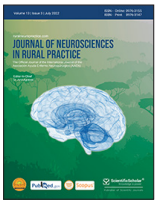
ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

https://ruralneuropractice.com

Journal of Neurosciences in Rural Practice



Original Article

Hydatid disease of central nervous system, a clinicopathological study of 33 cases

Nasir Ud Din[1], Muhammad Raza[1], Shabina Rahim[1], Wasim Memon[2], Zubair Ahmad[1]

Departments of [1]Pathology and Laboratory Medicine and [2]Radiology, Aga Khan University Hospital, Karachi, Pakistan.

ABSTRACT

Objectives: Involvement of central nervous system (CNS) by Hydatid cyst is rare comprising 0.5–4% of all hydatid cysts and principally affecting those younger than 20 years, giving rise to cystic masses mostly in the cerebral hemispheres. To report the clinicopathological findings of CNS hydatid cysts, we diagnosed and review the findings of the previous studies.

Materials and Methods: All cases reported in our Section between January 1, 2001, and June 30, 2022, were included in the study. By searching our files, cases were retrieved, and diagnosis was confirmed. Follow-up was received on telephone. Ethical exemption was obtained.

Results: Thirty-three cases were diagnosed. Almost all were received from rural areas. There were 17 females and 16 males. Mean and median age were 20 and 19 years, respectively. Over 60% were younger than 20 years of age. All 33 involved the cerebral and cerebellar hemispheres. Seventy six percent were supratentorial while 24% were infratentorial. The most common signs and symptoms included weakness, headaches, and seizures. All appeared as solitary cystic masses on imaging. Almost 67% were clinically suspected to be hydatid cysts. Grossly, thin-walled transparent unilocular or multilocular cysts filled with viscous material were received intact in 52% and in multiple pieces in 48% cases. Intact cysts measured 7 cm on average. All demonstrated typical histology. Of the nine patients whose follow-up was available, one died from unspecified acute surgery related complications. Four patients were asymptomatic at the time of follow-up, whereas four developed recurrent cysts. All eight received albendazole therapy.

Conclusion: Cerebellum/posterior fossa location was common. Several cases were received in multiple pieces with increased risk of recurrence. Clinicopathological features were similar to those reported in literature. This series will hopefully serve to increase awareness regarding CNS hydatid disease.

Keywords: Central nervous system, Hydatid cyst, Cerebrum, Cerebellum

INTRODUCTION

Hydatid disease (Echinococcosis) occurs globally but is endemic in sheep and cattle breeding countries. It is especially common in the Middle East, South America, Central and South Europe, Mediterranean region, Australia, and New Zealand. The etiological agent is the tapeworm Taenia, genus Echinococcus, most commonly Echinococcus granulosus. Various carnivores, especially dogs, are the definitive hosts (dog tapeworm). The larva of the parasite resides in the gut of the definitive host. Humans are intermediate hosts and get infected accidentally via the feco-oral route or by direct contact with dogs. The ova migrate through the portal vessels to the liver. Liver and lungs are the organs most affected, accounting for 75% and 15% of cases, respectively. Passage through these first filters or bypassing of these filters by the larvae using lymphatics can lead to blood-born or lymphatic spread. Thus, any part of the body including brain, bones, spleen, kidneys, gallbladder, soft tissue including skeletal muscle may be affected by the cyst form of this parasite. Intracranial hydatid disease is rare and comprises 0.5–4% of all cases.[1-9]

Pakistan is an agricultural country. Hydatid disease is relatively common. We aim to report cases involving central nervous system (CNS) diagnosed over a 22-year period. We describe the clinical and histological features and discuss findings of the previous studies.

MATERIALS AND METHODS

We searched our files for cases involving the CNS diagnosed between January 1, 2001 and June 30, 2022. Cases were reviewed by two senior authors and histopathological

***Corresponding author:** Muhammad Raza, Department of Pathology and Laboratory Medicine, Aga Khan University Hospital, Karachi, Pakistan. raza.sajjad@aku.edu

Received: 11 November 2022 **Accepted:** 14 February 2023 **EPub Ahead of Print:** 16 March 2023 **Published:** 03 May 2023 **DOI**: 10.25259/JNRP_51_2022

diagnosis was confirmed. Clinical information was retrieved from computer files. Follow-up was obtained through telephone. Exemption was obtained from the Institution's Ethical Review Committee.

RESULTS

Thirty-three cases were diagnosed. Eleven (33.3%) were received from Baluchistan province in Southwest Pakistan. Seventeen patients (51.5%) were females and 16 (48.5%) were males. Age range was 3–50 years. Mean age was 20 years and median age was 19 years. Twenty patients (60.6%) were younger than 20 while 15 (45.5%) were younger than 15 years of age. All 33 cases involved the cerebral or cerebellar hemispheres. Twenty-five (75.8%) were supratentorial while 8(24.2%) were infratentorial. The location-wise breakup is shown in [Table 1].

The most common signs and symptoms included weakness, paralysis, headache, and seizures. Loss of vision, headache, and vomiting were also reported. On imaging, all 33 cases appeared as solitary cystic hemispheric masses [Figures 1a, b, and 2a, b]. Hydatid cyst was suspected clinically in 22 patients (66.7%). In 4 patients (12%), clinical diagnosis was arachnoid cyst. Grossly, thin-walled transparent unilocular or multilocular cysts filled with viscous, sticky, gelatinous, and grey-white material were received intact in 17 patients (51.5%) and in multiple pieces in 16 (48.5%) [Figure 3a]. Intact cysts on average measured 7.0 cm in largest dimension while those received in multiple pieces measured on average 8.5 cm in aggregate. Sizes of cysts ranged from 1 cm to 22.5 cm. Histologically, all demonstrated the characteristic lamellated, chitinous material, and scolices [Figure 3b-d]. Calcification was seen in 4 cases (12.1%). None of the patients had any radiological evidence of concomitant cysts in the liver. No history of cyst rupture, spillage of cyst contents, or anaphylactic shock was provided.

Almost all cases were received from rural areas and follow-up proved extremely difficult to obtain. It was available in nine patients only despite repeated attempts. None were treated at our institution. The follow-up period in these nine patients ranged from 8 to 216 months. One patient died of unspecified surgery-related complications, a week after undergoing resection. The remaining eight were alive. Four had no history of recurrence and were asymptomatic. The other four had recurrent symptoms such as loss of vision in both eyes, facial palsy, hemiplegia, etc. and underwent repeated surgeries for excision of recurrent/residual cysts. All eight received 10–15 mg/km/day of albendazole not exceeding 800 mg/day, in two doses, for 3-month post-resection. Use of the drug in the four patients with multiple recurrences was associated with visible improvement in symptoms.

Table 1: Sites of central nervous system involvement.

S. No.	Location	Number of cases	Percentage
1.	Frontal lobe	3	9.1
2.	Frontoparietal lobe	4	12.1
3.	Parietal lobe	8	24.2
4.	Frontotemporal lobe	1	3
5.	Temporal lobe	3	9.1
6.	Temporoparietal lobe	1	3
7.	Parietooccipital lobe	3	9.1
8.	Temporoparietooccipital lobe	1	3
9.	Occipital lobe	1	3
10.	Cerebellum (posterior fossa)	8	24.2

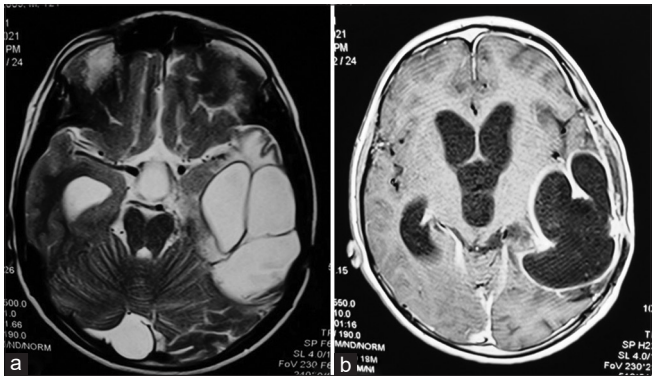


Figure 1: (a) T2WI showing septate lobulated cystic lesion with surrounding edema in the left temporoparietal lobe, (b) T1WI with contrast showing enhancement of thick walls of the cystic lesion.

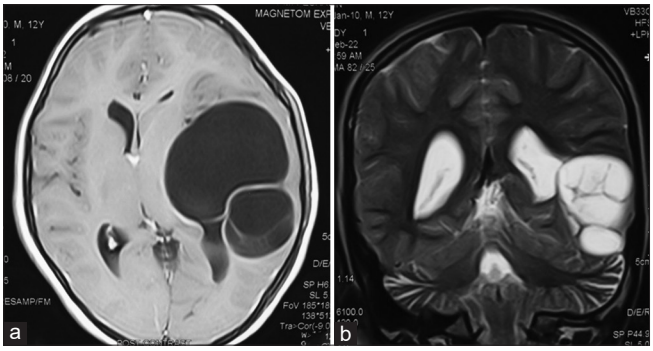


Figure 2: (a) T1WI with contrast showing large multiloculated cystic lesion with internal septa/daughter cysts in the left parietal lobe with significant edema and mass effect, (b) T2WI showing cystic lesion with multiple internal septa in the left parietal lobe.

DISCUSSION

Studies report incidence rates of 0.5–4%. In regions where hydatid disease is common, it may comprise as much as 10% of all intracranial expansile mass lesions. Intracranial hydatid cysts are mostly supratentorial involving the cerebral hemispheres and mostly affect patients younger than 20 years of age and mimic cystic cerebral masses both clinically and

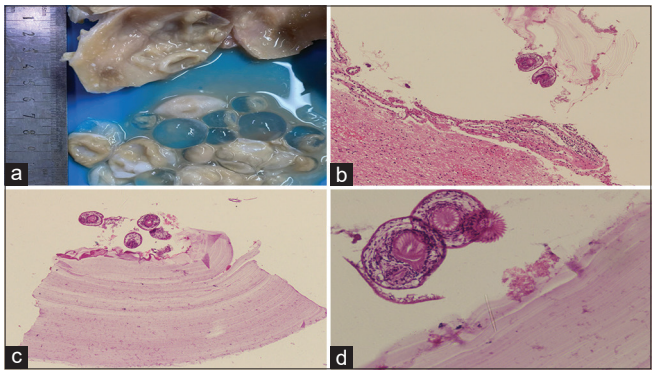


Figure 3: (a) Specimen shows fibrotic cyst wall with variably sized daughter cyst. (b) Reactive glial tissue covered by thin layer of inflammatory cells along with fragments of chitinous layer and protoscolices. (c and d) low- and high-power full thickness images of cyst wall with acellular laminated layer, nucleated layer, and protoscolices.

radiologically[1-4,9-21]. Headache, vomiting, seizures, loss of vision, papilledema, etc. are common clinical features.[3,9,13] In a recent series, the mean age was 11.5 years, seizure was the commonest symptom, majority of patients had a single cyst, and recurrence was seen in two patients.[3] In countries where hydatid disease is endemic, a high degree of suspicion is needed when confronted with intracranial cystic lesions on imaging (computerized tomography, CT scan and magnetic resonance imaging, MRI), to ensure early diagnosis.[2,8,13,19,22-28] On imaging, cysts may be non-complicated or complicated (pericystic edema present owing to rupture and leakage).[18] Non-complicated cysts are well-defined, isointense, and do not show pericystic edema or rim enhancement on MRI. Complicated cysts with superadded infections show hyperintense pericystic edema and complete or incomplete rim of enhancement which results from cyst rupture and these cysts are likely to recur.[18,28] In an MRI study of 16 cases by El-Shaman *et al.*, 10 cysts were simple and were excised intact. They did not show pericystic edema or contrast enhancement. However,[6] three solitary and three multiple were complicated (rupture occurred during surgery). All these six patients subsequently developed recurrence. All recurrent cysts showed pericystic edema and ring enhancement on MRI.[28] Primary CNS hydatid cysts are usually solitary and unilocular.[1,10] However, multiple cysts have been reported.[12,21,26]

In endemic regions, neurosurgeons must keep it in mind when confronted by cystic masses of the cerebral hemispheres mimicking abscess, arachnoid cyst, etc. in children and young adults.[13,23,26,29,30] Complete surgical excision is curative.[2,9,13,31,32] As prognosis following excision is good, early diagnosis is crucial[7] Incomplete excision and rupture at the time of surgery with spillage are the most common causes of recurrence and poor outcome. It is critical to excise the cyst un-ruptured and avoid any spillage as it can lead to anaphylaxis and disseminated infection. Pre-operative diagnosis is therefore extremely important.[2,8] To an experienced radiologist, CT findings are usually pathognomonic in the majority of cases.[25] Pre-operative diagnosis is more difficult in non-endemic areas.

Dowling's method is the most used resection procedure. It is based on wide exposure, meticulous cortical dissection, and excision of the cyst by hydro-dissection. Patient is placed in left lateral position. A frontoparietotemporal inverted-U incision is performed, and dura is opened. Head of the operating table is tilted slightly downward and by hydrodissection with saline, the cyst is carefully separated from adjacent brain parenchyma until it is removed entirely with intact capsule. Cavity is filled with isotonic saline and duroplasty is done.[33,34] The gap resulting from removal of the cyst is washed with sodium chloride[1,9,22] Apart from rupture and spillage, other surgery-related complications may rarely occur. Therefore, follow-up with MRI is essential.[35]

Post-operative medical treatment for 6–12 months is given in most cases.[1,11,12,14,16,18,22,28,36-38] Albendazole is the medical treatment of choice and is effective in recurrent cases or in cases where cysts are ruptured. Pre-operative albendazole has been advocated by some authors to prevent infection and reduce the risk of anaphylactic shock and recurrence.[1] Recommended albendazole treatment is 10–15 mg/kg/day in divided doses.[39-41]

CNS hydatid cysts are mostly supratentorial and posterior fossa is a rare location.[11,12,16,28] Hydatid cysts in the posterior fossa may be confused with arachnoid cyst on MRI.[42] In our study, posterior fossa involvement was common. Although the average size of cysts in our study was 7.5 cm, the largest cyst measured 22.5 cm in largest dimension. There are several published reports documenting giant CNS hydatid cysts.[1,9,10,21,35,43,44] Calcification was seen in four of our cases. There are several reports of calcification occurring in CNS hydatid cysts.[15,17,24,45,46] Even rarer reported locations include pons,[47,48] brainstem,[49] periventricular area,[50] cerebellopontine angle, and middle fossa.[51]

CONCLUSION

Cerebellum/posterior fossa location was common. Several cases were received in multiple pieces with increased risk of recurrence. Clinicopathological features were similar to those reported in the literature. This series will hopefully serve to increase awareness regarding CNS hydatid disease among neurosurgeons, radiologists, and pathologists.

Acknowledgment

Dr. Mujtaba Haidari, for providing gross pathology images.

Declaration of patient consent

The authors certify that they have obtained all appropriate consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Gök H, Başkurt O. Giant primary intracranial hydatid cyst in child with emiparesis. World Neurosurg 2019;129:404-6.
2. Padayachy LC, Dattatraya M. Hydatid disease (*Echinococcus*) of the central nervous system. Childs Nerv Syst 2018;34:1967-71.
3. Tanki H, Singh H, Raswan US, Bhat AR, Kirmani AR, Ramzan AU. Pediatric intracranial hydatid cyst: A case series with literature review. Pediatr Neurosurg 2018;53:299-304.
4. Gun E, Etit D, Buyuktalanci DO, Cakalagaoglu F. Unusual locations of hydatid disease: A 10-year experience from a tertiary reference center in Western Turkey. Ann Diagn Pathol 2017;29:37-40.
5. Taslakian B, Darwish H. Intracranial hydatid cyst: Imaging findings of a rare disease. BMJ Case Rep 2016;2016:bcr2016216570.
6. Senol MG, Tekeli H, Kendirli MT, Kaya S, Turhan V, Sonmez G, *et al*. Intramedullary hydatid cyst of the cervical spine. Indian J Med Microbiol 2012;30:480-1.
7. Kamath SM, Mysorekar VV, Rao SG, Varma RG. Intraventricular hydatid cyst in a child. Indian J Pathol Microbiol 2009;52:571-2.
8. Taratuto AL, Venturiello SM. Echinococcosis. Brain Pathol 1997;7:673-9.
9. Çavuş G, Açik V, Çavuş Y, Bilgin E, Gezercan Y, Ökten AI. An extraaxially localized intrasellar giant hydatid cyst with hypophyseal insufficiency. Childs Nerv Syst 2018;34:1391-6.
10. Kandemirli SG, Cingoz M, Olmaz B, Akdogan E, Cengiz M. Cerebral hydatid cyst with intraventricular extension: A case report. J Trop Pediatr 2019;65:514-9.
11. El Saqui A, Aggouri M, Benzagmout M, Chakour K, Chaoui ME. Cerebral hydatid cysts in children: About 15 cases. Pan Afr Med J 2017;26:205.
12. Lakhdar F, Arkha Y, Bougrine M, Derraz S, El Ouahabi A, El Khamlichi A. Posterior fossa extradural and extracranial hydatid cyst. Neurochirurgie 2010;56:391-4.
13. Ali M, Mahmood K, Khan P. Hydatid cysts of the brain. J Ayub Med Coll Abbottabad 2009;21:152-4.
14. Anvari M, Amirjamshidi A, Abbassioun K. Gradual and complete delivery of a hydatid cyst of the brain through a single burr hole, a wrong happening! Childs Nerv Syst 2009;25:1639-42.
15. Abderrahmen K, Aouidj ML, Kallel J, Khaldi MM. Calcified cerebral hydatid cyst. Neurochirurgie 2007;53:371-4.
16. Braham E, Bellil S, Bellil K, Chelly I, Mekni A, Haouet S, *et al*. Hydatid cyst of the posterior fossa. Med Mal Infect 2007;37:281-3.
17. Bouaziz M. Calcified cerebral hydatid cyst: A case report. Sante 2005;15:129-32.
18. Erşahin Y, Mutluer S, Demirtaş E, Yurtseven T. A case of thalamic hydatid cyst. Clin Neurol Neurosurg 1995;97:321-3.
19. Hamdi A, Ayachi R, Gargouri R, Mourad A. Hydatid cyst of the brain. Apropos of a series of 14 cases. Ann Chir 1990;44:226-30.
20. Gökalp HZ, Erdoğan A. Hydatid cyst of the aqueduct of Sylvius. Case report. Clin Neurol Neurosurg 1988;90:83-5.
21. Basarslan SK, Gocmez C, Kamasak K, Ceviz A. The Gigant primary cerebral hydatid cyst with no marked manifestation: A Case report and review of literature. Eur Rev Med Pharmacol Sci 2015;19:1327-9.
22. Duishanbai S, Jiafu D, Guo H, Liu C, Liu B, Aishalong M, *et al*. Intracranial hydatid cyst in children: Report of 30 cases. Childs Nerv Syst 2010;26:821-7.
23. Acha-Sánchez JL, Yaya-Loo HJ, Chipana-Sotomayor ME. Cerebral hydatid cyst: Clinical case and review of the literature. Neurocirugia (Astur) 2016;28:207-10.
24. Köktekir E, Erdem Y, Gökçek C, Karatay M, Yılmaz A, Bayar MA, *et al*. Calcified intracranial hydatid cyst: Case report. Turkiye Parazitol Derg 2011;35:220-3.
25. Abbassioun K, Rahmat H, Ameli NO, Tafazoli M. Computerized tomography in hydatid cyst of the brain. J Neurosurg 1978;49:408-11.
26. Popli MB, Khudale B. Primary multiple hydatid cysts of the brain. Australas Radiol 1998;42:90-1.
27. Pedrosa I, Saíz A, Arrazola J, Ferreirós J, Pedrosa CS. Hydatid disease: Radiologic and pathologic features and complications. Radiographics 2000;20:795-817.
28. El-Shamam O, Amer T, El-Atta MA. Magnetic resonance imaging of simple and infected hydatid cysts of the brain. Magn Reson Imaging 2001;19:965-74.
29. Bahloul K, Ouerchefani N, Kammoun B, Boudouara MZ. Unusual brain edema caused by an intracranial hydatid cyst: Case report and literature review. Neurochirurgie 2009;55:53-6.
30. Isikay S, Yılmaz K, Ölmez A. Two cases of rare cerebral hydatid cyst. Turkiye Parazitol Derg 2012;36:41-4.
31. Gezen F, Baysefer A, Köksel T, Gönül E, Akay KM, Erdogan E. Hydatid cysts of the brain. Clin Infect Dis 1995;21:938-42.
32. Guney O, Ozturk K, Kocaogullar Y, Eser O, Acar O. Submandibular and intracranial hydatid cyst in an adolescent. Laryngoscope 2002;112:1857-60.
33. Hage P, H Salle H, Ibrahim I, Khalil W. Hydatid cyst excision using Dowling's technique of hydrodissection. Acta Neurochir (Wien) 2022;164:2851-4.
34. Izci Y, Tüzün Y, Seçer HI, Gönül E. Cerebral hydatid cysts: Technique and pitfalls of surgical management. Neurosurg Focus 2008;24:E15.
35. Polat G, Ogul H, Sengul G. Hydatidosis following giant cerebral hydatid cyst operation. World Neurosurg 2018;118:14-5.
36. Yilmaz N, Kiymaz N, Etlik O, Yazici T. Primary hydatid cyst of the brain during pregnancy. Neurol Med Chir (Tokyo) 2006;46:415-7.
37. Sharma T, Khosla VK, Brar R. Primary extradural hydatid cyst associated with nephrotic syndrome in a pediatric patient. Childs Nerv Syst 2010;26:1247-9.
38. Tuzun Y, Solmaz I, Sengul G, Izci Y. The complications of cerebral hydatid cyst surgery in children. Childs Nerv Syst 2010;26:47-51.
39. Bildik N, Cevik A, Altintaş M, Ekinci H, Canberk M, Gülmen M. Efficacy of preoperative albendazole use according

to months in hydatid cyst of the liver. J Clin Gastroenterol 2007;41:312-6.
40. Arif SH, Shams-Ul-Bari, Wani NA, Zargar SA, Wani MA, Tabassum R, *et al*. Albendazole as an adjuvant to the standard surgical management of hydatid cyst liver. Int J Surg 2008;6:448-51.
41. Dehkordi AB, Sanei B, Yousefi M, Sharafi SM, Safarnezhad F, Jafari R, *et al*. Albendazole and treatment of hydatid cyst: Review of the literature. Infect Disord Drug Targets 2019;19:101-4.
42. Belfquih H, Azami MA, Akhaddar A. Rare case of hydatid cyst in the cerebellopontine cistern. World Neurosurg 2021;145:48-50.
43. Onal C, Erguvan R. Gigantic intracranial mass of hydatid cyst. Childs Nerv Syst 2002;18:199-200.
44. Ghaemi K, Masoudifar MA, Mehdi M, Solgi R, Kareshk AT. Giant brain hydatid cyst in an adult: A new case report. Turkiye Parazitol Derg 2021;45:76-9.
45. Safavi M, Dabiri S. Calcified cerebral hydatid cyst Presenting with seizure. Arch Iran Med 2019;22:102-3.
46. Bouali S, Boubaker A, Bouhoula A, Abderrahmen K, Said IB, Kallel J, *et al*. Calcified cerebral hydatid cyst. Tunis Med 2015;93:579-80.
47. Abbassioun K, Amirjamshidi A, Moinipoor MT. Hydatid cyst of the pons. Surg Neurol 1986;26:297-300.
48. Yilmazlar S, Aksoy K. Approach via the floor of the fourth ventricle for hydatid cyst of the pons. Pediatr Neurosurg 1999;31:326-9.
49. Muthusubramanian V, Pande A, Vasudevan MC, Ravi R. Surgical management of brainstem hydatid cyst--an unusual site. Surg Neurol 2009;71:103-6.
50. Yuceer N, Gökalp HZ. Periventricular hydatid cyst. J Neurosurg Sci 1998;42:173-5.
51. Taghipour M, Saffarrian A, Ghaffarpasand F, Azarpira N. Dumbbell-shape hydatid cyst of meckel cave extending to cerebellopontine angle and middle fossa; Surgical technique and outcome of rare case. World Neurosurg 2017;104:1049.e7-10.

How to cite this article: Din NU, Raza M, Rahim S, Memon W, Ahmad Z. Hydatid disease of central nervous system, a clinicopathological study of 33 cases. J Neurosci Rural Pract 2023;14:293-7.