U. Agrillo
M. N. Tirendi
P. V. Nardi

Symptomatic cystic dilatation of V ventricle: case report and review of the literature

Received: 19 April 1996
Revised: 26 July 1996
Accepted: 17 August 1996

U. Agrillo · M. N. Tirendi · P. V. Nardi (✉)
Neurosurgery Division,
S. Pertini Hospital, Via Monti Tiburtini,
I-00157 Rome, Italy

Abstract We report a case of V ventricular cystic dilatation, presenting with specific neurological symptoms including low back pain, bilateral sciatica, weakness of dorsiflexion, and urinary retention. MRI showed a large cystic dilatation of the ventriculus terminalis. Surgical

fenestration of the cyst allowed complete relief from symptoms and remission of the neurological deficit.

Key words Spinal cord · Conus medullaris · Ventricular cyst

Introduction

The ventriculus terminalis is a small cavity located inside the conus medullaris. The cavity is lined with ependyma and usually rudimentary. A pathological study by Choi et al. [2] demonstrated that the ventriculus terminalis was always detectable by histologic examination of the adult or pediatric conus medullaris, while neuroimaging techniques can hardly detect it.

The genesis of the ventriculus terminalis is related to the embryonic development of the neural tube, more specifically to the period between the 4th and 6th weeks of fetal life. During this period, the neural epithelium (caudal to the posterior neuropore) fuses with the notochord forming a caudal cell mass. Later, multiple vacuoles are formed inside this mass: they coalesce until they form an ependyma-lined canal which subsequently joins the central canal of the rostral neural tube. At 5.5 weeks the upper part of this ependyma-lined cavity, at the 32nd somite, dilates, thus producing the ventriculus terminalis within the conus medullaris. The lower part of this canal and the caudal cell mass undergo complete regression resulting in the formation of the filum terminale, though in some cases the ventriculus terminalis may extend into the filum terminale itself. The ventriculus terminalis reaches its final size after the 2nd year of life [6].

So far, neither the physiologic role of the ventriculus terminalis nor the cause of its cystic dilatation has been

defined [2]. The studies by Gardner and Williams studies [4, 5] and Aboulker's theory [1] do not seem applicable to this lesion. We prefer to agree with the hypothesis of Sigal et al. [13] who suggest that the dilatation could be the result of a lack of communication between the ventriculus terminalis and the central canal of the spinal cord. Consequently, cystic dilatation of the ventriculus terminalis has to be considered a congenital malformation and should be differentiated from other cystic lesions of this area, especially tumors.

Case report

S.P., 34 years old, was admitted to our division at the end of November 1993, complaining of having suffered from bilateral sciatica for 4 years and urinary retention for 3 years. Neurological examination showed bilateral paresis of dorsiflexion of the feet, mild bilateral atrophy of the tibialis anterior muscles, hypoesthesia of the L4, L5, and S1 dermatomes, and loss of knee and ankle reflexes.

Urodynamic analysis documented marked postmicturitional urinary retention, while the electromyographic study showed bilateral radiculopathy of the L4, L5, and S1 nerve roots. Finally, the patient underwent MRI of the dorsal and lumbar spine, which showed an intradural cystic lesion situated intraspinally at the level of the conus, compressing the conus itself and the cauda nerve roots. The lesion had the same signal intensity as CSF in all sequences and there was no change after Gd-EDTA administration (Figs. 1, 2). The dilated conus showed a normal position within the spinal canal, ruling out a tethered cord. Brain MRI showed no co-existing congenital malformations.

Fig. 1A, B Preoperative sagittal T1- and T2-weighted MR images, showing a thin-walled intraspinal cyst extending between T12 and L2. Gd-EDTA administration did not lead to contrast enhancement of the cyst wall or its fluid portion

Fig. 2 A Preoperative axial T1-weighted MR image after Gd-EDTA infusion, showing a marked ballooning cystic dilatation of the conus medullaris. **B** Postoperative collapse of the lesion after opening and fenestration

Fig. 3A, B Postoperative (14 months) sagittal T1- and T2-weighted MR images, showing partial obliteration of the dilated ventriculus terminalis

The patient was therefore operated upon. A bilateral T11–L2 laminectomy was performed. After opening of the dura, the conus was exposed. There was marked cystic dilatation of the conus extending caudally between the roots of the cauda equina. The posterior surface of the cyst was opened, releasing CSF and showing an internal lining of the ependymal type, surrounded by a thin layer of neural tissue. The cyst was fenestrated by suturing the arachnoid and pial layers of the lesion to the inner side of the dural envelope. The dura was subsequently closed in a water-tight manner.

The post-operative course was uneventful and the patient was discharged after 9 days. Neurological examination at discharge showed almost complete regression of the preoperative paresis and hypoesthesia. Urinary function improved with a postmicturition residual volume of 200 ml. The follow-up examinations at 3 and 6 months showed complete regression of the neurological deficits and absence of urinary retention. Fourteen months after surgery, the electromyographic examination was normal, while spinal MRI

confirmed obliteration, albeit partial, of the cystic dilatation of the ventriculus terminalis (Fig. 3).

Discussion

Our review of published data reveals a homogeneous presentation and clinical evolution in patients with symptomatic cystic dilatation of the ventriculus terminalis [9, 13]. Nassar et al. [9] reported three cases of cystic symptomatic lesions of the conus medullaris. All the patients complained of low back pain with radiation to the legs. Pain was always associated with slowly increasing paresis of the legs, involving most frequently the quadriceps, tibialis anterior, and peroneal muscles. In addition, sphincteric disturbances were always present. After laminectomy and fenestration of the cysts, the authors obtained resolution of the symptoms and almost complete regression of the neurological deficits. A biopsy of the lesion was obtained in two cases, identifying the cyst as a congenital cystic malformation of the filum terminalis, and ruling out more probable diagnosis of symptomatic cystic dilatation of the ventriculus terminalis.

A series published by Sigal et al. in 1991 [13] consisted of four patients aged between 35 and 62. All patients showed the same neurological deficits and symptoms as those reported by Nassar et al. Sigal et al. identified the lesions of the conus with cystic dilatation of the ventriculus terminalis and described their MR characteristics.

MRI allows differentiation of a dilated ventriculus terminalis from other, more frequently observed, cystic neo-

plastic lesions, especially ependymomas, glial neoplasms, or dermoid cysts. MRI shows the lesion to be strictly localized inside the conus medullaris. This is important for the differential diagnosis with syringomyelic lesions [3]. In the rare cases of conus medullaris syringomyelia, the syrinx will always extend cranially. Neoplastic cystic lesions usually show solid portions with contrast enhancement after Gd-EDTA administration. In a case of cystic ventriculus terminalis there is no uptake of Gd-EDTA, and the signal intensity on MRI is always exactly the same as that of CSF in all sequences [13].

Finally, Sigal et al. recommend a complete neuroradiological study of these lesions with MRI to exclude the coexistence of other associated malformations [7, 10, 11, 14], reporting that one of their patients had a coexistent Chiari I malformation.

To conclude our brief review, we wish to emphasize that dilatation of the ventriculus terminalis has only rarely been described. We feel that this type of congenital lesion has to be considered in the differential diagnosis of more frequent neoplastic lesions. Usually spinal MRI will allow differentiation between these lesions.

In patients who undergo surgical treatment, we do not recommend biopsy of the lesion wall, because this will show histological characteristics just like those of normal ependyma. In published cases where surgical biopsy of the lesion was performed, the histological examination always showed an ependymal cell layer or normal neural tissue. Moreover, there is always the risk that surgical biopsy could cause a neurological deficit.

References

1. Aboulker J (1979) La syringomyélie et les liquides intrarachidiens. *Neurochirurgie* 25 [Suppl 1]: 1-144
2. Choi BH, Kim RC, Suzuki M, Choe W (1992) The ventriculus terminalis and filum terminale of the human spinal cord. *Hum Pathol* 23: 916-920
3. Enzmann DR (1990) In: Enzmann DR, De LaPaz RL, Rubin JB (eds) *Magnetic resonance imaging of the spine*. Mosby, St Louis, pp 318-324
4. Gardner WJ (1965) Hydrodynamic mechanism of syringomyelia: its relationship to myelocoele. *J Neurol, Neurosurg Psychiatr* 28: 247-259
5. Gardner WJ, Angel J (1975) The mechanism of syringomyelia and its surgical correction. *Clin Neurosurg* 6: 131-140
6. Kernohan JW (1924) The ventriculus terminalis: its growth and development. *J Comp Neurol* 38: 107-125
7. Kochan JP, Quencer RM (1991) Imaging of cystic and cavitary lesions of the spinal cord and canal. *Radiol Clin North Am* 29: 867-911
8. Mairui F, Gambardella A, Trinchillo G (1989) Congenital lumbosacral lesion with late onset in adult life. *Neurol Res* 11: 238-244
9. Nassar SI, Correl JW, Housepian EM (1968) Intramedullary cystic lesions of the conus medullaris. *J Neurol Neurosurg Psychiatr* 31: 106-109
10. Robertson DP, Kirkpatrick JB, Mawad ME (1991) Spinal intramedullary ependymal cyst. *J Neurosurg* 75: 312-316
11. Rogers MA (1992) Congenital, hereditary and developmental spinal cord disease. *Curr Opin Neurol Neurosurg* 5: 537-539
12. Russel DS, Rubinstein LJ (1989) *Pathology of tumors of the nervous system*, 5th edn. Arnold, London, pp 94-95
13. Sigal R, Denys A, Halimi P, Shapeero L, Doyon D, Boudghéne F (1991) Ventriculus terminalis of the conus medullaris: MR imaging in four patients with congenital dilatation. *Am J Neuroradiol* 12: 733-737
14. Steinbock P, Cochrane DD (1991) The nature of congenital posterior cervical or cervico-thoracic midline cutaneous lesions. *J Neurosurg* 75: 206-212
15. Towfighi J, Housman C (1991) Spinal cord abnormalities in caudal regression syndrome. *Acta Neuropathol* 81: 458-466