The Journal of Laryngology & Otology (2010), ***124***, *456–459*.

doi:10.1017/S0022215109991927

Clinical Record

Infratemporal hydatid cyst: a case presenting with blindness

N Yazdani*, A Basam*, M Heidarali*, S Sharif Kashani*†, M Hasibi‡, Z Mokhtari*

Abstract

Objective: We report a very rare case of a hydatid cyst in the infratemporal fossa, causing visual loss over a 10-day period, which disappeared with rapid surgical and medical treatment.

Case report: A 14-year-old girl presented with right exophthalmos and visual loss. Over a 10-day period, her visual acuity had decreased to detection of hand motion only, due to pressure on the optic nerve caused by a parapharyngeal cyst pressing through a inferior orbital fissure on the right side. A craniotomy had previously been performed for a right frontoparietal hydatid cyst. The patient had been treated intermittently with albendazole. The patient was primarily diagnosed with hydatid cyst, on the basis of her previous medical history and radiological findings, and underwent surgery. Three cysts were carefully removed from the right maxillary sinus, via a standard Caldwell–Luc approach, and the surgical area was irrigated with hypertonic saline.

Conclusion: Infratemporal hydatidosis is very rarely reported in the world literature, although hydatid cysts are endemic in many countries, including Iran. We discuss the common presenting features, investigation and treatment options for infratemporal hydatosis. Constant evaluation of adjacent organs is necessary, with treatment as required, due to the propensity of hydatidosis to recur in essential organs. Immediate surgery is recommended, both to prevent the development of disease and to improve the prognosis.

Key words: Hydatid Cysts; Echinococcus Granulosus; Infratemporal Fossa

Introduction

The occurrence of a hydatid cyst in the infratemporal region is rare, with only a few cases reported in the English language literature.[1–3] Hydatidosis occurs all over the world, but it is especially common in the cattle-raising communities of Africa, Australia, New Zealand and South America, and is endemic in Turkey and the Middle East.[3,4]

The majority of hydatid cysts appear in the liver (65 per cent) and lungs (25 per cent). The presence of hydatid cysts in other organs is less common, and only 2 per cent of cases involve the maxillofacial region.[5] Such cases commonly involve cystic lesions located in the mandible, maxillary sinus, orbit, infratemporal fossa, pterygopalatine fossa, parapharyngeal space, tongue, parotid gland or submandibular salivary gland.[1,2,6–8]

Hydatid disease of the infratemporal fossa is extremely rare; to our knowledge, only a few cases have been reported.[7,9–11] Primary involvement of the maxillary sinuses is very rare, and very few cases have been reported.[12]

We report a unique case of a 14-year-old girl in whom a hydatid cyst developed in the right infratemporal region, with the only presenting sign being visual loss over a 10-day period; the patient's vision recovered rapidly following surgical removal of cysts.

Case report

A 14-year-old girl presented with right exophthalmos and a two weeks history of visual loss.

Due to pressure on the optic nerve caused by a parapharyngeal cyst pressing through a inferior orbital fissure on the right side, Ophthalmological examination showed limited medial and inferior oblique muscle movement.

The patient's visual acuity of right eye had decreased to detection of hand motion only while the left eye was normal. The anterior, vitreal and fundus examinations of both eyes were normal.

The patient had a history of seizures treated with carbamazepine for two years (from five to seven years of age). During the most recent investigation, multiple hydatid cysts had been noted in the patient's brain. She had been referred to our otolaryngology centre in April 2006 by a neurosurgeon who had performed craniotomies on the patient seven, five and three years earlier. At time of referral, the right frontoparietal lobe had been affected. The patient had been treated intermittently with albendazole.

Examination of the patient's nervous system revealed normal and intact cranial nerves.

While the patient had been receiving treatment with albendazole, a computed tomography (CT) scan of the paranasal sinus had shown multiple cystic masses in the right infratemporal fossa, which were eroding the posterior wall of the maxillary sinus and inferior orbital wall and compressing the orbit (Figure 1).

A parasagittal, T2-weighted magnetic resonance imaging (MRI) scan showed a well defined, homogeneous, high signal, multilocular lesion in the right infratemporal fossa, with some extension within the right maxillary sinus antrum. Intracranial extension was not seen in this scan (Figure 2).

From the *Otorhinolaryngology Research Centre, and the Departments of †Radiology and ‡Infectious Diseases, Amir-Alam Hospital, Tehran University of Medical Sciences, Iran.
Accepted for publication: 23 July 2009. First published online 27 November 2009.

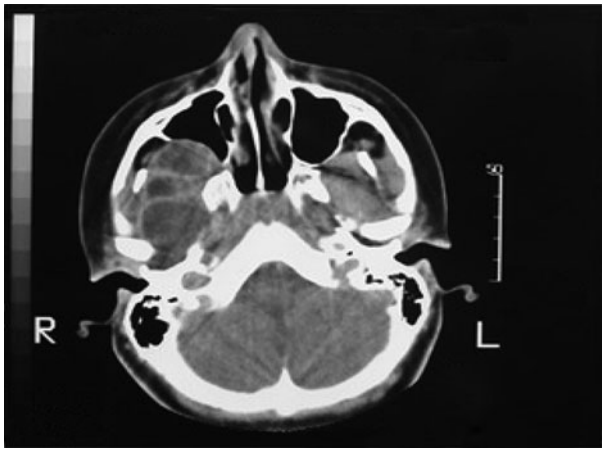

FIG. 1

A multilocular, hypodense lesion in the right infratemporal fossa, with expansion of the fossa and compression of the pterygoid muscles. Erosion of the right maxillary sinus posterior wall is also seen, with some extension within its antrum.

An axial, T2-weighted MRI scan showed a well defined, homogeneous, extraconally located, multilocular lesion in the right infraorbital fissure, which was seen to be expanding and exerting a mass effect (Figure 3). Proptosis was not seen.

Computed tomography scans of the patient's brain showed the impact of previous craniotomies, together with hypodense defects in the right frontoparietal region.

Laboratory investigations were undertaken, revealing a positive enzyme-linked immunosorbent assay for anti hydatid antibody and 5 per cent eosinophils in the peripheral blood smear. Surprisingly, chest X-rays and liver sonography were normal.

The patient received albendazole 400 mg for three weeks, ceasing one week before surgery.

The patient underwent surgery with a primary diagnosis of a hydatid cyst, on the basis of her previous medical history and radiological findings. Three hydatid cysts were found (2, 1 and 1 cm in diameter); all were encased in a capsule, and extended superiorly from the posterior wall of the maxillary sinus to the infratemporal fossa. These cysts were removed easily and completely from the right maxillary sinus, without any spillage, via a standard Caldwell–Luc approach. During surgery, gauze soaked with 2 per cent formalin was placed around the cysts and iodoform gauze was packed into the antrum and oral cavity. After cyst removal, the surgical area was irrigated with hypertonic saline.

Macroscopically, the yellow-white colour and elastic nature of the cyst walls suggested parasitic disease.

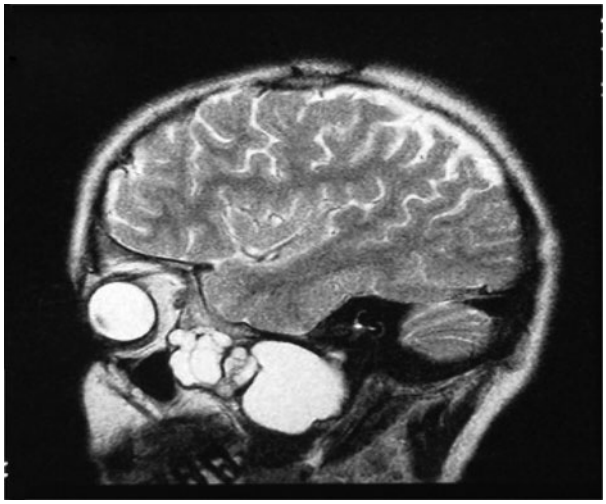

FIG. 2

Parasagittal, T2-weighted magnetic resonance imaging scan, showing a well defined, homogeneous, multilocular lesion in the right infratemporal fossa, with some extension within the right maxillary sinus antrum. Intracranial extension is not seen in this scan.

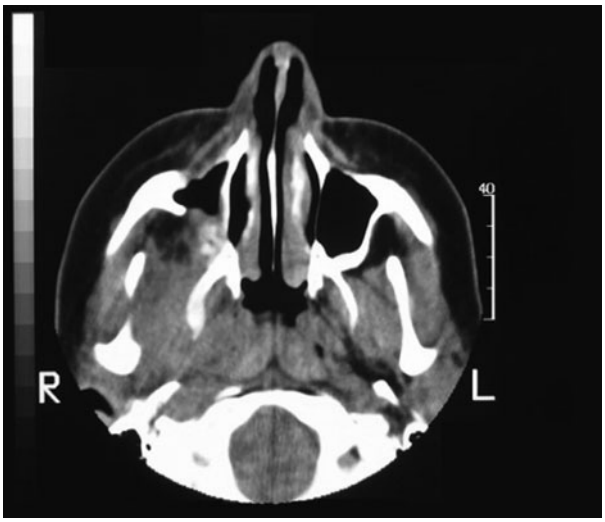

FIG. 4

Axial, paranasal sinus computed tomography scan taken after surgery. No evidence of the lesion is seen in the right infratemporal fossa. Some post-operative inflammation is seen. A residual defect is present in the right maxillary sinus posterior wall, due to the lesion.

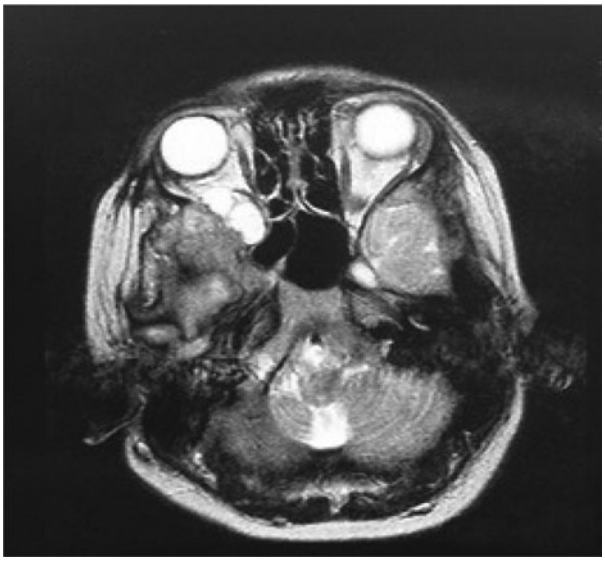

FIG. 3

Axial, T2-weighted magnetic resonance imaging scan (orbital section), showing a well defined, homogeneous, extraconally located, multilocular lesion in the right infraorbital fissure, which is seen to be expanding and exerting a mass effect on the contents of this anatomical area.

https://doi.org/10.1017/S0022215109991927 Published online by Cambridge University Press

Histopathological examination revealed a basophilic cuticular membrane consistent with a hydatid cyst.

Post-operatively, the patient's visual acuity improved considerably over just two weeks.

A six-month course of albendazole (15 mg/kg/day) was prescribed to prevent recurrence of hydatidosis.

Computed tomography scans were performed four, 12 and 18 months later. No evidence of any lesion in the right infratemporal fossa was seen. Some post-operative inflammation was noted. A residual defect in the right maxillary sinus posterior wall, due to the lesion, was noted (Figure 4).

At the patient's last follow-up consultation, two years after surgery, she showed no evidence of hydatidosis.

Discussion

Hydatid disease is a parasitic infestation in humans caused by the larval form of *Echinococcus granulosus*. Infratemporal hydatid disease can only occur when parasitic embryos succeed in passing through the hepatic and pulmonary filtering systems.[9,13,14]

Hydatid cysts in the maxillofacial region are extremely rare, comprising only 2 per cent of reported cases of hydatidosis. They commonly appear as cystic lesions located in the mandible, maxillary sinus, orbit, infratemporal fossa, pterygopalatine fossa, parapharyngeal space, tongue, parotid gland or submandibular salivary gland.[4,7,9,10,15]

At the time of presentation, our patient showed no evidence of hydatid cysts elsewhere in the body, other than the brain. A maxillofacial hydatid cyst had been present during the period when the patient had received intermittent medical therapy for brain hydatosis.

- **Hydatid disease is a parasitic infestation in humans caused by the larval form of *Echinococcus granulosus***
- **This paper describes a case of a hydatid cyst in the infratemporal fossa, with visual loss**
- **The most effective treatment for hydatidosis is surgical removal**
- **Immediate surgery is necessary when visual loss occurs**

The growth rate of hydatid cysts is highly variable, ranging from 1 to 5 cm a year. The body's tolerance of hydatid cysts, and their effect on function, depends on their growth rate and size.[7] In our patient, a slow-growing cyst had displaced the zygomatic arch, mandibular ramus and maxillary bone, causing facial asymmetry. The cysts had also eroded the inferior wall of the orbit, due to a mass effect, leading to visual loss. During surgery, three hydatid cysts (2, 1 and 1 cm in diameter) were removed completely from the right maxillary sinus and infratemporal fossa, without spillage, via a standard Caldwell–Luc approach. Silver nitrate (0.5 per cent), hypertonic saline (20 per cent) and other chemicals are often injected into hydatid cysts at the time of surgery to inactivate the protoscolices. We did not inject any chemicals into the cysts at the time of surgery.

In the present case, the diagnosis of echinococcus infestation was based on the clinical history, physical examination, and radiological and histopathological investigation. The use of serological tests (such as indirect haemagglutination, latex agglutination, enzyme-linked immunosorbent assay and immunoelectrophoresis) in the diagnosis of hydatosis is controversial. Both false positive and false negative results are common.[1,16] Traditional serological tests are not sufficiently sensitive to establish a diagnosis or to be useful for follow up of patients with hydatid disease. The Casoni intradermal test and the Weinberg complement fixation test frequently give false negative results.[17,18]

We did not depend on such tests but, rather, undertook CT and MRI scanning, which showed typical features of the hydatid lesion and its location, enabling more accurate diagnosis. The radiologist was able to determine the size and anatomical relations of the cystic lesion, and to inform the otolaryngologist of the possibility of hydatid disease. The most effective treatment for hydatid cysts is surgical removal; therefore, the extent of the lesion must be carefully evaluated to determine the best treatment strategy. Computed tomography and MRI scanning can be used to guide hypertonic saline infusion and cyst fluid aspiration (to decrease cyst size). The surgeon can then decide either to remove the cyst completely or to administer chemotherapy, in cases of inoperable disease due to anatomical factors. Appropriate treatment and ongoing evaluation of the adjacent organs are necessary, due to the risk of recurrence. Immediate surgery is recommended both to prevent disease development and to improve the patient's prognosis.

References

1 Gangopadhyay K, Abuzeid MO, Kfoury H. Hydatid cyst of the pterygopalatine-infratemporal fossa. *J Laryngol Otol* 1996;**110**:978–80
2 Hotz MA, Gottstein B. Cystic echinococcosis of the parapharyngeal space: case report with a 20-year follow-up. *J Oral Maxillofac Surg* 1999;**57**:80–3
3 Turgut S, Ensari S, Katirci H, Çelikkanat S. Rare otolaryngologic presentation of hydatid cyst. *Otolaryngol Head Neck Surg* 1997;**117**:418–21
4 Bouckaert MM, Raubenheimer EJ, Jacobs FJ. Maxillofacial hydatid cysts. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2000;**89**:338–42
5 Amr SS, Amr ZS, Jitawi S, Annab H. Hydatosis in Jordan: an epidemiological study of 306 cases. *Ann Trop Med Parasitol* 1994;**88**:623–7
6 Akyildiz AN, Ozbilen MS, Goksu N. Hydatid cyst of the pterygopalatine fossa. *J Oral Maxillofac Surg* 1991;**49**:87–8
7 Sennaroglu L, Önerci M, Turan E, Sungur A. Infratemporal hydatid cyst – unusual location of echinococcosis. *J Laryngol Otol* 1994;**108**:601–3
8 Zaidi M. An unusual case of orbital hydatid cyst: a surgical emergency. *Ann Plast Surg* 1999;**42**:327–9
9 Kalovidouris A, Gouliamos A, Andreou I, Ioannovits I, Levett J, Vlahos I *et al*. Primary hydatid disease of the infratemporal fossa and the parotid gland. *Radiologe* 1985;**25**:235–6
10 Sener NR. Hydatid cyst in the infratemporal fossa. MRI demonstration. *Clin Imaging* 1994;**18**:213–15
11 Shuker S. Hydatid cyst in the maxillofacial region. *J Oral Maxillofac Surg* 1982;**40**:171–4
12 Mortazavi SH, Motamedi MH. Primary cyst of the maxillary sinus: case report. *Gen Dent* 2008;**56**:444–6
13 Rudwan MA, Khaftaji S. CT of cerebral hydatid disease. *Neuroradiology* 1988;**30**:496–9
14 Sharma SC, Ray RC. Primary hydatid disease of the brain in adult: report of a case. *Neurosurgery* 1988;**23**:374–6
15 Onerci Turan E, Ruacan S. Submandibular hydatid cyst: a case report. *J Craniomaxillofac Surg* 1991;**19**:359–61
16 Schipper HG, Kager PA. Diagnosis and treatment of unilocular hydatid disease (*Echinococcus granulosus* infection) [in Dutch]. *Ned Tijdschr Geneesk* 1997;**141**:984–9
17 Dogăn R, Yüksel M, Cetin G, Süzer K, Alp M, Kaya S *et al*. Surgical treatment of hydatid cysts of the lung: report on 1055 patients. *Thorax* 1989;**44**:192–9

https://doi.org/10.1017/S0022215109991927 Published online by Cambridge University Press

18 Bret PM, Fond A, Bretagnolle M, Valette PJ, Thiesse P, Lambert R *et al*. Percutaneous aspiration and drainage of hydatid cysts in the liver. *Radiology* 1988;**168**:617–20

Address for correspondence:
Dr S Sharif Kashani,
Radiology Department,
Amir-Alam Hospital,
Tehran University of Medical Sciences,
Sadi St, Enghelab Ave,
Tehran, Iran.

Fax: 00982166704805
E-mail: Sh_Sh_md_rad@yahoo.com

Dr Nasrin Yazdani takes responsibility for the integrity of the content of the paper.
Competing interests: None declared