

Neurinoma of cauda equina associated with normal pressure hydrocephalus

Abad Cherif El Asri, Okacha Naama, Miloudi Gazzaz, Brahim El Mostarchid, Mohamed Boucetta

Abstract: Neurinoma of cauda equina associated with normal pressure hydrocephalus is rare with only 13 cases reported in the literature. A 65-year-old man was operated for lumbar neurinoma, revealed by cauda equina syndrome. On the 15th postoperative day, the patient presented neurological stagnation, with mental deterioration. Cranial CT-scan showed tetraventricular hydrocephalus. Following insertion of ventriculoperitoneal shunt, we observed good amelioration of neurological deterioration. Twelve months postoperatively, the patient regained full neurological function. Many hypotheses were presented but alteration of craniospinal compliance distribution and the increase in CSF protein content are the most widely accepted theories explaining this association. (p94-96)

Key words: Neurinoma, cauda equina and normal pressure hydrocephalus

Introduction

Hydrocephalus complicates approximately 1% of intraspinal tumours.⁶ In opposition association of normal pressure hydrocephalus (NPH) and an intraspinal tumour is rare. Only 13 cases of intraspinal neurinoma associated with NPH have been reported in the literature since 1973.^{1,3,5,7-15} Increased intracranial pressure is the most commonly revealed symptom of acute hydrocephalus, although root pain and motor deficit/s can be an isolated symptom of the association of NPH with spinal tumour. Epidemiologic, physiopathologic and therapeutic aspects are discussed in the light of the relevant literature.

Case Report

A 65-year-old man with diabetes and hypertension, complaining for 1 year of progressive gait disturbance and intermittent urinary incontinence, without memory disturbance or headache. He reported bilateral anterior thigh paresthesias. Physical examination revealed bilateral 2/5 weakness of the hip flexion and hypoaesthesia of the lateral side of thigh. Lumbar spinal magnetic resonance imaging (MRI) demonstrated a well-demarcated intradural tumour

in the cauda equina at L1-L2 level (Fig. 1). The tumour exhibited marked enhancement after intravenous administration of gadolinium and compressed the medullary cone and the cauda equina roots. The electromyography of the lower limbs detected bilateral crural suffering. The patient was operated and posterior L1-L3 laminectomy was performed. After the dural sac was opened, encapsulated process of the left L3 root was discovered and completely removed. The histological examination showed typical features of benign neurinoma. The immediate postoperative findings were marked by progressive recovery of motor and sensory deficits of lower limbs but with persistence of gait and urinary disturbance. After three weeks, the patient presented with progressive memory disturbance and headache, however, Adams-Hakim triad was well-distinguished and cranial CT- scan showed tetraventricular dilatation (Fig. 2). Lumbar puncture was performed, CSF analysis showed high protein level at 1g/l. Following lumbar puncture, some degrees of improvement of Adams-Hakim triad was observed, and CSF protein level was decreased one month later to 0.4 g/l. Ventriculoperitoneal shunting was performed 3 months after neurinoma removal. The patient showed a satisfactory postoperative course and within 2 months he regained normal cognitive and urinary function with persistence of slight left limb deficit.

Discussion

The symptom triad of NPH is usually described as gait disturbance, dementia, and urinary incontinence.⁴ It occurs in elderly patients, since the first report of intraspinal tumour and NPH by Neil-Dwyer in 1973, only 13 cases of neurinomas have been published to date.^{1,3,5,7-15} Table 1 illustrates the previously reported cases including our presentation. The mean age of occurrence is 65 years (45 -

Department of Neurosurgery
Military Hospital
Rabat
Morocco

Correspondence:

Dr. Abad Cherif El Asri
Department of Neurosurgery
Military Hospital
Rabat
Morocco
Tel: (212 6) 124 7544
Email: abad20031@hotmail.com

75 years). The site of the lesion is mostly cauda equina. A third of cases presented with radiculopathy or medullary symptoms as the major symptom of this association; hydrocephalus may be underestimated.^{1,3,5,7-15} However, gait and urinary disturbance were confined to signs of cauda equina root compression. In our case, signs of radiculopathy and urinary disturbance were related to the compression of cauda equina roots by neurinoma. No evident symptom of NPH was detected preoperatively and cranial CT scan was not performed at this time. Urodynamic studies are necessary to distinguish between

urinary disturbance secondary to root compression and those secondary to NPH. In this situation, it can demonstrate hyperreflexia and instability of the bladder detrusor muscle, but there is no evidence of defective bladder control. This suggests damage in the periventricular pathways to the sacral-bladder center and decreased inhibition of bladder contractions.² The pathophysiology of this association is unclear and many theories have been proposed. Postoperative spinal subarachnoid haemorrhage may decrease CSF absorption or circulation.¹² Compression of the spinal venous plexus and the venous medullary plexus by tumour invasion causes increased venous intracranial pressure and may interfere with CSF absorption.⁶ However, this explanation alone is not sufficient. One of the most widely accepted theories relates to the increase in CSF protein content, which may obstruct CSF absorption pathways or raise CSF flow resistance by increasing CSF



Figure 1 ← Sagittal lumbar T2 weighted MRI showing intradural tumour in the cauda equina at L1-L2 level



Figure 2 → Cranial CT scan showing markedly dilated ventricles with signs of periventricular CSF resorption

Table 1 - Summary of reported cases of intraspinal neurinoma associated with NPH

Authors (year)	Age	Sex	Chief symptoms	Tumour location	Treatment
Neil-Dwyer 1973 ⁹	66	M	Paraparesis	Cauda equina	Tumour removal and ventricular shunt
Luzecky 1974 ⁵	58	M	Papilloedema + NPH	Cauda equina	Tumour removal
Bamford & Labadie 1978 ¹	75	M	Cauda equina syndrome	Cauda equina	Tumour removal
Ridsdale 1978 ¹² 1 st case	49	F	Spinal cord compression	Conus medullaris	Tumour removal
Ridsdale 1978 ¹² 2 nd case	62	F	NPH	Cauda equina	Tumour removal and ventricular shunt
Messer 1980 ⁷	73	F	NPH	Cauda equina	Tumour removal
Feldmann 1986 ³	68	F	NPH	Cauda equina	Tumour removal
Yarnitsky 1987 ¹⁵	71	M	Cauda equina syndrome	Cauda equina	Tumour removal and ventricular shunt
Nichida 1990 ¹⁰	45	M	NPH	Cauda equina	Tumour removal
Ohta 1990 ¹¹	65	F	NPH	Cauda equina	Tumour removal
Tourniaire 1993 ¹⁴	76	M	NPH	Cauda equina	Tumour removal and ventricular shunt
Sugaya 1993 ¹³	69	F	NPH	Cauda equina	Tumour removal
Munakata 2002 ⁸	69	F	Parkinson's + NPH	Cauda equina	Tumour removal
Present case	65	M	Cauda equina syndrome	Cauda equina	Tumour removal and ventricular shunt

viscosity.⁶ All reported cases have a high level CSF protein, as in our patient.¹⁰ Moreover, the protein concentration rostral to tumour is usually 9-10 times less than in CSF caudal to tumour.⁶ After tumour removal, CSF rich in protein and snared caudally to tumour will be discharged and can decrease the mechanisms of periventricular CSF absorption. This is found in association to hypertension, subcortical and periventricular white matter abnormalities, and it suggests a possible cerebrovascular role in the development of NPH.² In our patient, the CSF protein level was decreased from 1 g/l at the third week after surgery, to 0.4 g/l one month later. When the mechanisms of periventricular CSF absorption are sufficiently deteriorated, hydrocephalus will evolve, in spite of intraspinal tumour removal.¹⁴ This will explain the mental, gait and urinary disturbances experienced 3 weeks after neurinoma removal in our patient. Morandi proposed a mechanical theory of hydrocephalus and hypothesized that hydrocephalus in benign intraspinal tumours is the result of alteration of craniospinal compliance distribution.⁶ Intraspinal tumours may decrease the compliance of the caudal spinal portion, thereby reducing overall CSF space compliance. In this condition, minimal changes to other implicated factors such as CSF protein concentration, intracranial CSF flow resistance, due to arachnoiditis or intracranial vascular compliance may have a greater effect on CSF dynamics.⁶ These theories were proposed to explain increased intracranial hypertension, which are well described in young people. In elderly patients, the excess ventricular fluid is compensated for by ventricular enlargement, the baseline CSF pressure remains unchanged but pressure increases with transient and repetitive events resulting from cardiac and respiratory pulsations. Ventricular enlargement may develop when the compensation phenomena are exceeded. Consequently, "compensation" induced by increased ventricular size and probably ependymal absorption of cerebrospinal fluid, instead of spinal compliance, occurs at the expense of normal brain function.⁴ After spinal tumour removal, hydrocephalus disappears rapidly due to spinal compliance recovery; as in most cases reported but in 4 cases, as in our patient shunting of CSF was necessary to resolve all symptoms of NPH.

Conclusion

Normal pressure hydrocephalus must be dreaded in elderly people with spinal tumours. Many hypothesis were well

proposed, alteration of craniospinal compliance distribution and the increase in CSF protein content are the most widely accepted theories explaining this association. Treatment consisting of only tumour removal can be sufficient to resolve all NPH symptoms. Rarely, ventriculoperitoneal shunting is necessary after spinal tumour removal.

References

1. Bamford CR, Labadie EL: Reversal of dementia in normotensive hydrocephalus after removal of a cauda equina tumor: Case report. *J Neurosurg* 1976, 45(1): 104-7
2. D'Abreau A: Normal pressure hydrocephalus. *Medicine and Health / Rhode Island* 2004, 87(12): 372-374
3. Feldmann E, Bromfield E, Navia B, et al: Hydrocephalic dementia and spinal cord tumor: report of a case and review of the literature. *Arch Neurol* 1986; 43: 714-718
4. Hamlat A, Sid-Ahmed S, Adn M, Askar B, Pasqualini E. Idiopathic normal pressure hydrocephalus: theoretical concept of a spinal etiology. *Med Hypotheses* 2006, 67:110-4
5. Luzecky M; Siegel BA; Cox WS; Berg L. Papilledema and communicating hydrocephalus. Association with a lumbar neurofibroma. *Arch Neurol*. 1974, 30(6): 487-489
6. Morandi X, Amlashi AFA, Riffaud L: A dynamic theory for hydrocephalus revealing benign intraspinal tumours: Tumoural obstruction of the spinal subarachnoid space reduces total CSF compartment compliance. *Med Hypotheses* 2006, 67(1): 79-81
7. Messer HD, Brinker RA: Hydrocephalus and dementia complicating spinal tumor: Case report. *J Neurosurg* 1980, 53: 544-547
8. Munakata S, Nagumo K, Tanno T, Kojima S: A case of Parkinsonian syndrome caused by normal pressure hydrocephalus accompanied by the cauda equina neurinoma. *Rinsho Shinkeigaku* 2002, 42(2): 131-5
9. Neil-Dwyer G: Tentorial block of cerebrospinal fluid associated with a lumbar neurofibroma. *J Neurosurg* 1973, 38: 767-770
10. Nishida K, Ueda S, Matsumoto K, Kusaka K, Takeuchi R: Cauda equina neurinoma associated with normal pressure hydrocephalus - case report. *Neurol Med Chir (Tokyo)* 1990, 30(4): 258-62
11. Ohta K, Gotoh F, Amano T, Obara K: Normal pressure hydrocephalus associated with cauda equina neurinoma. *Annals Neurol* 1990, 27(4): 441-443
12. Ridsdale L, Moseley I: Thoracolumbar intraspinal tumours presenting features of raised intracranial pressure. *J Neurol Neurosurg Psychiatry* 1978, 41: 737-45
13. Sugaya M, Noguchi M, Tsutsumi Y: Neurinoma of the cauda equina associated with normal pressure hydrocephalus. *No To Shinkei* 1993, 45(9): 871-5
14. Tourniaire D, Sankey R, Pages M, Privat JM, et al: Hydrocephalie a pression normale et neurinome lombaire. *Rev Neurol (Paris)* 1993, 149(12): 802-4
15. Yarnitsky D, Honigman S, Hemli JA, Bental E: Normal-pressure hydrocephalus associated with spinal cord tumor. *Acta Neurol Scand* 1987, 76(4): 302-305