Bernhardt-Roth Syndrome in young women. A rare case

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ABSTRACT

Originally known as the Bernhardt-Roth syndrome, meralgia paresthetica (MP) is a condition caused by entrapment of the lateral femoral cutaneous nerve of the thigh, (Greeke: meros-thigh, algos-pain). It is a purely sensory branch with contributions from L2-L3 nerve roots and it enters to the thigh though the opening between the inguinal ligament and its attachment to the anterior iliac spine.
Hydatid disease (Greek for 'watery cyst') is an uncommon cause of peripheral nerve compression. Progressive neurological and mechanical deterioration over the years was the most frequently reported disease course.
We present a patient with MP caused by a large echinococcal cyst-mass, localized in the pelvis. A woman was admitted on to our hospital with chronic mild lower abdominal pain, severe constipation, which had lasted for about a month, and pain on the left thigh. The diagnosis after all (blood, serological and radiological tests) was an echinococcal large cystic mass (9cm x 4cm).
Total cystectomy was performed en block. Postoperatively there were significant improvement of constipation and completed resolve of femoral cutaneous nerve compression. The pain on the left thigh has disappeared.

Key words: Echinococcus cysts, meralgia paresthetica, antihelminthic, hydatid disease.

INTRODUCTION

Bernhardt-Roth syndrome is a distinctive condition, characterised by paraesthesiae and often burning pain over the anterolateral aspect of the thigh. The disorder is caused by compression of the lateral femoral cutaneous nerve as it exits the pelvis. It more commonly occurs in men than women, and is generally found in middle-aged or overweight individuals (Pearce, 2006).

Hydatid disease or hydatidosis is the most widespread, serious human cestode infection in the world. It is a zoonosis caused by Echinococcus granulosus that is transmitted from domestic and wild members of the canine family.

The genus Echinococcus contains three species: E. granulosus, E. multilocularis, and E. vogeli. Man is an incidental intermediate host for all three species.

The dog and other Canidae are the primary definitive host of the adult worm, (adult parasite < 0.6cm). Ova in the thousands are shed in the dog feces and contaminate the grass eaten by sheep. After ingestion, the embryos hatch and the parasite burrows though the duodenal wall to gain hematogenous access to multiple organs (liver, lungs, heart, bone, brain). Dogs eat these infested organs and the parasite enters the intestine where it remains. Man is infected either by eating food contaminated with eggs, or by direct contact with dogs. Echinococcal cyst are mostly frequency (60%-70% of cases), followed by the lung (10%-25%), spleen, ovaries, kidneys, brain, bones and elsewhere in the body. Hydatid disease in extrahepatic locations usually remains asymptomatic unless and produces symptoms due to pressure, rupture to the pleural or peritoneal cavity, secondary infection reaction.Treatment is surgical removal of the intact cyst (Prousalidis et al,1998;) Tepetes et al,2007).

CASE REPORT

An 28-year-old woman presented to our institution complaining of severe burning dysesthesias in the lateral aspect of the upper thigh, occasionally just above the knee, usually with increased sensitivity to clothing (hyerpathia). Hip extensions also cause pain and she mentioned that she had occasionally low pack pain and vague abdominal pain. She did not report any other symptoms. The signs and symptoms begun one year ago but the last month they had worsted.

She underwent at blood and serological tests as well as radiological examinations (X-rays, abdominal-US, CT-scans, lumbar-abdominal-pelvis-MRIs). The diagnosis was a large retroperitoneal multiloculated echinococcal cystic mass (9cm x 4cm), being in contact with the left ovary and the left ureter.

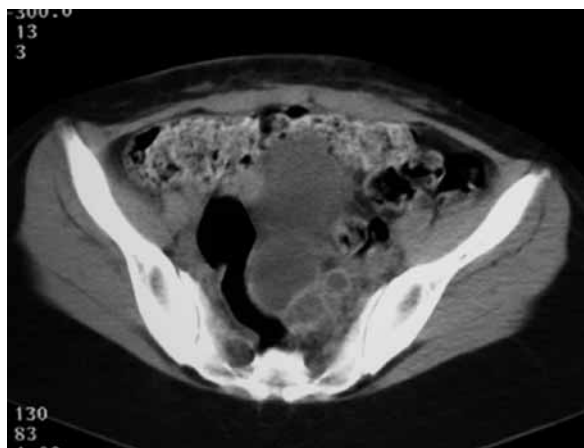
Total cystectomy was performed en block. Postoperatively there were significant improvement of constipation and completed resolve of femoral cutaneous nerve compression. The pain on the left thigh has disappeared.

DISCUSSION

Meralgia paresthetica is a clinical pain syndrome caused by entrapment, compression, or neurinoma of the lateral femoral cutaneous nerve (LFCN).The LFCN arises from the second and third ventral rami. It passes posterior to the psoas muscle, then at the lateral border emerges to run anterior to the iliacus. It continues a caudal course to the pelvic floor supplying the parietal peritoneum of the iliac fossa. To exit the pelvis the LFCN commonly passes beneath or through the inguinal ligamentmedial to the anterior superior iliac spine and through or anterior to the Sartorius muscle. This course of the nerve makes it very vurneable to stretching or compresing by big masses into the pelvis. Conservative nonoperative management is successful in relieving symptoms in the majority of cases 91% and should be tried prior to considering surgery (Williams et al, 1991). However the refractory case needs more aggressive treatment.

Once meralgia paresthetica is confirmed with history, examination, and electrodiagnostic testing (including the exclusion of a rare L2 radiculopathy), patients with bothersome symptoms may attempt a trial of oral medication prior to surgery. If this does not help or the side effects are problematic, then surgery is an option. For surgical candidates, the lateral femoral

Figure 1: A CT image of a large retroperitoneal multiloculated echinococcal cyst being in contact with the left ovary and the left ureter



cutaneous nerve at the anterior superior iliac spine is blocked with lidocaine, not only to confirm the diagnosis, but also to let the patient understand what a resected nerve would feel like.

Causes of the condition are numerous. A few of those reported in the literature include, tumors metastasis to the iliac crest, tumor involving the psoas, surgical positioning, rapid weight loss, seat-belt injury, open or laparoscopic hernia repair, surgical scars post-abdominal surgery, cardiac catheterization, sports trauma, tumor involving the lumbar vertebral bodies, pregnancy, ascites, obesity, metabolic neuropathies, and abdominal or pelvic mass (retroperitoneal tumors) (Amoiridis et al, 1993; Baldini et al, 1982; Rinkel et al, 1990) .

In the cases of retroperitoneal mass possible the large mass-size pressure and stretched the nerve at enter point to the thigh though the opening between the inguinal ligament and its attachment to the anterior iliac spine figure-1.

The condition can usually be diagnosed on clinical grounds. Differential diagnosis have to be done between femoral neuropathy, (sensory sings tend to be more anteromedial than MP), L2-L3 radiculopathy, (usually there are motor weakness- knee extension or/and thigh flexion), and nerve compression by abdominal or pelvic tumor, (it is very important to suspected if there are GI or GU symptoms).

Confirmation can be helped by tests, these tests are: EMG may be difficult, the electromyographer cannot always find the nerve, Magnetic Resonance Imaging, (MRI), somatosensory evoked potential (SEP)(Greenberg, 1997).

CONCLUSION

Although extremely rare, primary pelvic hydatid cyst-infection can cause meralgia paresthetica in man living in endemic countries and sought after with imaging, serology and clinical manifestation. Aggressive surgery combined with antihelminthic therapy is recommended to eradicate the disease and prevent recurrence.

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