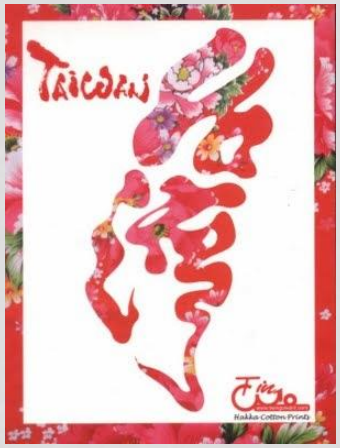

Jeff Hung-Wen Kao

Sandy Cheng-Yu Chen

臺北醫學大學

TAIPEI MEDICAL UNIVERSITY

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Taiwan
NATIONAL DEFENSE MEDICAL CENTER
1902
中華民國 R.O.C.
Google

69 y.o., Female

Dept. of Diagnostic Radiology,
Yamagata University, JAPAN

Masafumi KANOTO

69 y.o., Female

Chief Complaint:

Bilateral legs motor weakness.

Past History:

Right breast cancer, treated (more than 10 years ago).

History of Present Illness:

She had gradual bilateral legs motor weakness and consulted a doctor in our hospital.

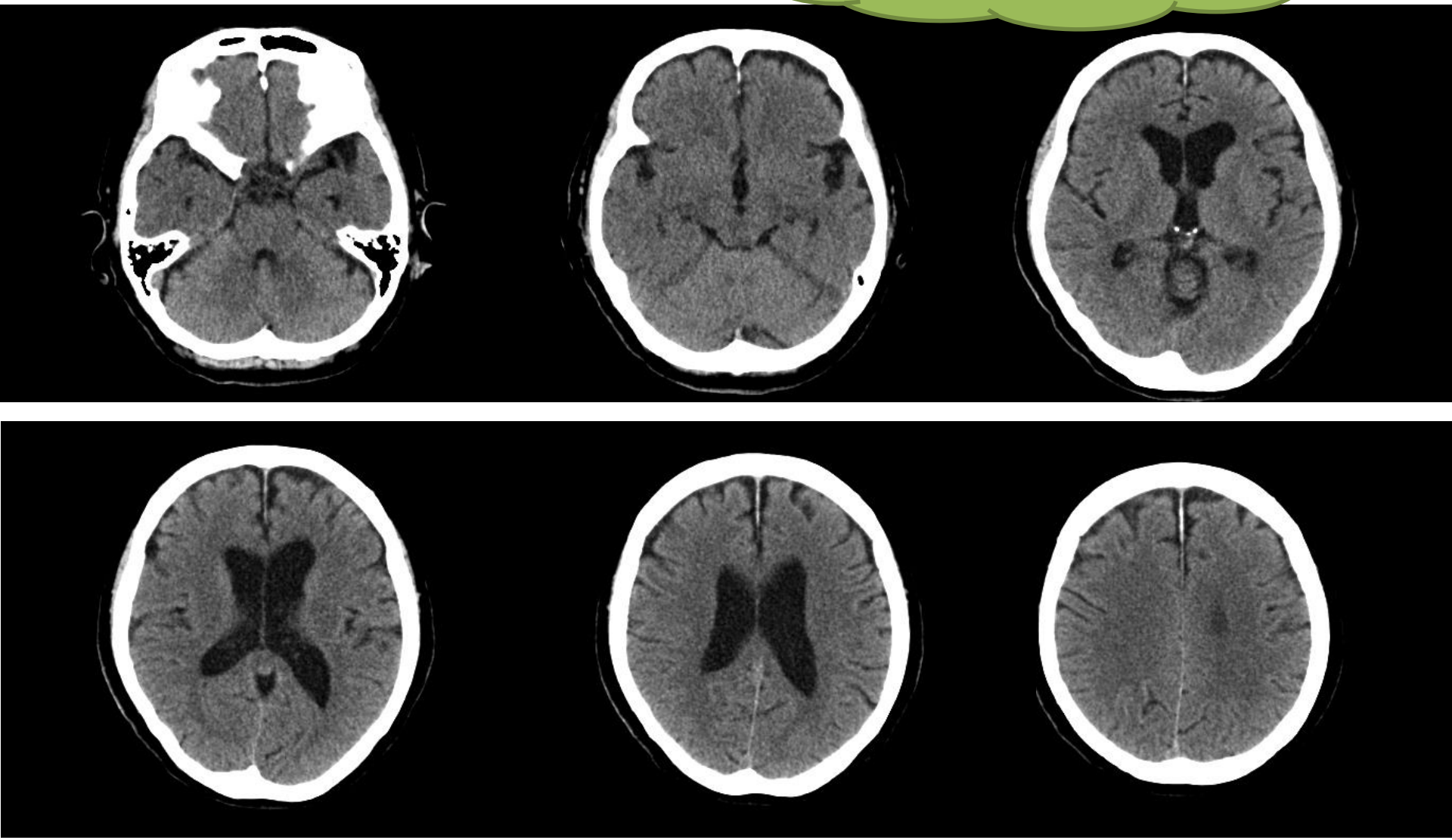
69 y.o., Female

Laboratory Data on Admission			
Total Protein	6.9 g/dl	Hb	11.4 g/dl
Albumin	3.5 g/dl	CRP	1.44 mg/dl
AST	19 U/l		
ALT	8 U/l	***CSF study***	
BUN	29 mg/dl	Total cells	3 /μl
Creatinine	1.11 mg/dl	Monocytes	3 /μl
Na	143 mEq/l	PN Leucocytes	<1 /μl
K	3.9 mEq/l		
Cl	104 mEq/l		
WBC	11990 /μl		
RBC	4.02 $\times 10^6$/μl		
Plt	227 $\times 10^3$/μl		

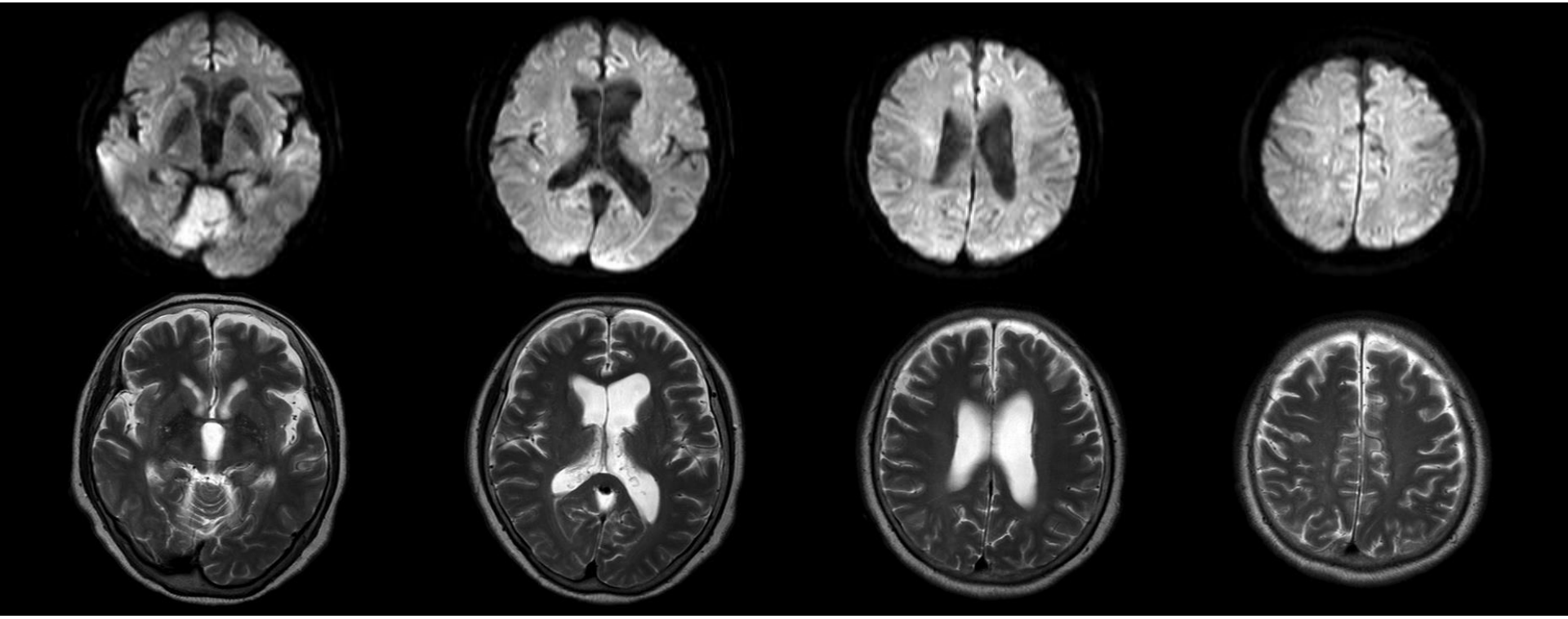
CT on admission (7, July)

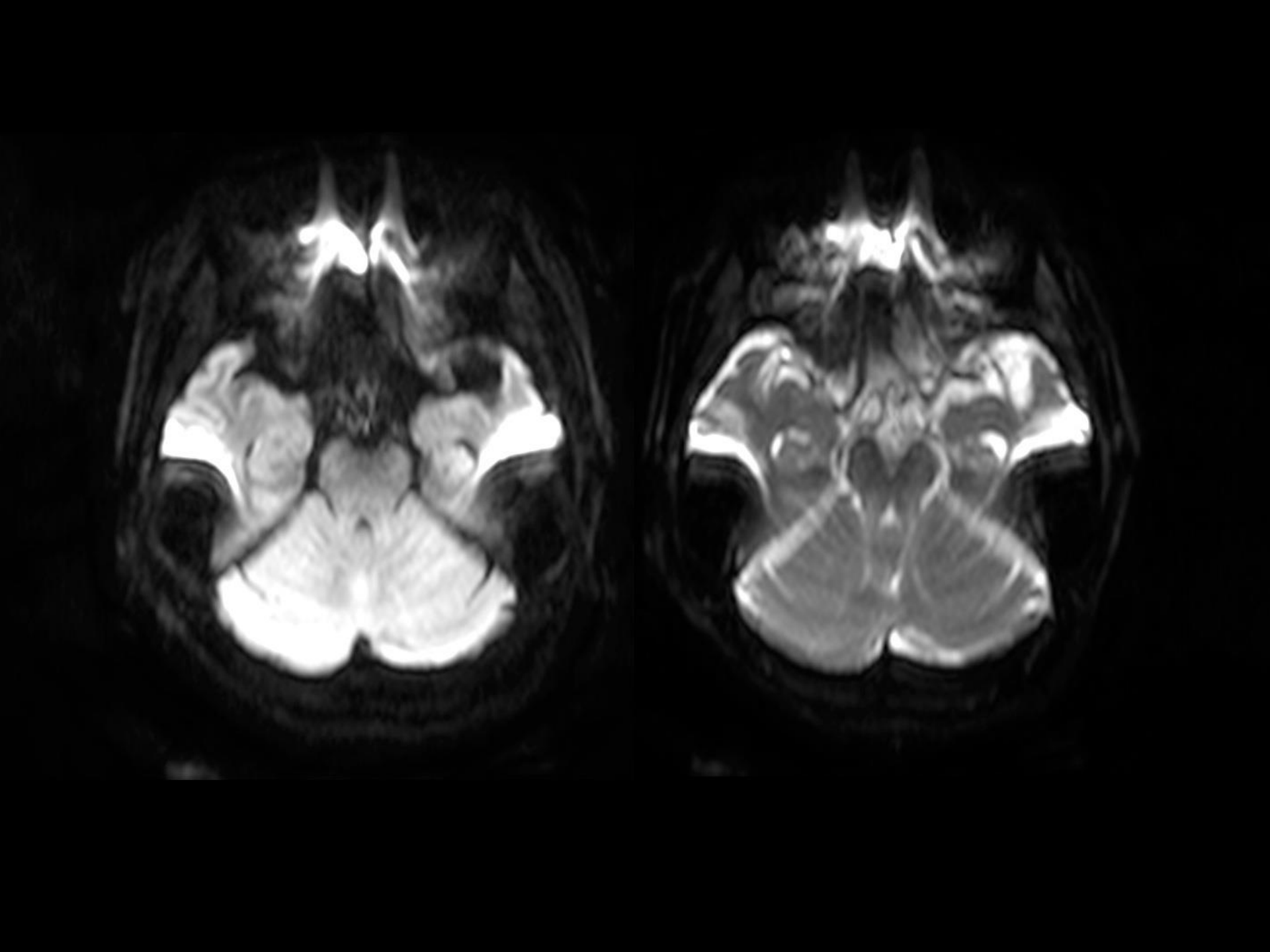
. . .

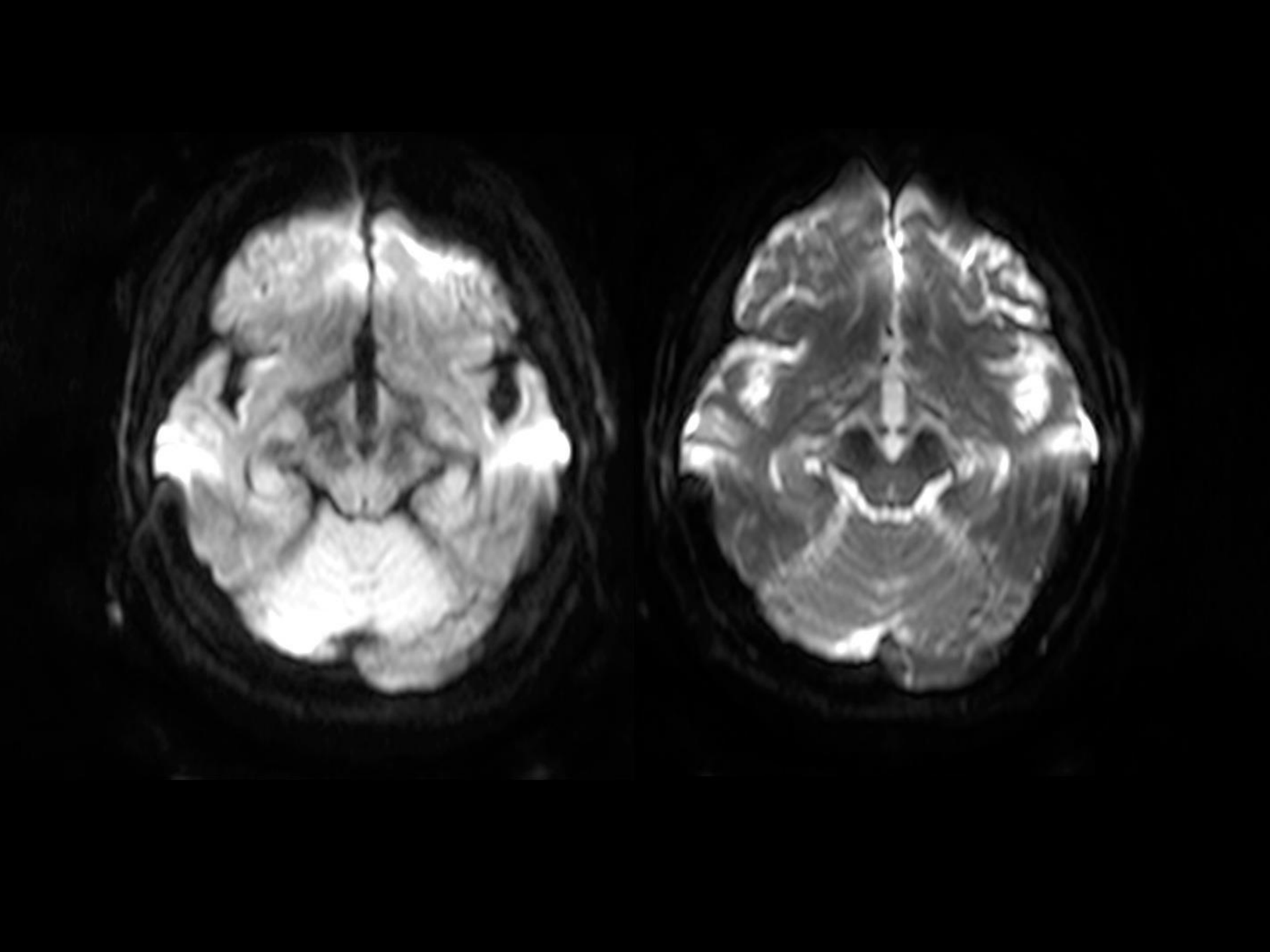
Unremarkable

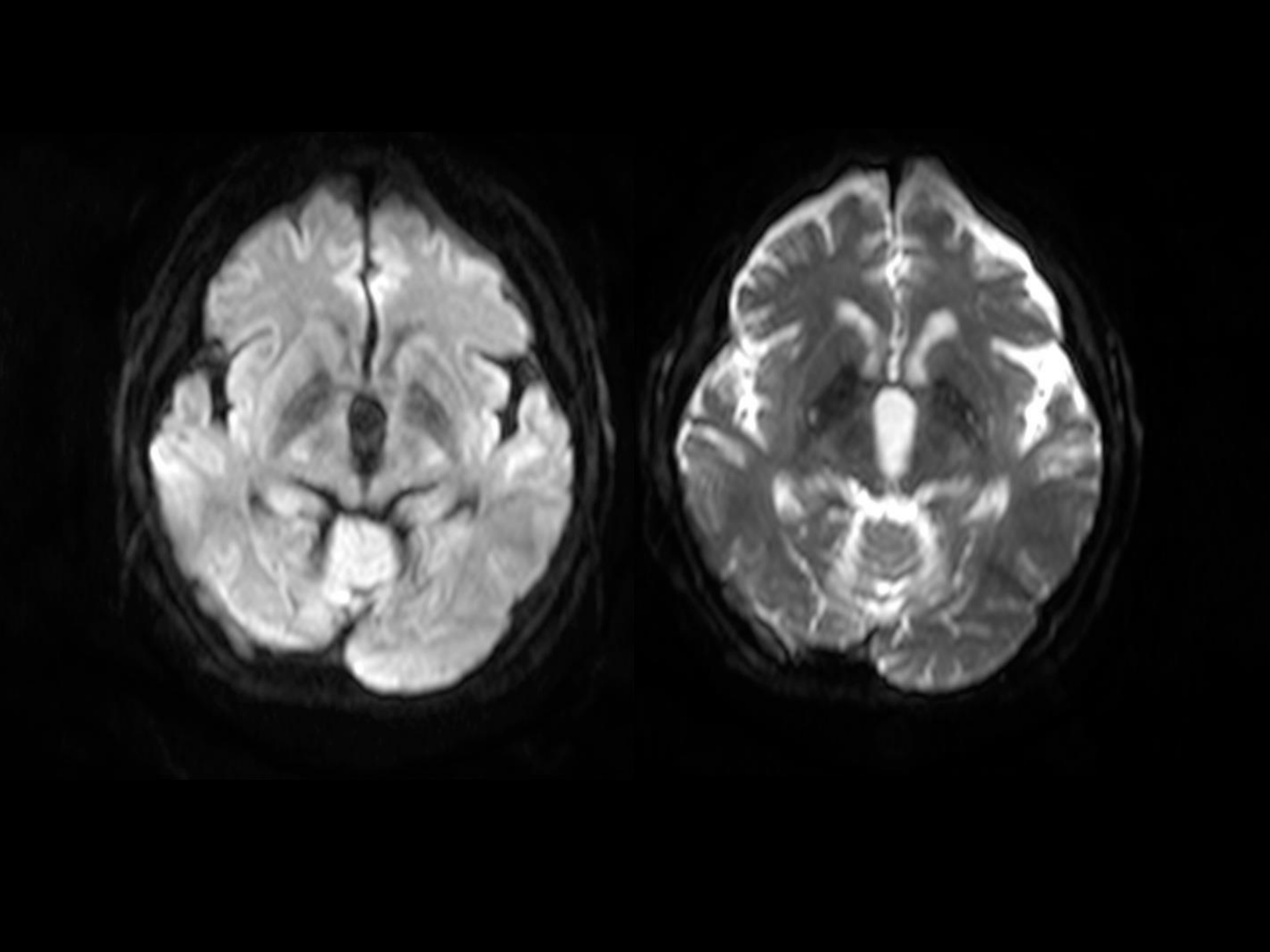


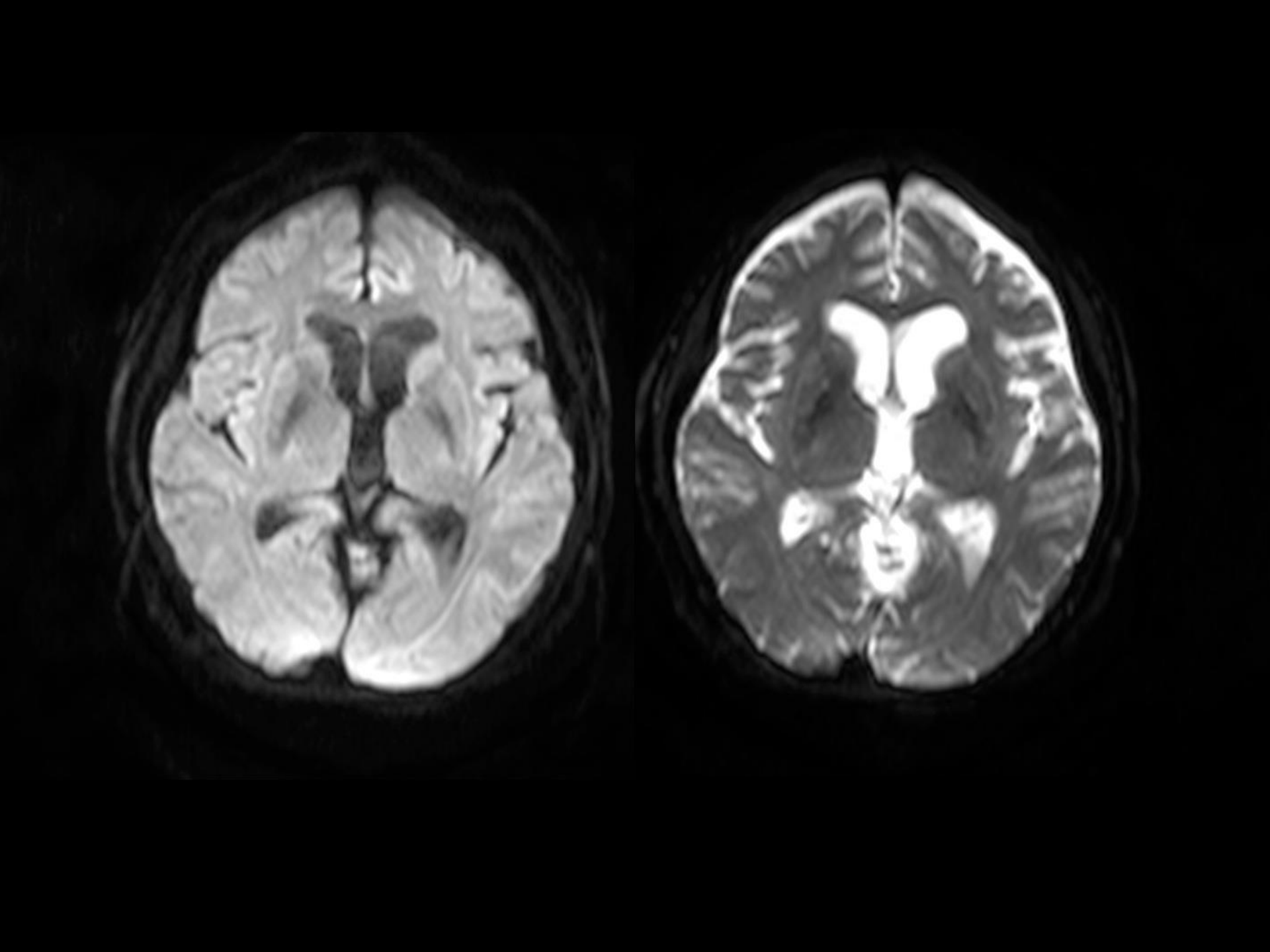
1st MRI study (13, July)

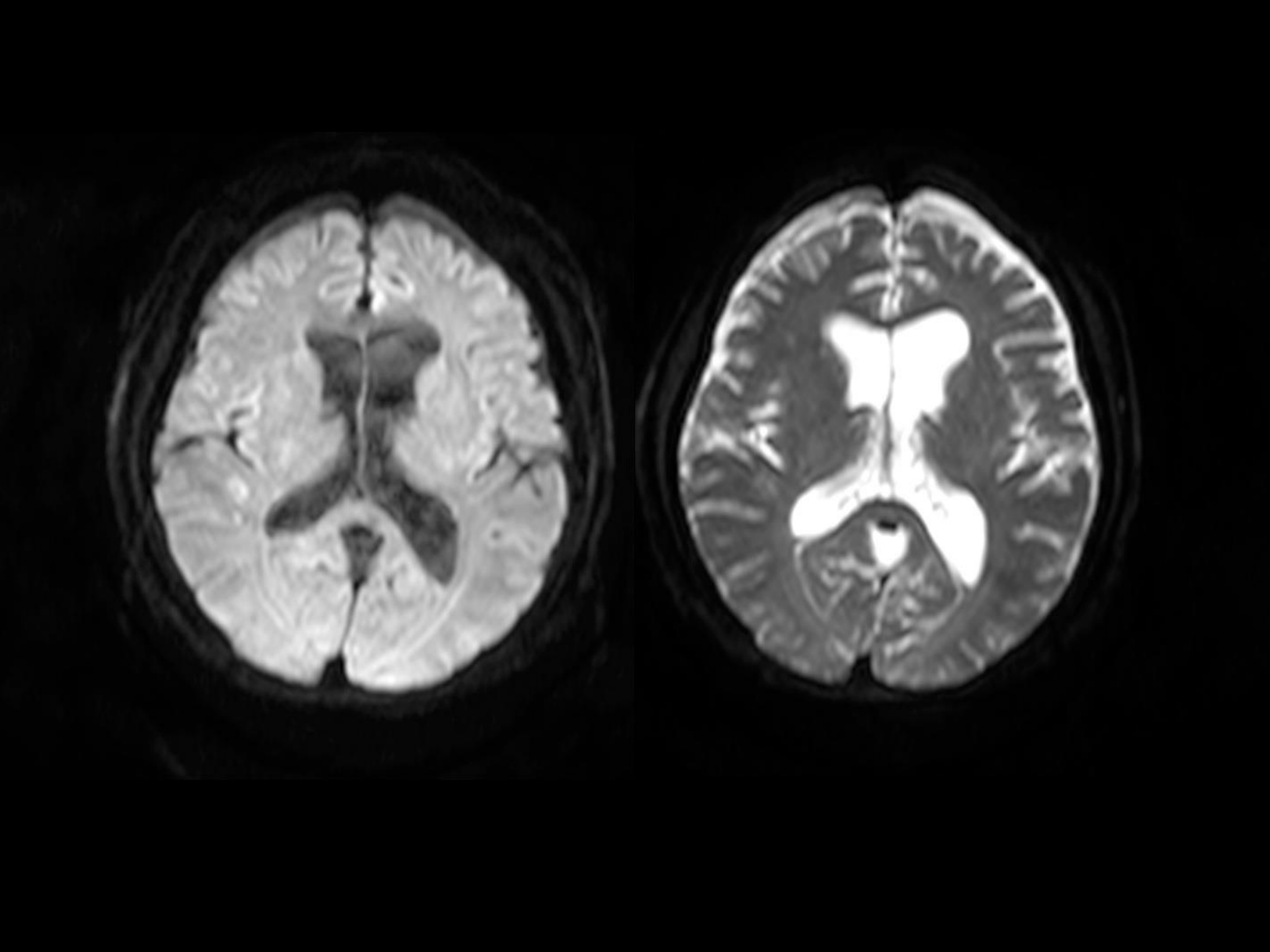


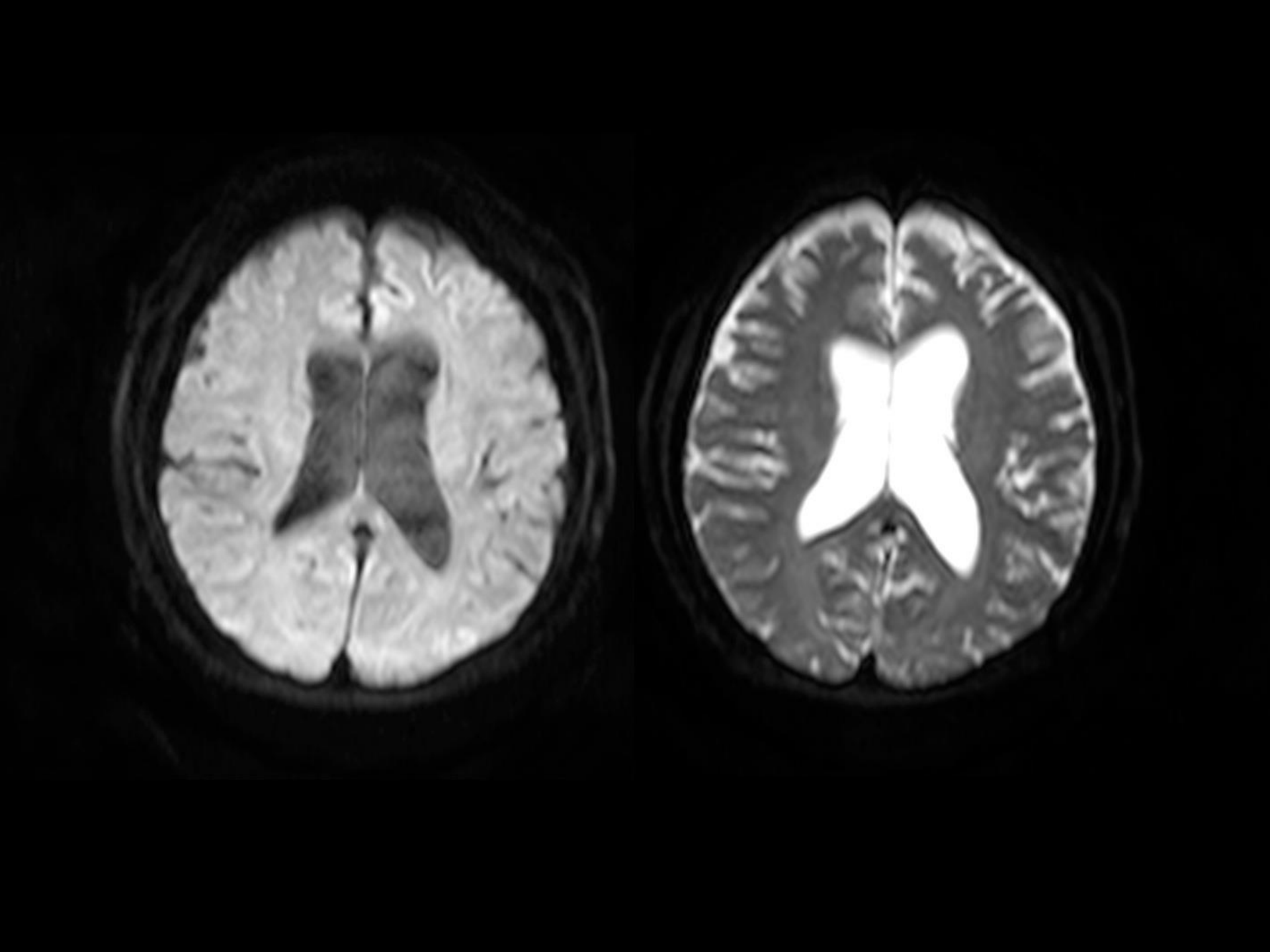


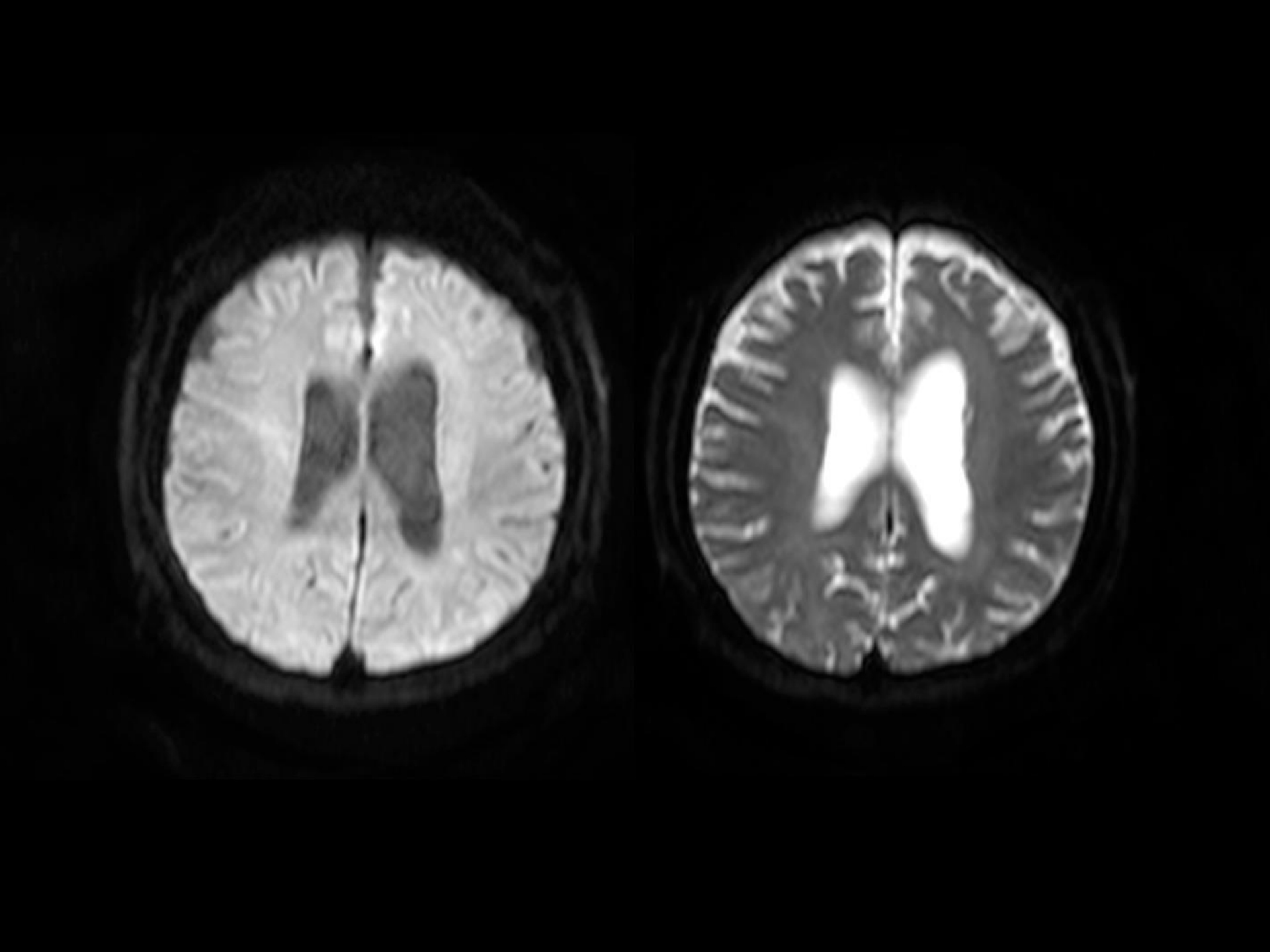


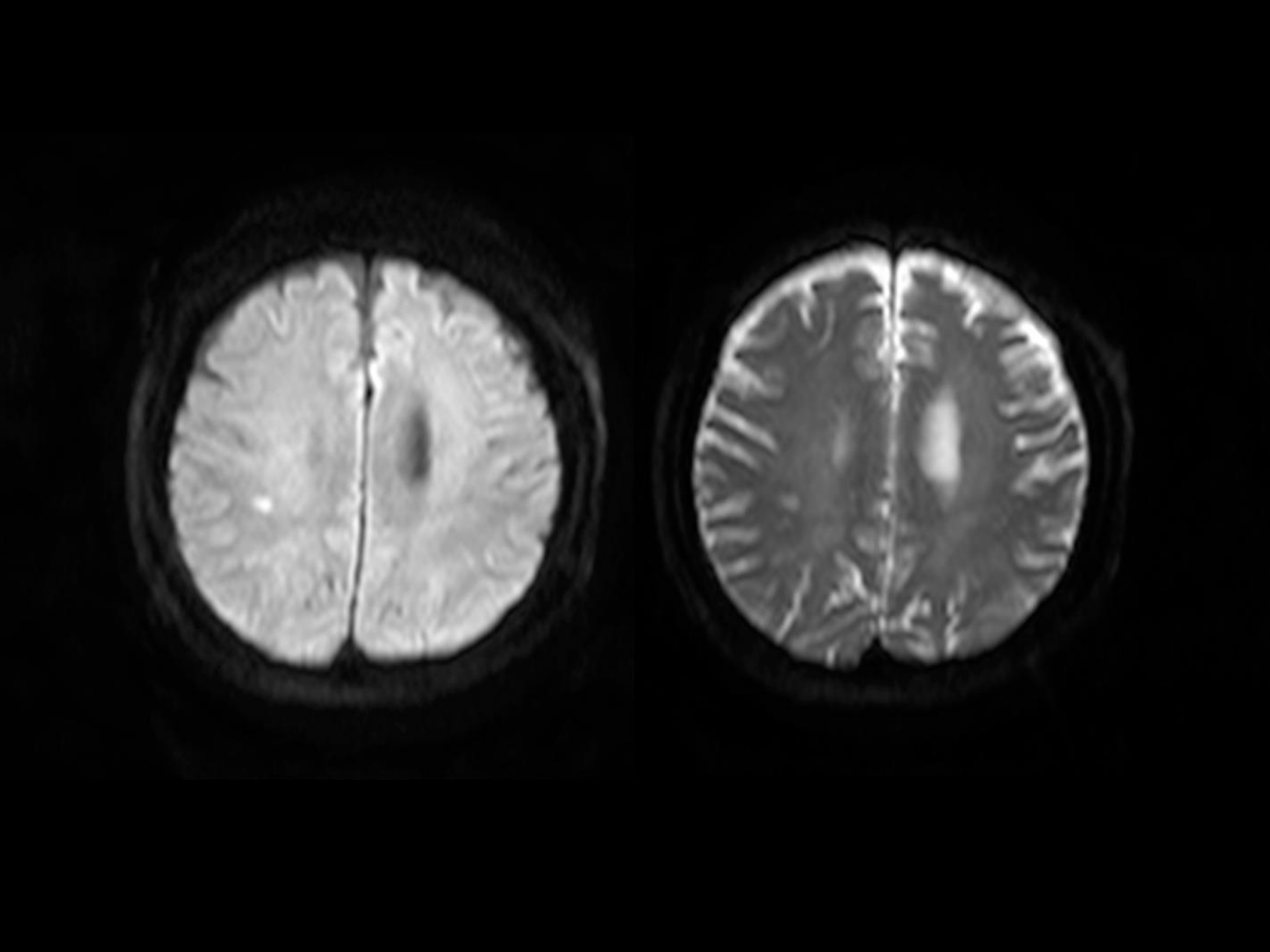


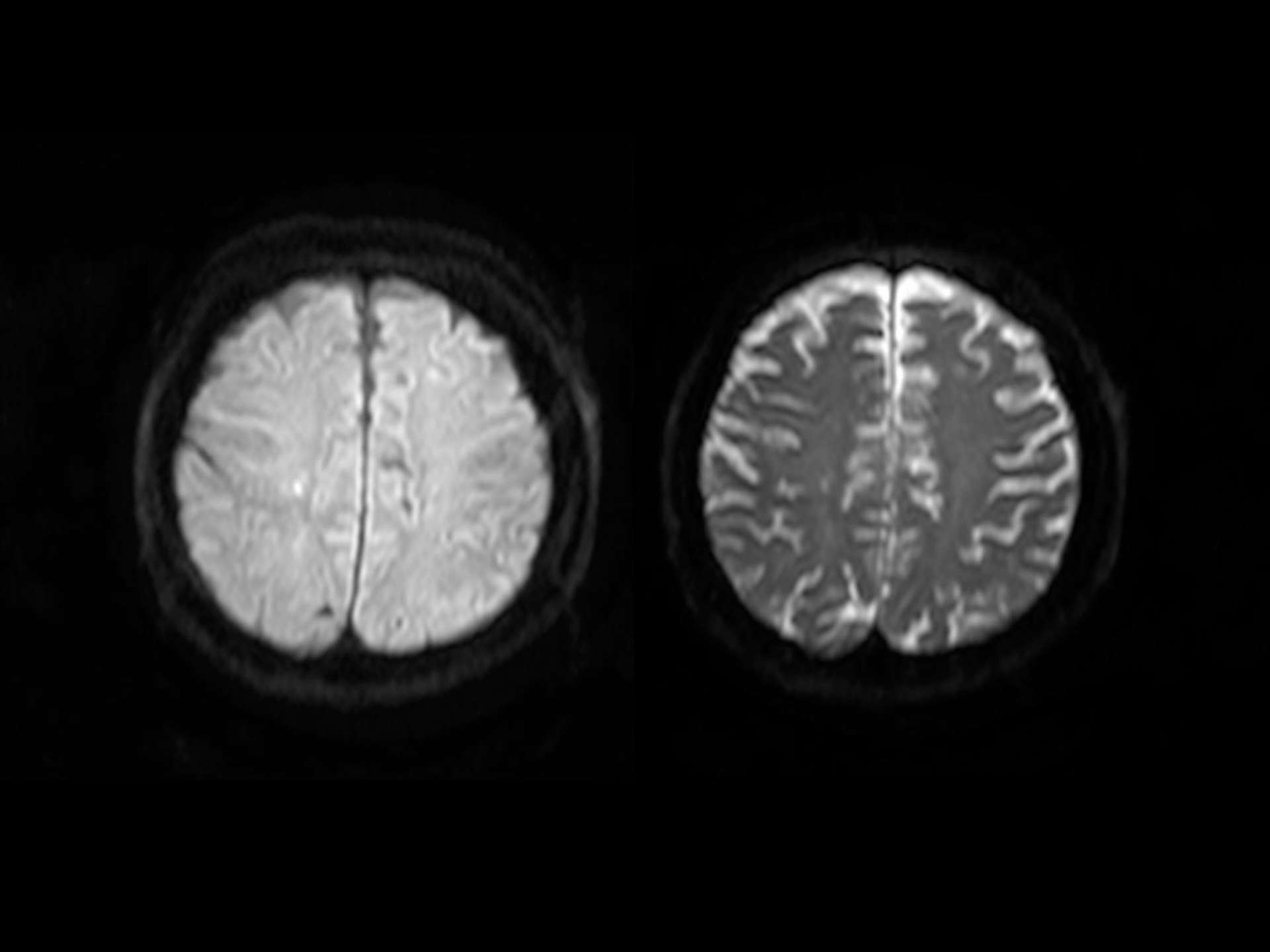


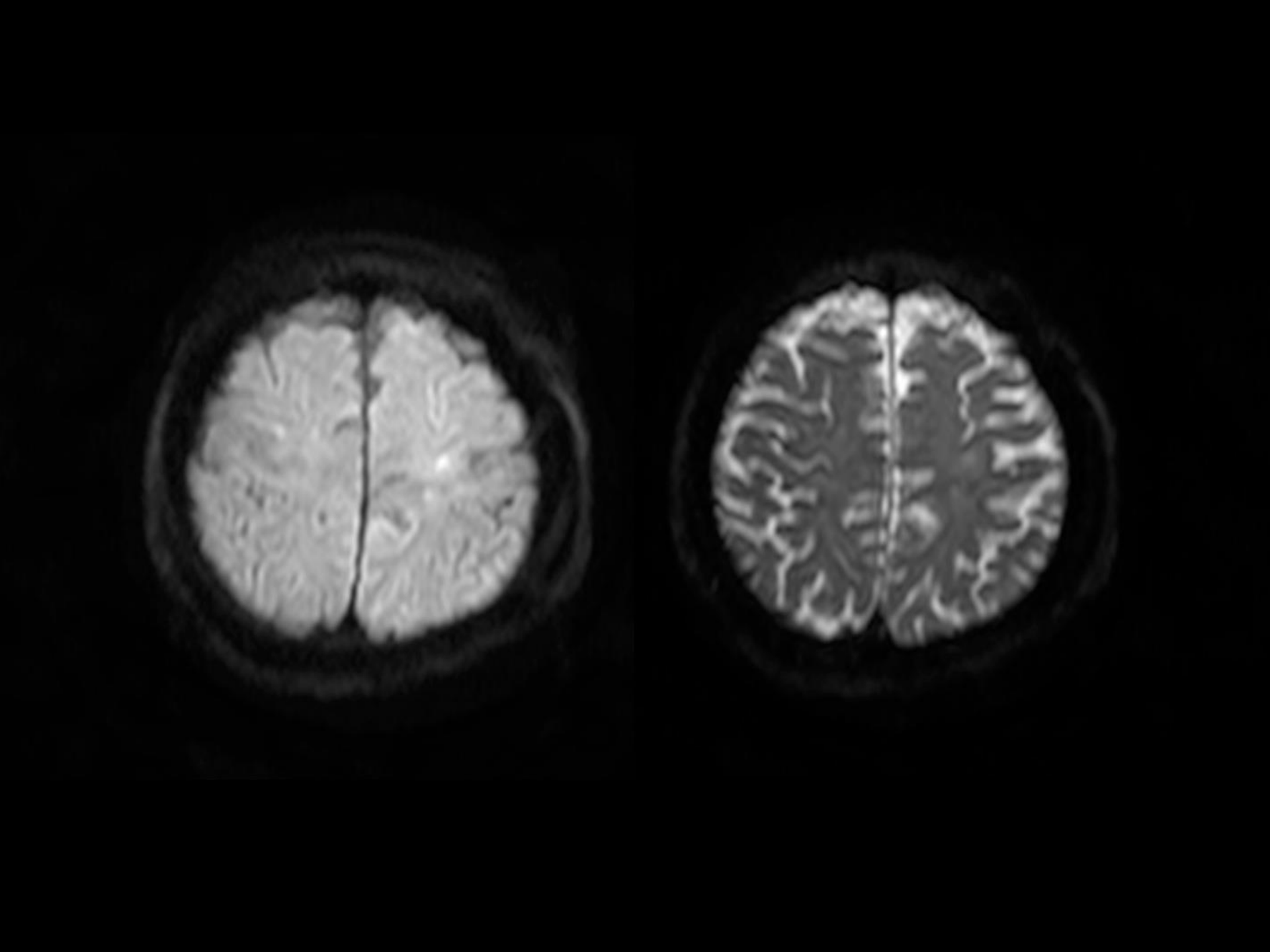


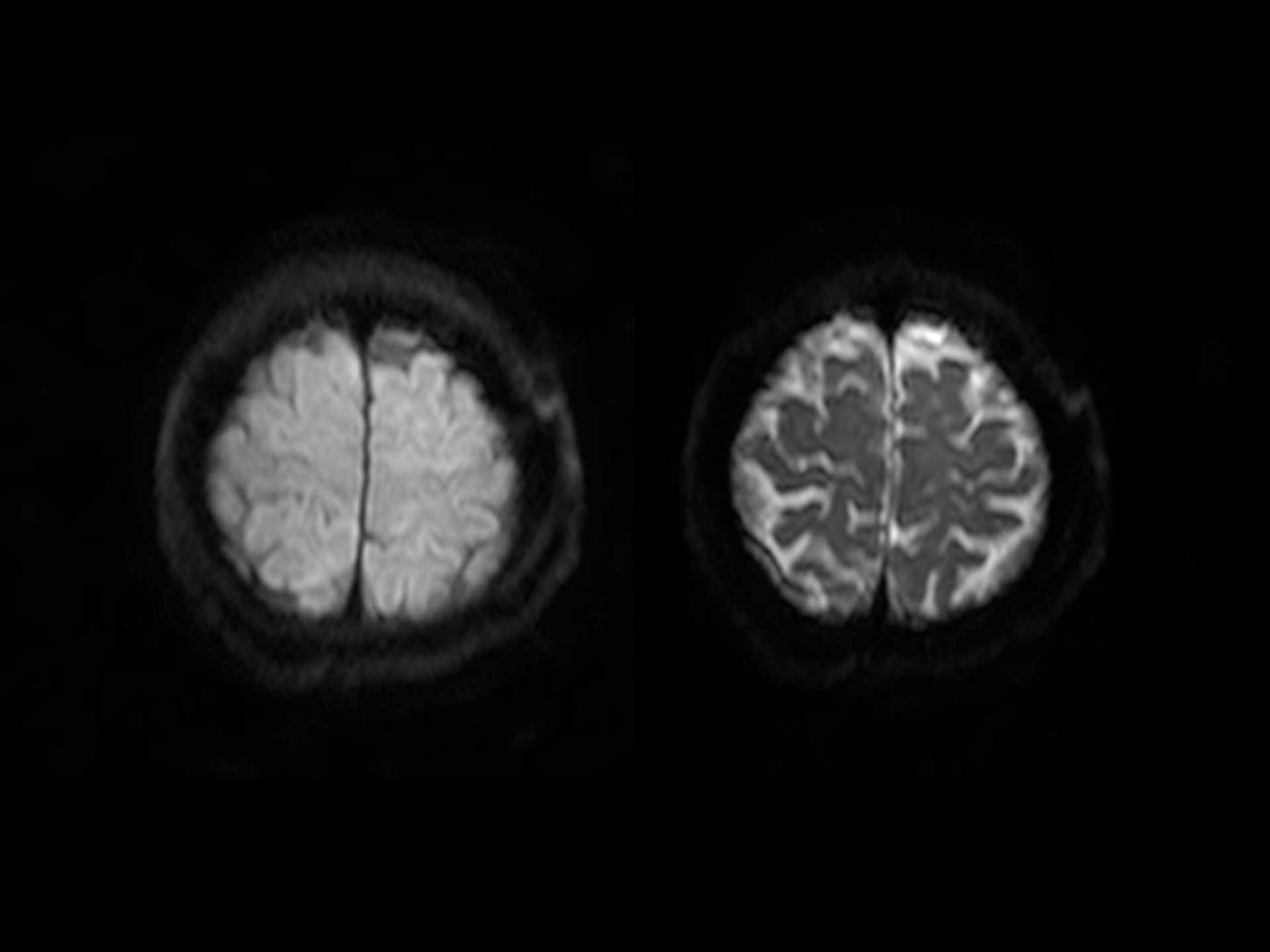




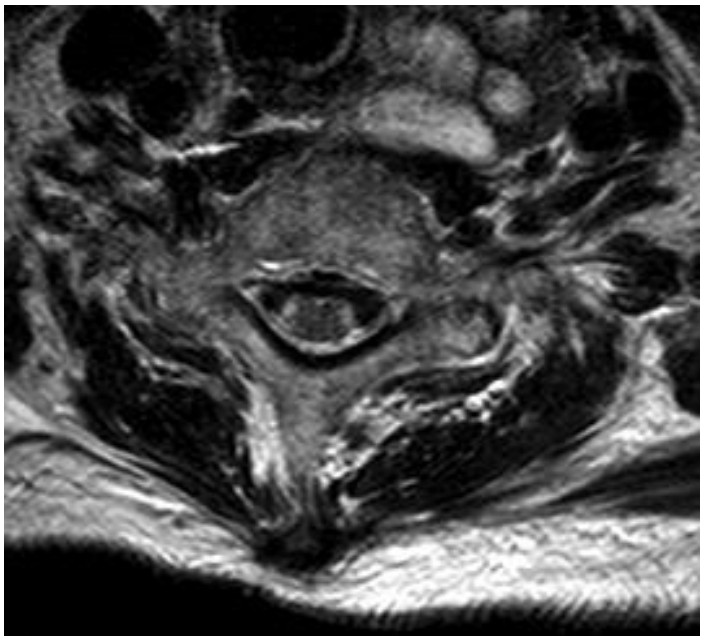


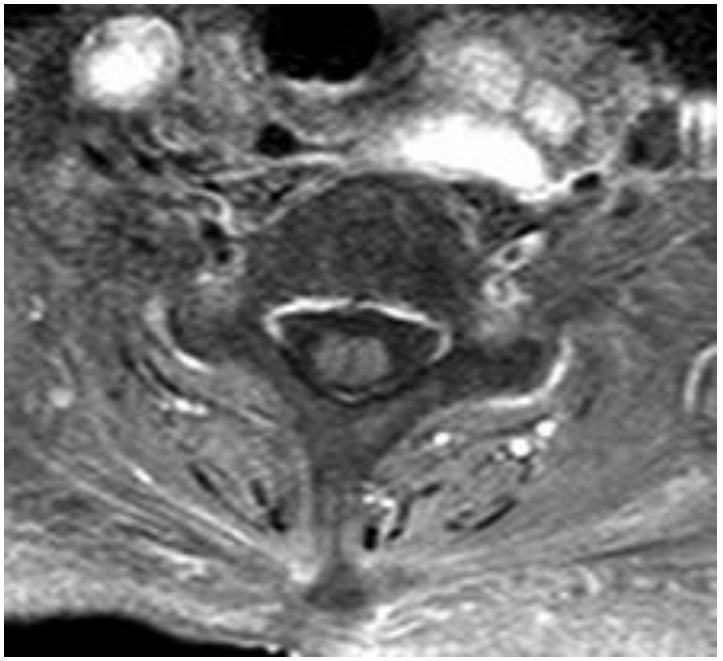


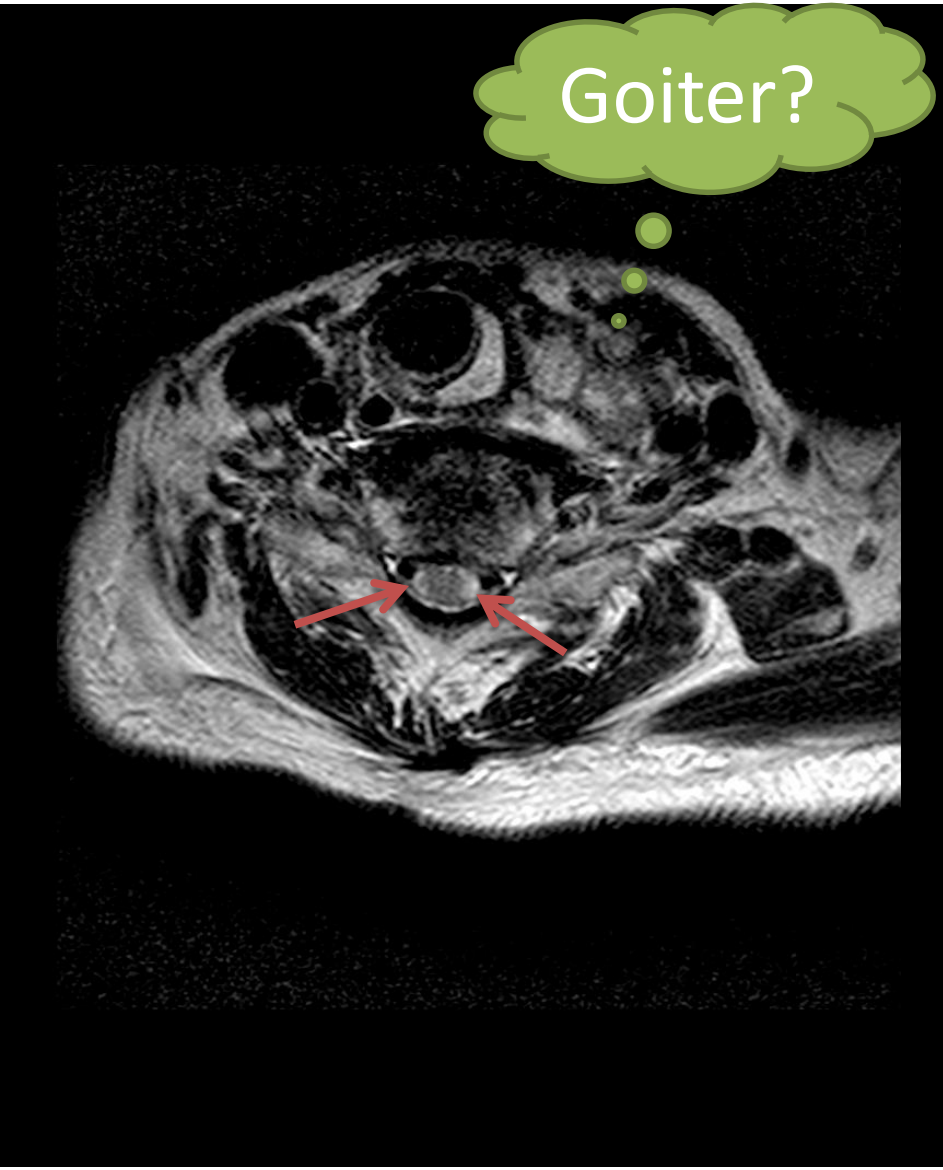


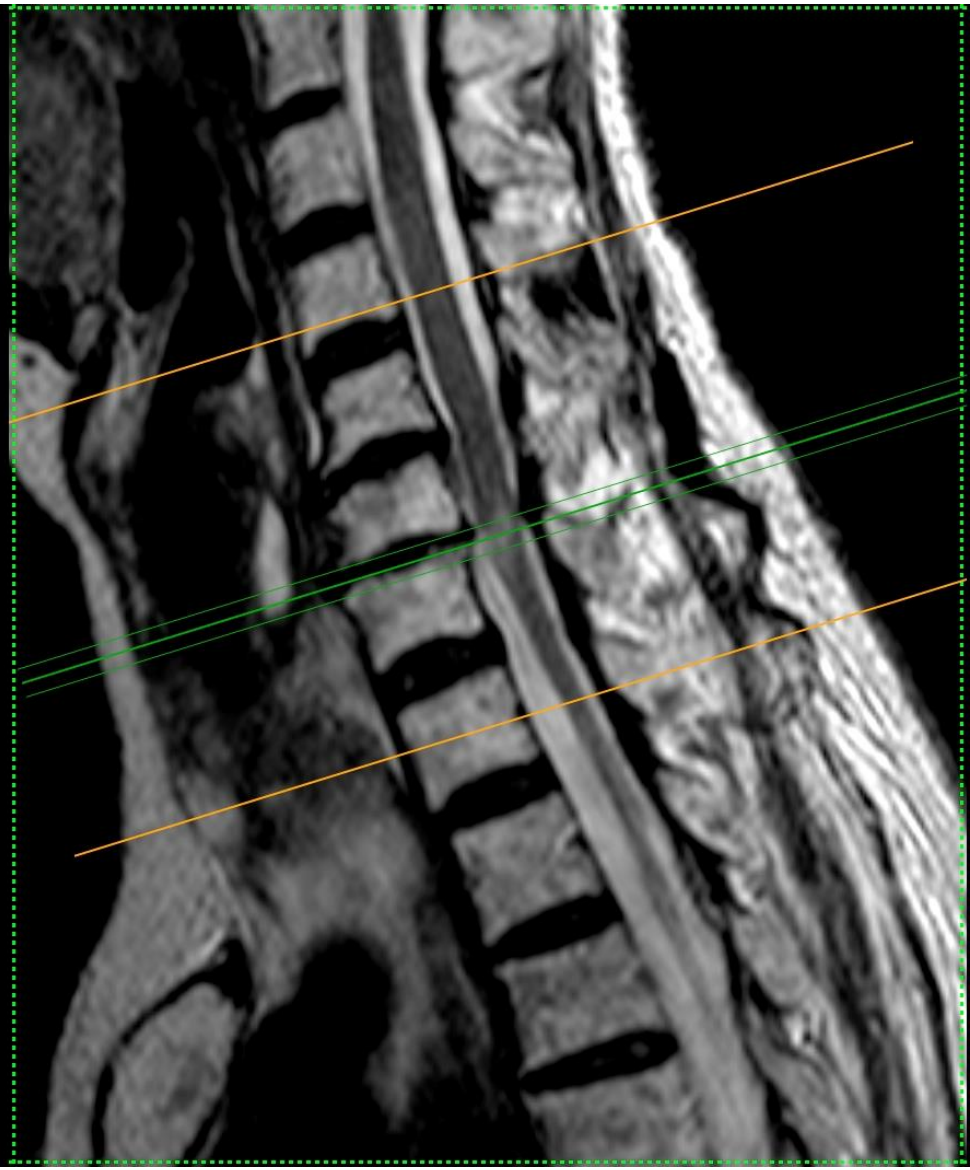


Spinal MRI (16, July)

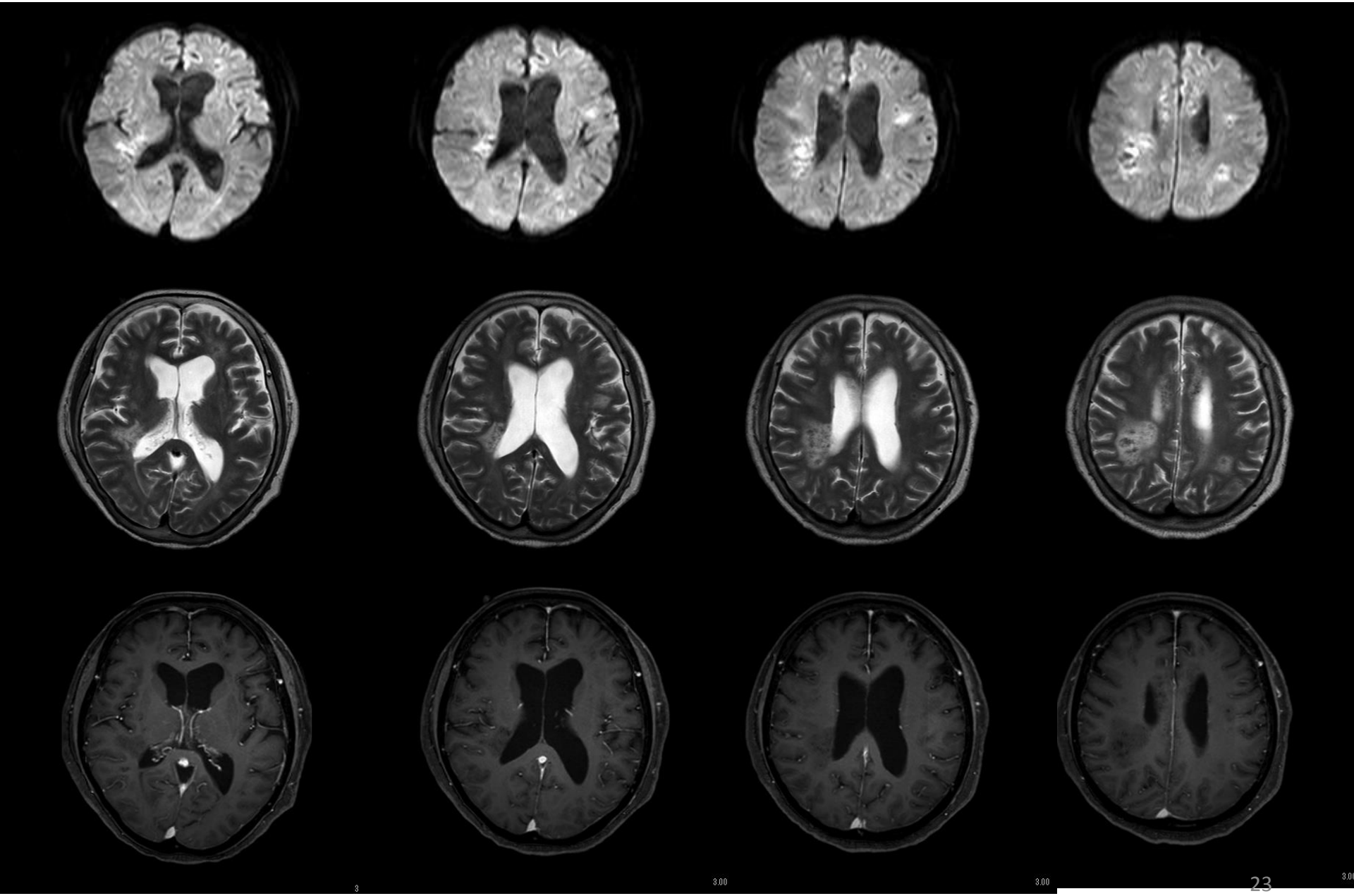


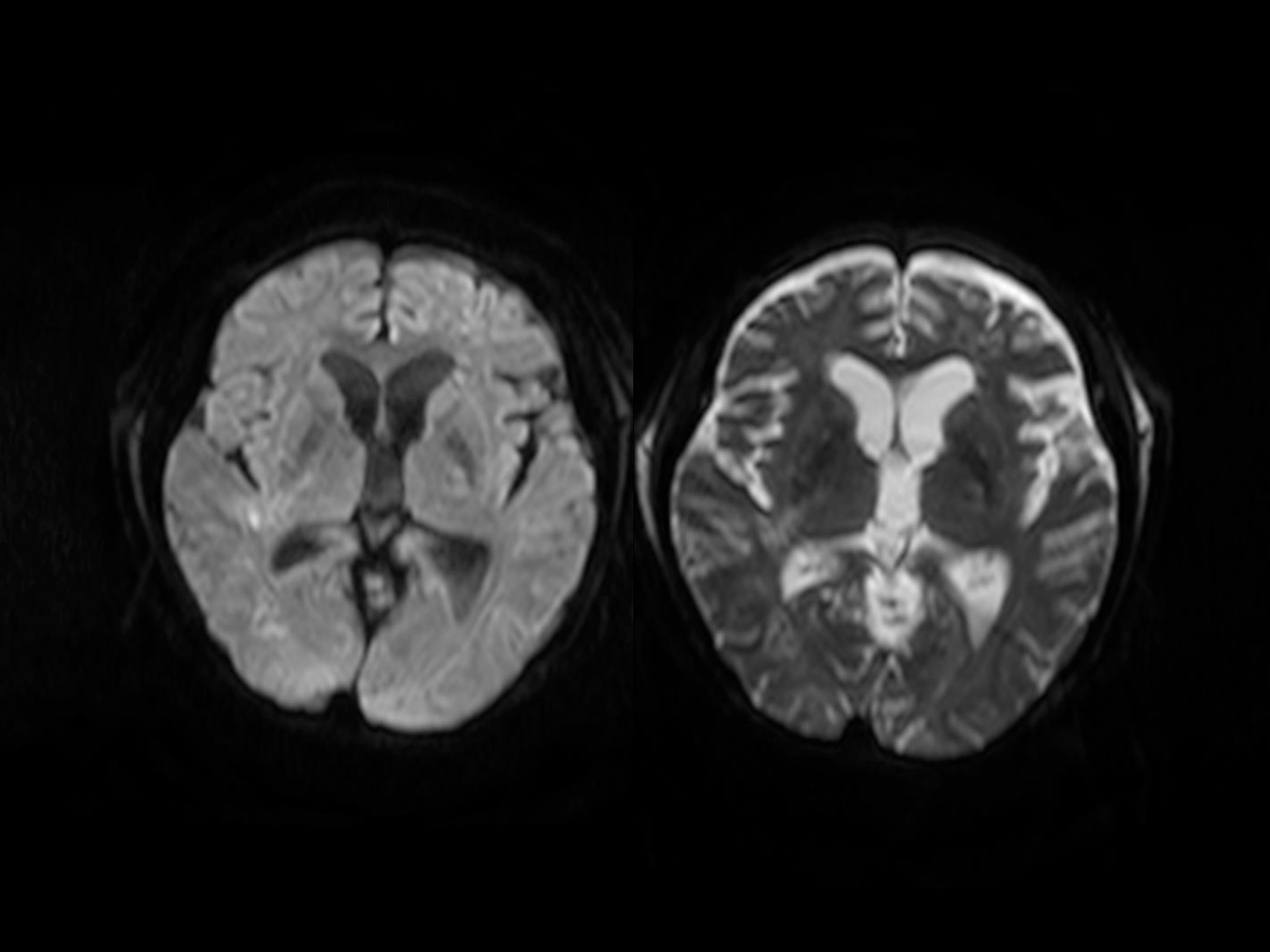


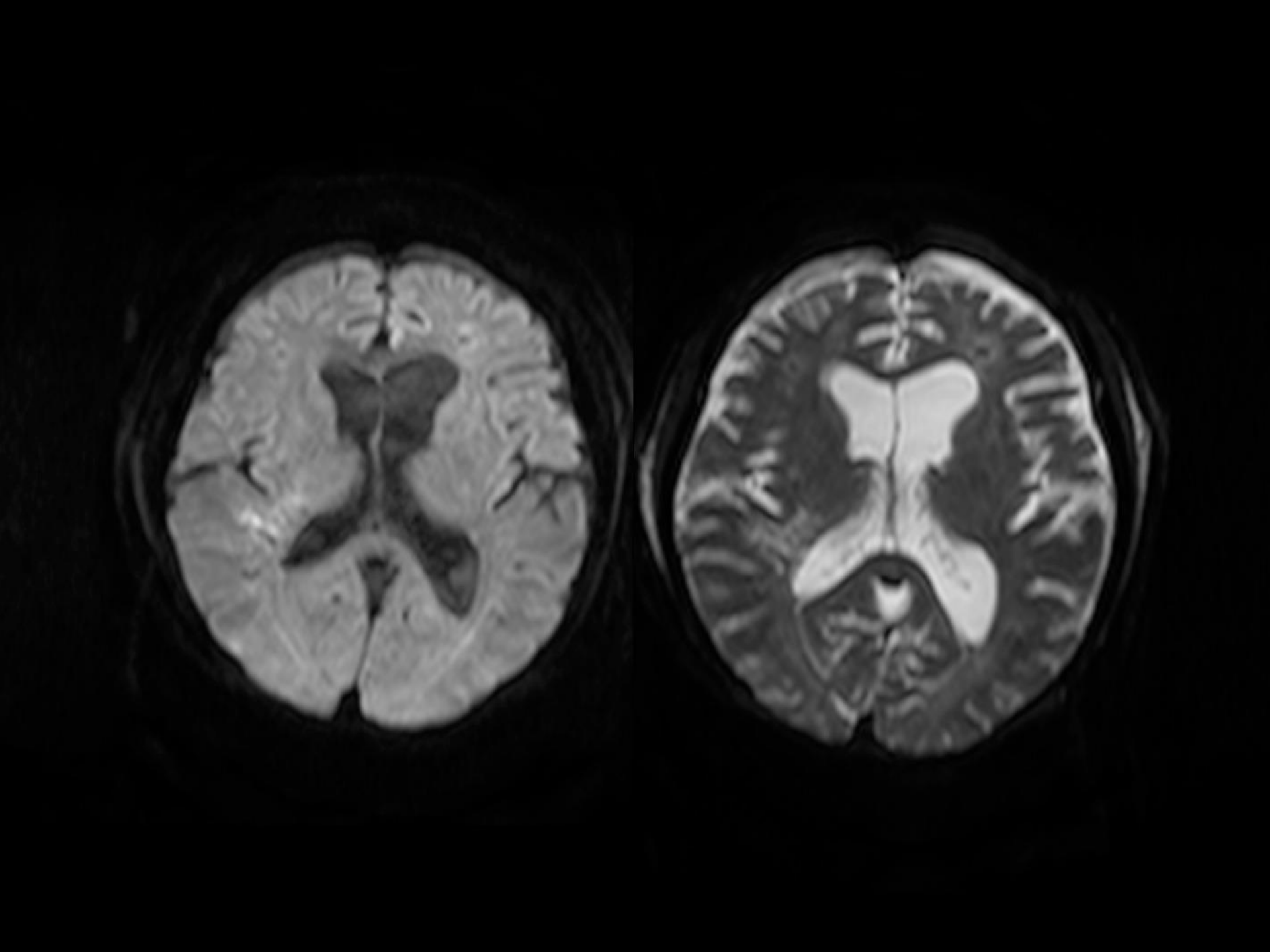

Goiter?

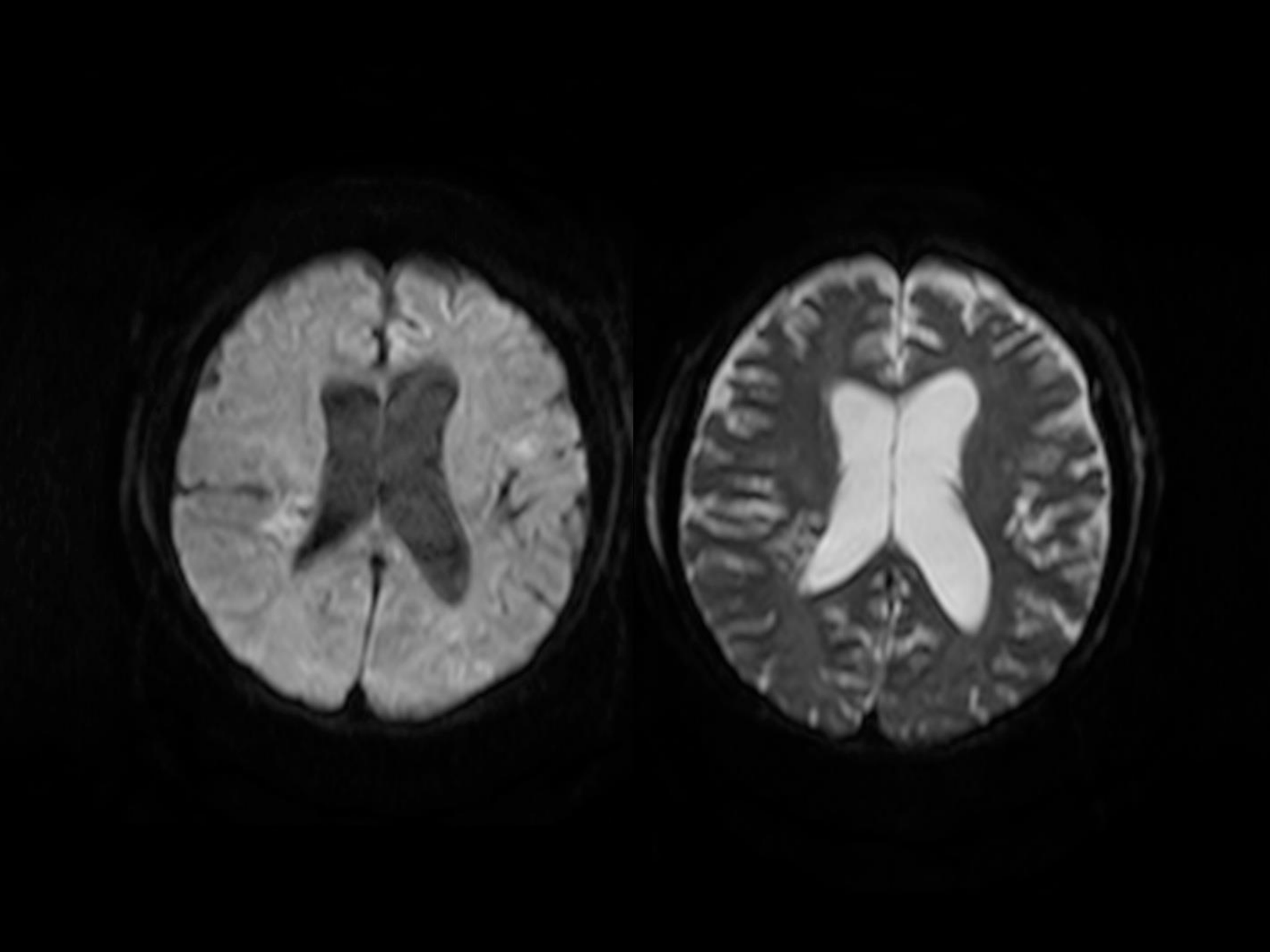


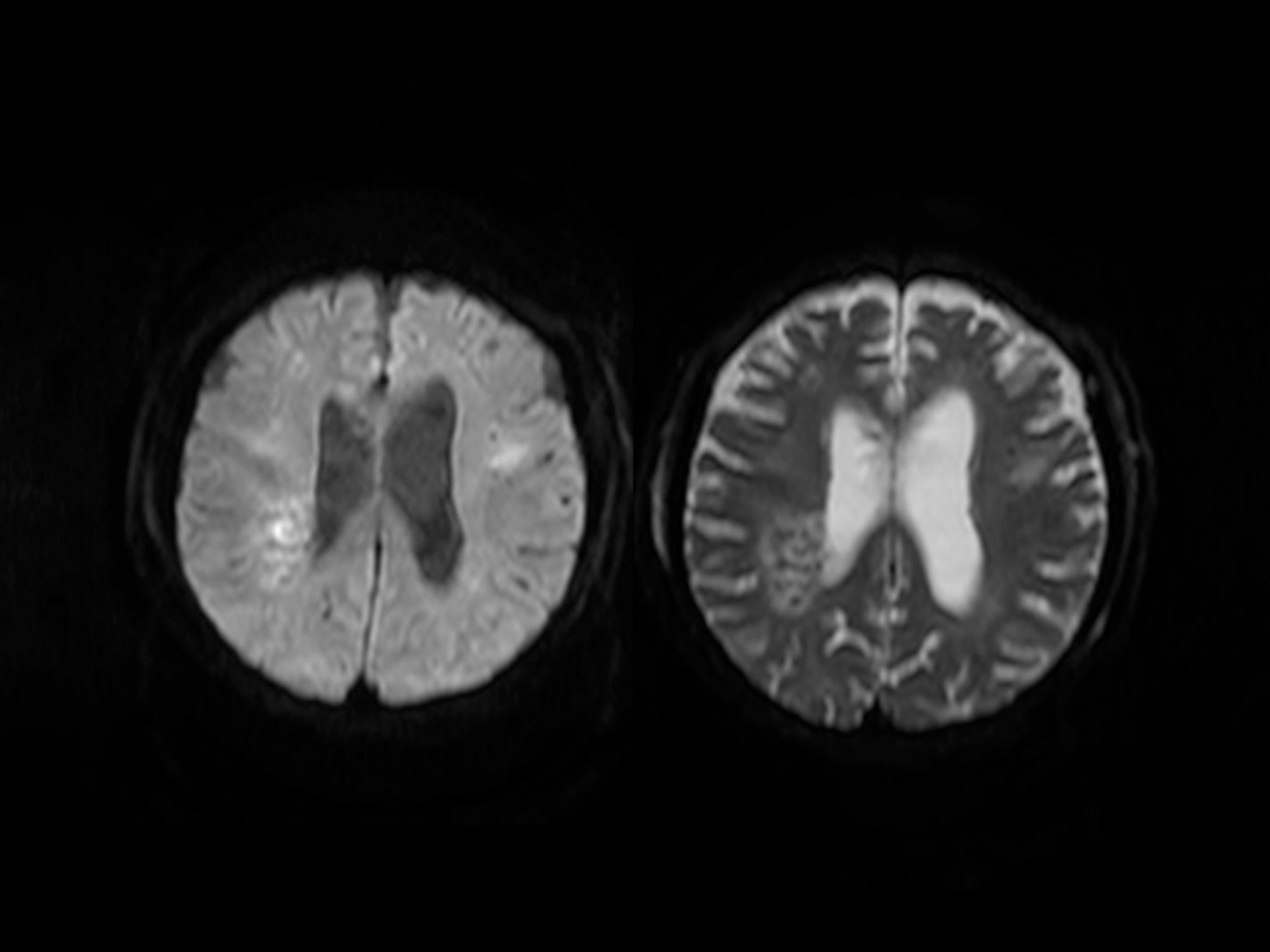
2nd MRI study (4, August)

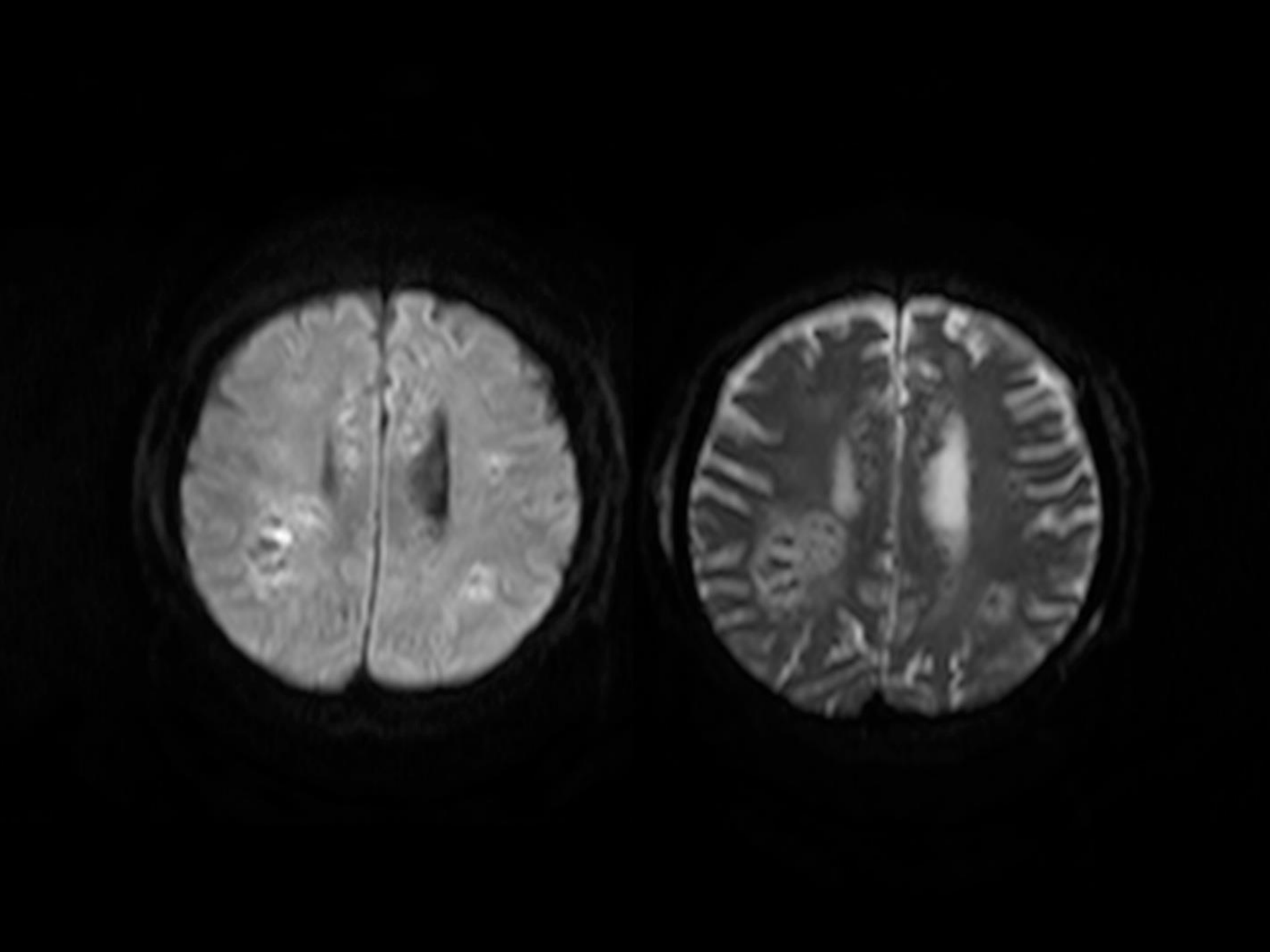


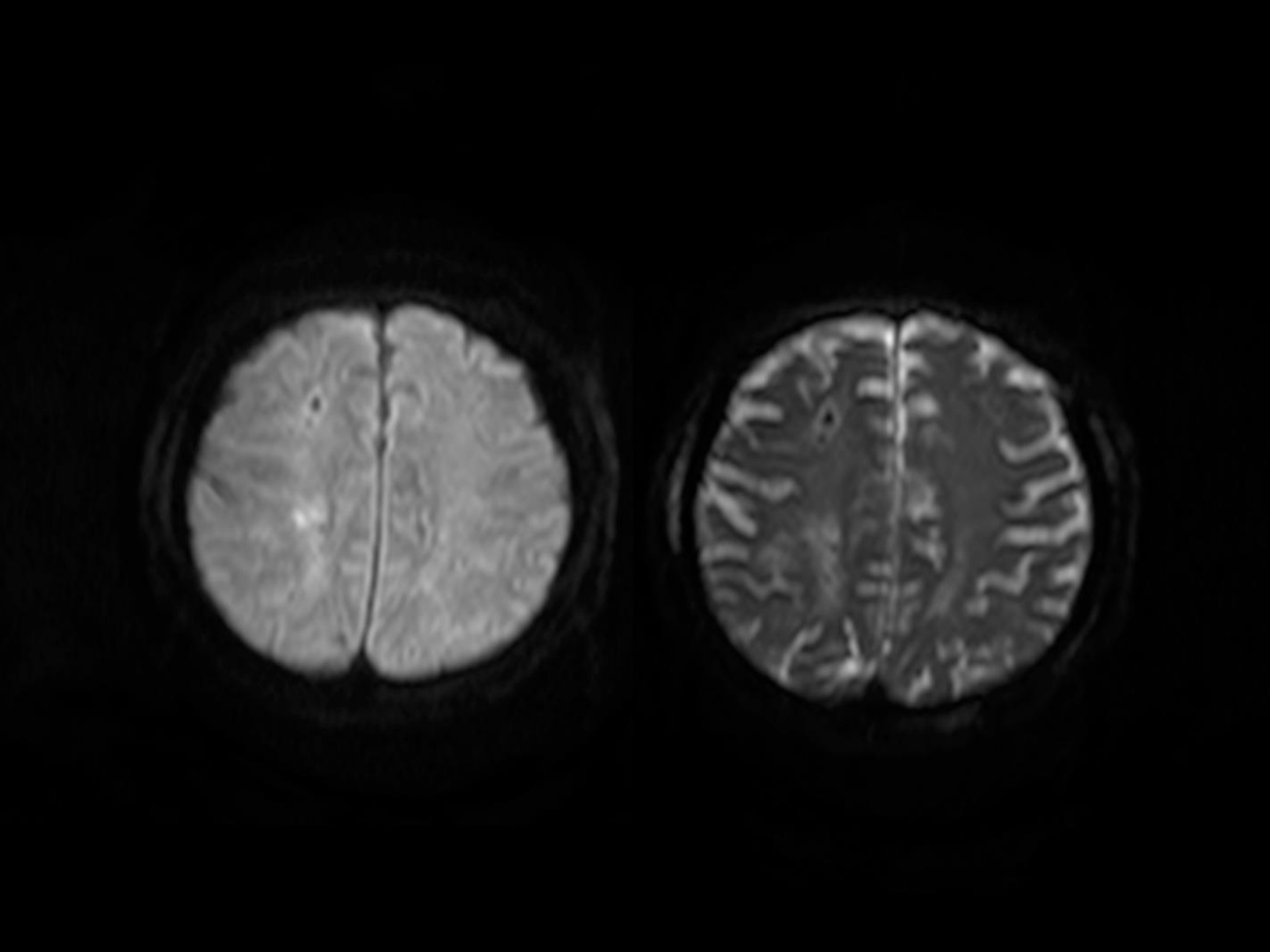


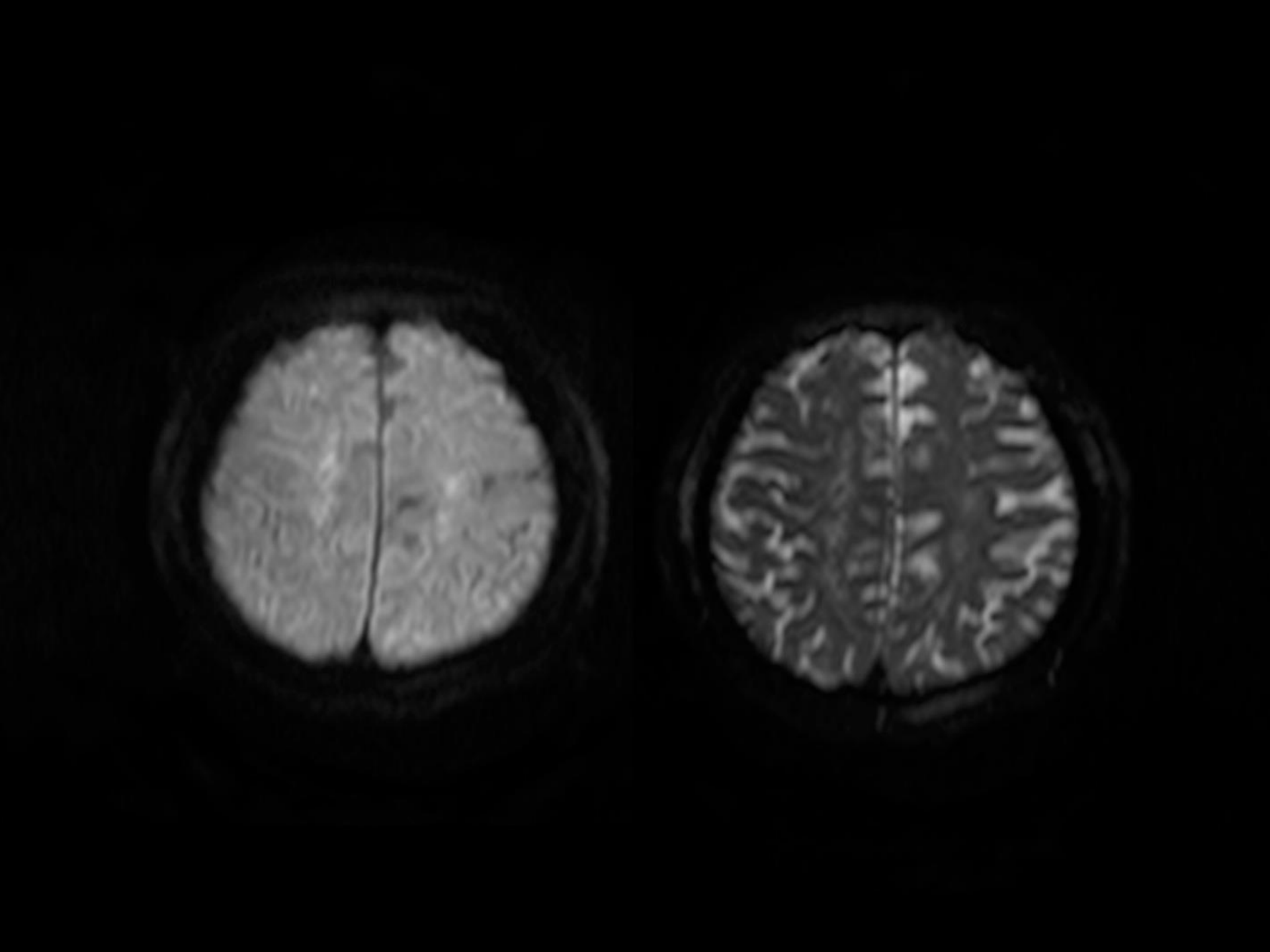








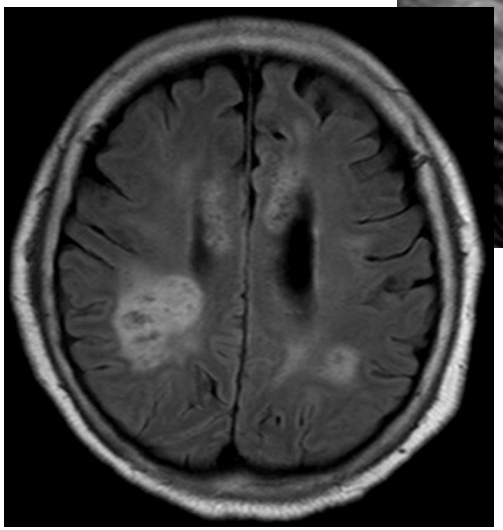


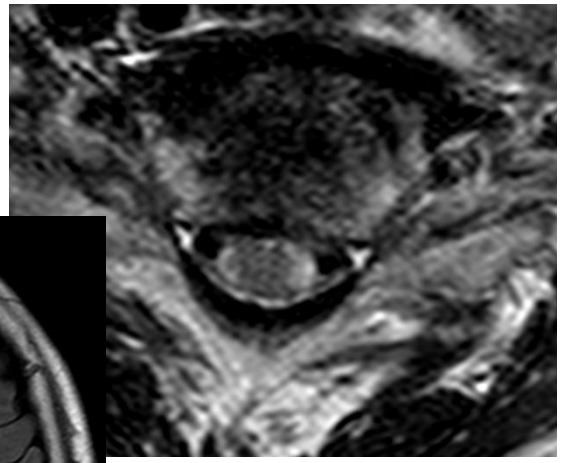


Please make a diagnosis !

Summary

- 69 y/o, Female
- Progressive vasogenic edema in the bilateral cerebral white matter and corpus callosum
  - petechial hemorrhage
  - non-enhancing
- Cervical cord lesion
  - In the periphery
- Mild leukocytosis
- Mild ↑ CRP



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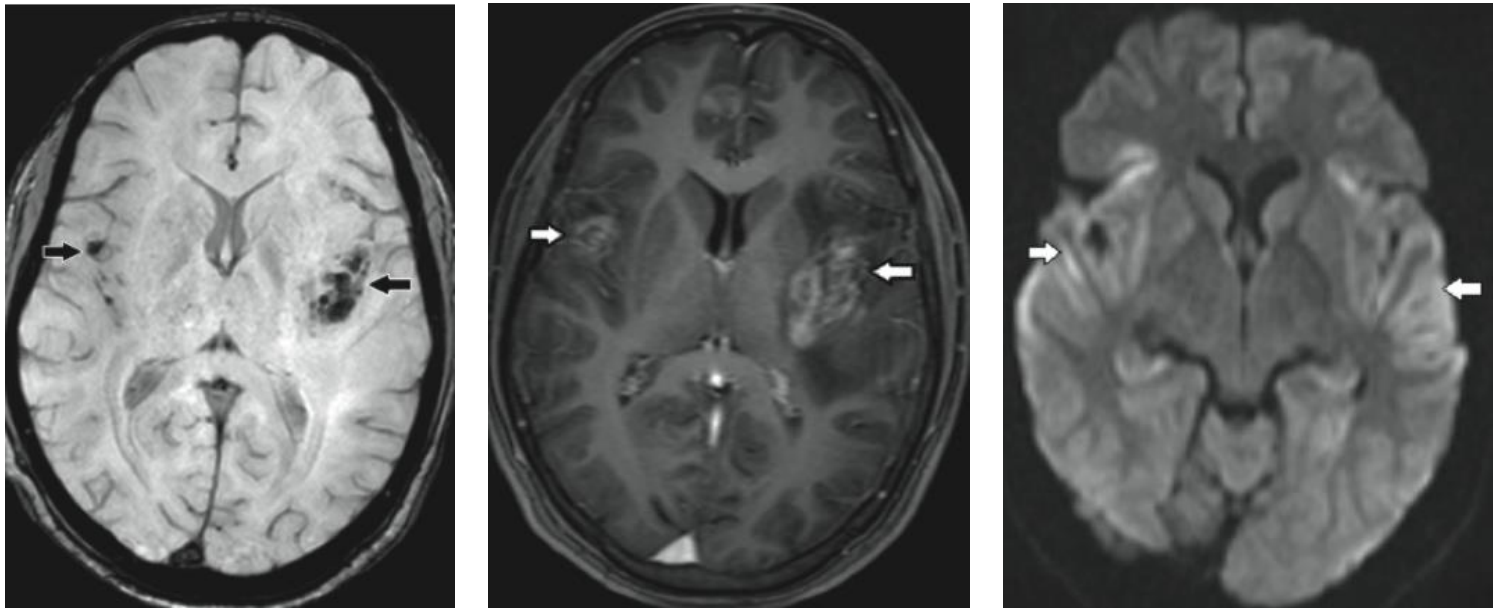
DDx

- Vascular
- Infection
- Neoplasm
- Drugs
- Inflammatory/Idiopathic
- Congenitial
- Autoimmune
- Trauma
- Endocrine/Metabolic

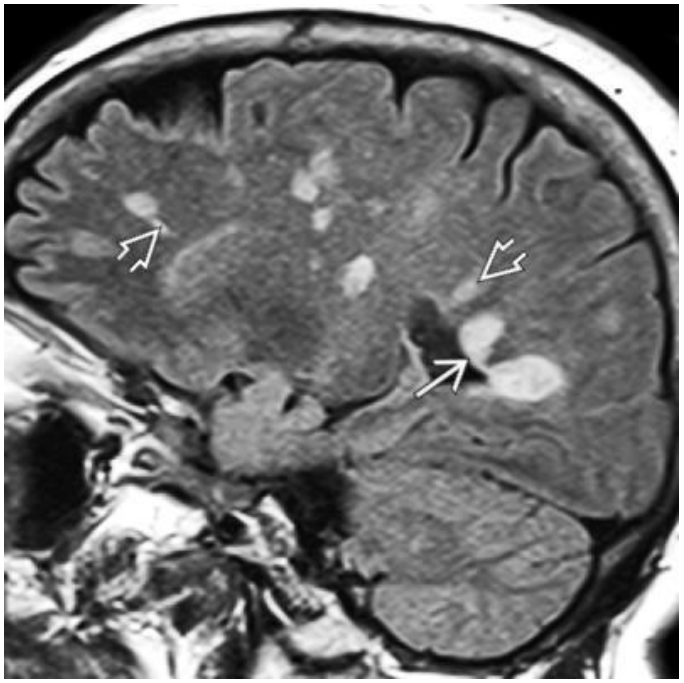
Infection

- Pyogenic infection
  - Abscess
- Viral infection
  - Herpes simplex encephalitis

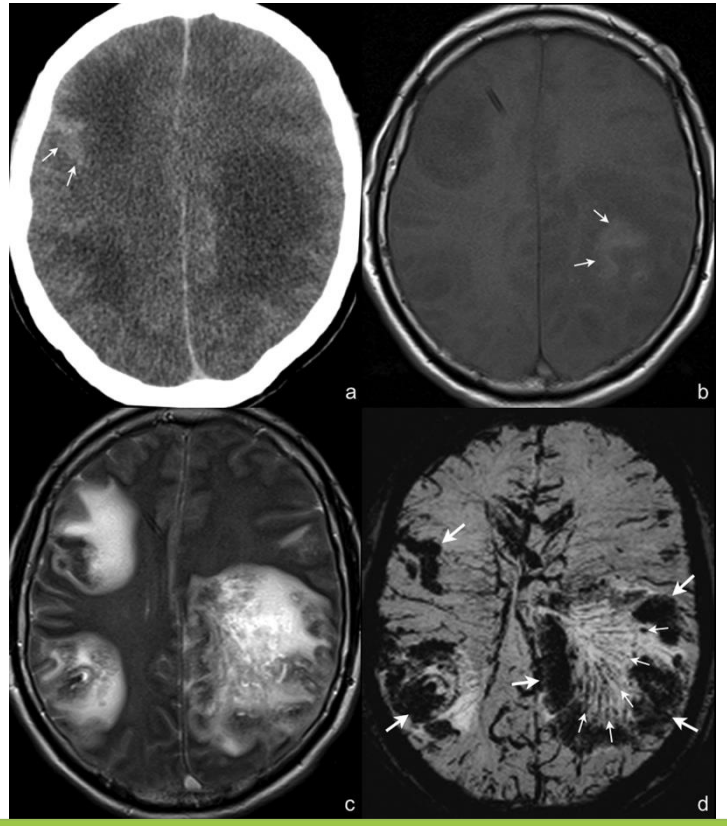


Sureka J, etal. Clinico-radiological spectrum of bilateral temporal lobe hyperintensity: a retrospective review. Br J Radiol. 2012 Sep;85(1017):e782–92.

Inflammatory/Autoimmune



Multiple sclerosis



Acute hemorrhagic encephalomyelitis

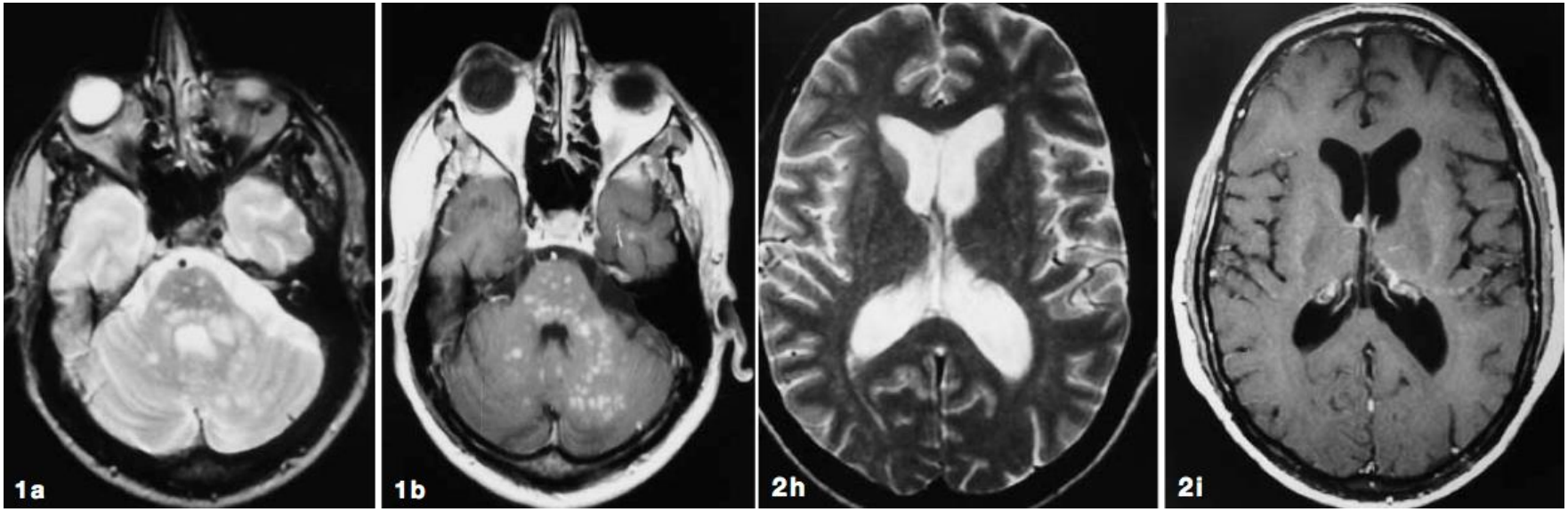
Osborn's Brain; Kao H-W, et al. Value of susceptibility-weighted imaging in acute hemorrhagic leukoencephalitis. J Clin Neurosci. 2012;19(12):1740–1.

Inflammatory/Autoimmune

- Vasculitis
  - SLE — Multiple infarcts
  - Polyarthritis nodosa — Small peripheral infarcts
  - Neuro-Behcet disease — Midbrain involvement & ulcers
  - Wegener's granulomatosis — Patchy dural thickening
  - Paraneoplastic vasculitis of CNS — Remote history
  - PACNS (primary angiitis of the CNS) — Diagnosed by exclusion
Brain & cord

PACNS

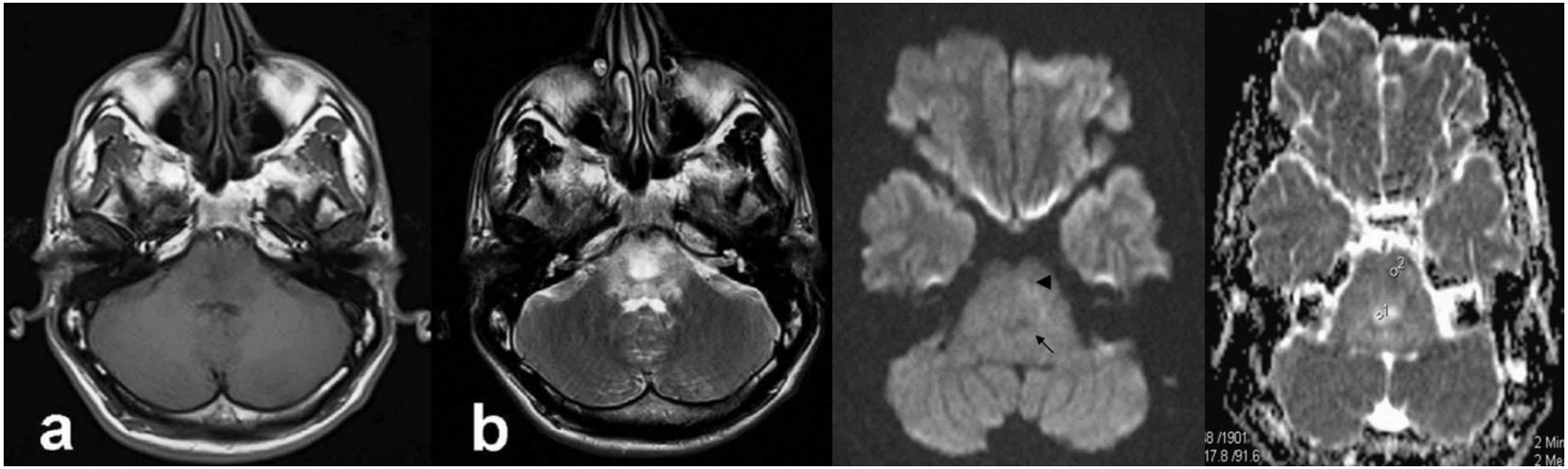
- Vasculitis exclusively in the CNS
- Highly variable MR imaging findings
  - Normal to diffusely abnormal
- Simultaneous enhancement of brain and spinal cord lesions and of perivascular spaces, at the onset of the disease, which resolves during follow-up



Campi A, et al. Primary angiitis of the central nervous system: serial MRI of brain and spinal cord - Springer. Neuroradiology. 2001.

Neuro-Behcet disease

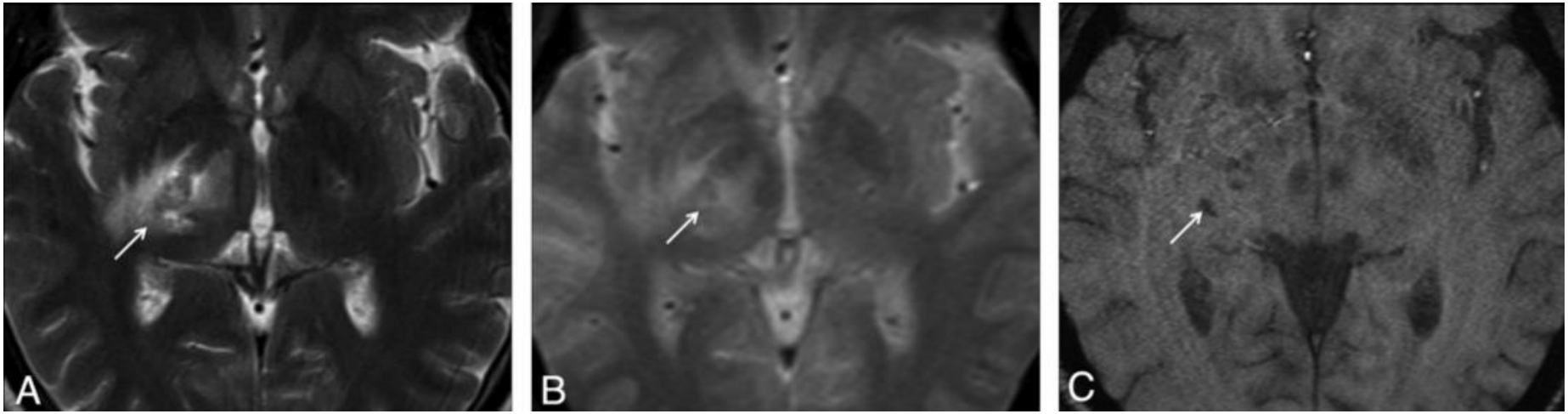
- Multisystemic inflammatory disease of unknown cause
- Vasculitis with perivascular infiltration of lymphocytes and neutrophils
- Triad of uveitis, oral ulcers, and genital ulcers
- CNS affected in 4%-49%
  - most commonly in the brainstem, basal ganglia, and thalamus
  - less commonly in the cerebral white matter and cervicothoracic spinal cord



Yildiz S, et al. Different diffusion-weighted MRI findings in brainstem neuro-Behçet's disease. BMJ Case Rep. 2013.

Neuro-Behcet disease

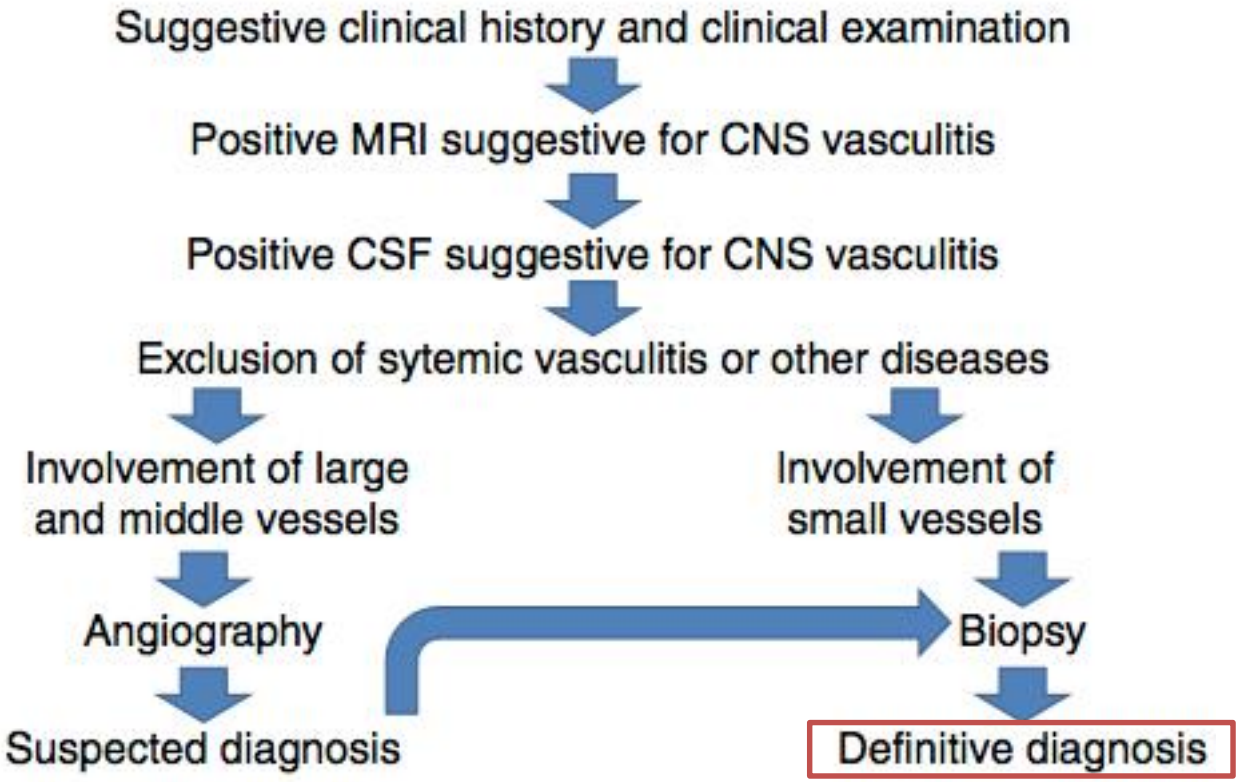
- Most of the lesions in parenchymal NBD were found to be hemorrhagic with SWI



Albayram S, et al. Evaluation of parenchymal neuro-behçet disease by using susceptibility-weighted imaging. American Journal of Neuroradiology. 2011 Jun;32(6):1050–5.

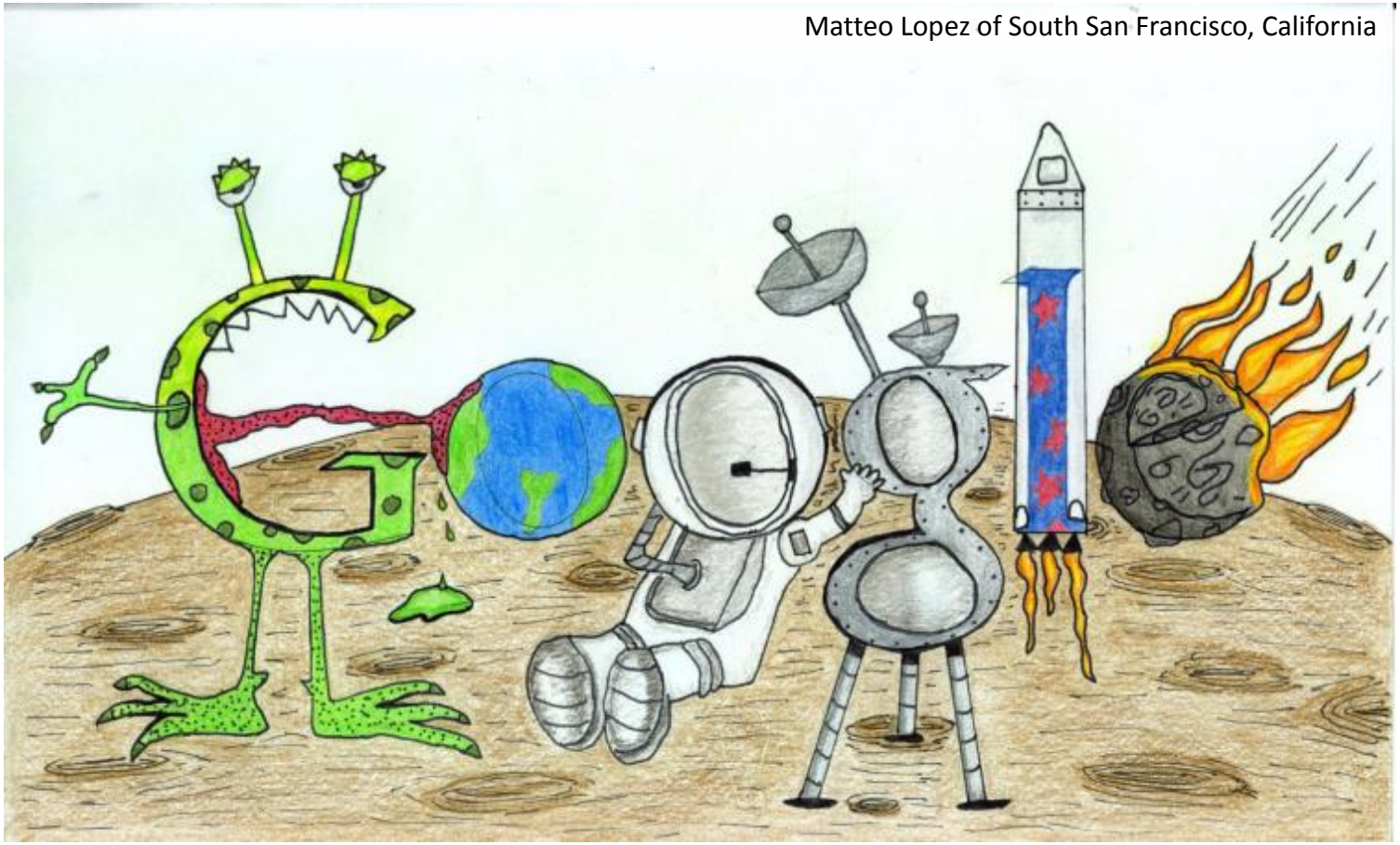
Clinical & Experimental Immunology
The Journal of Translational Immunology

Cerebral vasculitis in adults: what are the steps in order to establish the diagnosis? Red flags and pitfalls



Berlit P, et al. Cerebral vasculitis in adults: what are the steps in order to establish the diagnosis? Red flags and pitfalls. Clin. Exp. Immunol. 2014 Mar;175(3):419–24.

Kinkaku-ji, Temple of the Golden Pavilion



Masafumi KANOTO

Masafumi Kanoto

1. Kanoto, M., et al., Impact of superselective transarterial infusion therapy of high-dose cisplatin on maxillary cancer with orbital invasion. AJNR Am J Neuroradiol, 2010. 31(8): p. 1390-4.
2. Kuroda, Y., et al., Inverse-direction scanning improves the image quality of whole carotid CT angiography with 64-MDCT. Eur J Radiol, 2011. 80(3): p. 749-54.
3. Kanoto, M., et al., Focal deformity of the cranial nerves observed on multislice motion-sensitized driven equilibrium (MSDE) in patients with neurovascular compression. J Comput Assist Tomogr, 2012. 36(1): p. 121-4.
4. Hosoya, T., et al., Floating dural sac sign is a sensitive magnetic resonance imaging finding of spinal cerebrospinal fluid leakage. Neurol Med Chir (Tokyo), 2013. 53(4): p. 207-12.
5. Kanoto, M., et al., Delineation of malignant glioma by turbo spin echo multislice motion-sensitized driven-equilibrium (TSE-MSDE) with gadolinium-based contrast media: a case report. Magn Reson Imaging, 2013. 31(7): p. 1251-3.
6. Kanoto, M., et al., Brain stem and cerebellar atrophy in chronic progressive neuro-Behcet's disease. Eur J Radiol, 2013. 82(1): p. 146-50.
7. Kanoto, M., et al., Visualization of the trochlear nerve in the cistern with use of high-resolution turbo spin-echo multisection motion-sensitized driven equilibrium. AJNR Am J Neuroradiol, 2013. 34(7): p. 1434-7.
8. Kanoto, M., et al., Radiological Image Features of the Atypical Teratoid/Rhabdoid Tumor in Adults: A Systematic Review. Clin Neuroradiol, 2014.
9. Konno, Y., et al., Clinical Significance of Mammillary Body Enhancement in Wernicke Encephalopathy: Report of 2 Cases and Review of the Literature. Magn Reson Med Sci, 2014.

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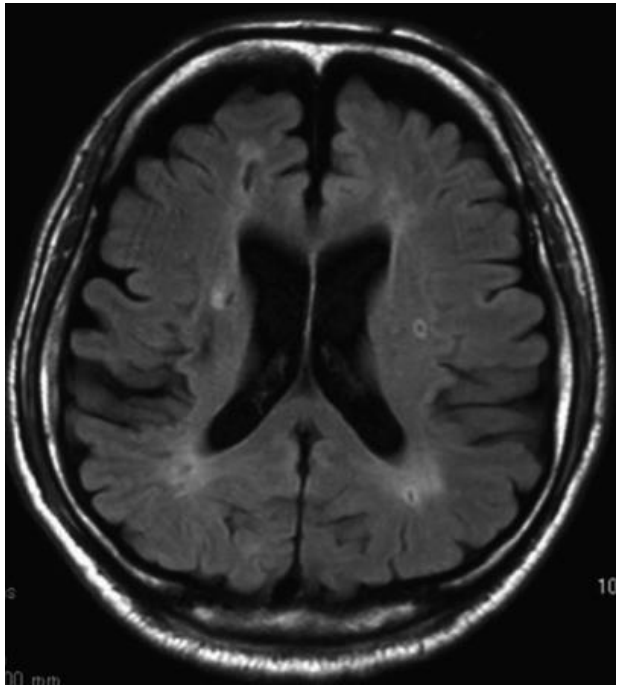
European Journal of Radiology

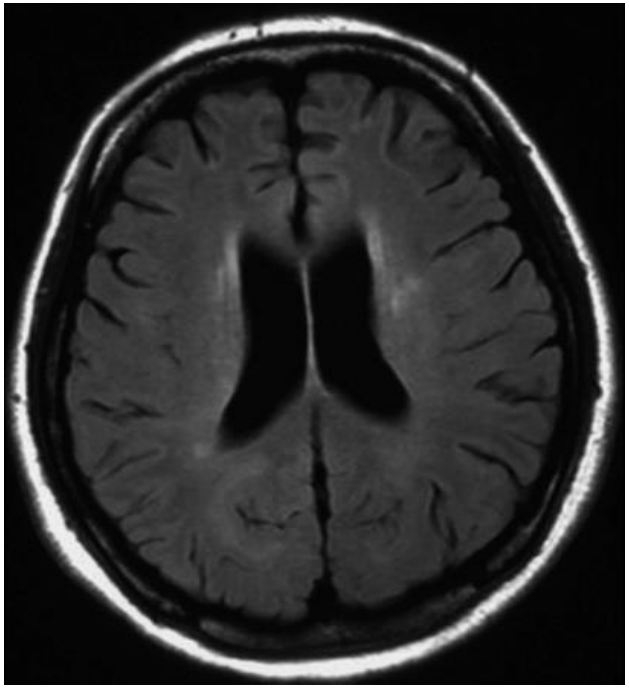
journal homepage: www.elsevier.com/locate/ejrad

Brain stem and cerebellar atrophy in chronic progressive neuro-Behçet's disease

Masafumi Kanoto*, Takaaki Hosoya[1], Yuuki Toyoguchi[1], Atsuko Oda[1]

Department of Diagnostic Radiology, Faculty of Medicine, Yamagata University, Iida-Nishi 2-2-2, 990-9585 Yamagata, Japan





Brain stem and cerebellar atrophy in chronic progressive neuro-Behçet's disease

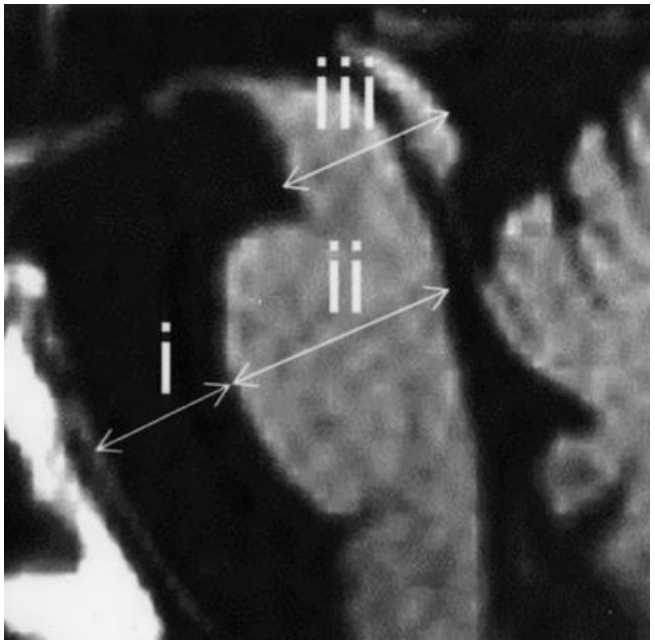
A B S T R A C T

Purpose: Chronic progressive neuro-Behçet's disease (CPNBD) resembles multiple sclerosis (MS) on patient background and image findings, and therefore is difficult to diagnose. The purpose is to identify the characteristic magnetic resonance imaging (MRI) findings of CPNBD and to clarify the differences between the MRI findings of CPNBD and those of MS.

Materials and methods: The subjects consist of a CPNBD group ($n = 4$; 1 male and 3 females; mean age, 51 y.o.), a MS group ($n = 19$; 3 males and 16 females; mean age, 45 y.o.) and a normal control group ($n = 23$; 10 males and 13 females; mean age, 45 y.o.). Brain stem atrophy, cerebellar atrophy, and leukoencephalopathy were retrospectively evaluated in each subjects. In middle sagittal brain MR images, the prepontine distance was measured as an indirect index of brain stem and cerebellar atrophy and the pontine and mesencephalic distance was measured as a direct index of brain stem atrophy. These indexes were statistically analyzed.

Results: Brain stem atrophy, cerebellar atrophy, and leukoencephalopathy were seen in all CPNBD cases. Prepontine distance was significantly different between the CPNBD group and the MS group ($p < 0.05$), and between the CPNBD group and the normal control group ($p < 0.001$). Pontine and mesencephalic distance were significantly different between the CPNBD group and the MS group ($p < 0.001$, $p < 0.01$ respectively), and between the CPNBD group and the normal control group ($p < 0.001$).

Conclusions: Chronic progressive neuro-Behçet's disease should be considered in patients with brain stem and cerebellar atrophy in addition to leukoencephalopathy similar to that seen in multiple sclerosis.



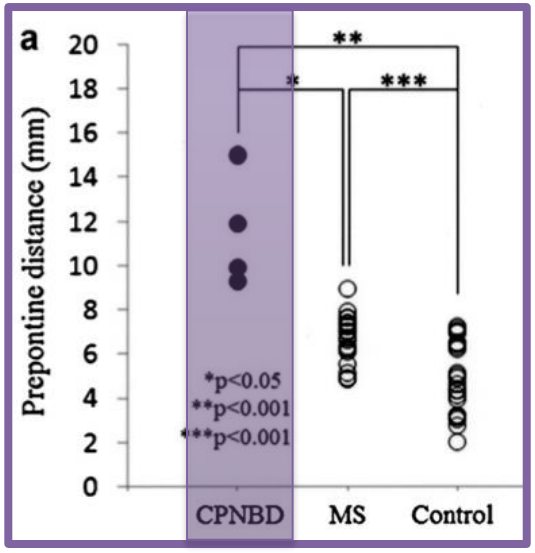
(i) prepontine distance (PPD)

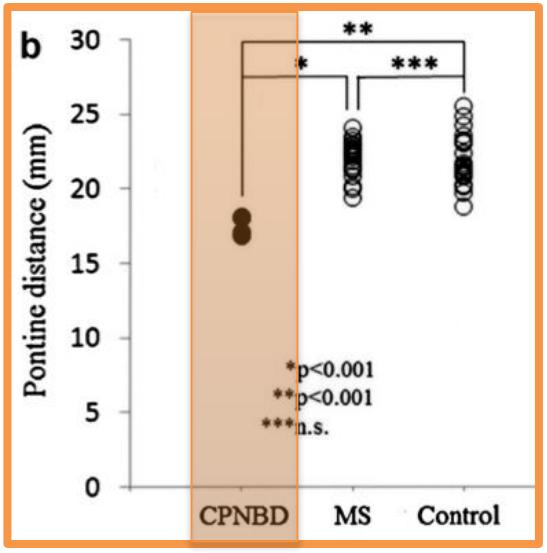
- index of cerebellar and brain stem atrophy

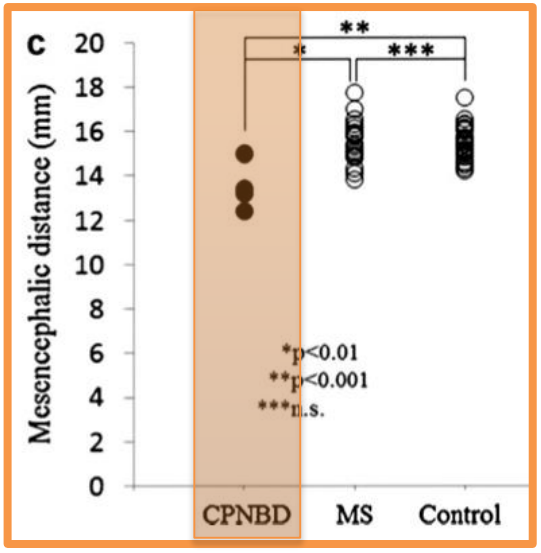
(i) pontine distance (PD)

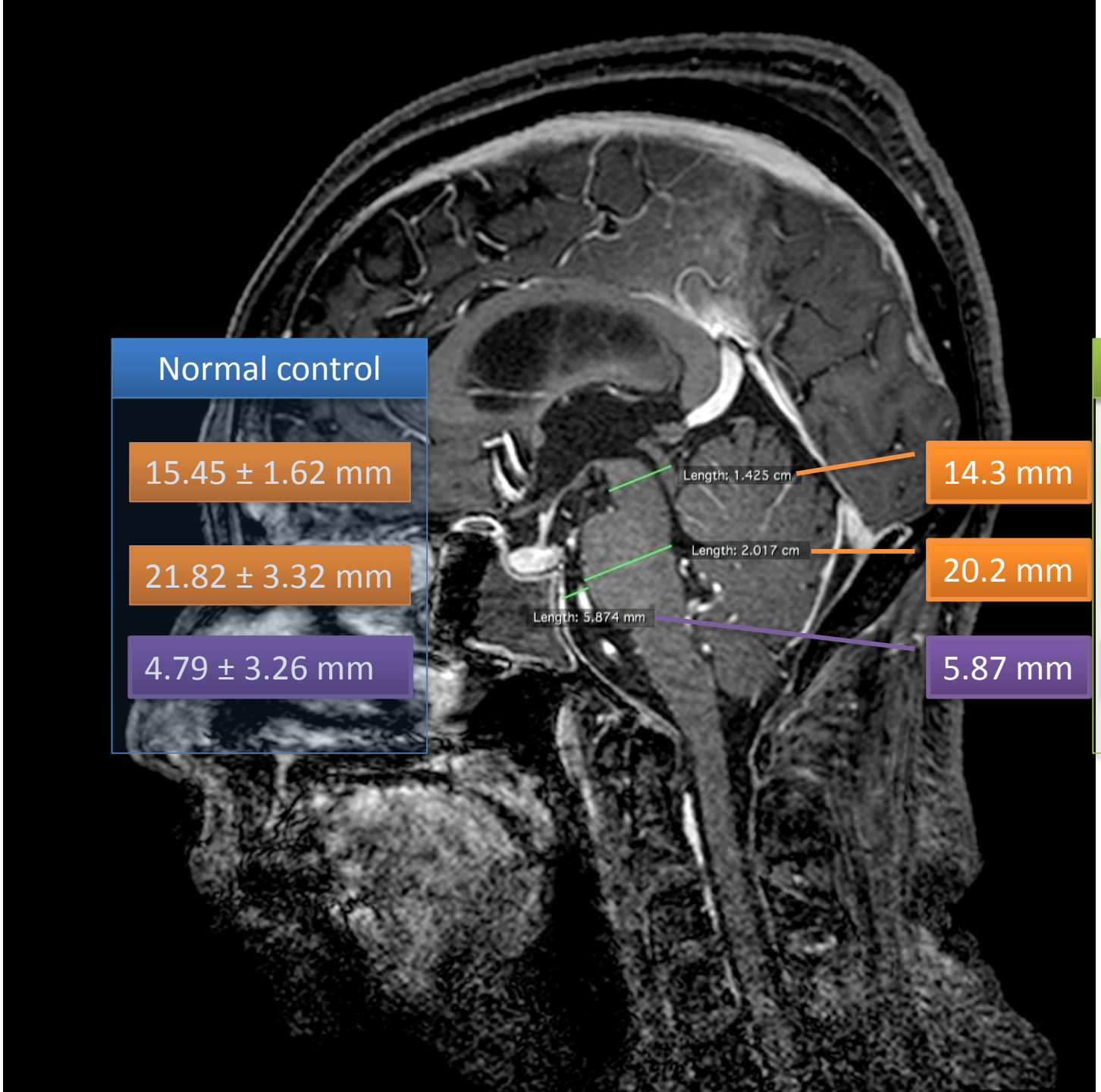
(ii) mesencephalic distance (MD)

- indexes for the evaluation of brain stem atrophy







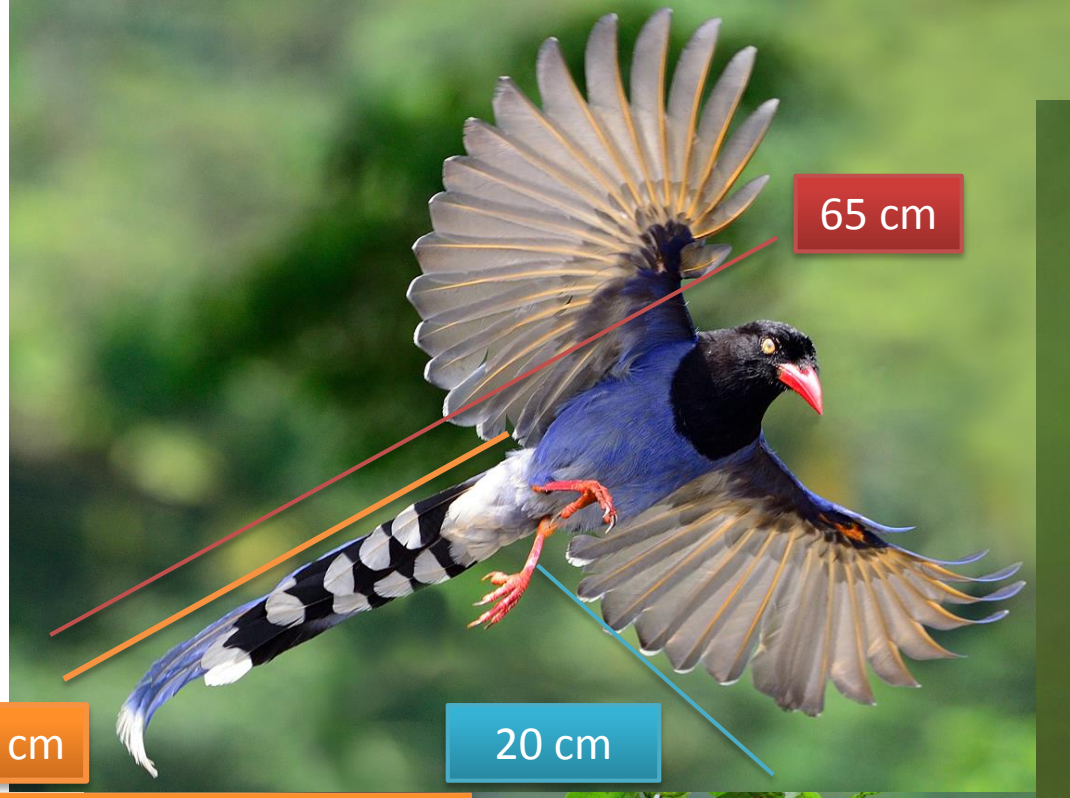


	Normal control	CPNBD
	15.45 ± 1.62 mm	13.50 ± 2.18 mm
14.3 mm		
	21.82 ± 3.32 mm	17.50 ± 1.30 mm
20.2 mm		
	4.79 ± 3.26 mm	11.53 ± 5.14 mm
5.87 mm		

Diagnosis

Neuro-Behcet disease

Encephalomyelitis of unknown origin



Taiwan blue Magpie

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謝謝