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RESEARCH ARTICLE

HYDATIDOSIS: A MYSTERY BOX AT VARIOUS SITES

Dr. Suwarna Patil[1], Dr. Dilip Sarate[2], Dr. Manjiri Khade[3], Dr. Swarada Kangate[3] and Dr. Pradeep Rudra[4]

1. Associate Professor, Department of Pathology, GMC, Akola, Maharashtra, India.
2. Professor and Head, Department of Pathology, GMC, Akola, Maharashtra, India.
3. Assistant Professor, Department of Pathology, GMC, Akola, Maharashtra, India.
4. Junior Resident, Department of Pathology,GMC, Akola.

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Abstract

Introduction: Hydatid disease caused by the larvae of the cestode Echinococcus, occurs worldwide. Although, hydatid can occur at any site, liver (59-75%), lung (27%) and spleen are the most commonly affected organs. Clinical suspicion along with radiological findings help in the diagnosis of hydatid. Fine Needle Aspiration Cytology (FNAC) plays an important role for initial diagnosis, while histopathology is confirmatory.

Aims: This study was carried out to know the various possible sites for hydatid and to understand the approach towards diagnosis.

Materials and Methods: The present study was a retrospective five years cross-sectional study, carried out at Tertiary Care Teaching Hospital, in Department of Pathology. Total of 24 cases were found to have hydatidosis.

Results: maximum of 12 cases (50%) were seen in 41-60 years of age. liver as most common site of hydatid cyst (11 cases – 46%), followed by lung (25%) and spleen (17%).

Conclusion: Hydatidosis occurs worldwide with human as the dead-end host. FNAC is the easy, feasible way to diagnose hydatidosis. Histopathology plays the key role in autopsy cases.

Introduction:-

Hydatid disease caused by the larvae of the cestode Echinococcus, occurs worldwide. The common species are E. granulosus and E. multilocularis. The former is more prevalent and causes unilocular cysts, while E. multilocularis is rare and causes multilocular cysts. Human echinococcosis was described in ancient times by Hippocrates, as "cysts full of water" in a human liver.[1] Echinococcus granulosus can reach any organ of the body where it develops into a small hydatid cyst. The parasite involves dogs as the definitive hosts and sheep as the intermediate hosts. Humans are the coincidental dead-end intermediate hosts. Hydatid disease occurs frequently when individuals handle infected dogs or ingest contaminated food or drink.[2] When infected ova are consumed by the human host, embryos develop in the proximal small intestine and reach the liver through the blood resulting in one or more echinococcal cysts. Some of the embryos may pass through the hepatic capillaries (first filter) and enter the pulmonary circulation (second filter).[3] A few may pass the pulmonary capillaries to enter the general stream and lodge in various organs, causing hydatid disease in other organs and sites. Dissemination through lymphatic channels has also been suggested as a possible mechanism and accounting for cases with solitary cysts in uncommon

Corresponding Author:- Dr. Swarada V. Kangate
Address:- Department of Pathology, Government Medical College, Akola, Maharashtra, India - 444001.

sites.[3,4] Another mechanism may be direct spread from adjacent sites.[5]Although, hydatid can occur at any site, liver (59-75%), lung (27%) and spleen are the most commonly affected organs.[1]

Clinical suspicion along with radiological findings help in the diagnosis of hydatid. Fine Needle Aspiration Cytology (FNAC)plays an important role for initial diagnosis, while histopathology is confirmatory.

This study was carried out to know the various possible sites for hydatid and to understand the approach towards diagnosis.

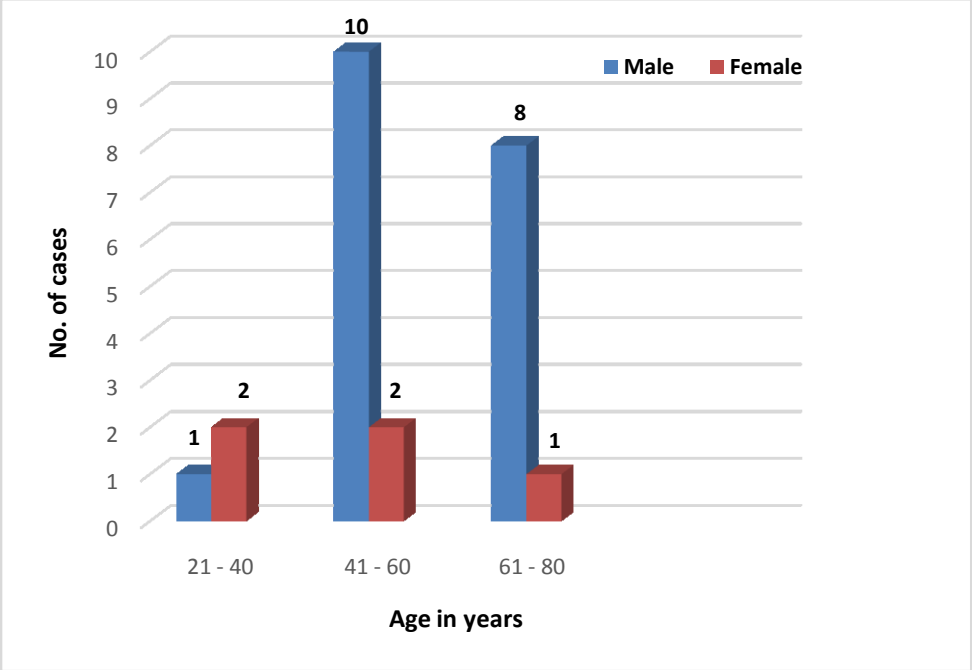
Materials and Methods:-

The present study was a retrospective cross-sectional study, carried out at Tertiary Care Teaching Hospital, in Department of Pathology, over a period of five years from January 2015 to January 2020. Total of 24 cases were found to have hydatidosis. Clinical history with age, presenting signs & symptoms, site, number (single or multiple) and radiological findings were studied in detail. Gross examination of 10% formalin fixed specimen was done and sections were taken from the representative areas. Further histopathological processing of sections was done and slides stained with Haematoxylin and Eosin (H&E) were studied. All the cases were referred to Department of Pathology from the Department of Surgery [either for Fine Needle Aspiration Cytology (FNAC) or for Histopathological examination], Department of Obstetrics and Gynecology and Department of Forensic Medicine (for Histopathological examination of the autopsy specimens).

Results:-

Total of 24 cases of Hydatid cyst were included in the present study.

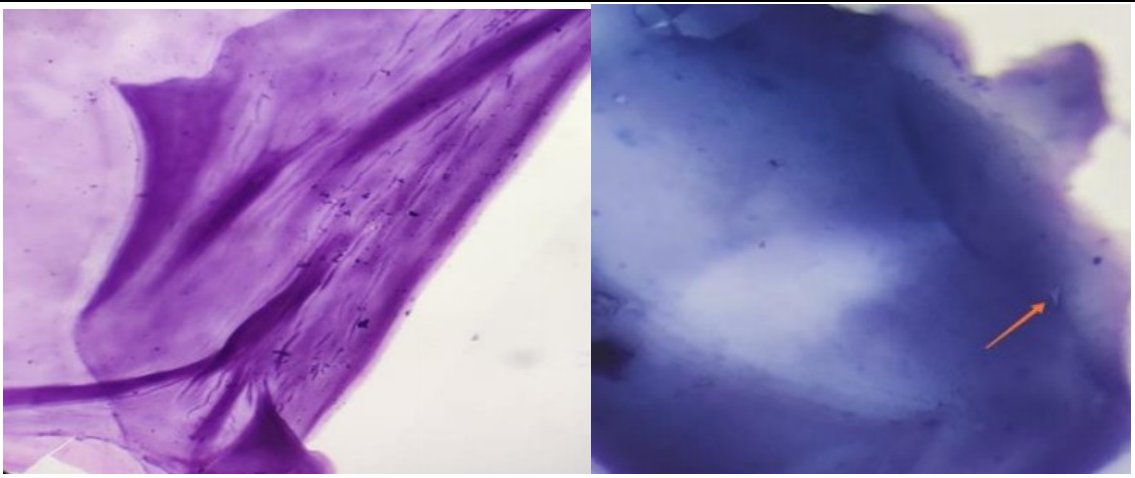
Figure 1:- Age-wise and Sex-wise distribution of cases (n=24).



As shown in fig 1, maximum of 12 cases (50%) were seen in 41-60 years of age. Youngest was 25-years male with hydatid in the liver, while oldest was 78-years male showing hydatid in spleen on autopsy. Males (79%) outnumbered females (21%), with the male to female ratio of 3.8:1.

In the present study, diagnosis of hydatid cyst was done on FNAC in one case (4%) (Fig 1), while 19 cases (79%) of routine histopathological specimen were diagnosed with hydatid cyst and four cases (17%) were found in the autopsy examination of the viscera.(Fig 2) (Table 1)

Figure 1:- Microphotographs demonstrating Hydatid cyst elements on FNAC.



Microphotographs showing laminated membrane, acellular material fragments with delicate parallel striations (H&E, 400x) and refractile, triangular hooklet (MGG, 400x)

Figure 2:- Microphotographs demonstrating Hydatid cyst showing acellular hyaline laminations on Histopathology(H&E 200x).

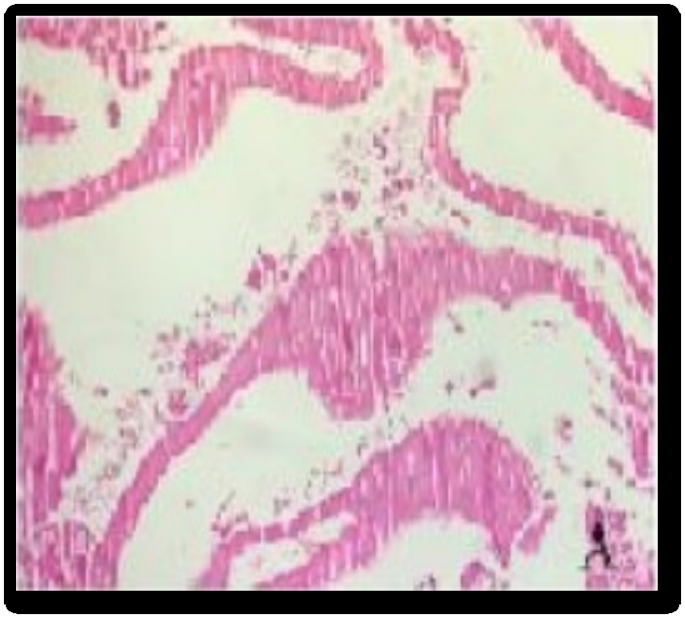


Table 1:- Site-wise distribution of diagnosis in the cases (n=24).

Site	Diagnosis			Total No. of cases
	FNAC	Histopathology	Autopsy	
Liver	-	09	02	**11**
Lung	-	05	01	**06**
Spleen	-	03	01	**04**
Ovary	-	02	-	**02**
Thigh	01	-	-	**01**
Total	**01**	**19**	**04**	**24**

Table 1 shows, liver as most common site of hydatid cyst (11 cases – 46%), followed by lung (25%) and spleen (17%). In present study, 8% cases of hydatid were found in ovary. While only one case (4%) of 68-years of female had hydatid in left thigh.

Discussion:-

Out of total 24 cases of hydatid cyst, included in the present study, maximum of 12 cases (50%) were seen in 41-60 years of age. Male (79%) preponderance was seen, with the male to female ratio of 3.8:1. Hydatid cyst was diagnosed on FNAC in one case (4%), while 19 cases (79%) on routine histopathological specimen and four cases (17%) were found in the autopsy examination of the viscera.

The adult E. granulosus is capped by a scolex at the anterior terminus and is 3 to 5 mm long. Four suckers and a double crown of 28-50 hooks emerge from the scolex. The cyst wall has a laminated, anuclear membrane with thickness of about 1 mm lined by a germinal layer. The germinal layer is 10 to 25 μm in thickness and contains nuclei. Cytologically, protoscolices, hooklets and fragments of the laminated membrane are commonly found in hydatid cysts. FNAC appears to be a safe and useful method in the diagnosis of hydatid cyst. Cytologic diagnosis of hydatid cysts has been reported in the lung, omentum, muscle, soft tissue,brain, orbit and joints.It may be difficult to find protoscolices in FNAC. Hydatid cyst should be considered when laminated membrane fragments are present on smears without evidence of protoscolices. Presence of protoscolices, refractile hooklets or fragments of a laminated membrane gives the definite confirmatory diagnosisof hydatid cyst.[6]

Three-fourth of infected individuals develop one or more hepatic cysts, predominantly in the right lobe. They may be multiple and can involve all lobes of the liver. A hydatid cyst is white, spherical cyst filled with fluid, few mm to many cms in diameter. The diagnosis may be delayed, due to its slow growth. Clinically, may present with hepatomegaly, obstructive jaundice and cholangitis, complicating to cholangitis and rupture. Hydatid cyst may prove to be fatal, if not treated. The liver is the most commonly involved organ. The differential diagnosis of hepatic hydatid cyst includes abscess, hemangioma, and non-parasitic cysts such as solitary bile duct cyst and hepatobiliary cystadenoma.Treatment of choice includes complete surgical excision of the intact hydatid cyst without rupture, to reduce the chance of seeding and recurrence.Early diagnosis of hydatid cyst by serology may provide opportunities for early treatment and more effective chemotherapy.[6,7] It is important to include hydatid cyst in the differential diagnosis of cystic lesions in the liver, particularly in those who have lived or travelled in endemic areas.[6] In the present study, liver as most common site of hydatid cyst (11 cases – 46%), out of which two cases were diagnosed as autopsy findings. Literature related to hepatic hydatid cyst are available in the form of case reports.[4,6]

Hydatid disease involves the lungs by various mechanisms. Most of the embryos are stuck in the liver sinusoids and, through the hepatic vein and the inferior vena cava (IVC) enter the right heart and finally, settle in the lungs. Embryos can also enter the thoracic duct via lymphatics of the small intestine and then through an internal jugular vein and right side of the heart, they enter the lungs. This is a major pathway by which the hydatid larva bypasses the liver and involves the lungs. Another possible lymphatic pathway is the lymphatics of the dome of the liver and the diaphragm, which ascend to the parasternal and intercostal lymph nodes. The next possible route is a veno-venous anastomosis in the liver and the space of Retzius. Another possibility is direct pulmonary exposure through the inhalation of air contaminated with Echinococcus eggs.[8] Borrie et al. has shown that eggs administered to sheep via a tracheostomy resulted in the development of lung cysts.[9] Transdiaphragmatic dissemination via a broncho-biliary fistula due to transdiaphragmatic rupture of liver hydatid (simultaneous hepatopulmonary involvement). Lungs may also become the site of secondary hydatidosis due to the rupture of a primary cyst resulting in dissemination of multiple daughter cysts and scolices. The secondary cyst may develop within the lung parenchyma, an adventitial layer of the primary cyst, intrabronchial, or metastatic.Metastatic lung lesions develop due to rupture of the cyst into right heart chamber, IVC or rupture of a bone cyst.[8]

The growth rate of the hydatid cyst is proportional to softness of the organ and surrounding tissue elasticity. Hence, lung cysts grow faster than the liver cysts. Negative pleural pressure may further accelerate the growth rate of the cysts.Due to their more elastic lung tissues; cysts in children grow at a faster rate and become larger than in adults. Bloomfield et al. reported a doubling time of 16–20 weeks. Borrie reported that a hydatid cyst usually achieves 1–2 cm in diameter at the end of 6 months and up to 6 cm within a year.[10]

Complications of pulmonary hydatid cysts include rupture, secondary infection, pneumothorax and suppuration. Patients may develop sudden onset of chest pain, cough, fever and hemoptysis after rupture of cyst. Perforation of the cysts into a bronchiole, resulting in expectoration of the germinative membrane or the hooklets of the parasite are called hydatoptysis. Sometimes patients may also complain of a salty taste in the mouth or development of hypersensitivity reactions (from urticaria and wheezing to anaphylaxis), indicating ruptured pulmonary hydatid cyst.

Screening for the presence of hepatic hydatid cysts should be done in all patients with PHCs because of the high incidence of the coexistence and also due to the asymptomatic nature of cysts in this location.[11]Surgery is the treatment of choice of pulmonary hydatid cyst.In inoperable cases or where surgery is contraindicated, medical therapy with benzimidazoles compounds may be tried.[8]

Present study includes, six cases of lung hydatid, including one autopsy finding. Case reports for pulmonary hydatid are being documented in the literature.

Although splenic hydatidosis has been reported as early as 1790 by Berlot as an autopsy finding[12], its occurrence even in endemic areas is less than 3%. Spleen is usually secondary to systemic dissemination or intraperitoneal spread from a ruptured hepatic cyst. Treatment and diagnosis of splenic cysts is troublesome for many physicians and surgeons.[13]Signs and symptoms are related to splenomegaly, like abdominal distension and compression of adjacent structures[14], local or referred pain also reported in majority cases. Known complications include infection of the cyst, intraperitoneal rupture of cyst and fistulization into hollow viscera especially colon, leading to upper or lower gastrointestinal bleeding.[15]

Tarccoveanu E. reported 38 cases of splenic Echinococcosis and abdominal pain was the most common symptom among these patients.[16] Teke et al.[17,18]reported a splenic hydatid cyst perforating into the left colon and causing massive gastrointestinal bleeding.

Chemotherapy and Puncture, Aspiration, Injection, and Re-aspiration (PAIR) technique using hypertonic saline or 0.5% silver nitrate solutions before opening the cavities tends to kill the daughter cysts.Diagnosis of splenic hydatid cyst is made by complete blood profile showing eosinophilia, radiological modalities like Ultrasonography (USG) and CT-scan and specific serum immune-electrophoresis tests. At present USG and CT, are the most valuable imaging techniques for the diagnosis and evaluation of focal splenic diseases.The intradermal (Casoni) test is a valuable diagnostic procedure.[19]The standard treatment of hydatid splenic cysts is splenectomy as it has low morbidity and mortality rate and complete resection removes all parasitic and pericystic tissues.[20] Gil-Grande et al. reported that albendazole sterilizes up to 72.3% of cysts by the end ofthe first month and 94% atthe end of 3 months of treatment.

In the present study, four cases of splenic hydatid were included, which was comparable to the reported splenic hydatid. One, out of four cases, was an autopsy finding.

Primary pelvic hydatid cyst is an extremely rare condition and occurs in approximately 0.7% of patients. Hydatid disease in pregnancy is a very rare condition with incidence of 1/20000 pregnancies. The pelvic hydatidosis in women could be either primary or more frequently secondary to hydatid cysts in liver or spleen.[2] Approximately 80% of all pelvic cases involve the genital area, the ovary being the most common, followed by uterus. In primary pelvic hydatid echinococcosis, exclusively confined to genital organs that are considered to be the primary site of inoculation via the bloodstream. There are no specific symptoms of pelvic hydatid disease. The mode of presentation is determined by the size of the cyst and any complication. Obstetric problems complicating the hydatid disease are abdominal pain, dystocia, obstruction of labour and uterine rupture. Anaphylactic shock may occur when cysts rupture during the second stage of labour. USG is the gold standard for diagnosis of hydatid cyst in pregnancy, showing the pregnancy status and the number and locations of cysts as well as their relationship with other organs. Serological tests are less reliable in pregnancy because of the usual immunological change. Surgery is the most effective treatment.[1]

Present study, included two cases of ovarian hydatid. Both the females were in reproductive age group, with the complaint of vague abdominal pain. Specimen received included cystic enlarged ovary, which was diagnosed as ovarian hydatid on histopathology. This was comparable to the data available in the literature.

Incidence of musculoskeletal echinococcosis including involvement of subcutaneous tissue is 1-5.4%. Soft tissue hydatid cysts occur in 2.3% of cases reported from endemic areas. The mechanism of the primary subcutaneous localization is unclear. A possible dissemination through lymphatic channels has also been reported. Muscular and subcutaneous area is supposed to be an unfavourable site for infestation because of its high lactic acid concentration. The primary soft tissue involvement is very rare, causing a diagnostic challenge.In majority of cases with atypical locations, the disease is asymptomatic. Usually it presents as painless, non-inflammatory, slow growing soft tissue

masses,mimicking other pathological conditions such as soft tissue tumours in some cases. Hydatidosis can be diagnosed preoperatively by FNAC. In patients coming with a soft tissue cystic lesion, possibility of hydatidosis should be kept in mind as a differential diagnosis.FNAC can be considered as a safe and effective modality for the diagnosis of hydatid cyst. Minimal complications can be managed by anti-anaphylactics.The best treatment option is complete surgical excision of the intact cyst which avoids leakage of cyst contents. If it is not possible, the cyst contents can be removed intraoperatively and the cyst pouch irrigated with scolicidal solutions. The combination of adjunctive chemotherapy with anti-helminthics pre and post operatively is also recommended to cover the risk of dissemination.[21,22,23]

One case of hydatid in left thigh was reported in the present study. The lady was referred to the cytology section with the complain of soft, painless swelling in the posterior aspect of left thigh, increasing in size. 01 ml of brownish fluid was aspirated following the uneventful fine needle aspiration. This was diagnosed as a subcutaneous hydatid, thus explaining the importance of FNAC for the diagnosis of the hydatid. Bothale KA et al[21] also reported similar case in 2015. Bagga PK[3] also found subcutaneous hydatid in the inguinal region.

A case report documenting primary hydatid cyst in thyroid gland is also found.[24]Thus, hydatid can occur at any site, either primary or secondary.

Conclusion:-

Hydatidosis occurs worldwide with human as the dead-end host. It can infect any organ or tissue in the humans, proper clinical work-up and approach towards diagnosis is necessary. Clinical, radiological and pathological approach helps in appropriate diagnosis and treatment. FNAC is the easy, feasible way to diagnose hydatidosis. Histopathology plays the key role in autopsy cases.

Although a small sample study, present study gives the probable spectrum of sites for hydatidosis and the differential diagnosis to be thought of. Still, studies with large sample size needs to undergone for better views.

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