A life-threatening brainstem compression by cerebral *Echinococcus granulosus*

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### INTRODUCTION

Cystic echinococcosis (CE) is a zoonotic disease caused by *Echinococcus granulosus*. It is of worldwide importance, and is widespread in the Mediterranean region and Middle East. This tapeworm shows great intraspecific variation in relation to host specificity, epidemiology, and morphology. This variability led in previous years to the identification of ten (G1-G10) different genotypes of the parasite. Cerebral localization of *E. granulosus* is not common: it especially affects children and is more frequently located in the supratentorial region. It can be life-threatening due to its localization in eloquent areas especially in the posterior fossa. Despite the benign nature of hydatid cyst, invasion of critical areas may cause significant mortality and morbidity in some patients. Urgent surgical decompression and adjuvant medical treatment must be employed as soon as possible in these patients. We present a clinical case of life-threatening brainstem compression in a child due to a rare form of CE which was confirmed with biomolecular techniques. She presented with respiratory distress and progressive quadriplegia. All cysts were removed by microsurgical technique and albendazole was given postoperatively for one year with regular follow-ups.

Keywords: *Echinococcus granulosus*, cerebral, respiratory distress, brainstem compression.

### SUMMARY

Cystic echinococcosis (CE) is a zoonotic disease caused by the metacestode of the dog tapeworm *Echinococcus granulosus*, whose larval stage causes CE in livestock, wild animals, and humans. CE is acquired by ingesting eggs, originating from the faeces of definitive hosts (dogs, wolves, and other carnivores), that harbour the adult *E. granulosus* worms in their small intestine [1].

The disease is of worldwide importance, and it is widespread in the Mediterranean region and Middle East [2]. This tapeworm shows a great intraspecific variation in relation to host specificity, epidemiology, morphology, developmental biology, biochemistry, physiology, and genetics [3]. This variability led in the past years to the identification of ten (G1-G10) different genotypes and genetic variants of the parasite [4]. The taxonomy of the genus *Echinococcus* is presently undergoing major changes; new data on epidemiology and genetic provide sufficient evidence that several of the previously identified “strains” of *E. granulosus* (G1 to G10) have to be regarded as species. Using these criteria, the following species will substitute the previous taxonomic classification: *E. granulosus sensu stricto* (G1-G2-G3), *Echinococcus ortleppi* (G5) and *Echinococcus equinus* (G4). And in addition, the closely related and apparently
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Monophyletic group of genotypes from G6 to G10 will be probably included in one species, named *Echinococcus canadensis*, even if the taxonomy of this group is not yet resolved [5, 6]. Several human cases have been reported in the Mediterranean region, Middle East, and South America but also Australia, New Zealand and USA [7]. Cerebral localization of *E. granulosus* is not common and it especially affects children and it’s more frequently located in the supratentorial region [8]. It can be life threatening due to its localization in eloquent areas especially in the posterior fossa. We present a clinical case of life threatening brainstem compression in a child due to a rare and uncommon form of cystic echinococcosis.

**CASE REPORT**

A 14-year-old girl who was living in Iraq came to Turkey for care and was referred to our hospital with respiratory distress and progressive quadriparesis during the last two weeks. She had previous surgery for posterior fossa cystic mass four years ago in her country. She had no written medical information for previous surgery which she underwent in her country.

The complete blood count, including the eosinophilic count was in normal range. The serological tests (indirect hemagglutination, enzyme immunoassay) for *E. granulosus* were negative. Neurologic examination revealed right hemiplegia. Cranial MRI showed thorough compression to pons, medulla and fourth ventricle by a multicystic lesion at right cerebellopontine angle. Also, there was another cystic lesion at left cerebellopontine angle. T2-weighted images showed a large, well-defined, round, hyperintense cysts with a hypointense rim (Figures 1 and 2). Abdominal and chest imaging, cardiac echocardiography didn’t show visceral involvement. Urgent surgical removal of the cyst was performed as the patient had respiratory distress symptoms. Suboccipital craniotomy was realized with right paramedian surgical incision. Cystic lesions were removed with the microsurgical technique at right cerebellopontine angle. Right 5, 6, 7, 8 and lower cranial nerves, both vertebral arteries and basilar artery were observed under microscope. After opening the prepontine cistern, a cystic lesion was found and removed on the left cerebellopontine angle. All cystic lesions were removed intact. All surgical area was washed with 3% saline after cysts were removed. Parasitological examination (Figure 3) of the isolates at light microscopy showed the presence of hooks and cystic structures compatible with a cestode infection, but not sufficient to determine the specie. Same results were obtained by histopathological examination (Figure 4), so the formalin-

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**Figure 1** - T2 weighted images of cyst in axial, coronal and sagittal section.
fixed paraffin-embedded parasite was examined with biomolecular techniques in Department of Veterinary Medicine, University of Sassari as follows.

Based on the alignment of NADH dehydrogenase subunit 1 (ND1) gene sequences chosen as representative of species belonging to *Cestoda*, primer NAD2 was designed (5′GGTTCTRTAAGGTTT-GARGC3′). NAD2 was coupled to primer JB12 to develop a PCR test targeting 213 bp of the ND1 gene of most of the species belonging to *Cestoda*.

Briefly, 100 to 150 ng of DNA extractions were used in a 50-μL PCR mixture containing 200 μM deoxynucleoside triphosphates (dNTPs), 1 μM concentrations each of the two primers, and 1.25 U of *Taq* DNA polymerase (Qiagen, Italy). PCR amplifications were performed with an initial denaturation at 95°C for 2 min, followed by 35 cycles of denaturation at 95°C (30 s), annealing at 50°C (30 s), and extension at 72°C (30 s), followed by a final extension at 72°C for 10 min. All DNA samples extracted from formalin-fixed paraffin-embedded parasites tested positive in PCR and showed a band of the correct size about 220 bp.

An ABI Prism BigDye terminator cycle sequencing ready reaction kit (Life Technologies, Italy) was used for direct cycle sequencing of PCR prod-

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**Figure 2** - T2 Axial images one month after surgery.

**Figure 3** - Wet mount preparation of *E. granulosus*. 
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Products according to manufacturer’s protocol. Ambiguous nucleotide positions were resolved by cloning amplicons into the vector pCR™4-TOPO® TA (Life Technologies, Italy), transformation of Top10 cells (Invitrogen, Italia), and by universal M13 primers sequencing. At least three clones were selected and sequenced for each transformation. Sequences were edited with Chromas 2.2 (Technelysium, Helensvale, Australia) and deposited in GenBank using the National Center for Biotechnology Information (NCBI, Bethesda, MD) BankIt v3.0 submission tool (http://www3.ncbi.nlm.nih.gov/BankIt/). Sequences were checked against the GenBank database with nucleotide blast BLASTN [9].

Sequences obtained from DNA samples obtained from formalin-fixed paraffin-embedded parasites were 100% identical and shown 100% identity with *E. granulosus sensu stricto* (GenBank accession number: KJ556994.1).

Postoperatively albendazole 15 mg/kg/day was given for one year with regular follow-up.

**DISCUSSION**

Cystic echinococcosis occurs worldwide and usually *E. granulosus sensu stricto* (G1-G3) and *E. canadensis* (G6-G7) are the most involved parasites in human infections [10]. Cerebral hydatid disease remains rare and 2-3% of the systemic disease is observed in pediatrics [11-14]. The most common site is parietal lobe; posterior fossa location is very rare [15, 16].

The clinical presentation in cerebral hydatid disease varies to its location. Nausea, headache, vomiting, seizure, weakness, visual disturbances are the most common symptoms [15, 17]. Brainstem compression is life-threatening and urgent surgery can be life saving. Our case presented with respiratory distress which indicated an urgent surgical intervention.

MRI and CT imaginings are satisfactory for the diagnosis of hydatid cyst of brain; MRI is especially helpful. Cerebral hydatid disease is generally solitary but may be multiple when it ruptures due to surgery, trauma or spontaneously. In differential diagnosis; arachnoid cyst, brain abscess, epidermoid tumors should be taken into consideration [18, 19].

Surgery with the medical treatment is the preferred treatment for the symptomatic cerebral *E. granulosus*. Dowling’s technique is the most useful method for intact cyst excision [15, 20, 21]. All cysts were removed by microsurgical technique under microscope from the posterior fossa. Dowling’s techniques may be helpful especially supratentorial cysts. But infratentorial area has important and delicate structures. Therefore, Dowling’s technique is not useful particularly in this area. Medical treatment with albendazole pre and post operatively reduces recurrence, sterilize cyst wall [22, 23].

Despite the benign nature of hydatid cyst, invasion of critical areas may cause significant mortality and morbidity in some patients. Urgent surgical decompression and adjuvant medical treatment must be employed immediately in these patients.

**REFERENCES**


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