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LETTER TO THE EDITOR

Radiologically atypical paraganglioma of the filum terminale as a rare cause of superficial siderosis of the central nervous system

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Introduction

Superficial siderosis (SS) is an uncommon but potentially devastating disorder characterized by linear deposits of hemosiderin within the leptomeninges and subpial layers of the brain, infratentorial regions, and spinal cord [1]. The most common underlying causes are dural pathologies (such as traumatic injury or neurosurgical procedures), tumors, and vascular malformations, but the source of bleeding remains frequently undetermined despite extensive imaging [1]. Spinal tumor is, however, a rare cause of SS [1]. Here, we present a case of diffuse SS of the central nervous system caused by a paraganglioma developing in the filum terminale.

Case presentation

A 62-year-old man was assessed in our hospital because of a 3-year history of gait difficulties. He also reported profound bilateral sensorineural hearing impairment over a span of 6 years and complained of lower back pain without radiculopathy for several years. He denied any heavy traumatism, intradural surgery, or severe headache. Neurological examination showed a wide-based gait with marked difficulty on tandem gait. There were no corticospinal tract signs or urinary symptoms. Cognitive functions were normal.

The patient underwent brain magnetic resonance imaging (MRI) with a time gradient echo sequence which showed a diffuse linear hypointensity indicating extensive deposition of hemosiderin along the pial surface of cerebellum and brainstem, in cerebral convexities and sylvian and interhemispheric fissures, consistent with SS (Fig. 1a, b). A full spine MRI was carried out for further evaluation and was remarkable for diffuse T2-weighted hypointensity of the pial surface of the cord (Fig. 1c, d). It also revealed the presence of an intradural extramedullary tumor located at the L5-S1 level, arising from the filum terminale. This lesion was hypointense on T2-weighted images (T2WI) and isointense on T1-weighted images (T1WI), and showed heterogeneous mild enhancement after gadolinium injection; a draining vein was identified at the cranial pole (Fig. 1e, f). Although the nature of the tumor was not identified on MRI, we assumed that SS was caused by the intrathecal tumor. Surgery was performed. Durotomy revealed a soft, well-encapsulated mass, engulfing the filum terminale and non-dissectible from it. A large draining vein was identified at its cranial pole, consistent with the preoperative MRI observation. The feeding artery was not identified on MRI and was not visible intra-operatively due to the presence of arachnoiditis at the caudal pole. Areas of petechial hemorrhages were observed at the surface of the tumor. The highly vascularized tumor was carefully dissected to preserve the surrounding rootlets and avoid capsular rupture, which would have caused bleeding of the tumor body. Then, vascular clipping and sectioning of filum terminale at the cranial and caudal poles of the tumor allowed its complete removal. Histopathological examination (Fig. 2) demonstrated a

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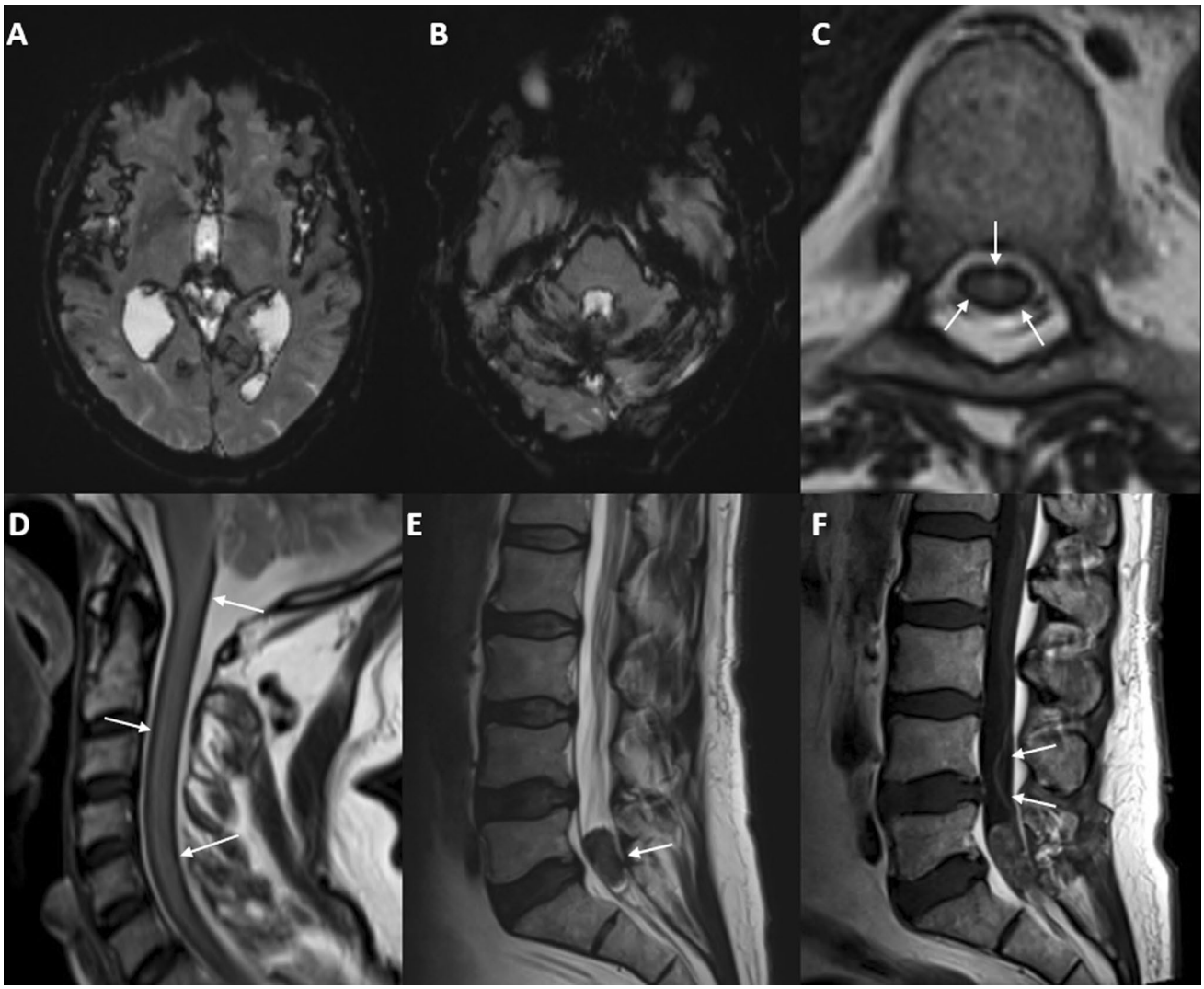

Fig. 1 Brain and full spine MRI. **a**, **b** Axial gradient echo T2*-weighted images showing a rim of hypointensity all around the cerebellum, brainstem, and interhemispheric and sylvian fissures. **c**, **d** Axial (**c**) and sagittal (**d**) T2-weighted images revealing hypointense coat along entire cord surface (arrows). **e**, **f** Sagittal T2-weighted images (**e**) and post-contrast T1-weighted images (**f**) showing a well-circumscribed intradural mass at the L5-S1 level with a draining vein at its cranial pole (arrows)

characteristic neuroendocrine architecture consisting of a population of tumor cells, with regular nuclei and stippled chromatin pattern, arranged in clusters, and enveloped within a prominent vascular network. A few hemosiderin-laden macrophages were present, providing the evidence of prior hemorrhage. There was no evidence of mitotic activity or necrosis. Tumor cells were negative for epithelial membrane antigen and GFAP. Immunopositivity for neuroendocrine markers—chromogranin A and synaptophysin—and positive reaction for S-100 protein supported the histological diagnosis of paraganglioma that was considered to have probably caused SS. At the last follow-up (5 months after surgery), the neurological symptoms were stabilized and lower back pain was significantly improved.

Discussion

Superficial siderosis is a rare condition resulting from a prolonged or recurrent low-grade bleeding into the subarachnoid space, leading to linear deposits of hemosiderin within the leptomeninges and subpial layers of the brain, infratentorial regions, and spinal cord [1]. Slowly progressive ataxia and sensorineural hearing loss are the commonest neurological manifestations and relate to a selective vulnerability of the cerebellum and vestibulocochlear nerves [1, 2]. Other clinical manifestations frequently observed in patients with SS are pyramidal signs, bladder disturbances, and cerebellar dysarthria. Cognitive decline may be present, but is less commonly noted. Early

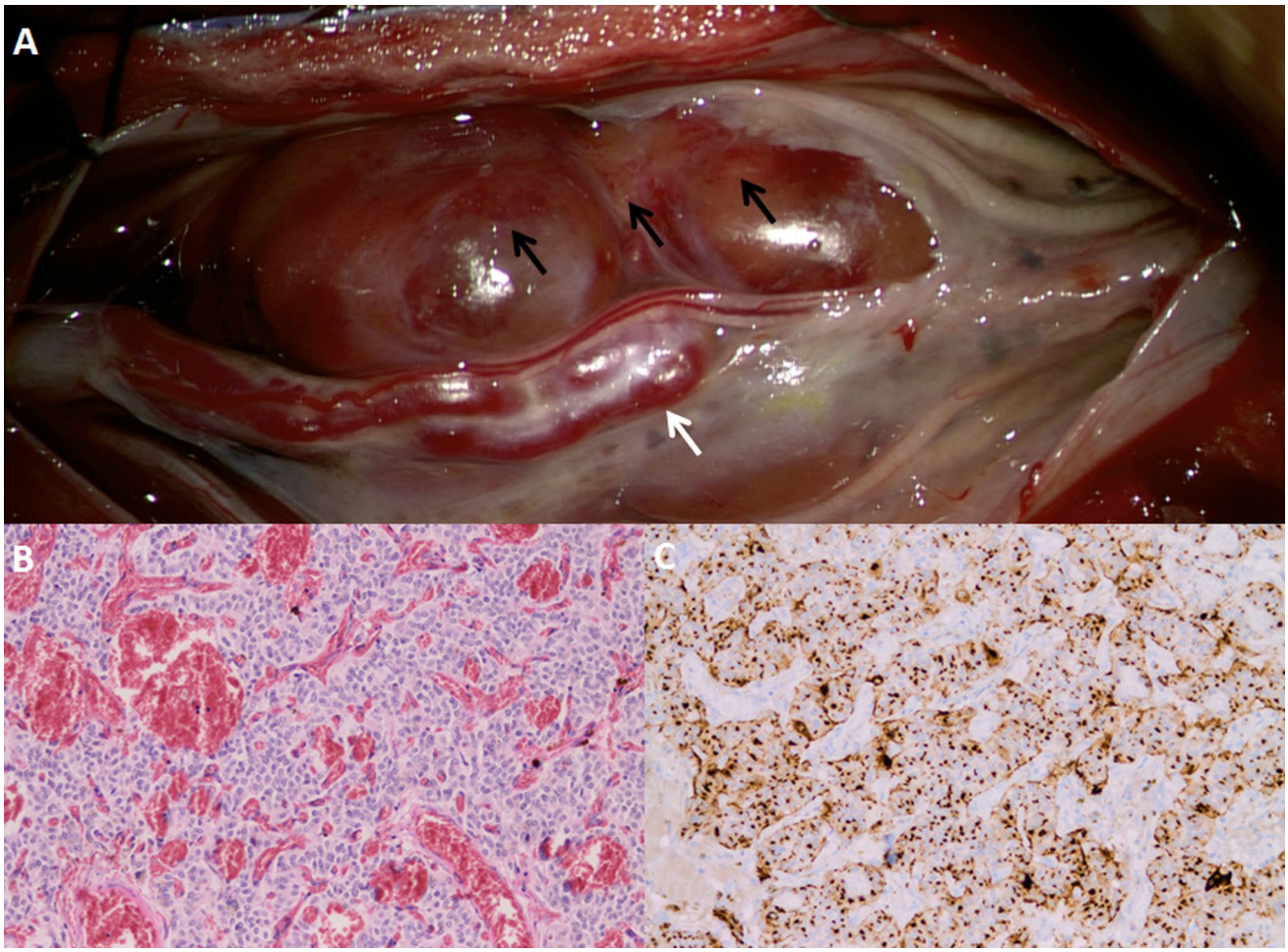

Fig. 2 Macroscopic and microscopic appearance of the tumor. **a** Intraoperative photographs of the intradural tumor arising from the filum terminale. A feeding vessel can be observed at the caudal pole of the tumor (white arrow). Small petechial hemorrhages were observed at the surface of the tumor (black arrows). **b** Haematoxylin and eosin stain (original magnification ×20) showing growth of tumor cells within highly vascularized network. **c** Cells showed strong and diffuse cytoplasmic reactivity with chromogranin A (×20), which is typical of paraganglioma. In panel **b**, the nested architecture is more subtle, but is conspicuous in panel **c**

diagnosis is likely to be missed due to the slow progression of these symptoms, and early identification of the origin of bleeding is crucial to halt the symptom progression and avoid significant disability.

Spinal paraganglioma (SP) is a rare, benign, and slow-growing neoplasm that manifests with non-specific chronic back pain and radiculopathy. This tumor has been reported as a cause of SS only in very rare cases [1, 3–5]. In our patient, a definitive radiological diagnosis of SP before surgery was not possible given that there was no pathognomonic radiological sign. Due to its rich vascular nature, this tumor has particularly high propensities for spontaneous hemorrhage, so that identification of some radiological features reflecting hypervascularity can help to improve preoperative diagnosis accuracy. The observation of intra- and peritumoral flow-voids and of a characteristic salt and pepper-like appearance on T2WI may be indicative of hypervascularity and, therefore, raise the possibility of SP [5]. Another radiological feature which may be helpful in differentiating paraganglioma from other hemorrhagic filum terminale lesions is the 'polar sign' which represents hypointense signals on contrast T1WI and hyper intensities on T2WI within the lesion's superior and inferior poles [5]. However, we did not observe any of these radiological features in our patient. Apart from SS, the observation of a drainage vessel was the sole imaging finding suggesting a vascular tumor, and MRI did not demonstrate intense gadolinium contrast enhancement as frequently observed in paraganglioma [5].

In summary, a full spine MRI should be performed in case of unexplained SS. In cases of SS caused by a spinal tumor, one should consider the possibility of paraganglioma and take every precaution to prevent avoidable bleeding during surgery.

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Compliance with ethical standards

Conflict of interest The authors declare no conflict of interest regarding this case report.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed consent For this type of study, formal consent is not required.

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