

Supraclavicular Hydatid Cyst: An Unusual Cause of Neck Swelling

Dilasma Ghartimagar^{*}, Arnab Ghosh, Manish Kiran Shrestha, Talwar O. P.

Department of Pathology, Manipal College of Medical Sciences, Pokhara, Nepal

Abstract

Hydatid disease, also known as echinococcosis or hydatidosis, is a zoonotic infection caused by the larval forms (metacestode) of *Echinococcus granulosus*. Hydatid disease is an important public health problem and is endemic in sheep and cattle raising areas worldwide. Musculoskeletal or soft tissue hydatidosis accounts for about 0.5%-5%. Even in regions where echinococcosis is endemic, hydatidosis of cervicofacial region is extremely rare. We present an unusual case of a hydatid cyst in 60 years old male who presented as supraclavicular swelling for a duration of 1 and half years. The swelling was slow on growth and there was vague pain for the last 8 months duration.

Keywords

Hydatid Cyst, Neck Swelling, Supraclavicular Region, Unusual Location

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1. Introduction

Hydatid disease is an important public health problem and is endemic in sheep and cattle raising areas worldwide.^{1, 2} Endemic foci are seen in Eastern Europe, the Mediterranean countries, Australia, New Zealand, India, Russia and South Africa.^{3,4} In different studies, the incidence of hydatid disease is found to range from 1 - 220 cases per 100,000 in endemic areas.^{1, 2}

Echinococcus is a zoonotic infection caused by tapeworm of genus Echinococcus including mainly 3 species, namely Echinococcus granulosus, Echinococcus multilocularis, Echinococcus vogeli.

The cyst wall is thin walled and consists of three layers: an outer layer of host origin, a middle cuticle layer and an inner germinal layer to which are attached brood capsules and scolices. In all sites, hydatid cysts have three layers except for those of the bone which do not have outer or host layer. ⁵ The life cycle of Echinococcus granulosus includes dogs (and other canines) as the definitive host, and a variety of species

of warm blooded vertebrates (sheep, cattle, goats and humans) as the intermediate host. The adult worms are very small usually consisting of only three proglottids and they live in the dogs small intestine. Eggs are liberated in the hosts faeces and when these eggs are ingested by the intermediate host they hatch in the hosts small intestine. The larvae can be distributed throughout the intermediate host's body (although most end up in the liver) and grow into a stage called hydatid cyst. ⁶

2. Case Summary

A 60 year old man presented with the history of pain and swelling in the right supraclavicular region since 1.5 years. Enlargement was associated with pain and tenderness. On examination, a firm to hard immobile mass of about 7 x 2 cm in size was seen in right supraclavicular region with ulcerated skin and whitish discharge. Patient was given courses of antibiotics but it did not help the lesion to heal the ulcerated part. Laboratory investigations showed the eosionphil count to be 8% and the rest blood parameters were within normal

* Corresponding author

E-mail address: dilasmagm@hotmail.com (D. Ghartimagar)

limit. HIV and HBsAg status was non-reactive. Contrast enhanced computed tomogram showed heterogenously enhancing soft tissue density mass lesion with areas of necrosis in right supraclavicular region with minimal fat stranding (Fig 1). Differential diagnoses were given as inflammatory lesion, soft tissue sarcoma and metastasis.

Fine needle aspiration cytology smears were cellular and showed sheets of neutrophils along with numerous singly scattered histiocytes, several multinucleated giant cells (Fig 2) and necrosis. Histiocytic cells showed marked reactive atypia. There was no epithelioid granuloma. A diagnosis of acute inflammatory lesion with giant cell and histiocytic reaction was given and biopsy was advised.

Surgical excision was done and a skin covered mass measuring 8 x 5 x 3.5 cm was received. Cut section showed gelatinous yellowish areas with lamellated structure (germinative membrane) (Fig 3). Microscopic examination showed a hydatid cyst with lamellated eosinophilic structure along with features of leakage of its content into the surrounding fibrofatty tissue eliciting marked panniculitis and a dense chronic inflammation associated with sclerosis and giant cell reaction (Fig 4). After pathological diagnosis of hydatid cyst, CT scan of abdomen and thorax was performed which did not reveal any hydatid cyst in other organs including lungs and liver.



Fig. 1. CT scan showing heterogenously enhancing soft tissue density mass lesion in right supraclavicular region.



Fig. 2. FNAC picture showing multinucleated giant cells (Giemsa 100x).



Fig. 3. Gross view of the resected cyst with its germinative membrane.



Fig. 4. Histopathology picture showing lamellated eosinophilic structure of hydatid cyst. b) Dense histiocytic and giant cell reaction of surrounding area (Hematoxylin and Eosin 100x).

3. Discussion

Hydatid disease produced by Echinococcus granulosus remains an important sanitary problem in many regions of the world. Moreover migrating current population is the reason why new cases of hydatid disease are being observed in areas with no previous prevalence.⁷ Although hydatid disease is known to affect liver and lung commonly, it also may affect brain, heart, kidney, ureter, spleen, uterus, fallopian tube, mesentery, pancreas, diaphragm and muscles.³

Patients with echinococcus infestation must undergo thorough systemic investigations because 20-30% have multi-organ involvement. Hydatid cysts in neck, in the absence of disease in lung and liver, may be caused due to systemic dissemination through lymphatic route, which is a strong possibility in case of unusual presentation sites.⁸ Although the disease is generally asymptomatic, it may exhibit clinical symptoms depending on the size and location of the cyst, and the pressure of the growing cyst.⁹ In our patient, the lesion was slowly growing for one and half years and there were no symptoms except a lump with vague pain which ulcerated and was discharging for 15 days in the left supraclavicular area.

The diagnosis of Echinococcus infection mainly depends on the clinical history of the patient, diagnostic radiological findings and serologic tests. ELISA, Casoni skin tests, latex agglutination, immune electrophoresis and direct hemagglutination are serological methods, used for the diagnosis of hydatid disease. An increase in titer indicates recurrence of disease and a decrease in titer indicates resolution.^{3, 8} Molecular study over hydatid cyst shows that Echinococcusgranulosus exhibits substantial genetic diversity that has important implications for the design and development of vaccines. DNA approaches like use of DNA probes are useful for the accurate identifi cation of this genus.¹⁰ Abdominal x-rays and chest x-rays, ultrasonography and CT scans should be performed to investigate other organs involvements, particularly liver and lungs.¹¹ In our patient, CT scan and FNAC were done, both of which could not give the definite diagnosis of hydatid cyst.

Though FNAC is beneficial for the evaluation of any mass lesion in the cervical region, in hydatid disease, there is potential threat to precipitate acute anaphylaxis and spread of daughter cysts. Therapy with nontoxic scolicidal agents or combination chemotherapy with mebendazole is of therapeutic value in the treatment of patients with recurrence or a high risk of contamination.⁸

From the period of infection to clinical examination, echinococcosis may display the characteristic symptoms of benign tumor that grows slowly without pain. Clinical symptoms depend on the anatomic location. It may imitate benign and malignant tumors, cysts, abscess, hematoma, pseudocyst and congenital cysts.^{12,13}

The common differential diagnosis in neck swelling includes thyroid, salivary gland and lymphnode all of which can be non-inflammatory, inflammatory and malignant.

Soft tissue sarcoma especially rhabdomyomatous differentiation may present as neck swelling.¹⁴ In this patient due to firm to hard neck swelling not responding to antibiotics, the clinical impression was metastatic lymphnode or soft tissue sarcoma.

At present the definitive treatment for hydatid cyst is surgical removal of the cyst which remains the gold standard. The prognosis is excellent in hydatid cyst cases who are treated by removal of cyst totally without rupture.⁹ Besides surgery, non conventional treatment like PAIR (Puncture, Aspiration, Injection, Reaspiration) had been widely studied recently and was found safe and effective.¹⁰ Total excision of the cystic mass was performed surgically in our patient (Fig 5) and on post operative follow up of 1 year he was doing well.



Fig. 5. Post-operative scar mark with sutures in the right supraclavicular region.

4. Conclusion

Since hydatid disease is one of the important public health problems and it can arise in parts of the body other than liver and lungs, it should be diagnosed correctly with the help of appropriate clinical history, radiological findings and histopathological correlations which will prevent future complications and improves the survival of the patients. The prognosis is excellent in hydatid cyst cases treated with total removal of the cyst without rupture.

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