

with IVIg (Patient 6). In our patient, similar to others in the axonal group, steroid therapy dramatically improved the neurological symptoms, although IVIg did not adequately improve symptoms.

To the best of our knowledge, this is the first report of a sarcoidosis patient positive for anti-ganglioside antibodies. An IgG anti-GalNAc-GD1a antibody-positive illness is most commonly GBS (89%).⁷ GBS with IgG anti-GalNAc-GD1a antibodies is most commonly seen in young men and is associated with antecedent diarrhea and distal-dominant limb weakness (pure motor type).⁷ Our patient is distinct from these other patients with GBS with IgG anti-GalNAc-GD1a antibodies. Therefore, we consider that our patient does not have sarcoidosis coincident with GBS, but rather neurosarcoidosis with IgG anti-GalNAc-GD1a antibodies. Patients with axonal neuropathy with GBS-like onset neurosarcoidosis may be positive for IgG anti-GalNAc-GD1a antibodies.

In conclusion, we report a patient with acute-onset neurosarcoidosis who was positive for IgG anti-GalNAc-GD1a antibodies. Based on a review of reports, patients with GBS-like onset neurosarcoidosis can be divided into an axonal group and a demyelinat-

ing group, with patients in the axonal group exhibiting pleocytosis in the CSF and a good response to steroids.

References

1. Delaney P. Neurologic manifestations in sarcoidosis: review of the literature, with a report of 23 cases. *Ann Intern Med* 1977;**87**:336–45.
2. Burns TM, Dyck PJ, Aksamit AJ, et al. The natural history and long-term outcome of 57 limb sarcoidosis neuropathy cases. *J Neurol Sci* 2006;**244**:77–87.
3. Zajicek JP, Scolding NJ, Foster O, et al. Central nervous system sarcoidosis—diagnosis and management. *QJM* 1999;**92**:103–17.
4. Fahoum F, Drory VE, Issakov J, et al. Neurosarcoidosis presenting as Guillain-Barré-like syndrome. A case report and review of the literature. *J Clin Neuromuscul* 2009;**11**:35–43.
5. Saifee TA, Reilly MM, Ako E, et al. Sarcoidosis presenting as acute inflammatory demyelinating polyradiculoneuropathy. *Muscle Nerve* 2011;**43**:296–8.
6. Findik S, Bulbul R, Ozbenli T, et al. Sarcoidosis and Gullain-Barré syndrome. *Acta Neurol Belg* 2011;**111**:72–5.
7. Tatsumoto M, Odaka M, Koga M, et al. Clinical features of neuropathies in a group of patients associated with anti-GalNAc-GD1a antibody. *Clin Neurol* 2004;**44**:508–12.

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Agraphia caused by an infarction in Exner's area



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ABSTRACT

Sigmund Exner postulated in 1881 that lesions of the base of the medial frontal gyrus could specifically produce writing impairments and attributed the writing centre to this area. We report two patients who suffered from strokes in this area. These patients suffered from writing disturbances comprising both omitted words within a sentence or badly written words, as well as aphasia. These patients, in line with prior reports, illustrate the crucial role of the Exner area at the base of the medial frontal gyrus for the cerebral writing network; we suggest that this region plays an important role for phoneme–grapheme conversions.

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1. Case report

1.1. Patient 1

A 46-year-old right-handed man presented at our stroke unit with an acute writing deficit. He could not combine syllables to form real words and exchanged or even omitted entire words when writing. However, his speech was fluent and he had no speech comprehension deficiencies. A neurological examination detected no other deficits. His medical history included a small left thalamic ischaemic stroke with no residua in 2009 and polytrauma after a motorbike accident in 2007, with multiple organ injuries. A cerebral MRI detected a left cortical ischaemic lesion in Exner's area, which is located within the middle frontal gyrus (Fig. 1). Very small embolic infarcts were also observed in the mouth motor cortex and in the adjacent centrum semiovale. His writing ability was completely restored after 3 days of hospitalization with speech therapy.

1.2. Patient 2

A 61-year-old right-handed woman presented to our stroke unit with acute global aphasia and right hemiparesis. She also had an acute writing deficit. Her past medical history was unremarkable. Her only known stroke risk factor was smoking. The hemiparesis and aphasia improved shortly after admission and completely resolved after 1 day, although her inability to write remained. The initial CT scan and subsequent MRI detected an infarct in Exner's area (Fig. 2). A detailed neurological examination detected no deficits, except for the inability to write. The patient could not write dictated texts, but she was able to copy texts almost faultlessly (Fig. 3). She was capable of writing numbers and performing calculations. Her main dysfunction was the translation of spoken words into written words, that is, phonological agraphia. She exchanged syllables or failed to produce the correct syllables, exchanged letters, omitted or added letters, or could not write a word at all. These symptoms are described as phoneme–grapheme conversion disorder (Fig. 3). Her speech was normal and she did not suffer from aphasia or any type of word-finding disorder. She was discharged to a neurological rehabilitation programme after 3 days of hospitalization with speech therapy.

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Fig. 1. Coronal diffusion-weighted MRI showing a cortical infarct in Exner's area in Patient 1 (arrow).

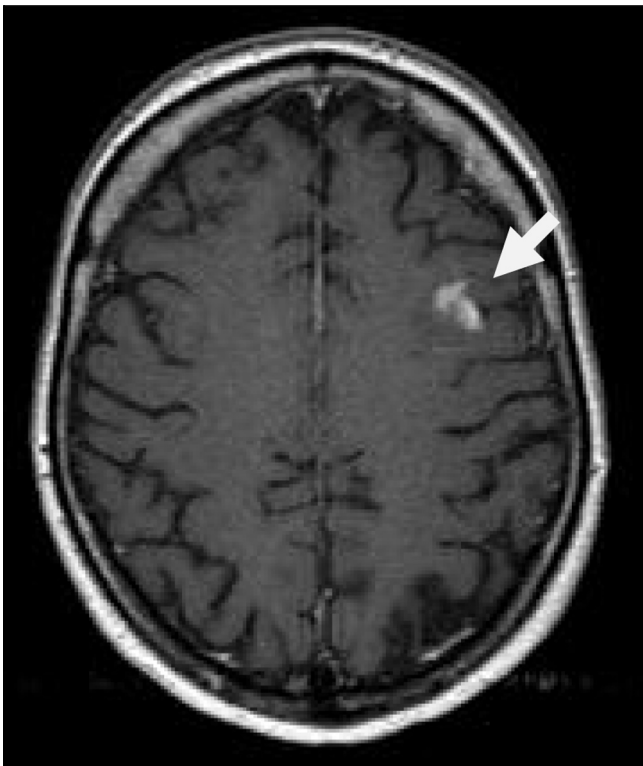


Fig. 2. Axial fluid attenuated inversion recovery MRI showing an ischemic lesion in Exner's area in Patient 2 (arrow).

2. Discussion

In 1881, Sigmund Exner postulated that specific lesions in the base of the middle frontal gyrus could produce writing impairments and attributed writing control to this area.¹ Currently, it is assumed that there are lexical and phonological writing systems and that these two systems are most likely located in different brain regions.^{2,3} A previous report⁴ described Japanese patients

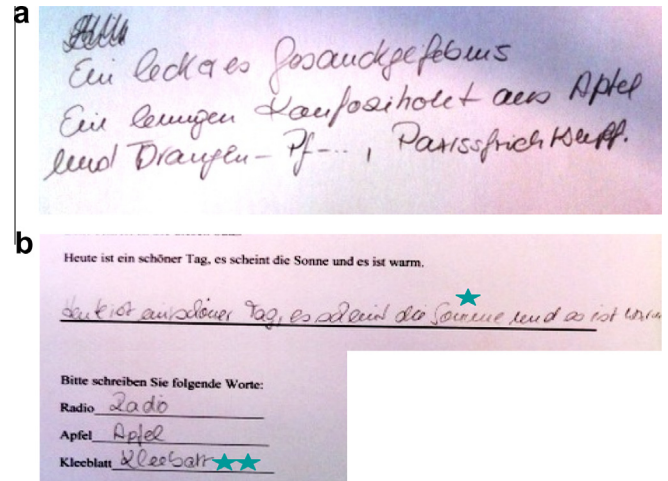


Fig. 3. (a) Photograph showing dictated sentences as written by Patient 2. Mistakes are in bold. Ein leckeres **Geschmackerlebnis**. Eine **gelungene Komposition** aus Apfel- und Orangen-, **Trauben-**, **Passionsfruchtsaft**. (b) Photograph showing words copied from another printed source by Patient 2. *One "n" too many: "Sonnne" instead of "Sonne". **One "l" missing: "Kleebatt" instead of "Kleeblatt".

with frontal lobe damage who could write kanji (ideographic characters, representing pictures, whole words, or meanings) but who were unable to write kana (sublexical characters, which represent vowels or combinations of consonants and vowels, such as ka, ke, ki, ko and ku). Patients with lesions in the middle and inferior frontal gyri are known to suffer from writing disturbances characterized by the omission of words in a sentence or badly written words,⁵ which suggests that these frontal areas may be critical for the lexical and phonological processes engaged during writing. This hypothesis is consistent with the clinical features of the patients in the present study. Patient 2 had a severe phoneme-grapheme disorder and she was unable to find the correct syllables, or even skipped whole words, because she did not know how to write them down. However, she could copy written sentences very well. Interestingly, it was much easier for her to copy written texts than to convert an aural input (dictated text) into written text. This suggests that different phonological and lexical conversion systems are responsible for writing, and they depend on the input modality.

3. Conclusions

Overall, these patients and previous reports support the hypothesis that Exner's area, located at the base of the middle frontal gyrus, is crucial in the cerebral writing network. We suggest that this region has an important role in phoneme-grapheme conversion. However, future studies are needed to disentangle the functional subnetworks in the brain that are involved in writing.

References

- Exner S. *Untersuchungen über die Lokalisation der Funktionen in der Großhirnrinde des Menschen*. Vienna: Wilhelm Braumüller; 1881.
- Lubrano V, Roux FE, Demonet JF. Writing-specific sites in frontal areas: a cortical stimulation study. *J Neurosurg* 2004;**101**:787–98.
- Roux FE, Dufor O, Giussani C, et al. The graphemic/motor frontal area Exner's area revisited. *Ann Neurol* 2009;**66**:537–45.
- Thogi H, Saitoh K, Takahashi S, et al. Agraphia and acalculia after a left prefrontal (F1, F2) infarction. *J Neurol Neurosurg Psychiatry* 1995;**58**:629–32.
- Shallice T. Phonological agraphia and the lexical route in writing. *Brain* 1981;**104**:413–29.