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Novel Multimodal Treatment Regimen for the Management of Primary Sacrococcygeal Cystic Echinococcosis

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Learning Point of the Article:

Multimodal treatment regimen comprising of oral neoadjuvant medical therapy (albendazole and praziquantel) along with PAIR therapy and surgical in toto excision followed by adjuvant medical therapy for 3 months has given excellent results in sacrococcygeal cystic echinococcosis without any recurrences.

Abstract

Introduction: Osseous hydatidosis is a rare condition most commonly involving the spine. Among spinal segments, sacrococcygeal involvement is even rarer. Moreover, the lesion is more prone to recurrence owing to the infiltrative nature of microvesicular lesions involving the spine. In this case report, we describe an effective multimodal management approach toward the management of primary sacrococcygeal cystic echinococcosis.

Case Report: A 56-year-old female presented with complaints of severe back pain and urinary incontinence for 3 months. She presented with a slow-onset cauda equina syndrome with radiating pain to both lower limbs. Radiographic evaluation showed an expansile lytic lesion affecting the right iliac wing with near-complete cortical bone destruction of the sacrum. Magnetic resonance imaging revealed neural involvement with sacral destruction by a multiloculated cystic mass, extending to the spinal canal. No coexisting lesions were noted anywhere. Echinococcosis was diagnosed with serum enzyme-linked immunosorbent assay. She underwent neoadjuvant therapy with albendazole and praziquantel, followed by ultrasound-guided percutaneous aspiration injection and reaspiration (PAIR) with hypertonic saline followed by sclerosant (95% ethyl alcohol) into the residual cyst cavity. Later, she open excision of the residual multiloculated cystic mass was performed. Adjuvant medical therapy was continued for 3 months post-surgery. The patient regained her neurological functions by 6 months without any residual sequelae or symptomatic recurrence until 4 years of follow-up.

Conclusion: Multimodal treatment regimen comprising of oral medical therapy by albendazole and praziquantel along with PAIR and surgical in toto excision of the cyst followed by post-operative oral medical therapy for 3 months has given excellent results in sacrococcygeal cystic echinococcosis.

Keywords: Spine, echinococcosis, sacrum, percutaneous aspiration injection and reaspiration therapy

Introduction

Hydatid disease or cystic echinococcosis is due to the parasitic infestation of tapeworm larvae belonging to the Echinococcus family [1]. Echinococcus granulosus is prevalent in most parts of the world where sheep and cattle farming are profound like areas of the Mediterranean Sea, Asia, North, and East Africa, South America, Australia, and the Middle East [2, 3]. Humans remain an accidental intermediate host, acquiring infection from infected dog through direct contact or by ingestion of contaminated food [4]. Echinococcal embryos find their way through the intestinal mucosa and reach intestinal venules and lymphatics to gain access to the liver in 60–70% of cases [5]. Later, they enter the systemic circulation and reach various parts of the body such as lungs, brain, and so on carried by the

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Author's Photo Gallery


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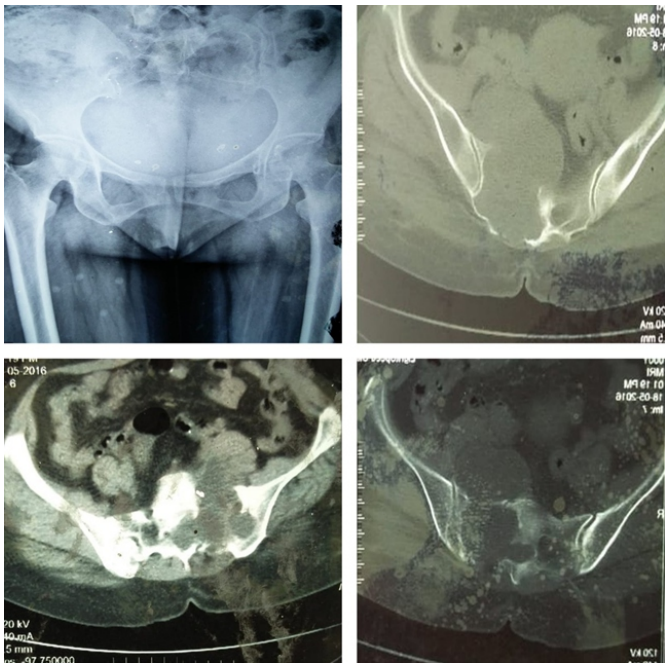

Figure 1: The X-ray and computed tomography features of the lesion extending from the posterior margin of the iliac crest to the sacrum.

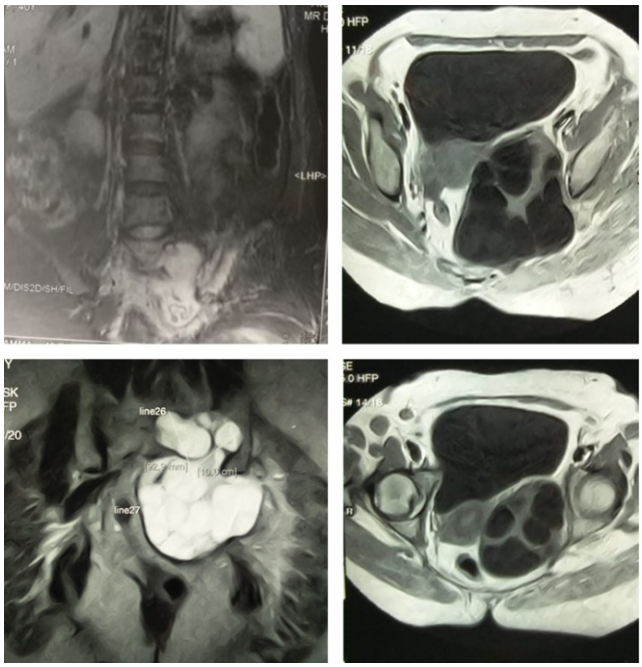

Figure 2: Magnetic resonance imaging features of the lesion with daughter cysts in situ.

bloodstream [6].

Osseous hydatidosis is not so common and is seen in only 0.5–3.1% of the total cases and of the total osseous involvement, the spine takes the major burden of the disease [7, 8]. Spinal hydatidosis may result in spinal cord compression syndromes depending on the location of the lesion [9]. Of the spinal regions, the thoracic spine is most frequently involved (46–50%) followed by the lumbar (20–29%) and sacrococcygeal involvement is very rarely noted [10]. Moreover, recurrence of the lesion is a common complication of the infection due to the infiltrative nature of the microvesicular lesions involving the spine [11].

In this case report, we describe an effective multimodal management approach toward the management of primary sacrococcygeal cystic echinococcosis combining medical, surgical, and percutaneous treatment methods described for the condition without any symptomatic recurrences.

Case Report

A 56-year-old female came with complaints of severe back pain and urinary incontinence of 3 months duration. There was no history suggestive of trauma or tumor or infection. On examination, bilateral radiculopathy with saddle anesthesia was noted. Motor power was comparable with plantar flexor response.

Urinary flow studies were carried out and the patient was found to have neurogenic overflow urinary incontinence. Other than leukocytosis and eosinophilia, all her baseline blood investigations were within normal limits. Plain radiographs showed an expansile lytic lesion affecting the right iliac wing with near-complete cortical bone destruction of the sacrum and pelvic computed tomography (CT) visualized a well-localized cystic swelling in the right pelvis over the region of lumbosacral plexus roots, as shown in (Fig. 1). Magnetic resonance imaging (MRI) revealed neural involvement, sacral destruction, and replacement by a multiloculated cystic mass extending to the

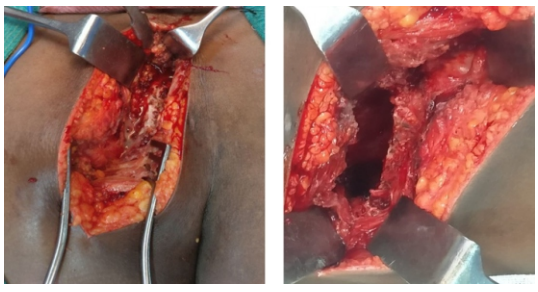

Figure 3: The exposure and complete evacuation of the cyst.

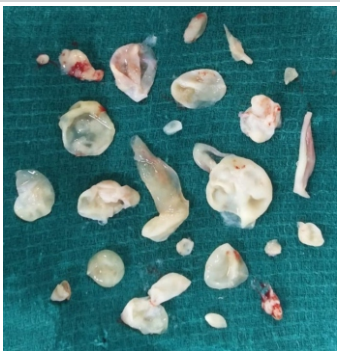

Figure 4: The surgical of hydatid cyst contents with daughter cysts and a thick cuticular layer of the cyst wall evident.

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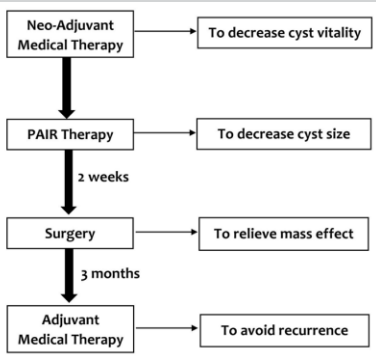

Figure 5: The multimodal approach toward themanagement of sacrococcygeal echinococcosis.

pinal canal, as shown in (Fig. 2), suggestive of infection or tumor such as echinococcosis or chordoma. The case was labeled as slow-onset cauda equina syndrome and further investigation to diagnose the etiology of this compressive radiculopathy was ascertained.

The etiology of the compression was diagnosed to be due to echinococcosis with serum enzyme-linked immunosorbent assay (ELISA). Total body survey with whole-body CT was normal without any coexistent lesions elsewhere in the body. Although the lesion presented as a slow-onset cauda-equina syndrome considering its chronic nature, we resorted to combined medical and surgical management to combat the lesion.

Neoadjuvant medical therapy in the form of tablet albendazole 400 mg twice daily and tablet praziquantel 40 mg/kg once weekly was started. Following the medical therapy, the patient underwent ultrasound-guided percutaneous aspiration injection and reaspiration (PAIR) therapy [12] of the residual cyst cavity with scolicidal agent (hypertonic saline) and sclerosant (95% ethyl alcohol). Follow-up radiological evaluation showed successful aspiration of the cyst. The following morphologic changes were noticed: A gradual decrease in cyst size, thickening, and irregularity of the cyst wall due to separation of endocyst from pericyst, and development of a heterogeneous appearance of the cyst components. The cytological examination of the fluid showed brood capsules, free scolices, and scattered hooklets which are pathognomonic of hydatid disease.

After 2 weeks of neoadjuvant medical therapy, she underwent open excision of the residual multiloculated cystic mass and lower sized hydatid vesicles to relieve the nerve root compression, as shown in (Fig. 3). The macroscopic examination of the surgical specimen showed a hydatid cyst containing clear fluid and daughter cysts with the cuticular layer of the cyst wall evident, as shown in (Fig. 4). The patient was discharged and advised to continue her adjuvant medical therapy for 3 months.

After successful completion of the multimodal treatment regimen, as shown in (Fig. 5), the patient not only recovered from her radicular pain and sensory disturbances but also had a dramatic regain of her bladder control. There was no evidence of recurrence clinically at 4 years follow-up.

Discussion

Although the most common location of hydatid disease is the liver, followed by the lung and spleen, whereas sacrococcygeal involvement of the disease is a very unusual localization [1, 10]. Bone hydatid cysts give clinical symptoms ranging from pain to pathological fractures and paraplegia [9, 13]. The severe neurological symptoms encountered, may fail to reverse after treatment mostly due to the late presentation [14]. The high rate of neurological symptoms and complications seen in bone cysts may be due to the high intracystic pressure of viable hydatid cysts [15]. The neurological symptoms are expected to regress after surgery if they are related to the mass effect of the lesion. Recurrence is more likely if bony destruction is present [16]. However, our patient despite having bony destruction did not show any sign of symptomatic recurrence until 4 years of follow-up which might be due to the early diagnosis of the disease.

Differential diagnoses of chordoma which has a lobulated and gelatinous structure are usually located in the sacrococcygeal area and infrequent presentation of tuberculosis of the sacrococcygeal area was confused with cysts on computerized tomography and MRI [17, 18]. The serological test such as ELISA throws a better light in diagnosing them as in our case [19]. Histology of the aspirated materials also reconfirms the diagnosis and can be used as a pre-operative diagnostic tool when serology remains inconclusive [20]. Pre-operative diagnosis of hydatid disease is essential because intraoperative recognition may lead to the inadvertent opening of the cyst and spillage of the cystic fluid and contents which may result in severe complications such as anaphylaxis and recurrence [21].

The most effective medical therapy appears to be the combination of albendazole and praziquantel rather than albendazole alone [22]. Hence, our regimen included both drugs. Although medical treatment can decrease the vitality of cysts, the definitive treatment is the complete surgical removal of the cyst [3]. Our multimodal treatment protocol with neoadjuvant medical management followed by ultrasound-guided PAIR initially with scolicidal agents such as hypertonic saline and sclerosant (95% ethyl alcohol) resulted in a significant reduction in the size of the lesion. After confirming the reduction of the size of the cyst, open excision for complete removal of the cysts and decompression of the nerve roots was done which was further followed by 3 months of continued adjuvant medical management. This novel approach ensured complete eradication of the hydatid cysts from the patient without any untoward complications.

Although many studies have shown that surgical approach with or without adjuvant medical therapy is the recommended treatment modality, there are still controversies regarding the correct modality of treatment [23, 24, 25]. We used PAIR therapy to downsize the lesion and to reduce the load of the cyst so that there is a decreased chance of spillover during surgery which, in turn, prevented per-operative allergic shock and the possibility of recurrence following surgery.

This novel multimodality approach toward the management of

the hydatid disease of the sacrococcygeal area adds up the advantages of all three modalities of management by medical therapy, PAIR therapy, and surgical excision. Literature review shows no such multimodal treatment protocol being followed anywhere in the management of sacrococcygeal hydatidosis [26, 27]. This combination therapy has provided excellent results on long-term follow-up in preventing recurrence and regaining the neurological loss incurred to the patient primarily. However, further studies are needed to validate the results obtained despite the disease being a rarity.

Conclusion

Multimodal treatment regimen comprising of oral medical therapy by albendazole and praziquantel along with PAIR and surgical in toto excision of the cyst followed by post-operative oral medical therapy for 3 months has given excellent results in sacrococcygeal cystic echinococcosis without any evidence of recurrence clinically.

Clinical Message

Multimodal treatment regimen comprising of oral medical therapy by albendazole and praziquantel along with PAIR and surgical in toto excision of the cyst followed by post-operative oral medical therapy for 3 months has given excellent results in sacrococcygeal cystic echinococcosis without any symptomatic recurrences.

Declaration of patient consent : The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's parents have given their consent for patient images and other clinical information to be reported in the journal. The patient's parents understand that his names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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