INTRASACRAL PERINEURAL CYST ASSOCIATED COMMUNICATING SYRINGOMYELIA

Zeki Şekerç, M.D., Ömer İyğün, M.D., Bedri Kandemir, M.D., Fahrettin Çelik, M.D., Sancar Barış, M.D.

University of Ondokuz Mayıs School of Medicine, Department of Neurosurgery (ZŞ, Oİ, FC) and Pathology (BK, SB). Samsun - TÜRKİYE

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SUMMARY:
In this report an unusual case of intrasacral perineural cyst associated syringomyelia diagnosed with metrizamide myelography and spinal computed tomography is presented and the pathogenesis is discussed.

KEY WORDS:
Intrasacral, perineural cyst, communicating syringomyelia.

INTRODUCTION:

In 1938, Tarlov gave first detailed description of cystic lesions of spinal nerve roots and introduced the term “Perineural cyst” (16). These cysts are usually asymptomatic and discovered during a delayed film of myelography with water soluble contrast medium. investigation of the spinal canal with computed tomography (CT) or nuclear magnetic resonance imaging (NMR) (5,6,11.12,13,17).

Nishihara et al. could collect only 16 similar cases in addition to their own case that satisfied the criteria established by Tarlov (8). Perineural cysts usually involve the sacral nerve roots and are often multiple (1,2,4,15,19).

Associated conditions with perineural cysts are uncommon (3,5,8,21).

In this report an unusual case of intrasacral perineural cyst associated communicating syringomyelia is presented and the pathogenesis discussed.

CASE REPORT:
A 26-year-old man was admitted to the neurosurgery clinic complaining of weakness and numbness of the right arm and leg over a period of five years, and urinary urgency for one year. He had also a history of trauma to the right lumbar region nine years before admission.

Physical examination revealed scoliosis at the thoracic region.

Neurological examination on admission showed loss of temperature and pain appreciation of the right upper and lower extremities with preservation of touch and joint position. Motor examination revealed a mild degree weakness of the right hand forearm, right leg, complete loss of dorsal flexion of the right foot and a slight degree of weakness of the left forearm and hand. Deep tendon reflexes (DTR) of the right upper and lower extremities were hyporeflexic. Diffuse fasciculation and atrophy of the muscles of the right arm and leg were also observed.

Plain radiography of the spinal canal showed kyphoscoliosis of the thoracic region and widening of the interpedicular distance of the cervico-thoracic region. There was also scalloping and erosion of the right dorsal lamina of the sacral canal on plain radiography of the sacrum.
Early and delayed (30 minutes and 6 hours after instillation of metrizamide to the lumbar subarachnoidal space) films of metrizamide myelography confirmed an intrasacral cystic lesion between L5-S4 levels (Figure 1 a.b).

Early and delayed examination of the spinal canal with CT confirmed a cystic lesion in the sacral canal and showed a communicating syrinx cavity at the cervico-thoracic region (Figures 2, 3).

Fig 1 a: Myelographic view of the intrasacral perineurial cyst immediately after instillation of metrizamide to the lumbar subarachnoidal space.

Fig 1 b: Myelographic appearance of the intrasacral perineurial cyst 1/2 hour after instillation of metrizamide to the lumbar subarachnoidal space.

Fig 2: Delayed CT appearance of syrinx cavity at cervical region.

Fig 3: CT appearance of the intrasacral perineurial cyst immediately after metrizamide myelography.
CT examination of the foramen magnum did not show any cerebellar tonsil herniation.

Following sacral laminectomy, a dark blue cystic mass was encountered. The right side of the posterior wall of the sacral canal was eroded. The mass which occupied and enlarged the right side of the sacral canal, extended from the level of S1 to S5 and displaced the cauda equina toward the left side. The cyst possibly originating from the S2 root sheath was removed completely with the degenerated S2 root. The early post-operative period was uneventful. Microscopic examination of the specimen proved the cyst wall contained degenerated nerve tissue (Figure 4 a,b).

![Fig 4: a Photomicrographic view of the cyst wall showing degenerated nerve tissue. x 25 (HE)](image)

![Fig 4: b High magnification photomicrographic view of the degenerated nerve tissue in the cyst wall. x200 (HE)](image)
A second operation was performed fifteen days after the first. With C5.6 laminectomy a polyethylene tube was placed in the syrinx cavity for treatment of syringomyelia. The second postoperative period was also uneventful. Follow-up neurological examination of the patient three months after the second operation was stable.

DISCUSSION:

The spinal perineural cyst arises between the arachnoid that covers a nerve root (perineurium) and the outer surface of its pia (endoneurium), and the walls are formed of these elements (9,19). These cysts are usually asymptomatic and involve the sacral nerve roots (8,9,18). The characteristics and pathogenesis of perineural cyst was defined by Tarlov during an anatomical study of five human cadavers (16,17). According to Tarlov perineural cysts occur as a result of a traumatic haemorrhage within the nerve sheath (16). Which leads to a splitting of the sheath with cystic dilatation of the potential space between the endoneurium and the perineurium (5,12,14). Arachnoidal proliferation may also cause cystic dilatation between root fascicles (10).

The perineural cyst is filled with a clear fluid that is probably initially CSF in communication with CSF in the spinal subarachnoidal space. Later this communication may close (19). At least a part of the wall of a perineural cyst must contain nerve fibres or occasionally ganglion cells on histopathological examination (8).

Routine use of water soluble contrast medium for myelography, spinal CT and NMR for spinal canal examination has increased the chance of finding perineural cysts and associated conditions (6,8,11,13,15). The hydrostatic pressure and water-hammer effect of the CSF is responsible for enlargement of cystic dilatation (8,19). In this case CSF pulsations and delayed equilibration of intraspinal and intracranial pressure may be a causative factor for enlargement of the syrinx and the intrasacral cyst cavity (5,20).

Since most of the spinal perineural cysts are asymptomatic and are found incidentally, specific treatment is not necessary (7,19). Surgical treatment of associated conditions like this presented case can be planned as a second step.

REFERENCES