered for 3 to 6 months for medical treatment. Cycles of four weeks of therapy followed by two weeks of rest is recommended to prevent hepatotoxicity during albendazole treatment. Radiological findings and serological markers are useful in follow-up, but no signs of radiological and serological improvement can be demonstrated in some cases.²⁷⁻²⁹

It is not possible to predict whether a complete treatment response will be achieved in our immunosuppressed patient. The patient is undergoing long-term albendazole therapy and is currently under regular follow-up.

In conclusion, Echinococcosis may appear also at unusual locations, and when an individual is diagnosed for hydatid cyst disease, other body parts should be screened for the presence of multiple cysts, as carried out in our patient. Immunosuppression can be a predisposing factor for multiple organ involvement.

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