Prof. Dr. med. Daniela Mihic-Probst
Institute of pathology and molecular pathology
University Hospital Zürich
Switzerland

ORBITAL ECCHINOCOCCUS CYSTICUS

Clinical history

A 26-year-old patient has been suffering for two years from a massiveexophthalmos right. Allegedly, after steadily enlarging in first year, the tumor remained stable in size during last year. The patient wasborn in Eritrea and came to Switzerland three months prior to first visit in our hospital. On physical examination, she showedamaurosiswith atrophy of the optic nerve right and normal vision left.
Ultrasound and MRI showed an intraorbital cystic lesion.An echinococcus cystwas suspected butechinococcus serologic test was negative.
Three months prior to the examination she suffered from acute ulcerophlegmonous appendicitis. In addition, she was suffering fromchronic Hepatitis B infection with positivity for HBs antigen (1306.07 IU/ml) and positivity forHBV-DNA (1900IE/ml). The transaminases were not elevated and she had no signs of liver cirrhosisor a cystic liver lesion.

Macroscopy

Intraoperatively, the cyst bursted and could not be removed as a whole. Macroscopy therefore showed fragmented, partly membrane-like easily tearable tissue. The color of content was grayish, brownish of gelatinous consistency. Pushed together, the fragments had a size of 4.8 x 5.5 x 0.9cm.

Histology

Narrow strips of fibrosedpericystic soft tissue including mild to moderate chronic lymphoplasma cellular inflammation with giant cell reaction and focal hemosiderin deposits. In addition, laminated acellular material with clear striping and partly degenerated protoscolices.[1]

Special examinations

Laminated membrane positive for PAS
Immunohistochemistry positive for Escreen (mAbEg3) and negative for Ealv (mAbEm2G11)

Diagnosis

Orbital echinococcuscysticus

Follow up

Postoperatively, there was a significant increase in inflammatory parameters and eosinophilic granulocytes. This was attributed to the intraoperative bursting of the cyst. In the context of bursting cyst, anaphylactic reactions, local recurrences or systemic secondary infestation are well known. Therefore, preoperatively initiated combination therapy with Albendazol and Praziquantelwas continuedpostoperatively. The patient showedrapid improvement. To avoid a bacterial superinfection additionally Co-Amoxycillinwas given. Combination therapy was continued for two weeks and therapy withAlbendazol for 2 month.

Discussion

More than one million people are infected worldwide with echinococcus. The infection is listed by the World Health Organization among the neglected tropical diseases. Ecchinococcus is more prevalent in rural areas but recently more and more urban areas are involved as well.[21]

In Central Europe and United States the transmission from animalsis well documented and an increase in incidence is observed, especially for Alveolar Echinococcosis (AE).[3, 4]Humans represent the dead end of the parasitic infection cycle. They are infected by the eggs of contaminated feces of primary or intermediate host. This may happen by contaminated environment (hands-mouth transmission) or contaminated food.

After ingestion, eggs hatch and release larval oncospheres, which travel to liver via portal vein. Oncospheres develop into cysts, which enlarge slowly (~ 1 mm per month), producing protoscolices and daughter cysts that fill interior of cyst.

Ecchinococcosisis caused by different Echinococcus ssp., manifesting in a cystic, polycystic, or alveolar form. Cystic Echinococcosis (CE), as our case, is caused by the larval stage of Ecchinococcusgranulosus, AE is caused by Echinococcusmultilocularis, and polycystic echinococcosis by Ecchinococcusvogeli or Ecchinococcusoligarthra, both of which only affect humans. The most common organ affected is the liver and second most the lung. However, any organ may be involved.

My colleague Dr. Reinehr identified from our archives of the Department of Pathology and Molecular Pathology, University Hospital Zürich between 1997 and 2018 114 patients with Ecchinococcus infection (59 EA and 53 EC). Most primary manifestation was the liver with the exception of two lung and one gallbladder EA and 7 soft tissue and one peritoneal EC

infection [5]. He was able to demonstrate that Escreen (mabEg3) is positive in EC and EA. In contrast, mAbEm2G11 immunohistochemistry is specific for EA and therefore able to distinguish between the two different Echinococcus types. Our case was confirmed as EC by PCR and positivity for Escreen (mAbEg3) immunohistochemistry.

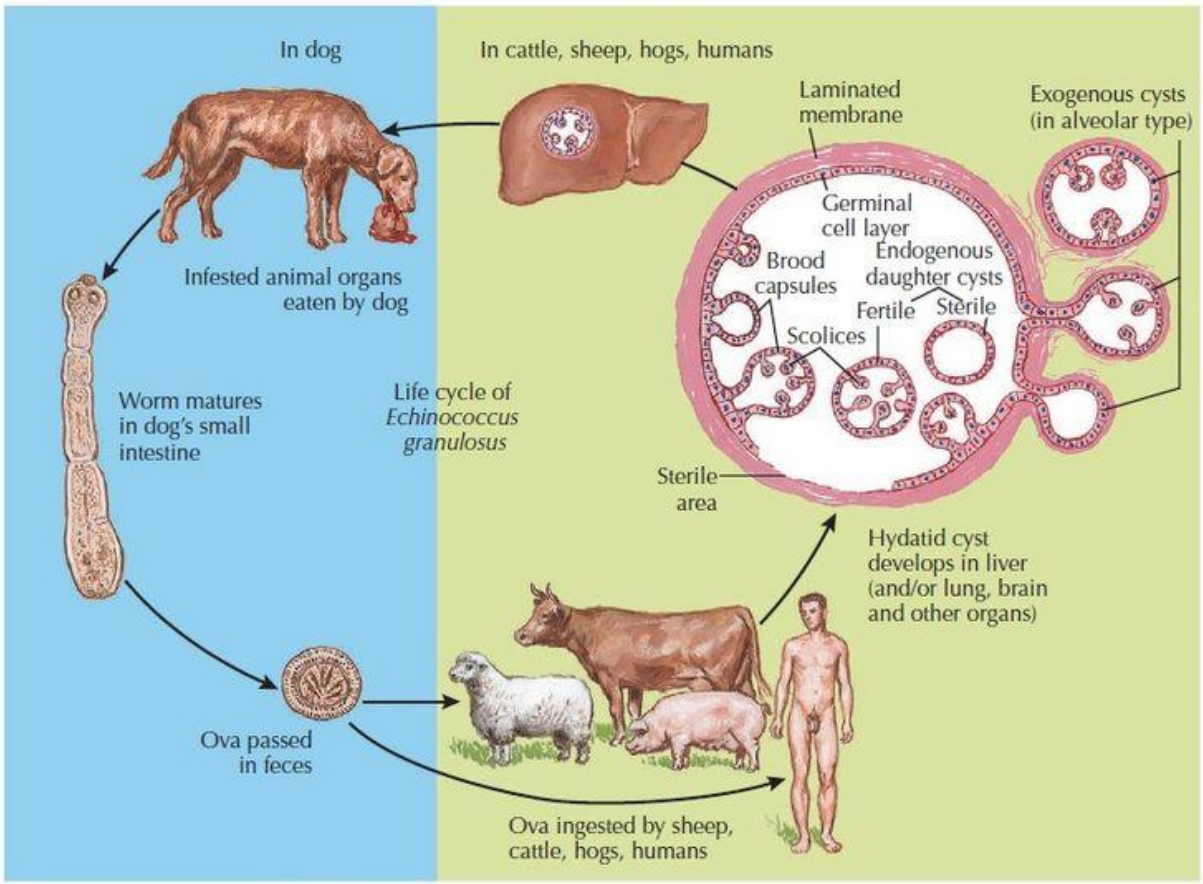
The distributed case is our first case of an orbital EC.Our patient has up to date no other EC manifestation. Therefore, a hand-eye infection is assumed.

However, by reviewing the *Pub Med* multiplecase reports of orbital EC can be found. The patients are children or young adults and people are mostly living in resource-poor countries [6, 78].

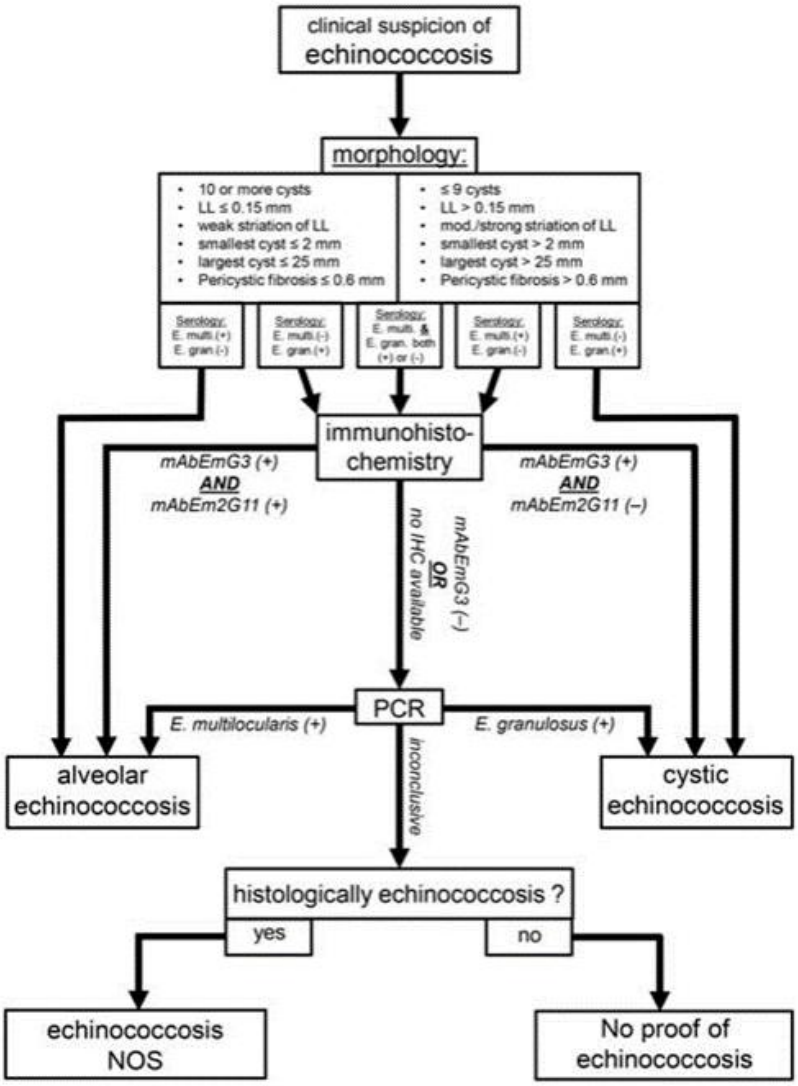
These findings correspond well with our patient.

Synonyms: Ecchinococcuscysticus, Ecchinococcusgranulosus, Dog Tapeworm

Echinococcusalveolaris; Ecchinococcusalveolaris, Fox Tapeworm



Ecchinococcuscysticus life cycle. Source: Wikipedia



Ecchinoccocusalgorhytm. Source: Reinehr M et al: *Am J Surg Pathol* 2020;**44**;43-54

1. Agudelo Higuita NI, Brunetti E, McCloskey C. Cystic echinococcosis. *J Clin Microbiol* 2016;**54**;518-523.
2. Deplazes P. Ecology and epidemiology of echinococcus multilocularis in europe. *Parassitologia* 2006;**48**;37-39.
3. Trotz-Williams LA, Mercer NJ, Walters JM *et al.* Public health follow-up of suspected exposure to echinococcus multilocularis in southwestern ontario. *Zoonoses Public Health* 2017;**64**;460-467.
4. Taxy JB, Gibson WE, Kaufman MW. Echinococcosis: Unexpected occurrence and the diagnostic contribution of routine histopathology. *Am J Surg Pathol* 2017;**41**;94-100.
5. Reinehr M, Micheloud C, Grimm F *et al.*Pathology of echinococcosis: A morphologic and immunohistochemical study on 138 specimens with focus on the differential diagnosis between cystic and alveolar echinococcosis. *Am J Surg Pathol* 2020;**44**;43-54.
6. Chtira K, Benantar L, Aitlhaj H, Abdourafiq H, Elallouchi Y, Aniba K. The surgery of intra-orbital hydatid cyst: A case report and literature review. *Pan Afr Med J* 2019;**33**;167.
7. Fasina O, Ogun OG. Hydatid cyst of the orbit in a young nigerian female: A case report. *Ghana Med J* 2017;**51**;204-206.
8. Assimakopoulos SF, Marangos M. Orbital hydatid cyst. *N Engl J Med* 2020;**382**;1352.