Clinical features- 12 year old female patient presented with back ache with progressively increasing paraparesis over a period of 2 months and acute onset of urinary problems.

Imaging finding: Multiple extra-dural intra spinal CSF intensity lesions extending from D5 – L3 vertebral level causing widening of spinal canal and scalloping of vertebrae. Extension of lesion is also seen along nerve roots with widening of neural foramina. Dorsal spinal cord displaced anteriorly.

Operative finding – D6 – L4 laminectomy done which revealed Multiple (five) well defined CSF filled cysts with dural defects at D5, D7, D9, D12 nerve roots sheaths. Widening of spinal canal was seen in affected segments. The cyst were excised, their ostia were closed. Post operative course – Post operative period uneventful. Patient discharged 6th post operative day with improved motor strength in lower limbs and improved bladder control. In two months follow up patient remained relatively asymptomatic.

Spinal arachnoid cysts were first described by Spiller in 1903, although the first reported case is traced back to 1898 by Nonne (an autopsy finding). Most of the reported cases are solitary ones. Extra-dural cysts occur most frequently in the thoracic spine (65%) followed by lumbar and lumbo-sacral (13%), thoraco-lumbar (12%), sacral (6.6%), and cervical (3.3%) regions. They could occur either dorsal or ventral to the cord, with the former being more common.

Regarding the reported case so far, multiple extra-dural spinal arachnoid cysts seem to occur with higher frequency in females. The peak age of presentation is the early second decade of life. Thoracic cysts usually occur in young adolescents whereas thoraco-lumbar and lumbar cysts usually appear in adults in the 3-4th decade of life.

SEACs is a rare disease entity accounting for 1% of all spinal tumors. SEACs are assumed to be the result of dural defects. Communication between the cysts and the intradural subarachnoid space has been reported in nearly all cases of SEACs. The cause of dural defect can be congenital or acquired. Trauma,
arachnoids or iatrogenic cause can result in small dural tear and subsequent CSF accumulation to develop SEACs. Some reports demonstrated an association with dural ectasia or Marfan syndrome. In this condition, a primary defect in the organization of collagen with decreased tensile strength weakens the ligamentous structures and other supporting tissues. Dural stretching can lead to dural thinning to such an extent that it becomes ectatic and even deficient in areas. Although there is still debate in determining the etiology of SEACs, the theory of congenital dural defect is widely accepted. Dural defects may also be found around the nerve. The explanation is that tension across the movable dural sac and relatively fixed roots can predispose such dural tears. If patients have underlying structural abnormality such as Marfan syndrome, the probability of such tears may be further increased.

The wall of a spinal extradural arachnoid cyst usually consists of fibrous connective tissue with an inner single cell arachnoid lining; however, this lining is sometimes absent on histological examination. A SEAC is a meningeal cyst that can be classified into three major categories (Nabors et al). Type I cysts are composed of extradural arachnoid cysts (EAC) without nerve root fibers. Type II cysts are composed of EAC with nerve root fibers, and Type III cysts are composed of intradural meningeal cyst. Type 1 meningeal cysts can be subcategorized as EAC (Type Ia) and sacral meningoceles (Type Ib).

Magnetic resonance imaging appears to be effective as an initial modality for diagnosing arachnoid cysts and does not require the intrathecal injection of contrast medium. It can define the anatomical relationship to surrounding structures. The imaging characteristics of arachnoid cysts are similar to those of CSF signal intensity. The presence of vertebral body scalloping and expansion of the neural foramina bilaterally from osseous remodeling suggests longstanding mass effect from the lesion. With use cinematic MRI (cine-MRI) small dural defect may be detected which were missed on routine MRI.

One important aspect of preoperative evaluation is to determine the size and location of the dural defect, since it facilitates its repair via minimal laminectomy. However, the preoperative identification of dural defects is difficult. Myelography and CT myelography may disclose the cystic nature of the lesion along with its communication with the subarachnoid space as well as determination of the communication between different cysts.

The location of the cyst within the spine and the severity of spinal cord and root compression affect the clinical presentation. Spastic tetraparesis and impaired sensory levels are indicative of cervical cysts, whereas Horner syndrome is a common presentation in patients with cysts that occur lower in the cervical spine. Patients with thoracic cysts tend to present with progressive spastic paraparesis, but back pain is generally uncommon; conversely, patients with lumbar and lumbosacral cysts classically present with low-back pain, radiculopathy, and bowel and bladder dysfunction. Overall, motor weakness is usually more predominant than sensory loss. Symptoms can be intermittent and exacerbated by Valsalva maneuvers or gravitational positional forces. Remissions and fluctuation in symptoms have been reported in approximately 30% of cases.

Total cyst excision, obliteration of the communicating pedicle, and repair of the dural flap is the treatment of choice of the symptomatic lesions. Postoperative kyphosis may be prevented by performing laminoplasty rather than laminectomy.

CONCLUSION

Spinal arachnoid cysts are a rare cause of spinal cord compression. MRI is the diagnostic procedure of choice as it is noninvasive and can demonstrate the nature of cyst, size, and the anatomic relationship with the spinal cord. Signs and symptoms of SEACs are due to chronic cyst expansion and compression of the neural structures and thus, SEACs need surgical treatment. Diverse surgical techniques have been introduced and complete microsurgical resection of SEACs with meticulous repair of dural defect has been advocated as treatment of choice for SEACs.

Figure 1 A sagittal T2W image showing large segment T2 hyperintense lesion extending from D3 – L4 level. Figure 1B T2 sagittal image showing thin T2 hypointense cyst walls. Image 1C T1W showing hypointense lesion in the spinal canal extending in the neural foramina. Image 1D T2W axial image showing T2 hyperintense lesion consistent with CSF intensity. Spinal cord is displaced anteriorly.

Figure 2 – Intra operative image showing CSF filled cystic lesion.

Reference

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