© Pol J Radiol 2018; 83: e229-e233
DOI: https://doi.org/10.5114/pjr.2018.76380

Received: 15.05.2017
Accepted: 27.07.2017
Published: 22.05.2018

Polish Journal of Radiology

http://www.polradiol.com

Case report

Spontaneous intracranial hypotension: two steroid-responsive cases

Russo Camilla[1,A,D,E,F], Buono Vincenzo[2B,D], Fenza Giacomo[3B,D], Zandolino Alexis[1B,D], Serino Antonietta[3B,D], Manto Andrea[3A,D,E,F]

[1]Department of Advanced Biomedical Sciences, University „Federico II", Naples, Italy
[2]Department of Neurosurgery, Umberto I Hospital, Salerno, Italy
[3]Department of Neuroradiology, Umberto I Hospital, Salerno, Italy

Abstract

Purpose: Spontaneous intracranial hypotension (SIH) is characterised by orthostatic headache, low cerebrospinal fluid pressure and diffuse pachymeningeal enhancement after intravenous gadolinium contrast administration. Magnetic resonance imaging (MRI) often plays a crucial role for correct diagnosis.

Case description: We described two similar cases of SIH, whose clinical and imaging features are typical for this pathology. At MRI brain scan, both patients showed diffuse and intense pachymeningeal enhancement and moderate venous distension and epidural vein engorgement. The two patients were treated with bed rest and oral steroid therapy, with complete and long-lasting symptomatic relief.

Conclusions: Orthostatic nature of headache is the most indicative clinical feature suggesting SIH; contrast-enhanced MRI provides definite imaging diagnostic findings. Conservative treatment coupled to steroid therapy is often sufficient to obtain complete disappearance of symptoms.

Key words: spontaneous intracranial hypotension (SIH), cerebrospinal fluid (CSF) pressure, myelopathy.

Introduction

Considered to be a benign condition, spontaneous intracranial hypotension (SIH) is defined as an idiopathic primary cerebrospinal fluid (CSF) pressure < 60 cmH_2O in patients with no history of penetrating trauma or dural punction. It must be distinguished from secondary intracranial hypotension, iatrogenic or traumatic, the incidence of which is significantly larger [1-4].

Patients generally have variable orthostatic symptoms such as postural headache, neck pain, vertigo, anorexia, weakness, nausea or vomiting, and aspecific visual and hearing disturbances; sometimes radicular symptoms involving the upper limb can be present. Complications are sporadic and generally secondary to subdural haematoma [5].

SIH is most commonly related to unapparent CSF leak along the neuraxis, whose exact site cannot be localised and primitive cause assessed. According to Monro-Kellie's hypothesis, SIH should be due to an alternate balance in the proportion of blood, CSF, and brain tissue in the central nervous system (CNS) [6].

Symptomatology usually disappears completely with conservative treatment and bed rest [7]. When spontaneous regression is not obtained with these measures, fluid restoration can be attempted in order to increase CSF volume by using intravenous or oral hydration, carbon dioxide inhalation, and steroid therapy [8]. In case of failure, epidural infusion of saline, epidural injection of fibrin glue, and epidural blood patches are considered an effective although more invasive treatment [9]. As a last

Correspondence address:
Camilla Russo, MD, Department of Advanced Biomedical Sciences, University "Federico II", Via Sergio Pansini 5, 80131 Naples, Italy, phone: 0039 333 7050711, e-mail: camilla_russo@hotmail.it

Authors' contribution:
A Study design · **B** Data collection · **C** Statistical analysis · **D** Data interpretation · **E** Manuscript preparation · **F** Literature search · **G** Funds collection

therapeutic chance, surgical strategies may be required only in the case of documented meningeal defects [10].

Common diagnostic features at magnetic resonance imaging (MRI) examination include pachymeningeal thickening and post-contrast enhancement, venous enlargement and engorgement (specially of intracranial dural sinuses and spinal peridural plexuses), intrasellar arachnoid diverticulum or pituitary enlargement, subdural fluid collections, and deep brain swelling [5,11-14].

Case description

Case 1

A 35-year-old woman presented to the emergency service with a headache that developed over a week and progressively grew in intensity until it became unbearable. The patient reported suffering from periodic attacks of transient headache, sometimes associated with nausea and/or dizziness, beginning within 30 minutes of standing without remarkable triggering events. No other comorbidity was reported; neither use of medicines or contraceptives nor alcohol or illicit drug intake was discovered in her medical history. Neurological examination, blood analysis, and computed tomography (CT) scan performed at the emergency service were normal. Due to the ineffectiveness of conventional therapies, a standard MRI brain and cervical spine examination before and after intravenous gadolinium administration was performed. MRI showed diffuse pachymeningeal enhancement with positive venous distention sign, and reduced mamillopontine distance (about 5 mm) and cerebellar tonsils ectopia (about 4 mm) [15]. No associated subdural effusion was highlighted; callosal and pontomesencephalic angles were normal. As an occasional finding, a single CNS capillary telangiectasia was observed in the right putamen (Figure 1). The patient was treated with bed rest and steroid therapy (oral treatment with prednisone, 50 mg/day, with a gradual withdrawal in 30 days) [16], with complete symptomatic relief in a few days. After two months, follow-up brain MRI showed almost complete resolution of the imaging pathological findings (Figure 2).

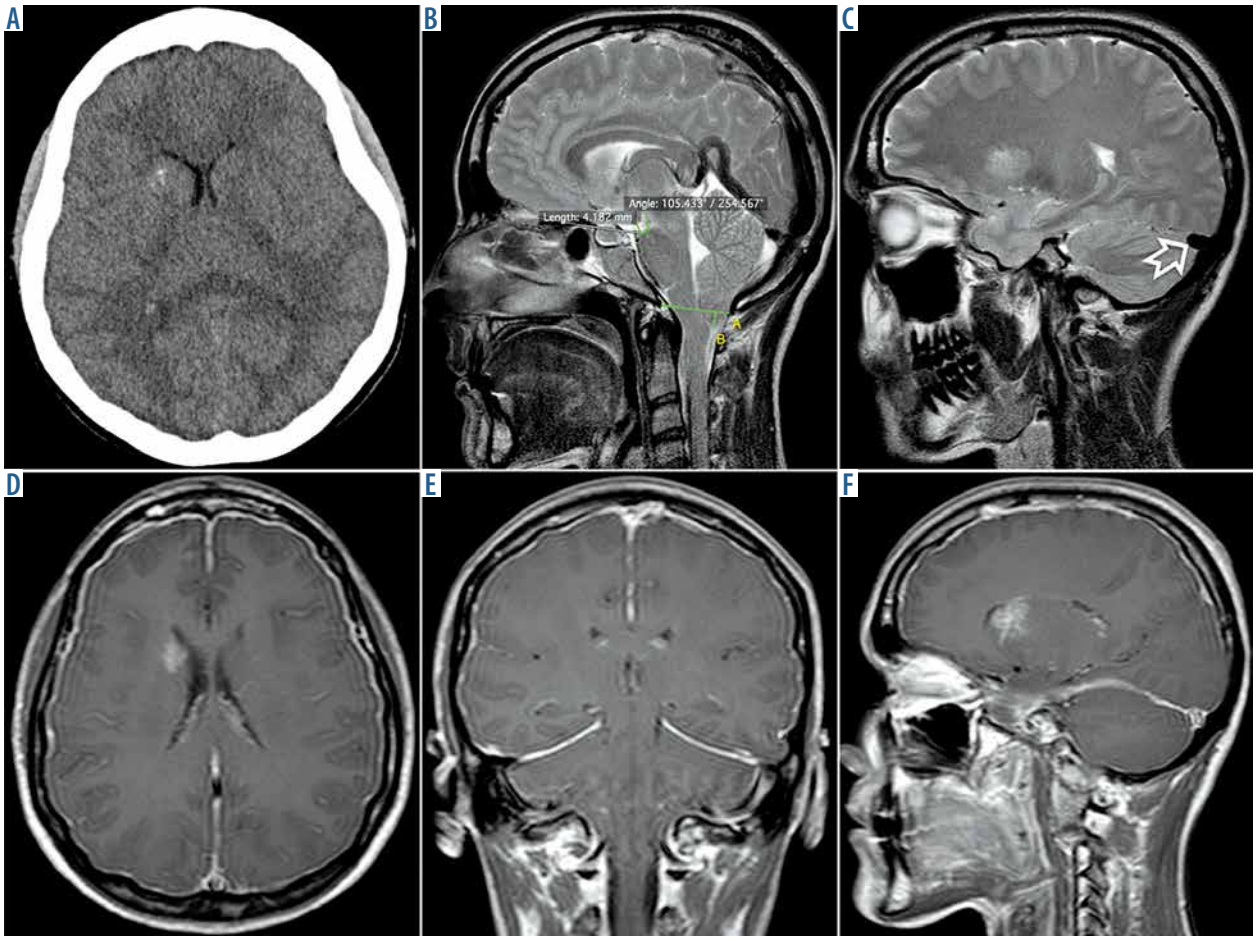

Figure 1. **A**) Normal axial un-enhanced computed tomography brain scan, showing as occasional finding a capillary telangiectasia in right putamen. **B**) Sagittal TSE-T2w magnetic resonance imaging (MRI) shows reduced mamillopontine distance and cerebellar tonsils ectopia, with normal callosal and pontomesencephalic angles; **C**) venous distention sign is positive (white arrow). **D**) Axial, **E**) coronal, and **F**) sagittal SE-T1w MRI after intravenous gadolinium administration shows diffuse pachymeningeal enhancement; right putaminal capillary telangiectasia is clearly visible

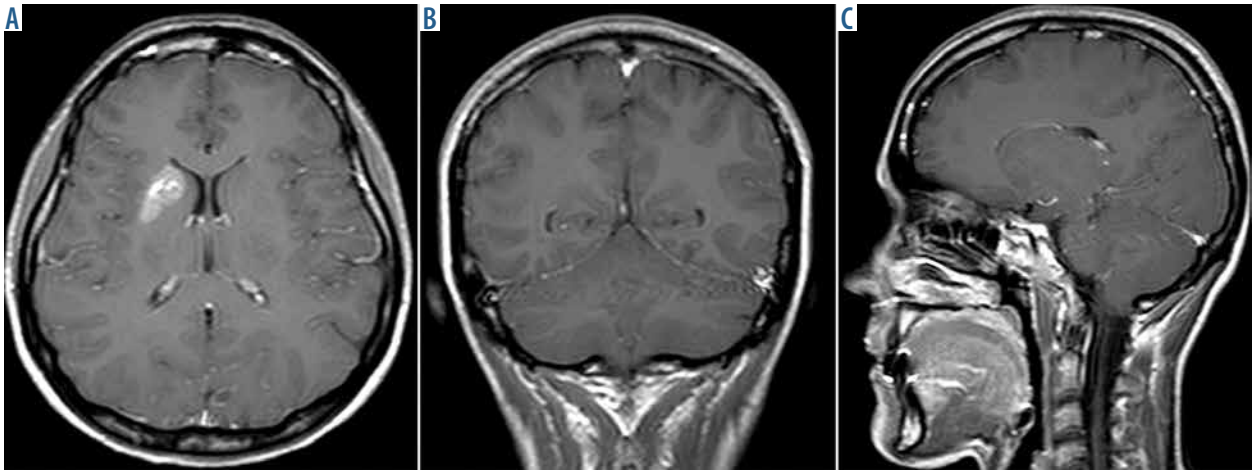

Figure 2. After 2 months, follow-up SE-T1w magnetic resonance imaging on the three orthogonal plans after intravenous gadolinium administration shows almost complete resolution of the pathological diffuse pachymeningeal enhancement

Case 2

A 66-year-old woman with personal history of orthostatic headache associated with dizziness and vertigo came to our attention to perform a contrast-enhanced brain and cervical spine MRI scan, because of the activity-impairing nature of her condition. She was under therapy for essential hypertension and dyslipidaemia. No other comorbidity was noticed at outpatient clinical anamnesis, and neurological examination was normal. A previous cerebellopontine and temporal bone CT scan (performed because of invalidating vertigo attacks) did not bring to light any anomaly. MRI examination showed diffuse and intense pachymeningeal enhancement, involving also spi-

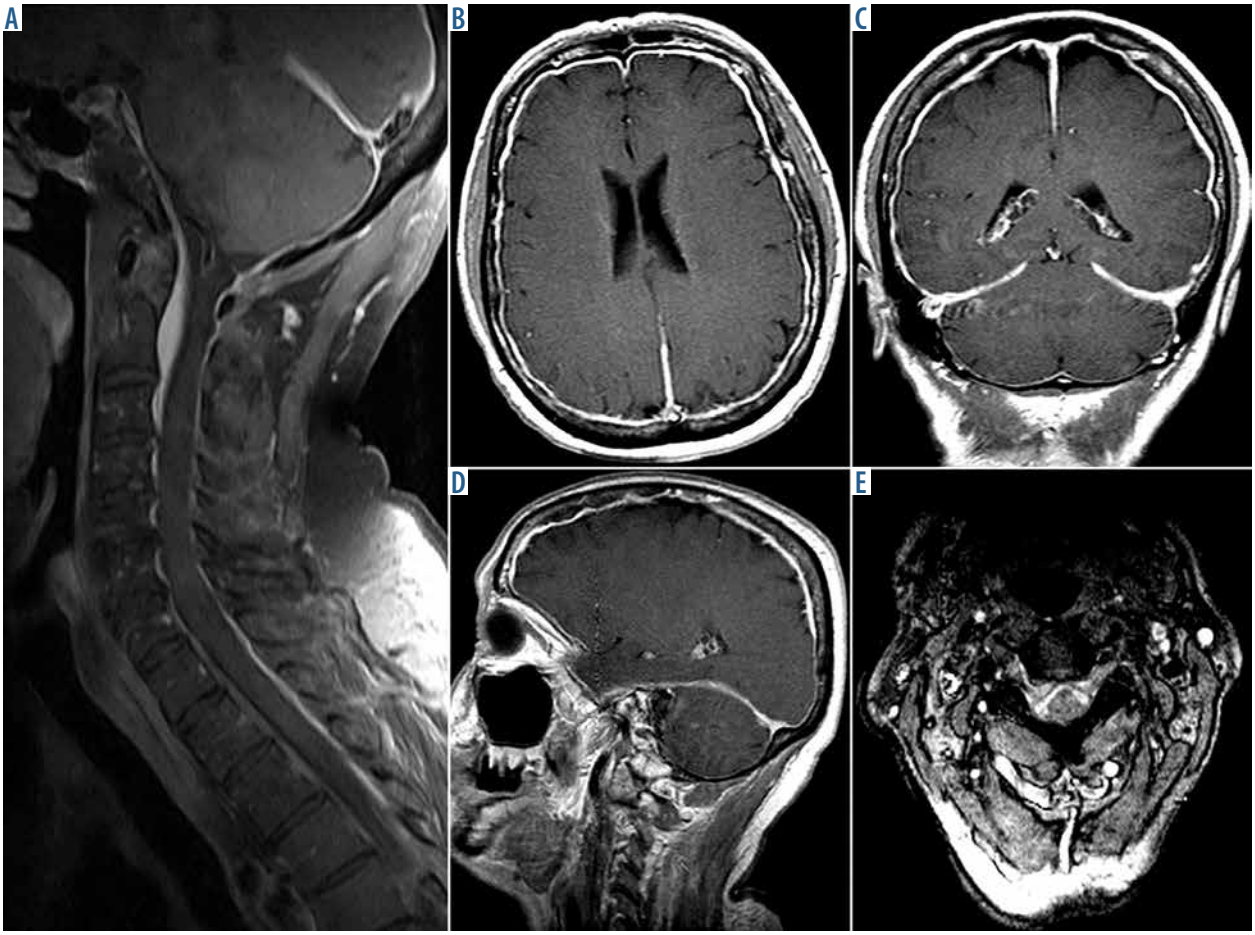

Figure 3. **A**) Cervical spine and **B-D**) brain SE-T1w magnetic resonance imaging after intravenous gadolinium administration shows diffuse and intense pachymeningeal enhancement, involving also spinal dura. **E**) Axial FE T2* shows moderate venous distension and epidural vein engorgement

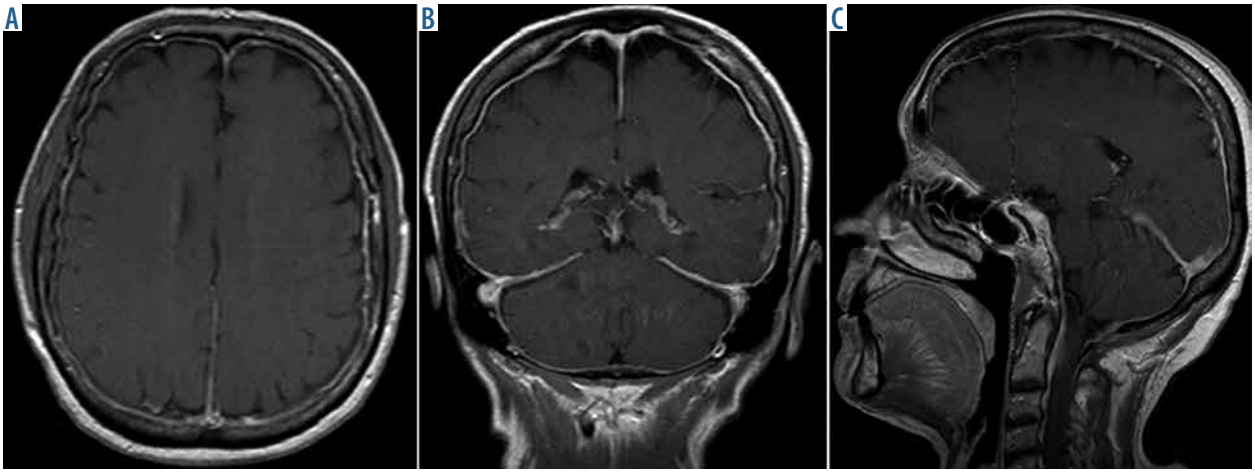

Figure 4. **A-C**) After 3 months, follow-up SE-T1w magnetic resonance imaging on the three orthogonal plans after intravenous gadolinium administration shows a partial reduction of the diffuse pachymeningeal enhancement

nal dura; neither cerebellar tonsils ectopia nor herniation was noticed (Figure 3). A moderate venous distension and epidural vein engorgement were present; callosal angle, mamillopontine distance and pontomesencephalic angle were normal, and subdural effusions were absent. The patient was treated with bed rest and steroid therapy (oral treatment with prednisone, 50 mg/day, with a gradual withdrawal in 30 days) [16], with a progressive and long-term clinical improvement. After three months, follow-up brain and spine MRI showed a persistent dural contrast-enhancement (Figure 4), but it was less pronounced compared to the previous MRI examination. After the above-mentioned conservative therapy, the patient reported long-lasting complete disappearance of the symptoms.

Discussion

We reported two cases of SIH with typical orthostatic symptoms and prompt relief of pain when the patients lay in a supine position, but symptoms recurred immediately when sitting or standing. The clinical picture is globally suggestive, but the diagnosis must be confirmed by contrast-enhanced brain and spine MRI examination. The exact incidence of this pathology is probably under-estimated because the diagnosis can be easily missed, especially when no history of dural puncture or trauma has been reported [10,17-19].

Among various endocranial findings, homogeneous and smooth pachymeningeal enhancement is one of the most common signs at brain MRI scan. Other signs may include increased blood volume in dural and epidural veins, subdural effusion, brain swelling and variation in CSF volume with consequent reduction of callosal angle, mamillopontine distance, and pontomesencephalic angle. Moreover, MRI could be useful to identify complications and possible sites of leak [4,5,10,18-20].

Similarly, spinal MRI can show a wide range of abnormalities, more inconsistent than endocranial findings, such as post-contrast dural enhancement and dilation of dural and epidural veins. In addition, it can lead to the identification of the probable site of leak, displayed as an area of increased signal in paraspinal soft tissue near the nerve roots [14,21-24].

When abnormal features are present, it is important to perform the differential diagnosis from conditions such as infections, vascular lesions, neoplasm, and immune-mediated disorders, distinguish axial from extra-axial enhancement pertinence and from disorders with spinal involvement [25-30].

As soon as the correct diagnosis is performed, an appropriate treatment can be put into practice. The most conservative therapeutic approach with bed rest and steroid administration should always be chosen in the first instance. If ineffective, epidural infusion of saline, epidural injection of fibrin glue, and epidural blood patches are considered as second options. Surgical strategies are generally required in the case of unsuccessful evolution, when clear meningeal defects have been documented. Complete resolution of symptoms after therapy is the final touchstone for the clinician [8-10,13,16].

Conclusions

In conclusion, we described two similar and relatively common cases of SIH, whose clinical and imaging features are typical of this pathology. Orthostatic nature of headache is the most indicative clinical element to suggest the hypothesis of SIH; contrast-enhanced MRI provides qualitative and quantitative findings, leading the radiologist and the clinician to the final correct diagnosis. Conservative treatment coupled to steroid therapy is often sufficient to obtain a complete disappearance of the symptoms.

Conflict of interest

The authors report no conflict of interest.

References

1. Paldino M, Mogilner AY, Tenner MS. Intracranial hypotension syndrome: a comprehensive review. Neurosurg Focus 2003; 15: ECP2.
2. Jacobs MB, Wasserstein PH. Spontaneous intracranial hypotension. An uncommon and underrecognized cause of headache. West J Med 1991; 155: 178-180.
3. Schievink WI, Dodick DW, Mokri B, et al. diagnostic criteria for headache due to spontaneous intracranial hypotension: a perspective. Headache 2011; 51: 1442-1444.
4. Mokri B. Spontaneous CSF leaks: low CSF volume syndromes. Neurol Clin 2014; 32: 397-422.
5. Savoiardo M, Minati L, Farina L, et al. Spontaneous intracranial hypotension with deep brain swelling. Brain 2007; 130 (Pt 7): 1884-1893.
6. Mokri B. The Monro-Kellie hypothesis: applications in CSF volume depletion. Neurology 2001; 56: 1746-1748.
7. Nosik W. Intracranial hypotension secondary to lumbar nerve sleeve tear. J Am Med Assoc 1955; 157: 1110-1111.
8. Goto S, Ohshima T, Yamamoto T, et al. Successful steroid treatment of coma induced by severe spontaneous intracranial hypotension. Nagoya J Med Sci 2016; 78: 229-236.
9. Raskin NH. Lumbar puncture headache: a review. Headache 1990; 30: 197-200.
10. Schievink W, Meyer FB, Atkinson JLD, Mokri B. Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension. J Neurosurg 1996; 84: 598-605.
11. Rando T, Fishman R. Spontaneous intracranial hypotension: report of two cases and review of the literature. Neurology 1992; 42 (3 Pt 1): 481-487.
12. Rabin BM, Roychowdhury S, Meyer JR, et al. Spontaneous intracranial hypotension: spinal MR findings. AJNR Am J Neuroradiol 1998; 19: 1034-1039.
13. Dillon WP, Fishman RA. Some lessons learned about the diagnosis and treatment of spontaneous intracranial hypotension. AJNR Am J Neuroradiol 1998; 19: 1001-1002.
14. Medina JH, Abrams K, Falcone S, Bhatia RG. Spinal imaging findings in spontaneous intracranial hypotension. AJNR Am J Neuroradiol 2010; 195: 459-464.
15. Arnett BC. Tonsillar ectopia and headaches. Neurol Clin 2004; 22: 229-236.
16. Gentile S, Giudice RL, Martino PD, et al. Headache attributed to spontaneous low CSF pressure: report of three cases responsive to corticosteroids. Eur J Neurol 2004; 11: 849-851.
17. Hoffmann J, Goadsby PJ. Update on intracranial hypertension and hypotension. Curr Opin Neurol 2013; 26: 240-247.
18. Mokri B. Spontaneous cerebrospinal fluid leaks: from intracranial hypotension to cerebrospinal fluid hypovolemia – evolution of a concept. Mayo Clin Proc 1999; 74: 1113-1123.
19. Mokri B. Spontaneous low pressure, low CSF volume headaches: spontaneous CSF leaks. Headache 2013; 53: 1034-1053.
20. Spears RC. Low-pressure/spinal fluid leak headache. Curr Pain Headache Rep 2014; 18: 425.
21. Ho JM, Law H, Yuen S, Yam K. Overshunting-associated myelopathy: report of 2 cases. Neurosurg Focus 2016; 41: E16.
22. Howard BM, Sribnick EA, Dhall SS. Over-shunting associated myelopathy. J Clin Neurosci 2014; 21: 2242-2244.
23. Moayeri NN. Spinal dural enhancement on magnetic resonance imaging associated with spontaneous intracranial hypotension. J Neurosurgery 1998; 88: 912-918.
24. Schievink WI, Chu RM, Maya MM, et al. Spinal manifestations of spontaneous intracranial hypotension. J Neurosurg Spine 2013; 18: 96-101.
25. Antony J, Hacking C, Jeffree RL. Pachymeningeal enhancement – a comprehensive review of literature. Neurosurg Rev 2015; 38: 649-659.
26. Yoo M, Lee C, Kim K, et al. A case of intradural-extramedullary form of primary spinal cysticercosis misdiagnosed as an arachnoid cyst. J Korean Neurosurg Soc 2014; 55: 226-229.
27. Elefante A, Caranci F, Del Basso De Carro ML, et al. Paravertebral high cervical chordoma. A case report. Neuroradiol J 2013; 26: 227-232.
28. Jeong SK, Seong HY, Roh SW. Extra-intradural spinal meningioma: a case report. Korean J Spine 2014; 11: 202-204.
29. Maroldi R. Metastatic disease of the brain: extra-axial metastases (skull, dura, leptomeningeal) and tumour spread. Eur Radiol 2005; 15: 617-626.
30. De Divitiis O, Elefante A, De Divitiis E. Historic background of spinal disorders. World Neurosurg 2013; 79: 91-94.