Case Report

Imaging of disseminated cysticercosis: Many faces to identify

Ravinder Kumar, Sahil Mehan, Shachee Zinzuvadia
Department of Radiodiagnosis, Geetanjali Medical College & Hospital, Geetanjali University, Udaipur, India

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ABSTRACT

Disseminated cysticercosis (DCC) is a very rare, infectious form of cysticercosis in which the cysticerci spread throughout the body. There are myriad clinical manifestations and imaging findings depending on the immune response mounted by the host on the parasite, site of larval encystment, stage and mass effect of the parasite. A 31-year-old immunocompetent male presented with symptoms of multiple palpable nodules and recurrent generalized seizures. After radiological assessment from whole body radiographs, ultrasound examination, MRI and pathologic biopsy, he was diagnosed as having disseminated cysticercosis involving the brain, lungs, eyes, skeletal muscles and subcutaneous tissue through the whole body. The patient was treated with a combination of antiepileptic medication and steroids. Follow-up imaging showed improvement, and the patient was asymptomatic 3 months after treatment. In this case report, we discuss and illustrate DCC in terms of epidemiologic features, disease course, pathophysiologic features, clinical presentation, diagnosis and radiologic findings, and treatment.

KEYWORDS: cysticercosis, disseminated cysticercosis, magnetic resonance imaging, neurocysticercosis, taenia solium

INTRODUCTION

Cysticercosis is a zoonotic disease seen in tropical countries which is caused by *Cysticercus cellulosae*, which is the larval form of the pork tapeworm, *Taenia solium*. It most commonly affects the central nervous system (subarachnoid space, ventricles, or spinal cord), subcutaneous tissues, lungs, eyes, liver, skeletal muscles, and occasionally the pancreas, thyroid, and heart[1]. Transmitted by the fecal-oral route, this disease is not only limited to the endemic zone; it has now spread worldwide due to globalization. One of the rare complications and uncommon manifestations of cysticercosis is its disseminated form. Diagnosis of disseminated cysticercosis (DCC) can be considered to be confirmed if there are multiple vesicular cystic lesions present in the brain and cysts are demonstrated in at least two other body parts[2]. However, widespread massive dissemination of the cysticercal infestation can result in the involvement of any organ in the body[3]. The main features of DCC include dementia, confusion, intractable epilepsy, enlargement of the subcutaneous and lingual nodules, muscle hypertrophy and a relative absence of focal neurological signs or obvious raised intracranial pressure, at least until late in the disease. Painless diffuse enlargement of all muscle groups (muscular pseudohypertrophy), a rare presentation, gives the patient a ‘Herculean appearance’[4]. Fewer than 50 cases of DCC have been reported worldwide, the majority being from India.

We report here an unusual rare case of DCC involving diffuse involvement of the brain, subcutaneous tissue and multiple organ systems accompanied by widespread muscle hypertrophy, treated with symptomatic management and antiepileptic medication.

CASE REPORT

Clinical presentation

A 31-year-old immunocompetent male, non-vegetarian, presented with a 2-month history of headache, vomiting and recurrent episodes of convulsions. He also complained of multiple palpable

Address correspondence to:
Dr. Ravinder Kumar, AG-1, Geetanjali Medical Campus, Mansakhera, Udaipur-313002, Rajasthan, India. Mob: +919571218953; E-mail: kundu19@yahoo.co.in
nodular lesions all over the body, progressive loss of memory and judgment for three months. A physical examination revealed several firm, well-circumscribed, non-tender subcutaneous nodules all over the body, predominantly seen over the neck, chest, back region, abdominal wall and extremities. The nodular lesions were well defined and varying in size from 0.5 to 3 cm. The typical “Herculean” appearance, associated with muscular pseudohypertrophy, was seen in all extremities. Neck rigidity and exophthalmos was also observed. He had stable vital signs and no focal neurological deficit was noted.

Routine hematological investigations showed hemoglobin to be 11.5 g/dl and the total leukocyte count was within normal limits. The differential leukocyte count showed borderline eosinophilia (6%). Human immunodeficiency virus, hepatitis B surface antigen and hepatitis C virus testing were non reactive. All other investigations were within normal limits.

**Diagnostic work-up**

Plain radiographs showed typical ‘rice-grain’ shaped or cigar shaped calcification in the soft tissues (Fig 1). Shoulder and abdominal X-Ray showed similar multiple calcified lesions in the muscles and subcutaneous tissues (Fig 1a & Fig 1b). Pelvic X-ray demonstrated innumerable small calcific densities projecting into the soft tissues of the right and left gluteus (Fig 1c). Ultrasound examination revealed multiple round to oval, thin walled, anechoic lesions of 8 to 10 mm with eccentric echogenic intralesional focus, representing scolex. These lesions were diffusely dispersed in subcutaneous tissue, superficial muscular plane of face and neck, chest, back and all extremities (Fig 2). Magnetic resonance imaging (MRI) of the brain revealed diffuse hyperintense cystic lesions with eccentric hypointense scolex in the parenchymal, scalp tissue and retroocular regions. T2W axial images showed multiple cystic lesions with hypointense foci within both cerebral hemispheres, cerebellum, extraocular muscles and soft tissues of the neck. These lesions had a “cyst with dot sign

![Fig 1a: Disseminated cysticercosis—anteroposterior view of right shoulder and posteroanterior view of abdominal X-Ray film (Fig 1b) revealing multiple rice grain or cigar-shaped calcifications in the soft tissue of shoulder, chest, abdomen, gluteal and iliopsoas muscle regions. Fig 1c: Lower pelvic and limb X-ray showing similar calcified lesions, lying parallel to muscle fibres, in gluteal, pelvic, and limb muscles.](image1)

![Fig 1(a) Fig 1(b) Fig 1(c)](image2)

![Fig 2: Ultrasound reveals a cystic lesion (white arrow) with hyperechoic scolex (black arrow) in the left calf region.](image3)
(starry sky)” appearance, or eccentric scolex (Fig 3a, b). There was evidence of mild hydrocephalus, brain edema and signs of raised intracranial tension (Fig 3c). The bilateral brain parenchyma, extraocular, facial and tongue muscles were involved (Fig 3 d, e). These imaging features, along with patient’s history of intractable epilepsy, are characteristic of encephalitic form of neurocysticercosis.

Similar numerous hyperintense lesions with longitudinal orientation along the muscle fibers were distributed in the limb muscles and adjacent subcutaneous tissues of the neck, chest wall, forearms and arms, back, abdominal wall, thighs, calves, gluteal, pelvic as well as the paraspinal muscles, suggestive of DCC (Fig 4). A cystic lesion was seen in right lung (Fig 5). The solid abdominal organs and cardiac muscles were normal. Perimetry and fundus examination were also within normal limits.

Fig 3(a): Axial T2W brain MR image showing multiple well defined round to oval shaped hyperintense cystic lesions (starry sky sign) seen scattered at places. Lesions were also noted in the scalp (indicated by arrow). MR Image shows cysts in different stages of the life cycle. Hypointense scolex (arrow) is also seen. Fig 3 (b): The perilesional T2 hyperintense image suggests edema or inflammatory changes (arrow). Fig 3(c): Post Contrast T1W sagittal section confirms multiple well-defined ring-enhancing lesions in the brain (white arrows). A few cystic lesions are seen in the subcutaneous tissues of the neck (black arrow). Fig 3(d & e): Coronal (Fig 3(d)) and Axial(Fig 3(e)) T2W Image showing multiple cysticerci lesions in the bilateral brain parenchyma, extraocular, facial and tongue muscles.
Serological investigation

Antibodies to cysticerci were detected in the serum and cerebrospinal fluid by means of an enzyme-linked immunosorbent assay. Other cerebrospinal fluid (CSF) findings include glucose: 42 mg/dl, 10 leucocytes per cu.mm, and protein: 96 mg/dl.

Histopathological examination

An oval translucent cyst was resected from the deltoid muscle, and histopathological examination of the cyst was consistent with the diagnosis of cysticercosis.

Treatment

This patient was symptomatically managed with mannitol and glucocorticoids to decrease edema and inflammation. Antiepileptics were continued. Follow-up imaging showed improvement, and the patient was asymptomatic 3 months after treatment.

DISCUSSION

Human cysticercosis is a tissue infection which occurs due to infestation by the encysted larval stage of the pork tapeworm, *Taenia solium*. With regard to clinical manifestation, the disease can present in two forms: taeniasis and cysticercosis. Taeniasis is acquired through the consumption of cyst-infected pork. In contrast, cysticercosis is developed through the ingestion of eggs from the feces of a tapeworm carrier, with little evidence of other forms of contamination (e.g. through the agency of air, water, or flies). Infective embryos (hatched from the ingested eggs) disseminate through the systemic circulation after actively crossing the intestinal mucosa. Some cysts are cleared by the liver. Lodged cysts in capillaries (mostly
in muscle and brain tissue) develop into immature cysts and later into larval cysts, taking up to 3 months to reach this stage[8]. When the parasite dies by natural processes or as a result of antihelminthic therapy, an inflammatory response with perilesional edema and focal enhancement ensues, followed by calcification[8]. DCC was reported as early as 1912 by the British Army medical officers stationed in India[3].

The clinical presentation of patients with DCC syndrome is highly variable and nonspecific and depends on the location, number, size, and stage of the parasites, as well as the degree of the host reaction[7]. Furthermore, individual responses to the parasite differ. Our patient was characterized by seizures, abnormal mentation, palpable subcutaneous nodules and pseudomuscular hypertrophy. These symptoms are typical for DCC.

The diagnosis of cysticercosis is based on a constellation of (a) clinical findings; (b) the cysticercus-specific IgG antibody level as determined with an enzyme-linked immunoelctrotransfer blot assay; (c) an enzyme-linked immunosorbent assay in either serum or CSF, with a specificity and sensitivity of 100% and 98%, respectively[8]; and (d) noninvasive imaging findings (CT and MRI)[8].

CT and MRI findings are better in diagnosing this entity as they demonstrate the number, anatomical localization and topography of lesions, their stage of involution, and the degree of inflammatory reaction of the host against the parasites and have largely replaced previous radiological procedures such as plain roentgenograms, pneumoencephalograms, cerebral angiography and myelography[9]. High resolution ultrasound is useful in noninvasively demonstrating subretinal cysticercosis and cysticercal cysts at other unusual sites. In our patient, ultrasonography of the eyes, color Doppler and MR studies revealed cystic lesions in both the orbits, diagnosing ocular cysticercosis.

In general, MR is superior to CT imaging in better image detection, higher resolution and definition, which makes for better lesion conspicuity. This higher contrast resolution is particularly helpful in the detection of inflammatory changes and the evaluation of ventricular involvement. However, its sensitivity for the detection of calcified lesions is poor. MRI is the imaging modality of choice for the evaluation of patients with brainstem cysts, intraventricular cysticercosis, small cysts located over the convexity of cerebral hemispheres, and in the follow-up of such patients whereas CT remains the best screening neuroimaging procedure for patients with suspected neurocysticercosis and small calcifications[1].

DCC is not a single disease entity that can be managed uniformly. Treatment of DCC is controversial and depends on the location and cyst burden, the symptoms, and associated complications[10]. Furthermore, cysticidal agents, such as albendazole and praziquantel, may complicate the treatment as they themselves initiate a host inflammatory response that may result in raised intracranial tension and more symptoms. In general, management of DCC is symptomatic (antiepileptics and steroids) and cysticidal (i.e., albendazole and praziquantel). Although surgical intervention (i.e., shunt placement or emergency decompression with parasite removal) may eventually become necessary, it is rarely used nowadays because the diagnosis is being made at earlier stages and pharmacologic therapy is usually sufficient[11,12]. The role of cysticidal drugs is controversial. Wilson et al recommended that all patients with multiple cysts should receive treatment with cysticidal drugs[13]. However, these drugs hasten the death of the cysticerci cysts, which may occur even in the absence of such treatment. Cysticidal therapy may be associated with a generalized anaphylactic reaction, which may be due to the demise of cyst and massive release of antigens. Hence, these therapies should be advised with high degree of caution and should be individualized. Cysticidal drugs have no role in the presence of inactive calcified cysts, because the parasites are already dead[14]. Our patient, a case of encephalitic form of disseminated cysticercosis, was well managed by antiepileptics and steroids. He did not receive albendazole and/or praziquantel, and was asymptomatic since last 3 months.

Recently, a combination of albendazole and praziquantel was found to be effective in comparison to a single drug therapy in patients with parenchymal brain cysticercosis with ≤ 20 viable cysts. Complete cyst clearance was seen in 75% (12 of 16) of the patients with a combination therapy whereas only 25% (4 of 16) patients had complete cyst clearance in the subgroup of patients who received only albendazole. The role of albendazole and praziquantel combination therapy is worth trying in patients with massively infected disseminated cysticercosis[15]. A randomized controlled study is urgently needed to assess the efficacy of the combination therapy and currently available drugs. The role of repeated courses of antiparasitic drugs also needs to be evaluated.

**CONCLUSION**

In DCC, diagnostic approach, treatment, and prognosis differ widely depending on the type of infection. It is important to recognize this pleomorphic disease clinically; early radiological evaluation is warranted in cases of disseminated condition. In most of the patients with disseminated cysticercosis, quality of life is often poor with sinister prognosis.
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REFERENCES