

ORIGINAL ARTICLE

CLINICAL FEATURES AND OUTCOME ANALYSIS OF
INTRACRANIAL HYDATID CYSTS

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Background: Hydatid cyst of the brain is serious zoonotic parasitic infections which have profound health consequences if left untreated. The surgical excisions of the cysts are rewarding for both the patient the neurosurgeon. **Methods:** The study was conducted prospectively at Department of Neurosurgery Hayatabad Medical Complex Peshawar from January 2013 to December 2014. Patients with a diagnosis of intracranial hydatid cysts were included, clinical and radiological features recorded, intervention and postoperative outcome were analysed. **Results:** Eleven patients with a male to female ratio of 1.7:1. Mean age was 12.4 (SD±6.5) years with median GCS on arrival of 10 (SD±2.5). Clinical features were headache (81.8%), vomiting (90.9%), seizures (36.4%), focal deficits (54.5%) and papilloedema (72.7%). The median GCS on discharge was 13 (SD±1.1) while GOS at 1 month follow up was 4 (SD±0.7). The bivariate analysis showed inverse correlation ($R^2=0.68$; $p=0.02$) between duration of symptoms and outcome while GCS on admission was positively correlated ($r_s=0.75$; $p=0.007$) with the outcome. There was no mortality. **Conclusion:** Despite its rarity the clinical features are non-specific while radiological features help in establishing diagnosis. Earlier diagnosis and prompt intervention is the key to favourable outcome.

Keywords: Hydatid disease, Paediatrics, Hydatid cyst, Echinococcus, Outcome

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INTRODUCTION

Hydatid disease is a common zoonotic infestation which occur in about 1-2% of cases intracranially.^{1,2} The causative organism is Echinococcus. E granulosus causes single large cysts while E multilocularis causes multiple cysts in a honeycomb appearance.³ Humans are accidental hosts where infection occur by faecal contamination or ingestion of the infected food. Dogs are known to be the primary hosts while sheep and cattle are the intermediate hosts.¹

The disease is endemic to rural areas of the Middle East, Southeast Asia, Australia and Africa. It is more common in individuals who live near animals or are involved in cattle farming, although the international travel may transfer it to the non-endemic areas.^{1,3,4}

The most common intracranial location is in the territory of middle cerebral artery (Figure-3) where the blood flow is abundant.^{4,5} There are infrequent reports of posterior fossa, brainstem and skull base hydatid cysts.^{6,7} Clinical features of intracranial hydatid cyst are non-specific to the condition and include symptoms of raised intracranial pressure (ICP), effects of local pressure, seizures, brain herniation syndrome and consequent death if untreated.^{8,9} Radiological investigations include CT scan of brain with

contrast enhancement and MRI of brain with gadolinium enhancement. There are various algorithms which neuro-radiologists have evolved in order to differentiate hydatid cyst from other intracranial cystic lesions.^{4,5,7,8} Eosinophilia on differential leucocyte count is only present in 25% patients.¹ The immunological tests include the historical Cassoni's test (now obsolete), indirect haemagglutination test, enzyme-linked immune-sorbent assay (ELISA) with overall sensitivity of 80%, immune-diffusion and immunoelectrophoresis.^{1,10-12}

Definitive treatment is surgical excision of the cyst(s).^{13,14} The most widely used technique is Dowling-Orlando technique (Figure-4) with high success rates and very low rates of neurologic adverse events.¹⁴ Surgical exploration is usually followed by 5 to 6 months of anthelmintic drugs such as albendazole or praziquantel or both.¹⁷⁻¹⁹

We conducted this study in order to observe the clinical features and their relationship to postoperative outcome.

MATERIAL AND METHODS

It was a prospective observational study for two years between January 2013 and December 2014 at Department of Neurosurgery Hayatabad Medical Complex Peshawar. All preoperative patients with

a diagnosis of intracranial hydatid disease were included from both genders and all age groups.

Permission from the hospital ethical committee was obtained prior to the commencement of the study. The data was collected on a pre-designed pro forma. All patients who were included or their guardians were approached and the pros and cons of inclusion in the study were explained to them. Informed consent was taken for their participation. All patients were subjected to detailed neurological history and examination which was followed by detailed neuro-radiologic examination of the brain and cranium. Radiologic diagnosis was confirmed by a consultant neuro-radiologists as well as a consultant neurosurgeon. Laboratory work was done for all patients from the hospital labs. Operative articles which were required during surgery were provided by the hospital pharmacy free of charge.

The Dowling-Orlando technique was used to excise the cyst by means of hydrostatic pressure after opening the skull at the site of the lesion with an osteoplastic flap, dura opened and the overlying cortical layer was dissected with bipolar cautery and blunt micro-dissectors. The cyst was delivered intact, taking care not to rupture it in situ. Haemostasis and irrigation was followed by water tight dural closure and wound was closed in layers.

Postoperatively the patient was followed up with vitals monitoring, frequent GCS (Glasgow coma score) monitoring and improvement noted on pro forma. Development of any morbidity after the surgery was regarded as a complication and was also recorded. Final GCS was recorded at the time of discharge and patient was sent home on Albendazole + Praziquantel regimen. Typical dosage for albendazole was 10 mg/kg twice daily for 30 days with an off period of 15 days, 4–5 cycles in total. The patients were followed one month postoperatively in the OPD and a CT scan of the brain with contrast was performed. Glasgow outcome score (GOS) was noted at this occasion and functional status of the patient was confirmed, as well as the radiologic resolution of the cyst site.

The data was entered in IBM SPSS Statistics version 20.0. Analyses regarding frequency and percentage calculations were done. Charts and graphs were also used for displaying the data. Chi Squared test was performed for all the categorical variables and their significance noted in terms of *p* value and coefficients.

Bivariate correlation parametric as well as non-parametric tests were applied to the predictor and outcome variables and their results interpreted. For continuous variables we performed Pearson correlation test while for ordinal variables we

performed Spearman rank test to see if any correlation exist between the dependent and independent factors. The correlation was also shown using scatter plots between predictor and outcome variables.

RESULTS

Eleven patients of cerebral hydatid cysts were included over the 2 years. There was a predominance of children and adolescents with mean age of 12.4 years±6.5 SD with only one patient who was a 28 years old female. (Table-1) There were 7 (63.6%) males and 4 (36.4%) females. 45.5% patients belonged to the rural areas of neighbouring Afghanistan. (Table-2) The mean duration of symptoms at presentation was 24 days±13.3 SD. (Table-1) The median GCS on admission was 10±2.5 SD while the mean size of the cyst on CT or MRI was found to be 59.6 millimetres±9.5 SD. (Table-1) The clinical features comprised of headache in 9 (81.8%), vomiting in 10 (90.9%), visual deficit in 5 (45.5%), seizures in 4 (36.4%), focal neurologic deficit in 6 (54.5%) and papilloedema in 8 (72.7%) patients. (Table-2)

The most commonly encountered location was temporal and parietal (both 4 cases; 36.4%) while the rest of cysts were found in frontal lobes in 3 (27.3%). No cyst in occipital or cerebellum was noted. The anti-ecchinococcus antibody test using ELISA was performed for patients and was found positive in only 7 (63.6%) of patients. (Table-2)

The Dowling-Orlando technique was used in all patients with no incidence of cyst rupture or significant haemorrhage. Median GCS at discharge was 13±1.1 SD while median GOS at 1 month follow up was 4±0.70 SD. (Table-1) The most common complications were hospital acquired chest infection in 3 (27.3%), wound infection in 1 (9.1%) while persistent focal neurologic deficit in 2 (18.2%) patients. (Table-2)

The functional outcome as assessed by GOS was favourable in 9 (81.8%) while in 2 (18.2%) patients the outcome remained unfavourable. There was no mortality at one month postoperatively. (Table-2)

The bivariate correlation tests were significant (Table-3) for the duration of symptoms before surgical excision in terms of GOS on 1 month follow up (Pearson Correlation: -0.68; *p* value=0.02). (Figure-1) The duration of illness was also strongly correlated to GCS on admission (Pearson Correlation Coefficient: -0.68). Similarly, as shown in table-4 the GCS on admission was strongly correlated to the outcome in terms of GCS on discharge (Spearman's Correlation Coefficient: 0.75, *p* value=0.007) and GOS at 1 month (Spearman's Correlation Coefficient: 0.83, *p* value=0.001). (Figure-2)

Table-1: Quantitative variables

Variable	Minimum	Maximum	Mean	SD	p value
Age	5	28	12.45	6.56	0.247
Duration of symptoms	10	60	24.09	13.38	0.020
GCS on Admission	6	14	10.00	2.56	0.001
Size of Cyst in mm	45	75	59.64	9.56	0.132
GCS at Discharge	12	15	13.27	1.10	
GOS at 1 month	4	5	4.27	.46	

Table-2: Clinical features & their frequencies

Clinical Variable	Frequency (n)	Percentage
Gender		
Male	7	63.6%
Female	4	36.4%
Headache	9	81.8%
Vomiting	10	90.9%
Seizures	4	36.4%
Visual deficits	5	45.5%
Focal Neurologic Deficits	6	54.5%
Papilloedema	8	72.7%
Cyst Location		
Frontal	3	27.3%
Temporal	4	36.4%
Parietal	4	36.4%
Complications		
Hospital acquired chest infection	3	27.3%
Wound Infection	1	9.1%
Persistent Neurologic Deficit	2	18.2%
Outcome		
Favourable	9	81.8%
Unfavourable	2	18.2%
Mortality	0	0%

Table-3: Bivariate correlation between duration of symptoms and outcome

Predictor	Outcome	p-Value	Pearson Correlation
Duration of Symptoms	GCS at Admission	0.02	- 0.68
	GCS at Discharge	0.075	- 0.55
	GOS at 1 month	0.02	- 0.68

Table-4: Bivariate correlation between admission GCS and outcome

Predictor	Outcome	p-Value	Spearman's Correlation
GCS on Admission	GCS at Discharge	0.007	0.75
	GOS at 1 month	0.001	0.83

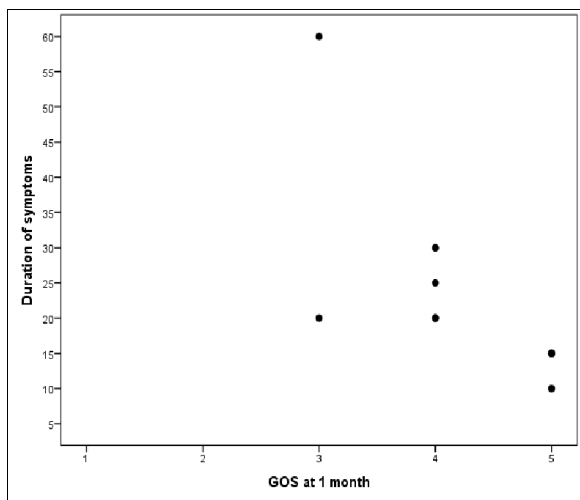


Figure-1: The inverse correlation between duration of symptoms and outcome

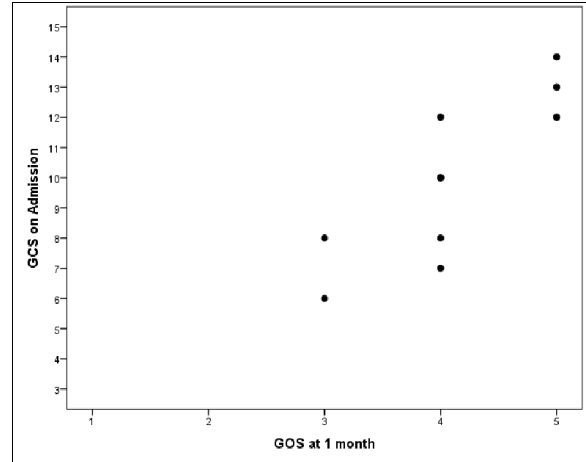


Figure-2: Positive linear correlation between admission GCS & Outcome



Figure-3: Axial MR of Brain: T1 + Contrast



Figure-4: A hydatid cyst after excision from a 6 year old female child

DISCUSSION

Intracranial hydatidosis is a serious infectious disease with significant morbidity. Its incidence varies globally with highest rates in Kenya (220 cases per 100,000 persons) and other Middle East regions.^{1,7} It accounts for up to 3.6% of all intracranial space occupying lesions.²⁰ Although mortality is low, the long term consequences of hydatid disease are high in terms of prolonged disability.²⁰

We included 11 patients with intracranial hydatid disease with 63.6% males and females were 36.4%. The mean age of presentation was 12.4 years with only one female patient aged 28 years. Many other studies have noted predominantly male pattern while other studies suggest that it is more common in female patients. Reason for the predominance of female patients is cited to be due to their close contact with animals in certain communities.¹ The average age of patients with intracranial hydatid cyst in majority of studies is predominantly paediatric although there are case reports which describe its occurrence in adult patients. The reason for predominance of this disorder in paediatric age groups is the earlier appearance of disabling symptoms for intracranial locations, while appearance of symptoms in sites like abdominal and thoracic cavities is frequently delayed due to the slow growth and late occurrence of local pressure effects. Ciurea *et al* found 95.7% patients in the paediatric age group which concur with our findings.²⁰

The clinical features of intracranial echinococcosis in our series included headache 81.8%, vomiting 90.9%, seizures 36.4%, focal neurologic deficits 54.5%, visual deterioration 45.5%, papilloedema 72.7% and/or progressive deterioration of consciousness. Symptoms of high intracranial pressure have been most frequently described in paediatric age group with variable range of papilloedema and focal neurologic deficits. We found three children who were treated for a week or two as cases of acute meningitis and encephalitis but on routine CT scan of brain the intracerebral cyst was identified. Another patient was on epilepsy treatment for a long duration before radiologic diagnostics were performed. Similarly one adult female patient presented with several weeks history of seizures, headache, vomiting and left hemiparesis. Ciurea *et al*²⁰ and Wani *et al*⁴ have pointed out that symptoms and signs of increased ICP are predominantly present in paediatric patients while focal neurologic deficits are more prevalent in adult patients with hydatid disease. Similarly studies by Duishanbai *et al*^{13,21} in large clinical series of intracranial hydatid disease have similar finding of clinical and radiological presentation as in our series. Duransoy *et al*⁹ has

reported a case of frank midbrain herniation syndrome with anisocoria and features of increased ICP.

Mean cyst diameter was found to be 59.6 millimetres where largest cysts were found in patients with increased intensity of neurologic symptoms. The mean duration of symptoms to the time of diagnosis was found to be 24 days while the mean admission GCS was found to be 10. It was observed that the longer the duration of illness the lower the GCS on admission would be. Ali *et al*¹⁴ has also detailed the radiological size of the cyst on presentation and found a higher average diameter (average 7.7 cm) of the cyst. They have stated delayed presentation (average length of symptoms 4.3 months) of patients to the tertiary care for the increased size of the cyst and higher levels of neurologic dysfunction.

The most prevalent intracranial location of the cysts were temporal 36.4% and parietal in 36.4% in our series. One cyst in a 28 year old female was located at temporal region in extradural location. There were no posterior fossa or skull base cysts in our series. In the clinical series of Ali *et al*¹⁴ the most common intracranial location was found to be frontal in 33.3% patients while there was also description of a case where they found a cyst in the infra-tentorial posterior cranial fossa. There are clinical reports of hydatid cysts located at deep cerebral structures like thalamus, skull base and the calvarium.^{7-9,22}

The most common complications which occurred postoperatively in our patients was hospital acquired chest infections. Unconscious patients and children are particularly at risk of respiratory complications where aspiration and lower immunity are known to play significant role. Persistent neurologic deficit in two patients remained static while the visual problems also did not respond promptly, probably due to the long term raised ICP leading to optic disc atrophy was the player. Ali *et al*¹⁴ in their series has found no postoperative morbidity and mortality except for the pre-existing neurologic deficit in the form of visual loss. However, Ciurea *et al*²⁰ in their long term follow up of patients have described a recurrence rate of 25% and 5.3% mortality within 6 months postoperatively. The most commonly employed surgical excision technique is the Dowling-Orlando method. The technique has been assessed in many clinical series with very successful outcome.^{6,11,13-15,20} Wang *et al*¹⁵ has also described a piecemeal resection technique for the removal of multiple cysts with good results.

In outcome analysis of postoperative patients we found that 81.8% patients achieved good functional outcome while 18.2% remained in the unfavourable functional outcome group at the end of 1 month follow up. The most significant factors

affecting the functional outcome in our series were the duration of symptoms at the time of presentation and the neurologic status of patient in terms of GCS. These both factors generally translate the overall advancement of the disease process intra-cranially and the resultant structural and functional damage. Ciurea *et al*²⁰ in their large series have provided the outcome data with a maximum of 22 years of follow up. They have found favourable outcome in 73.7% patients. The most significant factors affecting outcome in their series is described be a delay in diagnosis and intraoperative rupture of the cyst. Duishanbai *et al*²¹ assessed their outcome according to Karnofsky Performance Scale and found a 97.2% successful outcome postoperatively.

CONCLUSION

As could be concluded with any progressive disease process with local pressure effects on delicate structures like the brain, time to diagnosis is the most important factor. Clinical features are non-specific, though radiological evaluation with MRI and CT can narrow the diagnosis. Surgery is the mainstay of treatment with experience of the surgeon as the most important factor in order to excise the cyst without rupture in situ.

AUTHOR'S CONTRIBUTION

MMK: Concept & Manuscript Preparation. AA: Graphics and Photographs. AA: Data Collection. MAS: Follow up. MS, RUR: Statistical analysis. SA: Proof reading.

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