CLINICAL CASE - TEST YOURSELF | Neuro/Head and Neck Radiology

Patient with changes in mental status and motor dysfunction

Despoina Voultsinou[1], Triantafillos Gerukis[1], Panos Prassopoulos[2]

[1]*Department of Radiology, G.P.H "G. Papanikolaou", Thessaloniki, Greece*
[2]*Department of Radiology, AHEPA University Hospital, Aristotle University, Thessaloniki, Greece*

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PART A

A 29-years old male was evaluated for difficulty in initiation and control movement and changes in mental status. Medical history was unremarkable. Physical examination was indicative of basal ganglia dysfunction. Blood test results were within normal limits. A routine brain MRI imaging examination was performed (**Figs. 1-4**).

CORRESPONDING AUTHOR, GUARANTOR

Despoina Voultsinou, Department of Radiology, G.P.H "G. Papanikolaou", Papanikolaou Ave, 57010, Thessaloniki, Greece, Email: divou2@yahoo.gr

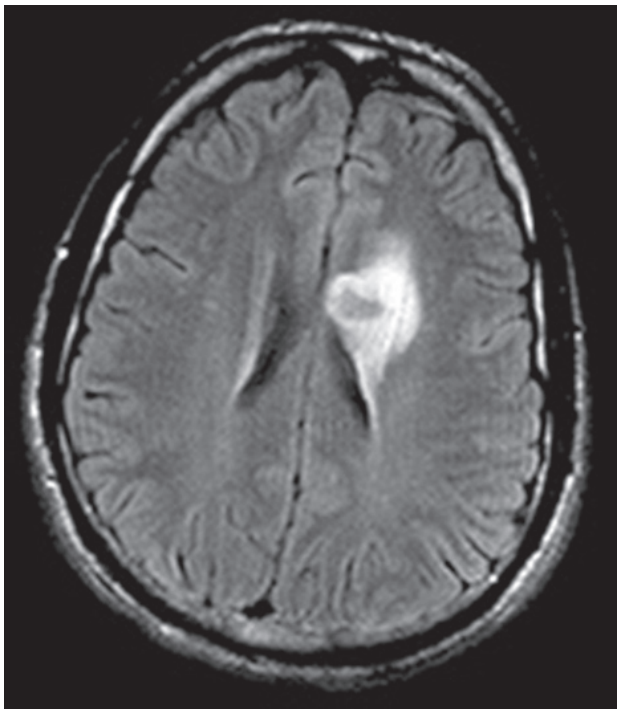

Fig. 1. *Axial brain MRI image at the level of the lateral ventricles, FLAIR sequence.*

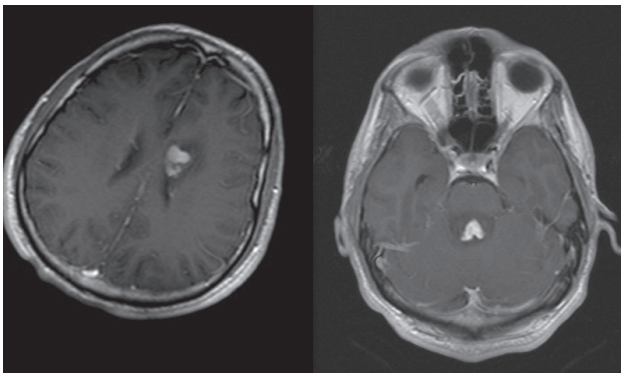

Fig. 2. *Axial post-contrast MRI images, at the level of the lateral ventricles and pons.*

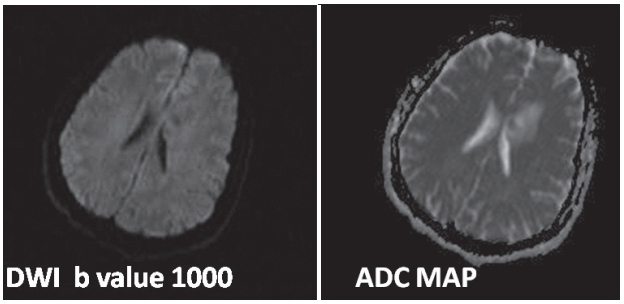

Fig. 3. *Diffusion Weighted Image (DWI) of b-value 1000 with corresponding Apparent Diffusion Coefficient (ADC) map.*

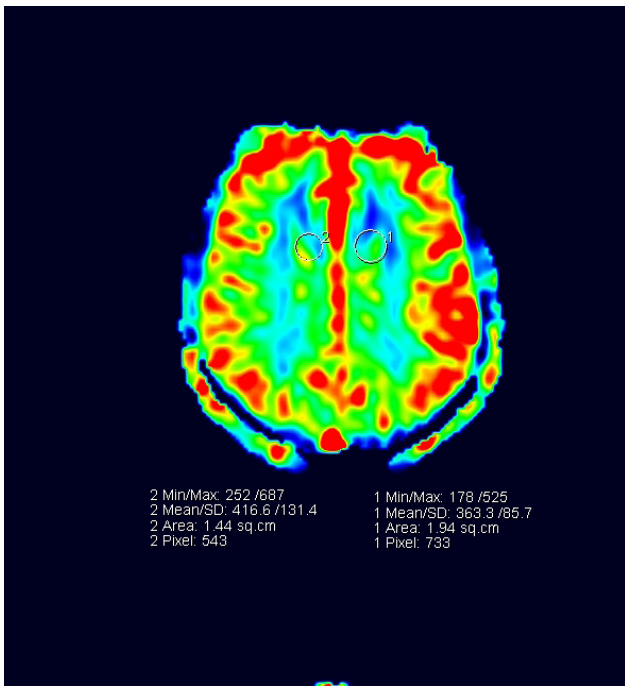

Fig. 4. *Perfusion Weighted Image (PWI), Cerebral Blood Volume (CBV) map.*

PART B

Diagnosis: Tumefactive Multiple Sclerosis

Imaging workup of the patient shows that on FLAIR MRI (**Fig. 1**) the intermediate signal intensity lesion demonstrates restricted peritumoural oedema and exerting minimal mass effect, same imaging features with the second lesion at the level of the pons. Both lesions illustrate intense homogeneous contrast enhancement (**Fig. 2**). The differential diagnosis includes a wide spectrum of neoplastic and non-neoplastic lesions. The restricted peritumoral oedema and the intense homogeneous contrast enhancement make the diagnosis of multicentric glioblastoma and cerebral metastases less possible. Sharp lesion margins exclude the rare case of brain gliomatosis, that usually develops in a diffuse pattern. The location and pattern of distribution of the lesions makes the diagnosis of secondary Brain Lymphoma (BL) possible [1]. However, there was no evidence of primary lymphoma elsewhere in the body in our patient. Whilst Primary Cerebral Brain Lymphoma (PCNSL) is presented as a solitary, well defined, round or oval lesion, it may also be multifocal, especially in immunosuppressed patients [2]. AIDS or any other cause of immunosuppression was excluded in our patient.

Non-neoplastic conditions such as vascular disease (brain infarct and vasculitis), infectious (tuberculosis, fungus, bacterial abscess) and inflammatory disease were excluded due to lesion distribution and enhancement pattern as well as the laboratory profile of the patient. Medical history was free (no trauma or medical intervention was mentioned). Demyelinating disease, namely Tumefactive Multiple Sclerosis (TMS), was also included in the differential diagnosis. Further imaging workup was performed in order to discriminate between PCNSL and TMS [3, 4].

Further imaging workup included Diffusion Weighted Image (DWI) and Perfusion Weighted Image (PWI). On DWI the lesions demonstrated increased signal intensity and mildly increased Apparent Diffusion Coefficient (ADC) (**Fig. 3**), T2- shine through phaenomenon, without restriction in diffusion. The mean minimum ADC value measured at the core of the larger lesion was 1.09 X 10^{-6} mm/sec^2, a borderline value between malignant and non-malignant brain lesions [5, 6].

Relative Cerebral Blood Volume (rCBV) ratio within the larger lesion (**Fig. 4**) as well as within the smaller lesion were significantly low. In TMS lower rCBV values have been recorded in comparison to CNSL. The rCBV values difference between tumefactive demyelinating lesions (TDL) and brain lymphomas are less pronounced but are statistically significant [7, 8]. However this finding may be inconclusive in some cases due to variability of different brain lymphomas subtypes [7, 8].

The considerable low rCBV and the mild increase in ADC values raised the possibility of TMS. A short echo time (TE=30 ms) in Magnetic Resonance Spectroscopy (MRS) and a TE of 135 ms (**Fig. 5**) were performed in the larger tumour-like lesion. The size of the second lesion was very small for accurate measurements. MRS showed elevation of myoinositol at 3.56 ppm, glutamate/glutamine (Glx) peaks at 2.1-2.5 ppm, marked elevation of lactate/lipid peaks at 0.8-1.3 ppm and mild choline elevation values compatible with demyelination [4]. There was also mild decrease of N-acetylaspartate, indicative of axonal pathology and reduced creatine, favouring an acute stage of the lesions [9]. Echo time of TE 135 ms showed increased lipid peaks.

No biopsy [3] was performed, since advanced imaging and laboratory tests were considered diagnostic of TMS with multiple lesions. Laboratory results were positive for the same monoclonal bands at both cerebrospinal fluid and blood serum, indicating the presence of multiple sclerosis. The patient was treated with corticosteroids and on follow up MRI examinations the lesions progressively resolved. Three years later the imaging findings were unremarkable (**Fig. 6**).

TMS is an unusual presentation of multiple sclerosis, seen in 0.1- 2.8% of MS patients [3]. It is a locally aggressive form of demyelination. The imaging appearance [4, 5] of TMS (well circumscribed lesion greater than 2 cm or sometimes a couple of lesions with minimal mass effect and oedema) may mimic neoplasm, abscess or stroke on imaging. Most of these lesions may not progress to multiple sclerosis plaques [6]. Peculiar imaging appearances on MRI, such as "open" or "broken" ring gadolinium enhancement, dilatation of cerebral veins either in or

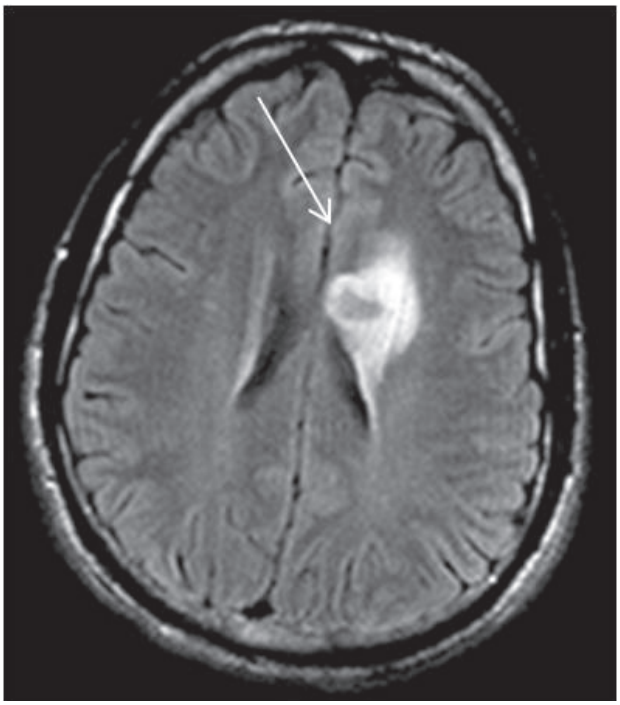

Fig.1. *Axial brain MRI image at the level of the lateral ventricles, FLAIR sequence. The intermediate density lesion (arrow) demonstrates minimal to moderate peritumoural oedema and exerts minimal to moderate mass effect.*

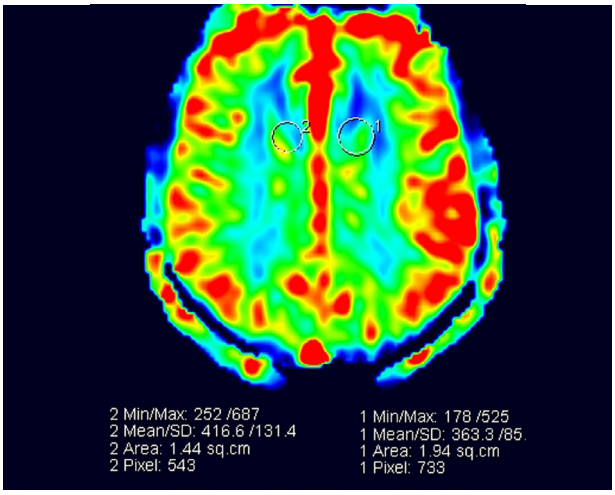

Fig.4. *PWI, CBV map. Regional perfusion measurements depict low rCBV values, even lower than the normal corresponding contralateral white matter (ellipse).*

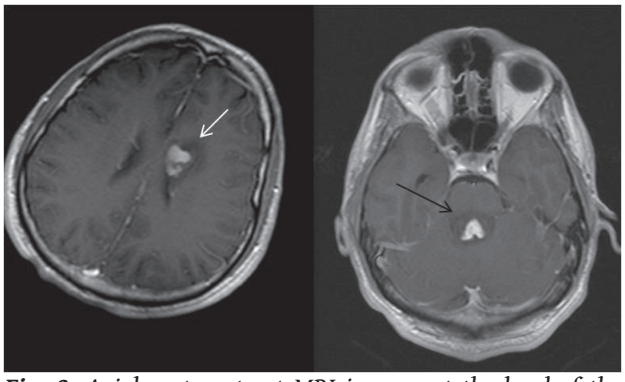

Fig. 2. *Axial post-contrast MRI images, at the level of the lateral ventricles (white arrow) and pons (black arrow). Both lesions show intense contrast medium enhancement.*

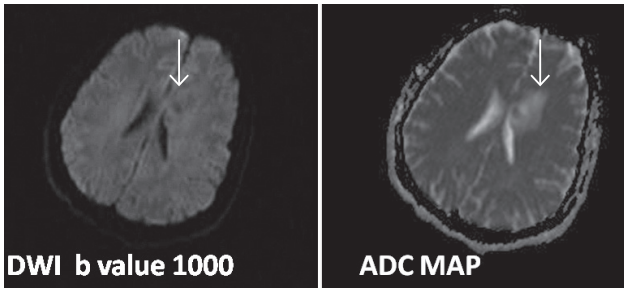

Fig. 3. *DWI of b-value 1000 with corresponding ADC map. DWI shows intermediate signal intensity (arrow on left image) with ADC values in corresponding ADC map, variable (arrow on right image). No restriction in diffusion is noted.*

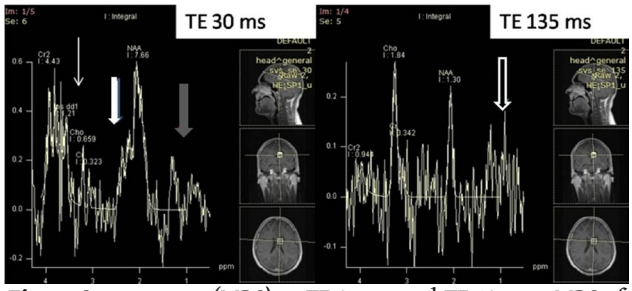

Fig. 5. *Spectroscopy (MRS) at TE 30 ms and TE 135 ms. MRS of the larger lesion at short echo time (TE=30 ms) reveals elevation of myoinositol at 3.56 ppm (thin white arrow), glutamate/glutamine (Glx) peaks at 2.1-2.5 ppm (solid white arrow), marked elevation of lactate /lipid peaks at 0.8-1.3 ppm (solid grey arrow) and at echo time TE 135 ms shows increased lipid peaks (open arrow). Note mild choline elevation and moderate NAA elevation. Cr, creatine; Cho, choline NAA indicates N-acetylaspartate.*

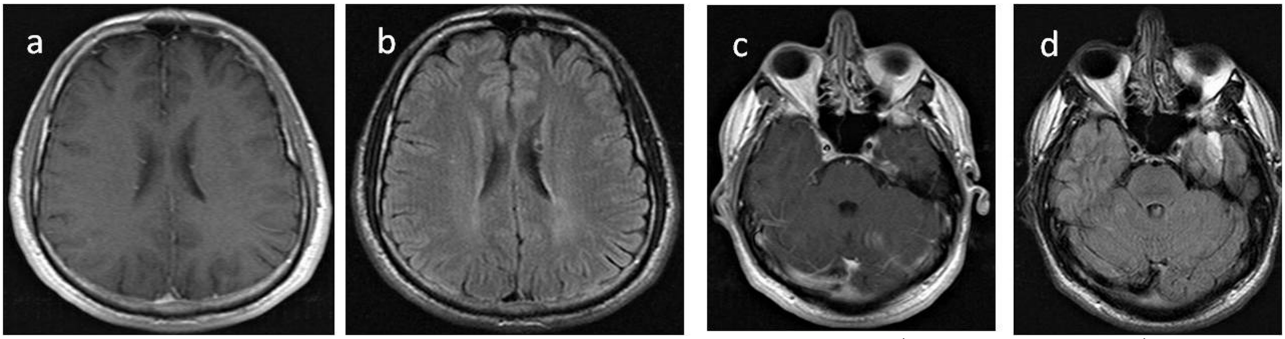

Fig. 6. *Follow up MRI examination after 3 years. At the level of the lateral ventricles* ***a)*** *Postgadolinium and* ***b)*** *FLAIR image. At the level of the pons at* ***c)*** *Postgadolinium and* ***d)*** *FLAIR image. Complete resolution of imaging findings is noted.*

around these tumefactive lesions may suggest the diagnosis of TMS [8-10], but were not present in our patient. Since diagnosis of TMS is difficult even on histology [10], a close follow up imaging strategy has been proposed as a tool to confirm diagnosis and avoid biopsy, with interval improvement of these lesions on MRI following steroid therapy [3]. Many unnecessary surgical operations and unnecessary complementary chemotherapy therapies due to false diagnosis in case of TMS have been reported.

TMS raises a diagnostic dilemma to neurosurgeons, radiologists and pathologists, since intracranial tumours and other pathologies share similar features. Minimal peritumoural oedema and mass effect, open-ring of enhancement, increased ADC values, centrally dilated veins within the lesion with decreased rCBV values and MR spectroscopy constitute a great aid in the diagnosis of TMS. **R**

Conflict of interest

The authors declared no conflicts of interest.

KEY WORDS

Central Nervous System, Magnetic Resonance Imaging, Perfusion Imaging, Diffusion, Lymphoma, Multiple Sclerosis

REFERENCES

1. Partovi S, Karimi S, Lyo JK, et al. Multimodality imaging of primary CNS lymphoma in immunocompetent patients. *Br J Radiol* 2014; 87(1036): 20130684.
2. Brandsma D, Bromberg JEC. Primary CNS lymphoma in HIV infection. *Handbook of Clinical Neurology* 2018; 152: 177-186.
3. Abddoli M, Freedman MS. Neuro-oncology dilemma: tumor or tumefactive demyelinating lesion. *Mult Scler Relat Disord* 2015; 4(6): 555-566.
4. Lin X, Yu WY, Liauw L, et al. Clinicoradiologic Features distinguish tumefactive multiple sclerosis form CNS neoplasms. *Neurol Clin Pract* 2017; 7(1): 53-64.
5. Hyland M, Bermel RA, Cohen JA, et al. Restricted diffusion preceding gadolinium enhancement in large or tumefactive demyelinating lesions. *Neurol Clin practice* 2013; 3(1): 15–21.
6. Mabray MC, Cohen BA, Villanueva-Meyer JE, et al. Performance of apparent Diffusion Coefficient Values and conventional MRI features in differentiating tumefactive demyelinating lesions from primary brain neoplasms. *AJR Am J Roentgenol* 2015; 205: 1075-1085.
7. Suh CH, Kim HS, Jung SC, et al. MRI findings in tumefactive demyelinating lesions: A systematic review and Meta –Analysis. *AJNR* 2018; (9): 1643-1649.
8. Parks NE, Bhan V, Shankar JJ. Perfusion imaging of tumefactive demyelinating lesions compared to high grade gliomas. *Can J Neurol* Sci 2016; 43: 316-318.
9. Cianfoni A, Niku S, Imbesi SG. Metabolite findings in tumefactive demyelinating lesions utilizing Short Echo Time Proton Magnetic Resonance Spectroscopy. *AJNR* 2007; (2): 272-277.
10. Algahtani H, Shirah B, Alassiri A. Tumefactive demyelinating lesions: A comprehensive review. *Mult Scler Disod* 2017; 72-79.

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