Superficial siderosis of the central nervous system in a patient with asymptomatic sacral paraganglioma as source of chronic bleeding

Ricardo Prat-Acín, Inma Galeano-Senabre, Daniel García-Sánchez, Lluís Morales, Ángel Ayuso-Sacido, Jaime Ferrer-Lozano

Introduction. Superficial siderosis of the central nervous system is an infrequent pathology secondary to chronic bleeding into the cerebrospinal fluid. Spinal tumors are infrequent cause of superficial siderosis being ependymoma the most common etiology.

Case report. We report the case of a woman with sensorineural hearing loss and cerebellar ataxia, diagnosed of superficial siderosis on brain MRI. She had no previous history of axial back pain or radicular leg pain or bowel or bladder incontinence. On spine MRI an intradural lesion was found at the S1 level. No signs of intratumoral hemorrhage were observed on MRI gradient-echo images. At surgery, an intradural soft mass with signs of chronic bleeding was completely resected. Based on microscopic examination and immunohistochemistry of the specimen, a diagnosis of paraganglioma World Health Organization grade I was made.

Conclusions. Since the only proven treatment able to prevent further deterioration from superficial siderosis is to stop chronic bleeding into subarachnoid space, is of paramount importance to establish an early diagnosis of the source of bleeding. Cases of unexplained superficial siderosis of central nervous system should include routine spinal MRI to rule out bleeding of spinal tumor even in asymptomatic patients. Due to severity of potential deterioration caused by superficial siderosis, any tumoral lesion observed on spinal MRI even without documented sings of bleeding should be considered for resection.

Key words. Central nervous system. Chronic bleeding. Hemosiderin. Magnetic resonance imaging. Paraganglioma. Superficial siderosis.

Brain Tumour Laboratory; Fundación Vithas; Madrid (A. Ayuso-Sacido). Department of Neurosurgery (R. Prat-Acín, I. Galeano-Senabre, D. García-Sánchez); Department of Neurology (LL. Morales); Department of Pathology (J. Ferrer-Lozano); Hospital Universitario y Politécnico La Fe; Valencia, Spain.

Corresponding author:
Dr. Ricardo Prat Acín. Department of Neurosurgery. Hospital Universitario y Politécnico La Fe. Avda. Fernando Abril Martorell, 106. E-46026 Valencia (Spain).

E-mail:
ricprat@hotmail.com

Accepted:
03.07.20.

How to cite this paper:
Prat-Acín R, Galeano-Senabre I, García-Sánchez D, Morales LL, Ayuso-Sacido A, Ferrer-Lozano J. Superficial siderosis of the central nervous system in a patient with asymptomatic sacral paraganglioma as source of chronic bleeding. Rev Neurol 2020; 71: 253-6. doi: 10.33588/rn.7107.2020215.

Versión española disponible en www.neurologia.com

Introduction

Superficial siderosis of the central nervous system (CNS) is an infrequent pathology secondary to chronic bleeding into the cerebrospinal fluid (CSF) and deposition of hemosiderin on the subpial and ependymal surfaces of the CNS [1]

Radiologically is characterized on brain and spine MRI by a layer of T_2 hypo-intensity surrounding the subpial and ependymal surfaces of the CNS Clinically it usually associates triad of progressive sensorineural hearing loss, cerebellar ataxia and myelopathy.

Frequent sources of chronic bleeding into the CSF include CNS tumors, prior neurosurgery, CNS trauma and vascular origin, mainly secondary either to arteriovenous malformations or brain aneurysms. Spinal tumors are infrequent cause of superficial siderosis being ependymoma the most common etiology. In this report we describe a unique case of superficial siderosis due to spinal paraganglioma as a source of unnoticed bleeding in the subarachnoid space.

Case report

A 53-year-old female patient presented in the outpatient clinic with a 2-year history of hearing loss and one year history of gait difficulty. According to the patient's family the patient presented also hoarse voice and slurred speech of 1 year's duration

On examination no cognitive deficits were observed. Motor examination revealed no spasticity, hyper-reflexia or Babinski sign. The patient had no previous history of axial back pain or radicular leg pain or bowel or bladder incontinence. No signs of dysmetria were found. Dysdiadokokinesia finger-nose and gait ataxia were present. Auditory examination demonstrated bilateral neurosensorial hearing loss.

MRI T_2-weighted images revealed hypointense superficial signal coating the surface of cerebral hemispheres, cerebellar hemispheres, brainstem and cisternal space. Gradient-echo images showed a pronounced hypointensity in the same location. Based on these findings on brain MRI the patient was diagnosed of superficial siderosis (Fig. 1). Mul-

Figure 1. Gradient-echo brain MRI images demonstrating a pronounced hypointensity in the surface of cerebral hemispheres, cerebellar hemispheres, brainstem and cisternal space consistent with hemosiderin deposition.

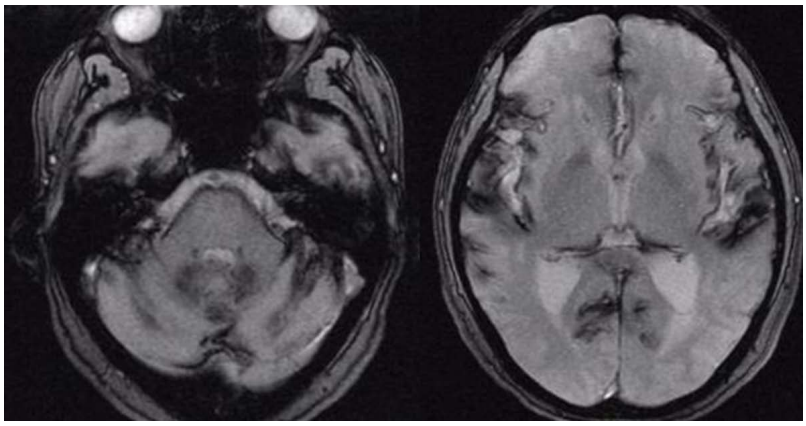
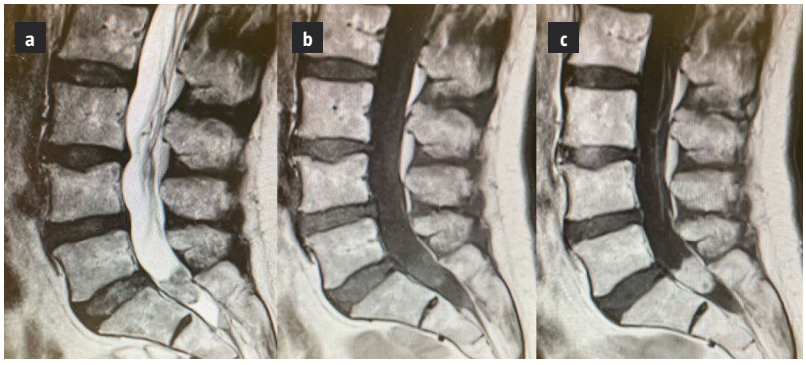


Figure 2. a) Sagital STIR sequence MRI showing intradural tumor at the S1 level; b) Sagital T_1 sequence MRI showing isointense intradural tumor at the S1 level; c) Sagital T_1 weighted imaging post-gadolinium showing homogeneous enhancement of the lesion.



tiple cerebral periventricular areas of demyelination were found but no areas of acute ischemia were observed on diffusion images. Further evaluation with angio-MRI failed to reveal any vascular cause for superficial siderosis. For this reason spine MRI was performed and showed a 21 × 15 mm intradural tumor at the S1 level. T_1 weighted imaging post-gadolinium showed homogeneous enhancement of the lesion and of lumbosacral roots (Fig. 2). No signs of intratumoral hemorrhage were observed on gradient-echo images. Somatosensory evoked potentials of upper and lower extremities were performed prior to surgery and considered as normal. Considering the likehood of intradural tumor to be the source of chronic bleeding into the CSF and after obtaining the informed consent from the patient, the decision was made to resect the tumor.

At surgery, a soft mass surrounding multiple sacral roots was completely resected after microsurgical dissection from nerve roots and rootlets. No dural attachment was observed. Intraoperative electromyography was utilized to identify nerve roots. On the caudal aspect of the lesion signs of chronic bleeding were observed intraoperatively. The patient had a quick recovery and left the hospital four days after operation.

Microscopic examination disclosed epithelioid cells arranged in solid and trabecular patterns, associated with a delicate stroma with thin-walled vessels. Fibrotic bands and areas of fresh and old hemorrhage were seen throughout the tumor, and were also present in the surrounding, thickened meningeal tissue. In those areas, hemosiderin deposition was observed. Tumor cells showed central and vesicular nucleus, with finely speckled chromatin, and wide, eosinophilic, granular cytoplasm, with focal clear change. Mitoses were rarely seen (less than one per 10 high-power fields). Cells with bigger, irregular, smudged nucleus of degenerative appearance were also observed. Immunohistochemistry revealed positive labelling of tumor cells with synaptophysin, chromogranin and cytokeratin (AE1/AE3), whereas no reactivity was seen against GFAP, EMA, and CD34. Sustentacular cells expressed S100 positivity. Proliferation index, as measured with Ki67 immunostain, was less than 1%. Based on these findings, a diagnosis of paraganglioma World Health Organization grade I was made (Fig. 3).

Discussion

Superficial siderosis of the CNS is a rare condition, being even less common to be observed as a result of spine tumor bleeding tumor. Less than 20 cases of superficial siderosis due to chronic bleeding of a spinal tumor have been reported in the literature, and only 5 cases were histopathologically diagnosed as paraganglioma [2-6].

Although asymptomatic cases of superficial siderosis have been diagnosed only based on MRI [7], superficial siderosis may be a severe clinical neurological condition. Superficial siderosis of the CNS has been associated with cognitive impairment, including dementia, in up to 24% of patients with this diagnosis. Superficial siderosis of the CNS usually associates triad of progressive sensorineural hearing loss, cerebellar ataxia and myelopathy. Since

the only proven treatment able to prevent further deterioration from superficial siderosis is to stop chronic bleeding into subarachnoid space, is of paramount importance to establish an early diagnosis of the source of bleeding.

At present case, our patient presented with progressive sensorineural hearing loss and cerebellar ataxia, but no signs or symptoms of myelopathy were found. The progressive nature of the hearing loss and the different moment of presentation of symptoms (cerebellar ataxia presented one year later) could explain the delay in performing a brain MRI that could have established the diagnosis.

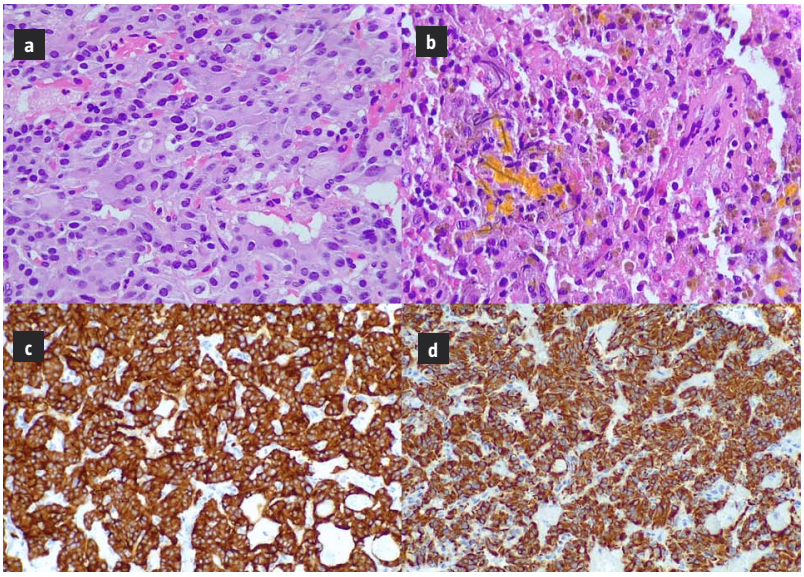
Paragangliomas are neuroendocrine tumors associated with autonomic structures most commonly (90% of cases) located within the adrenal gland where they are called pheochromocytomas. Only 10% of extra adrenal paragangliomas involve the spine, usually the filum terminale and cauda equina and the most common presenting symptoms are lower back pain and radicular leg pain.

In the present case neither superficial siderosis nor sacral paraganglioma were initially considered as the main hypothesis of diagnosis according to clinics. Although presenting with progressive sensorineural hearing loss and cerebellar ataxia, our patient had no cognitive impairment signs, interestingly these cognitive findings were observed in three out of four cases previously published. On the other hand, our patient did not present axial low back or radicular leg pain, or bowel or bladder incontinence, symptoms observed in three cases in the consulted literature. The present case is the first case combining the absence of these symptoms and also explains the delay and difficulty in establish a correct diagnosis. The absence of radicular leg symptoms in particular, was initially of great importance in the decision made not to consider performing a spine MRI.

In our case no clear signs of intratumoral bleeding were observed on spine MRI. In the consulted literature, bleeding of the tumor was only observed in two cases on spine MRI, whereas histopathological confirmation of intratumoral bleeding was reported in three cases. In the present case a major concern when the decision was made to resect the tumor, was the impossibility to assure the patient that the surgery would control the source of bleeding that had caused superficial siderosis [8].

As a conclusion, superficial siderosis due to chronic bleeding into the cerebrospinal space of sacral paraganglioma is a rare condition. Cases of unexplained superficial siderosis of CNS should include routine spinal MRI to rule out bleeding of spinal tumor even in asymptomatic patients. Due to severity of potential deterioration caused by superficial siderosis, any tumoral lesion observed on spinal MRI even without documented sings of bleeding should be considered for resection.

Figure 3. Histopathological analysis of the intradural tumor. a) Microscopic examination disclosed epithelioid cells arranged in solid and trabecular patterns (HE, ×200); b) Intratumoral deposition of hemosiderin (orange pigment; ×200); c) Synaptophysin strongly stained cells (×100); d) Positive labelling of tumor cells with cytokeratin (×100).



References

1. Dubessy AL, Ursu R, Maillet D, Augier A, Le Guilloux J, Carpentier AF, et al. Superficial siderosis of the central nervous system: a rare cause of dementia with therapeutic consequences. Age Ageing 2012; 41: 275-7.
2. Dillard-Cannon E, Atsina KB, Ghobrial G, Gnass E, Curtis MT, Heller J. Lumbar paraganglioma. J Clin Neurosci 2016; 30: 149-51.
3. Khan M, Lach B, Al Jishi A, Murti NK, Superficial siderosis of the central nervous system secondary to chronic bleeding from a lumbar paraganglioma. JSM Neurosurgery & Spine 2014; 2: 1046.
4. Kinge NG, Paliwal VK, Neyaz Z, Verma R. Superficial siderosis in a patient with filum terminale paraganglioma. Neurol India 2012; 60: 648-9.
5. London F, Mulquin N, Fervaille C, Lebecque O, Jankovski A. Radiologically atypical paraganglioma of the filum terminale as a rare cause of superficial siderosis of the central nervous system. Acta Neurol Belg 2020; Mar 10. [Epub ahead of print].
6. Sharma A, Gaikwad SB, Goyal M. Calcified filum terminale paraganglioma causing superficial siderosis. AJR Am J Roentgenol 1998; 170: 1650-2.
7. Fearnley JM, Stevens JM, Rudge P. Superficial siderosis of the central nervous system. Brain 1995; 118: 1051-66.
8. Mamourian AC. MR of superficial siderosis. AJNR Am J Neuroradiol 1993; 14: 1445-8.

Siderosis superficial del sistema nervioso central en una paciente con paraganglioma sacro asintomático como fuente de sangrado crónico

Introducción. La siderosis superficial del sistema nervioso central es una patología poco frecuente secundaria al sangrado crónico en el líquido cefalorraquídeo. Los tumores medulares son causa poco habitual de siderosis superficial, y el ependimoma es la etiología más común.

Caso clínico. Mujer con pérdida auditiva neurosensorial y ataxia cerebelosa, diagnosticada de siderosis superficial en la resonancia magnética cerebral. No tenía antecedentes de dolor raquídeo axial, dolor radicular ni incontinencia esfinteriana. En la resonancia magnética de la columna se encontró una lesión intradural en S1. No se observaron signos de hemorragia intratumoral en las secuencias de resonancia magnética en eco de gradiente. En la cirugía, se apreció una masa blanda intradural con signos de sangrado crónico que se resecó. Basado en el examen microscópico e inmunohistoquímico de la muestra, se alcanzó el diagnóstico de paraganglioma de grado I de la Organización Mundial de la Salud.

Conclusiones. Dado que el único tratamiento probado capaz de prevenir un mayor deterioro por la siderosis superficial es detener el sangrado crónico en el espacio subaracnoideo, es importante establecer un diagnóstico temprano de la fuente de sangrado. Los casos no justificados de siderosis superficial del sistema nervioso central deben incluir una resonancia magnética de la columna rutinaria para descartar el sangrado de un tumor medular, incluso en pacientes asintomáticos. Debido a la gravedad del deterioro potencial causado por la siderosis superficial, cualquier lesión tumoral observada en una resonancia magnética del raquis, incluso sin presentar signos de sangrado, debería ser objeto de indicación quirúrgica.

Palabras clave. Hemosiderina. Paraganglioma. Resonancia magnética. Sangrado crónico. Siderosis superficial. Sistema nervioso central.