Multiloculated disseminated Tarlov cysts: Importance of imaging and management options

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ABSTRACT

With technological advancements and wider availability of multimodality imaging, incidental lesions are frequently identified in patients undergoing various imaging studies. We report here a case of multiloculated disseminated perineural or Tarlov cysts (TCs). The primary aim of this case study was to (1) provide a comprehensive review of the clinical, imaging, and histopathological features of TCs (2) to draw attention to the fact that multiple lumbosacral and dorsal TCs can produce nerve injuries and serious movement disturbances, and (3) to document the usefulness of the magnetic resonance imaging and bone scan in non-invasive diagnosis and guiding management in such cases. These cysts are clearly identified by magnetic resonance imaging (MRI) and computerized tomography imaging of the lumbosacral spine. However, there are no reports on the scintigraphic findings of multilocular disseminated TC in literature. TCs are typically benign, asymptomatic lesions that can simply be monitored. To date, no consensus exists about the best surgical strategy to use when indicated.

Keywords: Magnetic resonance pelvis, perineural sacral cysts, tarlov cyst, technetium methylene diphosphonate bone scan

INTRODUCTION

Tarlov cysts (TCs) also known as Perineural cysts, are pathological dilatations, located in between the peri and endoneural spaces of the spinal posterior nerve root sheath.[1-3] They affect the sacral roots and cause a progressive painful radiculopathy. TCs usually are found in the spine and do not require surgical intervention unless symptomatic. The typical clinical presentation includes back pain, coccyx pain, low radicular pain, bowel or bladder dysfunction, lower limb weakness, sexual dysfunction, and infertility.[4] These cysts are usually diagnosed by magnetic resonance imaging (MRI) and can often be demonstrated by CT myelography to communicate with the spinal subarachnoid space.

CASE REPORT

Our patient is a 62-year-old Indian male who presented to orthopedics department with a vague history of low back ache, gradually increasing in intensity with no other associated symptoms. Bowel and bladder habits were normal. There is no history of trauma. Patient was otherwise healthy and well nourished. On local examination, no pelvic bone tenderness was elicited. No motor deficits were noted on neurological examination. A whole body bone scan was advised to rule out any skeletal pathology. 99mTc methylene diphosphonate (Technetium MDP) three-phase whole body bone scan was performed with 555 megabecqueral of 99mTc MDP intravenous injection. Initial dynamic images of pelvis followed by 3 hrs delayed high-resolution anterior and posterior whole body images were acquired using a dual head variable angle E Cam Gamma camera. An abnormal focal site of MDP uptake was noted in the sacrum S2 level indicating sacral osteoblastic activity of uncertain etiology. On further evaluation with contrast-enhanced MRI of lumbosacral spine, a cystic lesion was reported in right neural foramen of S2 of 25×15×25 mm size whose intensity was the same as cerebrospinal fluid (CSF) suggesting TC. No contrast-enhanced findings were observed. Similar lesions were also noted in S1, right T3-T4, right T7-T8, and left T6-T7 neural foramina but with no corresponding hot spots in MDP bone scan. Initial and a 6-h delayed CT myelography showed no free communication between cyst and subarachnoid space. However, there was surrounding sacral bony erosion around this cyst [Figures 1-3].

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DISCUSSION

Incidence of TCs in adult population is between 4.6% and 9%.[7] The incidence does not significantly differ between sexes but is more prevalent in younger people: 4.0% in people less than 50 years of age versus 1.3% in people over 50 years.[8] TCs are defined as CSF-filled saccular lesions located in the extradural space of the sacral spinal canal and are formed within the nerve root sheath at the dorsal root ganglion. Although the terminology of TC is synonymous for any cystic spinal lesions, the most common incidental lumbosacral lesions identified by MR are vertebral hemangiomas, perineural cysts, fibrolipomas, synovial cysts, and sacral meningoceles. TCs are distinctly different from various other benign etiologies. And their pathological confirmation rests on the fact that their cyst walls are composed of perineurium and neural tissue. The cysts show membranous tissue walls, with peripheral nerve fibers and ganglionic cells embedded into connective tissue.[7,9] Voyadzis, et al., found nerve fibers in the walls of the cysts in 75% of their patients.[10]

The cysts in TCs are usually formed by the dilated sheaths between the peri and endoneural spaces of the spinal posterior nerve root sheath having microconnections to the subarachnoid space. Thus, when pulsatile and hydrodynamic forces of CSF, through a ball-valve mechanism, cause these perineural cysts to fill and expand in size, they can begin to compress neighboring nerve fibers, resulting in neurological symptoms.[11] The ball-valve theory has been previously postulated as the reason why some large TCs cause symptoms that progress, whereas others cause only mild symptoms. Due to the valve-like mechanism and gravitational pressure within the cyst, these enlarge over time and cause neural compression. The cysts are often multiple and can erode surrounding sacral bony structures, causing irritation of the periosteal pain fibers and insufficiency fractures.[12] The other type of TCs, so-called un-valved cysts (with no CSF cross-connections) are usually asymptomatic.

Spinal meningeal cysts have been recently classified by Nabors, et al.,[11] into three different types:

• Type I: Extradural meningeal cysts without spinal nerve root fibers;
• Type II: Extradural meningeal cysts with spinal nerve root fibers (i.e., TCs); and
• Type III: Spinal intradural meningeal cysts.

Causative factors of TCs have been proposed especially leading to sacral TCs. A few of the important reasons put forward are inflammation of nerve root cysts followed by fluid collection, arachnoidal proliferation along and around the sacral nerve root, post-traumatic disruption of peri and epineural venous drainage secondary to hemosiderin deposition, other developmental or congenital factors apart from trauma have been reported in 40% cases.[7,10,12]

Patients can have a myriad of symptoms along with low back ache, which are typically accentuated by coughing, standing, and change of position explained by the increasing CSF pressure due to the ball-valve flow effect. The symptoms can be sudden or gradual. Symptomatic relief can usually be achieved by recumbent position.

MR is the gold-standard investigation in identifying TCs, and to study their relationship with surrounding structures.[13] Bone scan is highly sensitive and can easily identify early bony involvement and helps to plan surgical treatment when necessary. Conventional MR imaging shows the cyst to be a fluid-filled lesion with low signal on T1-weighted images and high signal on T2-weighted images (i.e., CSF signal). Sacral vertebral body involvement can be identified by planar or SPECT-CT (single photon emission computed tomography – computed tomography) bone scan apart from MR and CT images. Localization of MDP to sites of new bone formation is through chemisorption showing high degree of sensitivity for this imaging. There is limited role for CT here and is mainly advised for percutaneous aspiration treatment of the cysts. Development of CT myelography, an invasive imaging modality, has led to an improvement in our ability to diagnose “TCs” as a cause of sacral radiculopathy and to show any communication of these cysts with the spinal subarachnoid space. These cysts can enlarge due to inflow of CSF, ultimately producing symptoms due to distorting, compressing, or stretching of adjacent sacral nerve roots.

Optimal treatment for symptomatic TCs is still controversial despite advancements in diagnosis and imaging. Conservative management includes analgesics, physiotherapy, lumbar CSF drainage,[13] and CT-guided cyst aspiration, neither of which prevents symptomatic cyst recurrence. Neurosurgical techniques for symptomatic perineural cysts include simple decompressive laminectomy, cyst and/or nerve root excision,[14] and microsurgical cyst fenestration and imbrication.[15,16] Care must be taken in preserving nerve fibers of the parental nerve

Figure 1: (a) Lumbo-sacral contrast enhanced magnetic resonance imaging shows a cystic lesion in right neural foramen at S2 level, 25 × 15 × 25 mm in size (b) Tc MDP bone scan shows a hot spot corresponding to above lesion. The intensity of this lesion was the same as cerebrospinal fluid. Similar lesions were also noted in S1, right T3-T4, right T7-T8 and left T6-T7 neural foramina but with no corresponding hot spots in bone scan.
roots, which lie directly on the walls of the cyst. Although no consensus exists on the definitive treatment of symptomatic TCs, surgical methods have yielded the best long-term results to date. Based on patient symptoms, bone scan, and MR findings of sacral erosion, our patient underwent S2 cyst fenestration, partial cyst wall resection with myofascial cutaneous flap closure reinforcement surgery [Figure 4]. Patient has shown symptomatic improvement.

CONCLUSION

Sacral TCs are incidentally detected benign pathologies which may or may not require immediate management. However, this case is a reminder that multiple perineural TCs can be a cause of nerve roots injury, and their lumbosacral location can lead to cauda equina syndrome, without disc herniation or other cause of vertebral canal stenosis. Both MRI and bone
scan can be used as an effective screening tool to survey the whole body in a single sitting and can help in guiding early management.

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