Skeletal hydatidosis

Introduction:

Hydatid disease is a zoonotic parasitic disease in humans caused by infestation of tapeworms of genus Echinococcus. Developing countries, dealing with animal husbandries specially sheep are endemic zones for this disease. Human manifestation is common in liver (75%), followed by lungs (10-15%) and other organs (10%) like spleen, heart, brain, kidney, muscles. Skeletal manifestations are rare (0.5-4%) [1]. Spine is commonly involved. The pelvis, hip and long bones can also be affected. It lacks specific clinical features and radiographical characteristics. The radiograph mimics skeletal metastasis, tuberculosis, osteoclastoma and lytic cystic lesion. Bony lesions may or may not be accompanied by any organ involvement [2-5]. Treatment is usually difficult because of long latency period with delayed presentation and frequent recurrences even after medical and surgical management.

Etiopathogenesis:

Echinococcosis (Hydatid disease) is caused by larva stage of tapeworm Echinococcus granulosus and less commonly by Echinococcus multilocularis (does not affect bone). Dogs, being the primary host, the adult parasite reside in the intestine and release eggs in their stools. Sheep are the most common intermediate host. Humans become occasional intermediate hosts when they come in contact with infected dog faeces or contaminated plants or soil with parasite eggs by direct hand to mouth transfer. The eggs hatch to release Scolex (embryo) which is absorbed through intestinal circulation into hepatic portal circulation. This scolex is filtered in Liver and then lungs before reaching other organs and bone [1,2]. This explains its rare bone involvement. The embryo then forms the hydatid cyst in organs. The cyst consists of three layers; the outer pericyst, the middle laminated membrane and the inner germinal layer containing larva (endocyst). Bony involvement is slightly different because the outer pericyst layer is deficit. Embryos form microvesicles and expand. Metaphysis is involved initially and later spreads to the diaphysis. Daughter vesicles are produced and it expands, replacing the medullary cavity. Pressure effect on cortex causes erosion and spread to surrounding soft tissue and later to joint. Gradually multiplication causes ischemic necrosis of cortex and lesion looks like (multiloculated) bunch of grapes [3-5].

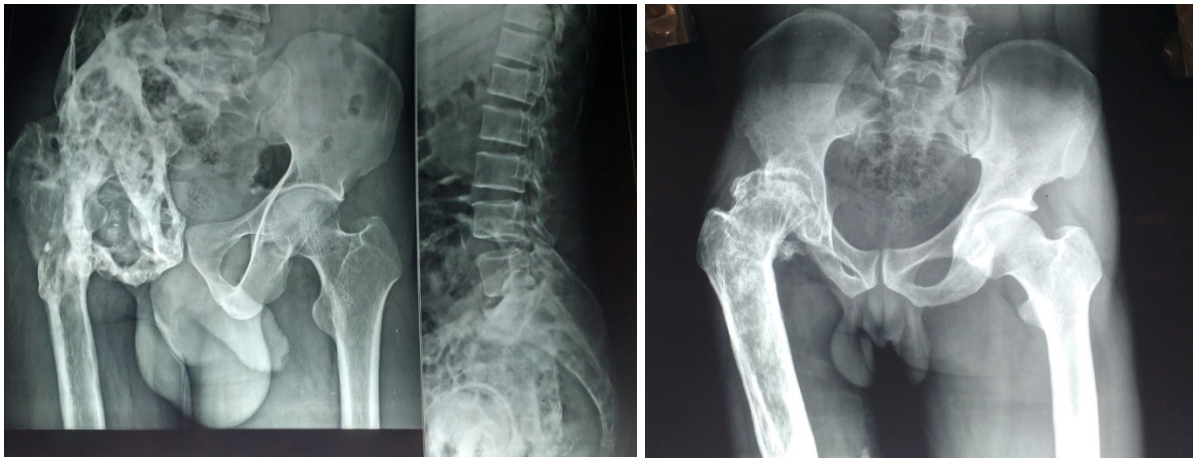
Clinical features:

Hydatid disease remains asymptomatic for a long period of time. The late appearance of clinical features is often non-specific; hence it usually manifests in fourth or fifth decade of life. It is not seen in children, but the parasite is acquired at a younger age of life. Presenting symptoms are according to the site of bony involvement apart from visceral symptoms. The spine involvement is common and it presents as back pain, radiculopathy, myelopathy or kyphotic deformity. Pelvis and hip involvement are second most common bone affection causing mass effect, antalgic gait, limp, restriction of hip joint movements. Long bones involvement causes pathological fractures, mass effect on nerves and paraesthesia [1-5]. There are no systemic manifestations but rupture of cyst may cause antigen exposure causing anaphylaxis that may be fatal.

Diagnosis:

Skeletal Hydatidosis is very difficult to diagnose. Plain radiography often reveals it in an advanced stage. The ill-defined multiloculated lytic lesions may give rise to moth-eaten appearance inside the medulla. There may be cortical thinning with intermediate zone of sclerosis and poor zone of transition. There is no periosteal reaction or calcification inside the cyst. Computer tomography shows the soft tissue extensions and uni- or multiloculated appearance of cysts and daughter cysts (“rosette” or “spoke wheel” appearance). Magnetic resonance imaging is the best modality of imaging which depicts extent, size, loculation, and type of cyst (parent or daughter cysts) [3-5]. It usually gives hyperintense signal intensity in T2 weighted images. Bone scan has no role.

Serological tests may be helpful in suspected cases. Antibodies assessment is more sensitive than detecting antigens. There is 25 to 50% sensitivity of ELISA, indirect haemagglutination and the complement fixation test in bone disease. The Casoni intradermal test is non-specific. 25 to 30% of cases show Eosinophilia. Fine Needle Aspiration is not advised in view of risk of cyst rupture which can lead to dissemination of the disease and anaphylactic reactions. Definitive diagnosis is by histopathological examination. Ultrasonography has no role except for muscular involvement. Differential diagnosis include: pyogenic skeletal infections, bone metastasis, benign cystic lesions of bone, osteoclastoma, multiple myeloma, tuberculosis [3-5].



Treatment:

There are no definitive guidelines of its treatment. Cure from the disease is difficult because of late presentation and complexity of involvement. Recommended protocol is a combination of wide surgical debridement, systemic medical therapy and local chemotherapeutic agents. Wide resection is usually difficult because of its complex location. Bony lesions are curetted, debrided and bone voids are filled with grafts, bone substitutes in combination with antihelminthics like albendazole. The treatment of pathological fractures is done as per standard protocol of fracture management of that particular bone. Polymethylmethacrylate has also been used in few cases; the exothermic reaction is helpful in eradicating local daughter cells. High doses of albendazole (10 mg/kg/day) and mebendazole (40 mg/kg/day) are effective. Albendazole having less toxicity and side effects is better tolerated. A minimum of six courses of albendazole (each course consisting of 28 days of treatment) is helpful. At least one course should be given pre-operatively. Local chemotherapeutics are also used during curettage. Most commonly used agents include: 3% hypertonic saline, 10% formalin, hydrogen peroxide, chlorhexidine, 80% alcohol, povidone-iodine alcohol solution or 0.5% silver nitrate. These agents destroy daughter cysts, but may not be completely effective and therefore do not necessarily prevent local recurrence. Despite all efforts there is a high chance of recurrence (40%) in this locally aggressive disease and hence few doctors usually skill-fully neglect the disease knowingly at an advance stage. Early diagnosis is important and hence there should be high degree of clinical suspicion in patients with similar bony lesions especially in endemic areas.

References:

1. Song XH, Ding LW, Wen H. Bone hydatid disease. Postgrad Med J. 2007;83(982):536-542. doi:10.1136/pgmj.2007.057166

2. Zlitni M, Ezzaouia K, Lebib H, Karray M, Kooli M, Mestiri M. Hydatid cyst of bone: diagnosis and treatment. World J Surg. 2001 Jan;25(1):75-82.
3. Inayat F, Azam S, Baig AS, Nawaz G, Ullah W. Presentation Patterns, Diagnostic Modalities, Management Strategies, and Clinical Outcomes in Patients with Hydatid Disease of the Pelvic Bone: A Comparative Review of 31 Cases. Cureus. 2019 Mar 5;11(3):e4178.
4. Johny J, Andrew D, Shyam K. Multimodality imaging in a patient with hydatid disease of the pelvic bone. BMJ Case Rep. 2020 Oct 30;13(10):e237830.
5. Kalinova K, Proichev V, Stefanova P, Tokmakova K, Poriazova E. Hydatid bone disease: a case report and review of the literature. J Orthop Surg (Hong Kong). 2005 Dec;13(3):323-5.

IOA Clinical record format (Google questionnaires format)

	Preoperative	Postoperative			
		3m	6m	12m	5yrs
Demography Age, sex Occupation Address					
Clinical details Age at the time of diagnosis Pain, swelling, tenderness, ROM etc. Bone(s) involved Patient Consent taken?					
Radiographic findings (upload image)					
MRI/CT (upload images) Any other Imaging					
Histopathology					
Treatment details					
Medications (dose, duration)					
Surgery details					
Radiation therapy details					
Follow up (recurrence/complications/sec surgery etc.)					