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Case Series

Pathology Section

# Unusual Presentation of Echinococcosis Causing Diagnostic Dilemma by Mimicking Neoplasm Clinically: A Case Series

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## ABSTRACT

Echinococcosis (hydatid disease) is caused by the cestode parasite belonging to genus Echinococcus (tapeworm). It is found frequently in rural areas where domestic livestock-raising is common. Globally, it is endemic in Middle East, Africa, South America, New Zealand, Australia, Turkey and Southern Europe including India. Even though echinococcosis can occur in any organ, it is very rare to see the disease at the sites reported in this article. Even at common sites, the way of presentation might be unusual causing diagnostic dilemma. Hence, current study was undertaken to evaluate the different unusual presentations of echinococcosis and to emphasise that it should be suspected in cystic lesions, especially in endemic areas. A series of six cases of echinococcosis over the period of three years (January 2017-December 2019) has been reported here. The correlation of clinical features, radiological with intraoperative findings (frozen section) wherever available and confirmatory diagnosis given on histopathology was attempted. All six cases which were suspected as neoplasm based on clinical and radiological findings turned out to be echinococcosis on cytopathology and histopathology. The sites involved were breast, brain, ovary, lung single case each followed by two cases in liver. Multisystem involvement was seen in only one case. Thus, the study conclude that echinococcosis can mimic cystic neoplasm clinico-radiologically and hence, should be considered as a differential diagnosis of cystic lesions irrespective of its endemicity, site and clinical presentation.

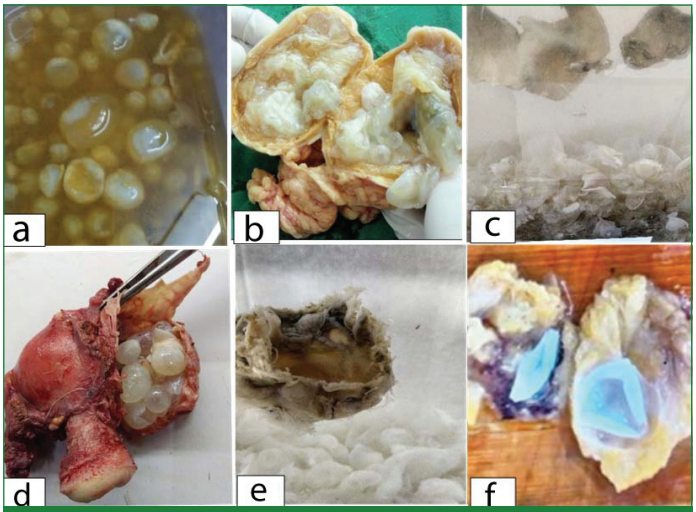

**Keywords:** Brain, Breast, Hydatid disease, Lung, Liver, Ovary

## INTRODUCTION

Echinococcosis was described by Hippocrates more than 2000 years ago who had used the term 'liver filled with water' [1]. Two forms of echinococcosis have been described: cystic (hydatidosis) and alveolar caused by E. granulosus and E. multilocularis respectively [2]. Today, despite of advances in its management, it still prevails a major public health problem. It is endemic globally in the Middle East, Africa, South America, New Zealand, Australia, Turkey and Southern Europe including India [3]. In endemic regions, human incidence rates for cystic echinococcosis can reach more than 50 per 100000 person-years, and prevalence levels as high as 5%-10% may occur [2]. The highest prevalence in India is found in the Saurashtra (now includes the state of Gujarat and Maharashtra), Andhra Pradesh, Tamil Nadu [4]. It is found frequently in rural areas where domestic livestock-raising is common causing serious health problems. The 2015 World Health Organization (WHO) Foodborne Disease Burden Epidemiology Reference Group (FERG) estimated echinococcosis to be the cause of 19300 deaths and around 871000 Disability-Adjusted Life-Years (DALYs) globally each year [2]. As the institute where these cases are reported is located in semi-urban area of Maharashtra, findings of cystic echinococcosis at unusual sites and with unusual presentations provoked us to undertake this study.

## CASE SERIES

Six cases of echinococcosis at various sites observed in the tertiary care hospital situated in Pune over the period of three years (January 2017-December 2019) are being discussed. Clinical features and radiological findings were taken from case record forms of these patients. Each and every case was suspected to be neoplastic based on clinic-radiological findings, the suspicion being cystic meningioma in brain, ovarian neoplasm for mass in peritoneum, pseudotumour appearance in liver, cystic neoplasm of unknown origin with metastasis for multisystemic involvement, cystic neoplasm of lung and solid-cystic mass suspicious of malignancy in breast. The gross findings of each case revealed multiple cysts with whitish, translucent 'tender coconut' appearance [Table/Fig-1a-f]. In all the six cases, the study performed cytological examination of wet mount and smears prepared from centrifuged deposits of fluid aspirated from cystic lesion followed by histopathological examination of formalin-fixed tissue sections stained with Haematoxylin and Eosin (H&E). In one case in addition to this, fluid aspirated from cystic lesion received intraoperatively was centrifuged and wet mount smears of the same were examined. Clinical and radiological comparison was done in each case individually and then histopathology confirmed the diagnosis of echinococcosis. Details of each case are presented in tabulated form [Table/Fig-2].

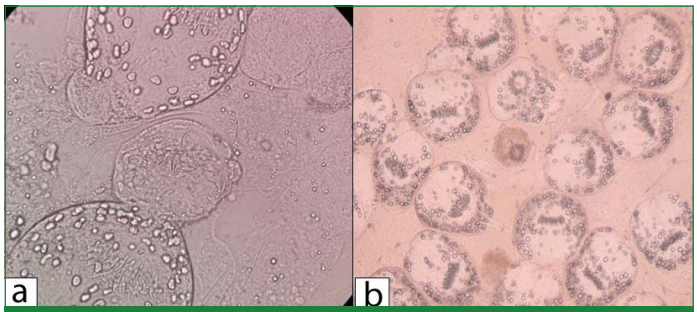



**[Table/Fig-1]:** a) Multiple whitish translucent cystic tissue pieces received in case of brain. b) Cystic mass in peritoneum showing multiple cysts within. c) Solitary cystic lesion in liver with thickened wall and multiple daughter cysts. d) Left adnexa showing single cystic mass with multiple daughter cysts within. e) Solitary cystic lesion of lung with multiple daughter cysts having tender coconut appearance. f) Solitary cystic lesion in breast parenchyma.

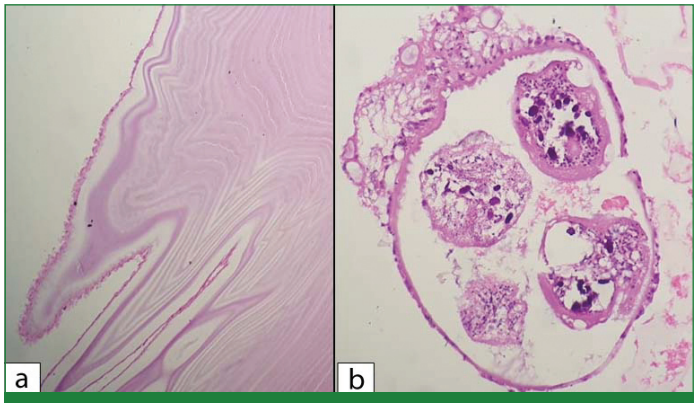
| Age (Years) | Sex | Clinical features | Radiological findings | Gross findings | Histopathology diagnosis |
|---|---|---|---|---|---|
| 20 | F | H/O recurrent episodes of convulsions and headache since four months. | CT scan: calcified cystic lesion was noted of size 10.9×10.8×8.4 cm in frontal region, reported as cystic meningioma. | Received multiple whitish translucent cystic tissue pieces [Table/Fig-1a] | Echinoccocosis -Brain |
| 65 | F | Lump in abdomen since three months | USG: Solid cystic mass in peritoneum, reported as ovarian neoplasm. | Received single soft to firm mass measuring 11×10×8 cm with intact capsule. On cutting open, approximately 200cc fluid oozed out. It was unilocular cyst containing multiple tiny grape like fluid filled vesicles within. Both fluid & tissue sections were examined [Table/Fig-1b] | Echinoccocosis -Peritoneum |
| 40 | F | Pain in right upper quadrant and epigastric region since five months. | USG: showed a solid-cystic hyper echogenic lesion of size 12×10×8 cm with heterogeneous pseudotumour appearance replacing the right lobe of liver. | Right lobectomy specimen of liver was received which showed single cystic lesion of 12×8×4 cm, almost involving entire lobe. On cutting approximately 150cc fluid oozed out which is clear and revealed single cyst with thick, whitish wall resembling tender coconut and had focal chalky white areas. Both fluid and tissue sections were examined [Table/Fig-1c] | Echinoccocosis -Liver |
| 45 | F | Fullness in abdomen, loss of appetite, weight loss since four months | CT scan: revealed a large multicystic lesion in left lobe of liver measuring 10.6×16.6×11.3 cm. Similar cystic lesions were seen in lesser sac and left adnexa. The findings were reported as cystic neoplasm of unknown origin with metastasis. | Clear fluid of approximately 20 mL was aspirated from cystic mass in left adnexa sent intraoperatively (frozen section). The fluid was cytocentrifuged and wet mount, HE, Leishman stain slides from sediment were prepared which on microscopy revealed typical hooklets and scolices of echinococcosis. Hence, further exploration was avoided and patient was managed conservatively [Table/Fig-1d] | Echinoccocosis -Left adnexa with multisystemic involvement |
| 32 | M | Progressive breathlessness since three months. | CT scan: well defined cystic lesion in left upper lobe of lung of size 3.0×2.5×2 cm suggestive of neoplastic aetiology. | Left upper lobectomy of lung was received which showed well defined single cystic lesion measuring 3×2.5×2 cm. Cyst was unilocular with tender coconut appearance [Table/Fig-1e] | Echinoccocosis -Lung |
| 65 | F | Painless slowly increasing lump in left breast since 2-3 months | USG: solid cystic mass (4.5×3 cm) in left breast suggestive of BIRADS IV | Lumpectomy specimen was received which showed solid cystic mass of size 4.5×3cm [Table/Fig-1f] | Echinoccocosis -Breast |

**[Table/Fig-2]:** Clinico-radiological and gross findings of all six cases.
M: Male; F: Female; H/O: History of; CT: Computed tomography; USG: Ultrasound sonography test; BIRADS IV: Breast Imaging-Reporting and Data System IV

The wet mount as well as smears prepared from centrifuged deposits of fluid in each case mentioned above were stained with Leishman's and H&E stain, the microscopy of which revealed typical hooklets with scolices [Table/Fig-3a,b]. The histopathological examination of tissue sections from cyst wall was followed by, which in all cases revealed cyst wall with an outer fibrous laminated layer and an inner germinative layer. The cyst cavity revealed numerous scolices and hooklets [Table/Fig-4a,b]. The surrounding host tissue showed infiltration of chronic inflammatory cells, including lymphocytes and eosinophils. In addition, only the third case revealed foci of calcification within the cyst wall. None of the cases revealed malignancy.




**[Table/Fig-3]:** a) Wet mount smear prepared from fluid showing typical scolices with hooklets (40X); b) Smear prepared from centrifuged deposit of fluid showing numerous scolices (H&E Stain, 10X).




**[Table/Fig-4]:** a) Lamellated acellular cyst wall (H&E stain10X); b) Daughter cyst with calcification (H&E Stain, 10X).

The patients were in the age range of 20-65 years, the mean age being 44.5 years. Marked female predominance was noted with male: female ratio of 1:5. None of the cases were clinically suspicious of echinococcosis, instead radiologically were suspected of neoplastic aetiology. However, histopathological examination revealed diagnosis of echinococcosis. Thus, none of the cases showed concordance between clinico-radiologically and histopathological diagnosis.

In one case (4th case), intraoperative cytological diagnosis avoided the further exploration of patient followed by conservative management.

## DISCUSSION

Human echinococcosis is a zoonotic infection with incidence in endemic areas ranging from 1-220 cases per lac [5,6]. Humans are occasionally accidently infected by oral ingestion of tapeworm eggs with contaminated food or water or direct contact with host [7]. The sites most frequently affected by echinococcosis in humans is liver (65%) followed by the lungs (25%) and the remaining 10% occur in muscle, spleen, bones, kidneys, brain, eye, heart, pancreas, peritoneum and breast [8]. The unusual clinical presentation even at the expected sites of involvement creates a diagnostic dilemma.

In this case series, the diagnosis of echinococcosis was made on examination of wet mount smears and cytology which enabled immediate diagnosis without the time lag that is required for histopathological diagnosis. In literature available, there are rarely any case series highlighting the significance of cytological examination of fluid for diagnosing echinococcosis, thus emphasising the need of the same in all the cystic lesions despite of clinical suspicion. This cytological examination was also found helpful in making correct diagnosis of echinococcosis intraoperatively in frozen section which further highlights the use of cytology especilally in resource limited facilities where cryostat is not available for processing of frozen section.

There are just a few published reports of separate cases on echinococcosis of brain based on radiological examination [6]. Yet, to the best of the knowledge, the study did not find any case of cerebral echinococcosis mimicking as cystic meningioma as was suspected in present case. Primary peritoneal involvement is seen only in 2% of all abdominal hydatid disease and its clinical presentation as suspicious of neoplasm is even rarer [9,10]. In the current study, the isolated peritoneal cystic mass was considered to be neoplastic

with possibly ovarian origin and turned out to be echinococcosis on histopathological examination. In liver, it can mimic neoplasm clinically as reported by Petrakis IE et al., which was similar to the finding in present case [11]. However, in case of cystic mass of liver with large dimensions without any additional symptoms, echinococcosis must be first ruled out. Mesenteric involvement by hydatid disease is very rare with this case being the third case reported in the literature so far. Multisystem involvement has been described and reported but simultaneous involvement of liver, lesser sac and ovary raising the suspicion of metastatic malignancy is very unusual. In case of echinococcosis of lungs, De Wilde C et al., has highlighted the presentation of complicated hydatid cyst mimicking neoplasm clinically leading to a diagnostic trap. Similar scenario was seen in present case where patient underwent surgical exploration and was diagnosed postoperatively [12]. Breast as a site of infestation for echinococcosis is very rare with only few single case reports being found in literature till date [13]. The rarity at this site makes it very less likely to consider the diagnosis of echinococcosis clinically, thus making it clinically suspicious of neoplasm [14].

Clinical details including patient's occupation, residence along with radiological findings of cyst with egg shell calcification, water lily sign and serpent sign should be looked for, to rule out echinococcosis, in cases of cystic lesions at any location. However, histopathology still remains gold standard diagnosis for echinococcosis. However in present study, the cytological examination of fluid in all cases revealed the diagnosis of echinococcosis immediately and was even used in frozen section to give stat report even before the histopathological examination. Thus, the study recommend the use of cytological examination of fluid aspirated from any cystic lesions, be it suspicious of neoplasm or not, to give rapid and confirmatory diagnosis. As this is reported within short period of time, it can help the treating physician to take appropriate decision for further management of the patient.

## CONCLUSION(S)

Echinococcosis should be considered in the differential diagnosis of cystic lesions at any site in endemic as well as non-endemic areas as it is curable with anti-helminthic drugs. The confirmatory diagnosis at early stage will prevent complications and unnecessary surgical exploration of the patients, thus reducing the morbidity and mortality of the disease. The cytological examination of fluids aspirated from cystic lesions should be mandatory as it confirms the diagnosis even prior to the histopathological examination.

## REFERENCES


[1] Kjossev K, Losanoff J, Velitchkov N, Belokonski E. Hydatid cyst of the diaphragm: A case report and review of the literature. The Internet Journal of Thoracic and Cardiovascular Surgery. 2003;6(1).
[2] World Health Organisation (2020, March 23). Echinococcosis. Retrieved from: https://www.who.int/news-room/fact-sheets/detail/echinococcosis.
[3] Gole S, Gole G, Satyanarayana V. Unusual presentation of hydatid cyst: A case series with review of literature. The Internet Journal of Parasitic Diseases. 2013;6(1):01-11.
[4] Rao SS, Mehra B, Narang R. The spectrum of hydatid disease in rural cental India: An 11- year experience. Ann Trap Med Public Health. 2012;5:225-30.
[5] Safioleas MC, Misiakos EP, Kouvaraki M, Stamatakos MK, Manti CP, Felekouras ES. Hydatid disease of the liver: A continuing surgical problem. Arch Surg. 2006;141:1101-08.
[6] Ashok KB. Old disease new location surgeons be alerted. International Journal of Collaborative Research on Internal Medicine & Public Health. 2011;3(4):328-32.
[7] Celik A, Turanli M, Kutun S, Delibasi T, Mengi N, Comert E, et al. Unusual location of hydatid cyst: Soft tissue mass in the neck. Eur Arch Otorhinolaryngol. 2006;263:1147-50.
[8] Sultana N, Hashim TK, Jan SY, Khan Z, Malik T, Shah W. Primary cervical hydatid cyst: A rare occurrence. Diagn Pathol. 2012;7:157.
[9] Pagaro PM, Chaudhari PK, Naphade NN, Patil T. Isolated peritoneal hydatidosisclinically mimicking ovarian tumour: A rare case report. Med J DY Patil Univ. 2014;7:78-81.
[10] Shangpliang D, Baishya P, Dey B, Raphael V, Wankhar B. Ovarian hydatid cyst mimicking an ovarian neoplasm. Autops Case Rep [Internet]. 2020;10(3):e2020177.
[11] Petrakis IE, Grysbolaki E, Paraskakis S, Lagoudis T, Filis D, Chalkiadakis G. Pseudotumoural hydatid cyst: Report of a case. J Hepatobiliary Pancreat Surg. 2009;2009:137956.
[12] De Wilde C, du Plessis AM, Goussard P. Unusual presentation of pulmonary hydatidosis mimicking thoracic malignancy in a paediatric south African Patient. J Belg Soc Radiol. 2018;102(1):70.
[13] Mujawar P, Suryawanshi KH, Nikumbh DB. Cytodiagnosis of isolated primary hydatid cyst of breast masquerading as a breast neoplasm: A rare case report. J Cytol. 2015;32:270-72.
[14] Taydas O, Kaya S, Ogul H, Kantarci M. Multisystem involvement of hydatid disease. Am J Trop Med Hyg. 2018;98(6):1557.



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