

Case Report

Orbital Hydatid Cyst with Diverse Locality in the Orbit and Review of Literatures

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Abstract

Purpose: to introduce five different type of orbital involvement by hydatid cyst in 8 consecutive patients.

Design: descriptive case series.

Methods: Herein 8 patients with slowly progressive proptosis from 2 to 4 months earlier at their first presentation are introduced. Complete ophthalmic examinations were done. Complete blood count, serologic test, ESR (erythrocyte sedimentation rate), CT scan and MRI were requested. Surgical removal of the lesions performed in all patients.

Result: The hydatid cysts found in the extraconal (2 patients) and intraconal space (3 patients), lacrimal gland (1 patient), medial rectus (1 patient) and intraosseous of orbital wall (1 patient). The diagnosis of the isolated orbital hydatid cyst without any lesion in other parts of the body was confirmed in all patients. The recurrence was not observed in any patients during 2 – 6 years follow up.

Conclusion: Although the most common site of involvement of orbital hydatid cyst is intraconal, it can invade various locations within the orbit.

Keywords: Extraconal, hydatid cyst, intraconal, orbit.

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Introduction

Hydatid cyst is caused by *Echinococcus granulosus* which can invade different sites of body and is prevalent in endemic areas. The most common site of involvement is liver and the orbital involvement, as an uncommon site, associates with 0.3 % – 2 % of prevalence.¹⁻⁴ However, the most frequent location in the orbit which can be invaded by hydatid cyst is the intraconal space but all other portions of the orbit may be involved either.^{1,5-8} Its most common clinical feature is progressive proptosis.⁶ In this paper five different types of orbital involvement by hydatid cyst are introduced.

Methods

This is a retrospective case series of 8 patients with orbital mass who had been referred to our clinic within 4 months and were diagnosed as orbital hydatid cyst. Their first presentation was slowly progressive proptosis from 2 – 4 months earlier. Complete ophthalmic examinations were done. Complete blood count, serologic test (Casoni test, enzyme linked immunosorbent assay and indirect hemagglutination assay), and ESR were assessed. Computed tomography (CT) and magnetic resonance imaging (MRI) were requested for all patients. Systemic investigation included Brain MRI, chest X ray and abdominal sonography was performed in all patients for recognition of possibility of other organs involvement. Orbital masses were noted in orbital imaging. The surgical

removal of lesions was performed in all patients with associated cyst rupture in 4 patients during removal. Orbital cyst removal was performed with medial, lateral, superior and inferior orbitotomy under general anesthesia. Their follow up was from 2 – 6 years. Post-operatively, Albendazole was prescribed for all our patients with the dosage of 10 mg/kg daily for 12 weeks.

Result

Demographic, clinical findings and surgical approach of patients are illustrated in table 1. All patients were living in rural areas of West Azarbayegan (4 patients), Kerman (2 patients), Fars (2 patients) and Zahedan (1 patient). Past medical history of all patients was unremarkable but all them had rural life-style and positive exposure to the cattle and dogs. Proptosis were present in all patients. The slit lamp examinations were normal in all patients except the presence of optic disc swelling in 2 of them with involvement of the relative afferent pupillary reflex (RAPD). The clinical features of patients concomitant with the cystic lesion pattern in orbital imaging resulted in the hydatid cyst diagnosis before surgical exploration. The masses were hypointensive in T1 and hyperintensive in T2 images of MRI without contrast enhancement or ring enhancement in peripheral of cyst consistent with cyst wall. The diagnosis of *Echinococcus granulosus* was confirmed by pathologic evaluation in all the patients after operation. Three layered cyst included acellular PAS-positive laminated membrane as outer wall of cyst, germinal layer and protoscolices, was reported in histological findings. The systemic evaluations for diagnosis of other possible sites of involvement were negative in all patients. Orbital cyst removal was performed with medial, lateral and superior orbitotomy. In 4 patients the cyst ruptured during surgical removal (3 with intraconal and one with intraosseous mass), but the cyst wall removed completely. The recurrence was not seen in

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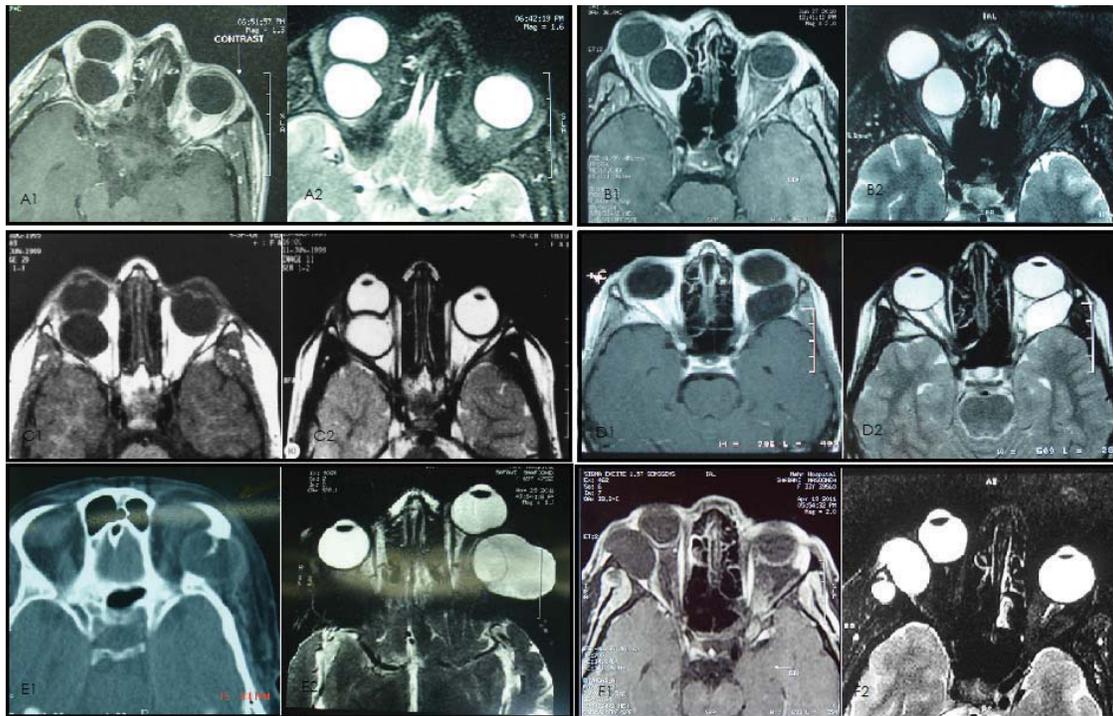


Figure 1. Different sites of orbital hydatid cyst in T1 and T2 weighted MRI. A) intraconal (A1: T1, A2: T2); B) medial rectus (B1: T1, B2: T2); C) extraconal with superior location (C1: T1, C2: T2); D) extraconal with lateral location (D1: T1, D2: T2); E) intraosseous (E1: CT scan, E2: T2); F) lacrimal gland (F1: T1, F2: T2).

Table 1. Demographic, clinical findings and surgical approach of patients

No. of case	Age (year)	Sex	Involved eye	Clinical feature	VA	Involved location of orbit	Methods of surgery and orbital approach and type of incisions
1	14	Male	OD	Proptosis, optic disc swelling, positive RAPD	5/10	Intraconal	Deep lateral orbitotomy, lateral lid crease incision
2	24	Male	OD	Proptosis	10/10	Medial rectus	Medial orbitotomy, trans caruncle incision
3	13	Male	OD	Proptosis	10/10	Extraconal (superior)	Superior orbitotomy, lid crease incision
4	18	Female	OS	Proptosis	10/10	Extraconal (lateral)	Deep lateral orbitotomy, lateral lid crease incision
5	62	Female	OS	Proptosis, optic disc swelling, positive RAPD	7/10	Intraosseous	Deep lateral orbitotomy, lateral lid crease incision
6	33	Female	OD	Proptosis	10/10	Lacrimal fossa	Deep lateral orbitotomy, lateral lid crease incision
7	44	Female	OS	Proptosis	10/10	Intraconal	Inferior orbitotomy, fornix incision
8	36	Male	OS	Proptosis	10/10	Intraconal	Deep lateral orbitotomy, lateral lid crease incision

OD: right eye; OS: left eye; RAPD: relative afferent pupillary reflex

any patients during follow up. Various locations of involvement in the orbit are displayed in radiologic images of patients with hydatid cyst in Figure 1.

Discussion

The hydatid cyst is one of the infrequent etiologies of cystic lesion in the orbital region, that is generated by *Echinococcus* which have four various strains consisting of *E. granulosus* (cystic lesion), *E. multilocularis* (alveolar lesion with more severe disease), *E. oligarthrus* (polycystic lesion) and *E. vogeli* (polycystic lesion).⁹⁻¹¹ Although the cystic echinococcus is the most common type of hydatid disease in the orbit,^{3,5,9} but also the polycystic

forms are reported in this location.⁹ The etiologic strain in all our patients was *E. granulosus*. Hydatid disease is widespread in some regions that is related to breeding cattle and dogs simultaneously such as Middle East, New Zealand, Australia, South America and Mediterranean region.^{12,13} The prevalence of hydatid cyst is estimated 0.61 – 2/100000 population.¹⁴ This disorder rarely involves orbit and has no gender predilection.^{15,16} However orbital hydatid cyst can involve patients at any age, as young as 3 years old to elderly, but it is most frequent in younger ages.^{6,17} The sluggish growth of hydatid cyst result in capsular fibrosis formation surrounding the two layered wall of cyst and clear antigenic fluid.¹⁸ Its 2 layers includes outer laminated ectocyst as a barrier layer in avoiding tissue invasion and inner germinal layer which con-

Table 2. Orbital imaging findings in hydatid cyst

Type of orbital imaging	Findings
Ultrasonography ²⁷⁻²⁹	<ul style="list-style-type: none"> • Double wall sign, attaching sand to the wall of the cyst • Numerous echoes within a anechoic cystic lesion • Multiseptate cystic lesion with anechoic appearance
CT scan ^{5,27-29}	<ul style="list-style-type: none"> • cystic lesion with distinct and regular border that may causes bowing and thinning of the bone
MRI ^{6,8,13,14,30,31}	<ul style="list-style-type: none"> • hypointense cystic lesion on T1 weighted • hyper intense on T2 weighted • marginal ring enhancement • fat suppression facilitates the diagnosis of cyst capsule and intraconal and retrobulbar lesions.

tains the daughter cysts.^{9,18} The daughter cysts create tape worm by producing scolices,¹⁸ so the cyst removal without its wall damage would prevent spread of scolices and it's possible recurrence. Although orbital involvement is so infrequent, it may involve different locations within the orbit.¹⁹ Intraconal and superior location were reported as the most frequent sites respectively, in contrary to low rate of invasion within the inferior orbital position, subretinal, extraocular muscle, vitreous and anterior chamber.^{5-8,13} In our patients, hydatid cyst was found within medial rectus muscle, intraosseous of lateral orbital wall, lacrimal gland, extraconal and intraconal. Orbital cavity has a limited volume and expansion as it is surrounded by bony walls and as the cysts grow larger by the rate of about 1 – 1.5 cm per year so they would become symptomatic less than 2 years.^{20,21} The location and size of the cyst determine the clinical features.¹⁵ Accordingly the intraconal location as the most common site in combination with growth rate of the cyst make the unilateral progressive proptosis as the most frequent presenting finding.^{6,12,20} Other findings of mass effect such as limited ocular motility, diplopia, optic disc swelling and optic atrophy with consequent abnormal papillary defect, palpebral mass, erosion of orbital bone, and periorbital pain or pain with ocular movement may be observed.^{1,17,18,22} Incomplete or total visual loss, conjunctival injection, chemosis, retinal vein engorgement, hypopyon, and eyelid edema might be its later manifestations.^{12,18,23-25} All our patients referred with complaint of proptosis and only two of them had clinical finding of visual impairment corresponding to their optic nerve swelling. The clinical findings in association of orbital imaging could bring up the diagnosis of orbital hydatid cyst before surgical exploration. Considering orbital abscess, dermoid and epidermoid cyst, congenital cyst with microphthalmos, mucocele, hematic cyst, lymphangioma, teratoma and other orbital cystic lesions as differential diagnosis of hydatid cyst in orbital imaging, the definite diagnosis is resulted by pathologic assessment.^{6-8,18,26} The double layered cyst with the existence of protoscolices that stuck to the inner layer and outer layer with specification of positive periodic acid Schiff (PAS) is reported in the pathologic investigations.⁹ Granulomatous reaction in association with eosinophilia was observed in a case of cyst content leakage.²⁶ The table 2 illustrate the findings of orbital imaging in the hydatid disease. Although the orbital imaging findings suggest the diagnosis of hydatid cyst, but the exact diagnosis is confirmed by pathologic evaluation. We surveyed all patients with CT and MRI.

The reliability of the hematologic tests is questionable due to high rate of normal reports especially in the presence of in-

tact cyst.^{5,17} On the other hand these hematologic tests including eosinophilia, elevated ESR and positive serologic test are often nonspecific findings with lower rate of positive results in orbital disease in comparison to the hepatic involvement.^{1,2,5} In our patients, only 2 of them (33 %) had positive serologic findings. The positive serologies may be found in only half of the patients with orbital involvement in spite of high positive rate of 98 % in existence of hepatic involvement.^{2,3} All our patients had isolated orbital hydatid cyst like the cases of Bagheri, et al.²⁸ The involvement of other organs may be existed in spite of normal imaging evaluation and more follow up time may be needed for detection of hydatid cyst in other site of body. Traditionally surgery is treatment of choice. The cyst removal by operation, PAIR (puncture-aspiration-injection-reaspiration), and medical treatment, separately or in combination are available for management of hydatid cysts.^{17,24,28-33} It should be emphasized that PAIR is not applicable in orbital hydatid cyst and the best option is surgical removal in these cases. Bagheri, et al.²⁸ have reported some success for PAIR method in orbital hydatid cyst while acknowledging that surgical removal remains the best option in these patients. The surgical removal of the hydatid cyst is the major method⁵ in treatment of this disease that different approaches (fronto-orbital, lateral orbitotomy, transcranial, transconjunctival, inferior orbitotomy, lateral rhinotomy, percutaneous, transmaxillary, enucleation),^{1,6,15,31,34,35} may be used from patient to patient according to several factors like the exact location of the cyst in the orbit, accessibility of lesion, and it's extension.

Albendazole (10 mg/kg) or Mebendazole (30 mg/kg) daily²³ are used as medical treatment which are more effective if combined with Praziquantel.^{16,36-38} Starting the medication 2 – 4 weeks before surgery is suggested,^{6,17} while it should be especially prescribed in patients with cyst rupture to prevent it's possible dissemination.^{17,39,40} Allowing for more penetration of Albendazole through the cyst wall in comparison to Mebendazole caused ensuing higher efficacy.^{15,17,41-45} Benzimidazole/carbamates is recommended for recurrence avoidance following cyst rupture.^{17,41,46}

We treated all patients with surgical approach concomitant with post operative oral medication of Albendazole. Although the most common site of involvement of orbital hydatid cyst is intraconal, it can involve any other location within the orbit.

References

1. Kiratli H, Bilgiç S, Oztürkmen C, Aydın O. Intramuscular hydatid cyst of the medial rectus muscle. *Am J Ophthalmol.* 2003; **135**(1): 98 – 99.
2. Flament J, Storck D. *Oeil et pathologie gé'nérale.* Paris: Masson,

- 1997; 694 – 695.
3. Xiao A, Xuelyi C. Hydatid cyst of the orbit in Xinjiang: a review of 18 cases. *Orbit*. 1999; **18**: 151 – 155.
 4. Chaabouni M, Ben Zina Z, Ben Ayez H, Tounsi R, Trigui A, Ben Mansour H. Hydatid orbital cyst: a unique intra-orbital locality. A case report. *J Fr Ophthalmol*. 1999; **22**(3): 329 – 334.
 5. Morales AG, Coxatto JO, Crovetto L, Ebner R. Hydatid cysts of the orbit: A review of 35 cases. *Ophthalmology*. 1988; **95**: 1027 – 1032.
 6. Somay H, Emon ST, Orakdogan M, Berkman MZ. A primary orbital hydatid cyst. *J Clin Neurosci*. 2012; **19**(6): 898 – 900.
 7. Mehta M, Sen S, Sethi S, Pushker N, Bajaj MS, Chandra M. Large orbital hydatid cyst presenting as subconjunctival mass. *Ann Trop Med Parasitol*. 2010; **104**: 601.
 8. Limaiem F, Bellil S, Bellil K, Chelly I, Mekni A, Khaldi M, et al. Primary orbital hydatid cyst in an elderly patient. *Surg Infect (Larchmt)*. 2010; **11**: 393 – 395.
 9. Azfar Siddiqui MA, Ali Rizvi SW, Ali Rizvi SA, Ahmad I, Ullah E. Atypical multifocal hydatid disease of cranial vault: simultaneous orbital and extradural meningeal involvement. *Emergency Radiology*. 2010; **17**(5): 427 – 430.
 10. McMannus DP, Zhang W, Bartley PB. Echinococcosis. *Lancet*. 2003; **362**: 1295 – 1304.
 11. Khuroo MS. Hydatid disease: Current status and recent advances. *Ann Saudi Medicine*. 2002; **22**: 56 – 64.
 12. Akata D, Bilgic S, Akata D, Kiratli H, Ozmen MN. Percutaneous treatment of an orbital hydatid cyst: A new therapeutic approach. *American Journal of Ophthalmology*. 1998; **125**(6): 877 – 879.
 13. Aksoy FG, Tanrikulu S, Kosar U. Inferiorly located retrobulbar hydatid cyst: CT and MRI features. *Comput Med Imaging Graph*. 2001; **25**(6): 535 – 540.
 14. Rokni MB. The present status of human helminthic diseases in Iran. *Ann Trop Med Parasitol*. 2008; **102**: 283 – 295.
 15. Gokhale SK, Sane VD, Ramanojam S, Gadre PK, Gadre KS, Deshmukh SD. Ipsilateral keratoconus associated with long-standing primary hydatid cyst of the orbit. *J Craniofac Surg*. 2012; **23**(4): e344 – e347.
 16. Onerci M, Turan E, Ruacan S. Submandibular hydatid cyst: A case report. *J Craniomaxillofac Surg*. 1991; **19**(8): 359 – 361.
 17. Jimenez-Mejías ME, Alarcón-Cruz JC, Márquez-Rivas FJ, Palomino-Nicás J, Montero JM, Pachón J. Orbital hydatid cyst: treatment and prevention of recurrences with albendazole plus praziquantel. *J Infect*. 2000; **41**(1): 105 – 107.
 18. Sevel D, Sapeika RJ. Hydatid cyst of the orbit. *Surv Ophthalmol*. 1977; **22**(2): 101 – 105.
 19. Ghosh A, Saha S, Jha A, Chandra S. Orbital hydatid cyst - A rare case report. *Indian J Otolaryngol Head Neck Surg*. 2008; **60**(2): 191 – 193.
 20. Rastogi A, Arora R, Chaturvedi K. Orbital hydatid cyst: an unusual presentation. *Orbit*. 1998; **17**: 107 – 111.
 21. Available from: URL: <http://misc.medscape.com/pi/android/medscapeapp/html/A216432-business.html>
 22. Diren HB, Ozcanli H, Boluk M, Kilic C. Unilocular orbital, cerebral and intraventricular hydatid cysts: CT diagnosis. *Neuroradiology*. 1993; **35**: 149 – 150.
 23. Benazzou S, Arkha Y, Derraz S, El Ouahabi A, El Khamlichi A. Orbital hydatid cyst: review of 10 cases. *J Craniomaxillofac Surg*. 2010; **38**(4): 274 – 278.
 24. Nahri GE. A simplified technique for removal of orbital hydatid cysts. *Br J Ophthalmol*. 1991; **75**: 743 – 745.
 25. Talib H. Orbital hydatid disease in Iraq. *Br J Surg*. 1972; **59**: 391 – 394.
 26. Betharia SM, Sharma V, FRCS (Edin), Pushker N, Ultrasound Findings in Orbital Hydatid Cysts. *Am J Ophthalmol*. 2003; **135**: 568 – 570.
 27. Karakas HM, Tokoglu F, Kacar M, Boyacigil S. Retrobulbar hydatid cyst: assessment of two cases. *Australas Radiol*. 1997; **41**: 179 – 180.
 28. Bagheri A, Fallahi MR, Yazdani S, Rezaee Kanavi M. Two different presentations of orbital echinococcosis: a report of two cases and review of the literature. *Orbit*. 2010; **29**(1): 51 – 56.
 29. Sharma R, Vashisht S, Berry M. Orbital hydatid cysts: sonographic and CT appearance. *Australas Radiol*. 1995; **39**: 246 – 248.
 30. Sanli M, Sabuncuoğlu H, Keskin T. Primary intraorbital hydatid cyst: an unusual location, case report and review of the literature. *Minim Invasive Neurosurg*. 2007; **50**(6): 367 – 369.
 31. Turgut AT, Turgut M, Kosar U. Hydatidosis of the orbit in Turkey: results from review of the literature 1963 – 2001. *Int Ophthalmol*. 2004; **25**: 193 – 200.
 32. Akhan O, Bilgiç S, Akata D, Kiratli H, Ozmen MN. Percutaneous Treatment of an Orbital Hydatid Cyst: A New Therapeutic Approach. *Am J Ophthalmol*. 1998; **125**: 877 – 879.
 33. Kahveci R, Sanli AM, Güler B, Sekerci Z. Orbital hydatid cyst. *J Neurosurg Pediatr*. 2012; **9**(1): 42 – 44.
 34. Al-Muala HD, Sami SM, Shukri MAR, Hasson HK, Alaboudy TA. Orbital hydatid cyst. *J Infection*. 2012; **2**: 197 – 199.
 35. Selçuklu A, Öztürk M, Külahlı I, Doğan H. Successful Surgical Management of an Intraorbital Hydatid Cyst through a Transmaxillary Approach: Case report. *Skull Base*. 2003; **13**(2): 101 – 105.
 36. Salto E, Juárez E, Roiz MP, Abad J. Quimioterapia combinada (mebendazol más praziquantel) en pacientes con hidatidosis. *Enf Infecc Microbiol Clin*. 1991; **9**: 527 – 529.
 37. Taylor DH, Morris DL. Combination chemotherapy is more effective in postspillage prophylaxis for hydatid disease than either albendazole or praziquantel alone. *Br J Surg*. 1989; **76**: 954.
 38. Yasawy MI, Al-Karawi MA, Mohamed AR. Combination of praziquantel and albendazole in the treatment of hydatid disease. *Trop Med Parasitol*. 1993; **44**: 192 – 194.
 39. Ergün R, Ökten A, Yüksel M, Gül B, Evliyaoğlu C, Ergünger F, et al. Orbital hydatid cysts: Report of four cases. *Neurosurg Rev*. 1997; **20**: 33 – 37.
 40. Cooney RM, Flanagan KP, Zehyle E. Review of surgical management of cystic hydatid disease in a resource limited setting: Turkana, Kenya. *Eur J Gastroenterol Hepatol*. 2004; **16**: 1233 – 1236.
 41. Morris DL, Richards KS. Hydatid disease: current medical and surgical management. Oxford: Butterworth-Heinemann Ltd, 1992.
 42. Davis A, Dixon H, Pawlowski Z. Multicentre clinical trials of benzimidazole carbamates in human cystic echinococcosis (phase 2). *Bull World Health Organ*. 1989; **67**: 503 – 508.
 43. Ellis M, Von Sinner W, Al-Hokail, Sieck J. A clinical–radiological evaluation of benzimidazoles in the management of Echinococcus granulosis cysts. *Scand J Infect Dis*. 1992; **24**: 1 – 13.
 44. Todorov T, Vutova K, Mechkov G, Georgiev P, Petkov D, Tonchev Z, et al. Chemotherapy of human cysts echinococcosis: comparative efficacy of mebendazole and albendazole. *Ann Trop Med Parasitol*. 1992; **86**: 59 – 66.
 45. Horton RJ. Albendazole in treatment of human cystic echinococcosis: 12 years of experience. *Acta Trop*. 1997; **64**: 79 – 93.
 46. Morris DL, Taylor DH. Optimal timing of post-operative albendazole prophylaxis in Echinococcus granulosis. *Ann Trop Med Parasitol*. 1988; **82**: 65 – 66.