

CASE REPORT

Dominant Hemisphere and Upper Cervical Cord Tumefactive Multiple Sclerosis in a Nigerian Teenager Initially Misdiagnosed and Managed

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DISCLOSURE

Memfys Hospital for Neurosurgery's Management gave waiver for the follow-up MRI.

A portion of this work was presented at the WFNS meeting (Istanbul 2017)

ABSTRACT

A 13-year old right handed Nigerian girl presented with eleven weeks history of progressive visual loss, three weeks right hemi-body weakness which progressed to quadriplegia, dysphasia, severe headache, tonic-clonic seizures, neck pain, vomiting and fever.

Examination revealed Glasgow Coma Score (E4M6V2), expressive dysphasia and depressed mood. Visual Acuity was counting finger bilaterally. Neck was supple. She had global hypotonia, grade 3 hyper-reflexia and extensor Babinski bilaterally. Power was 0/5 in the right upper limb and right lower limb, 2/5 in the left lower limb and 4/5 in the left upper limb. Other examinations were unremarkable.

Contrast enhanced brain MRI revealed three cystic ring enhancing masses at left fronto-parieto-occipital region, associated oedema, midline shift and C2-C4 intramedullary mass with cord oedema. The ring enhancement was incomplete towards the cortex.

Following initial suspicions of cystic brain tumour with spinal cord metastasis, she had decompression biopsy. Histology was inflammatory lesion, clinically assumed to be from brain abscess. Failure of antibiotic treatment and the deteriorating neurology prompted mini-craniectomy and biopsy of the cyst wall. Final diagnosis was tumefactive multiple sclerosis. She was successfully managed with methylprednisolone. Follow-up clinical condition has been satisfactory.

Keywords: TMS, cranial masses, multiple sclerosis, visual loss

INTRODUCTION

Tumefactive multiple sclerosis (TMS) is an atypical form of multiple sclerosis characterized by large lesion size with cerebral oedema and minimum mass effect.^{1,2} It is rare especially in the paediatric population and poses serious diagnostic and treatment challenge.^{3,4} We report a very rare case of TMS in a Nigerian, involving the dominant hemisphere and cervical spinal cord, which was initially misdiagnosed.

Case Report

A 13-year old right handed school girl who brought with eleven weeks history of progressive visual loss. She also had progressive right sided hemi-body weakness that was noticed three weeks after the onset of the visual complaint, and which gradually progressed to quadriparesis. There were associated dysphasia which progressed to aphasia, intermittent tonic-clonic seizures, neck pain, headache with vomiting and fever. About six months earlier, she had similar visual complaints that necessitated ophthalmologist review. Brain computed tomography scan done six months earlier did not reveal any structural lesion and the visual symptoms improved eventually.

Examination revealed an admission Glasgow Coma Score of 12/15 (E4M6V2) with expressive dysphasia. The visual acuity was counting fingers bilaterally. There were no signs of meningeal irritation. She had grade 3 deep tendon reflex globally with extensor plantar reflex bilaterally. She had right hemiplegia. Power was grade 4 in the left upper extremity and 2 in the left lower extremity.

Brain magnetic resonance imaging (MRI) done at presentation revealed three cystic

open ring enhancing masses with the base of the ring abutting the lateral ventricles in the left fronto-parieto-occipital region measuring 45x52x54mm, 34x43x58mm and 54x43x42mm, with associated oedema and midline shift. Cervical spine MRI revealed C2-C4 intramedullary mass with cord oedema (Figure 1). The erythrocyte sedimentation rate and C-reactive protein levels were elevated.

An initial burr hole aspiration of the lesion yielded straw coloured fluid and cytology showed inflammatory cells. A diagnosis of brain infection was then entertained as the most likely diagnosis. Failure of antibiotic treatment and the deteriorating neurology prompted a mini-craniectomy and biopsy of the cyst wall. Findings revealed features in keeping with multiple sclerosis (Figure 3).

She responded to high dose methylprednisolone which was administered at a dose of 800mg for three days and a maintenance dose of 40 mg before it was tailed off and was discharged home. She benefited from physiotherapy and other supportive care.

Follow-up care was continued in the clinic with a remarkable clinical improvement at 6 months. She had mild residual expressive dysphasia with dyscalculia. The visual acuity was 6/12 bilaterally. She regained full power in the left upper and lower limbs with grade 4 in the right upper and lower limbs. The follow-up brain and cervical spine MRI done at six months follow-up visit were satisfactory with resolution of the structural lesions (Figure 2).

Figure 1. Axial and sagittal Brain MRI of the patient done at presentation. There are hypointense areas (*) in the left fronto-parieto-occipital region (image A) with associated oedema and midline shift (*image B). The post contrast T1W image revealed three cystic open ring enhancing masses (image C). Images D and E revealed coexisting C2-C4 intramedullary lesion with cord oedema (*).

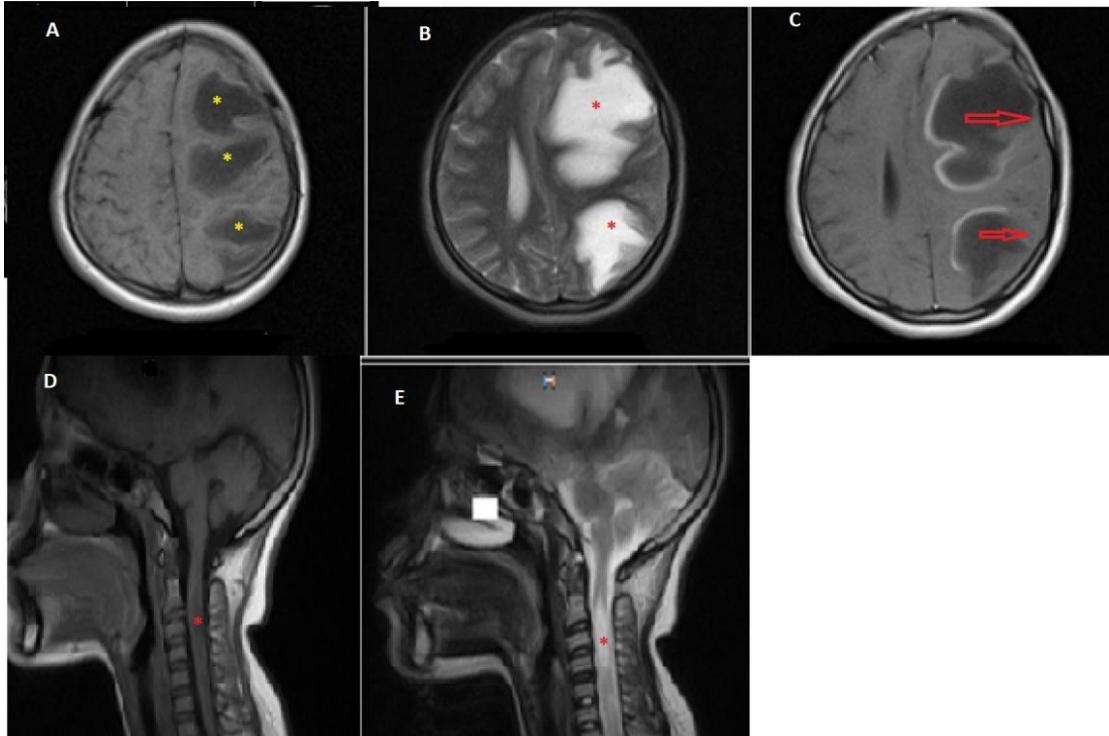


Figure 2: Follow-up Brain MRI done at time of patient discharge from hospital (images A and C) and at sixth month follow-up visit (images B and D) showing resolution of the structural lesions lesion. (See arrows)

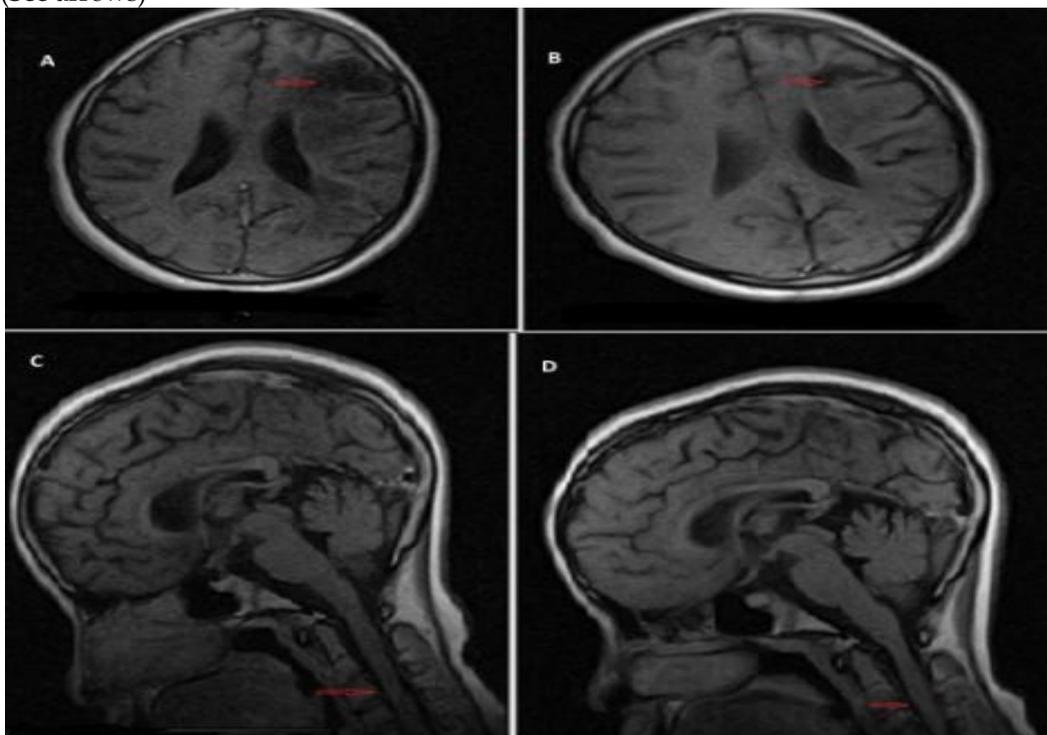
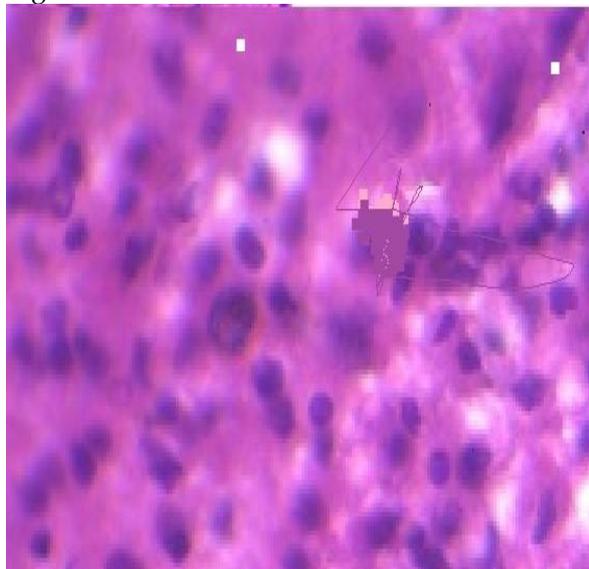


Figure 3. Post-operative Histopathology slide. Findings were inflammatory cystic or cavitory lesions with inflammation and focal white cell infiltrates as well as reactive gliosis and angiogenesis in keeping with multiple degenerative lesion.



DISCUSSION

Tumefactive multiple sclerosis is an unusual form of multiple sclerosis which can be misdiagnosed when the index of suspicion is not high. It can occur as the first attack or in a patient with a previously established multiple sclerosis.^{5,6} MRI is an invaluable tool for diagnosis. The most important diagnostic clue for TMS in the MRI is the incomplete ring enhancement.^{1,7} Other features include mild oedema or mass effect, T2Wi hypo intense rim and venular enhancement.^{2,4,8} However, a high index of suspicion is required since the MRI finding may still easily be dismissed as a glioma or an intracranial abscess. It is important to distinguish TMS from these potential differential diagnoses in order to get the treatment plan right. When TMS is misdiagnosed as high grade gliomas, administration of adjuvant radiotherapy may hasten the progression of TMS.⁹ Also comparing a CT scan attenuation of the MRI enhancing component of the lesion with

cortical grey matter or basal ganglia may help to differentiate TMS from gliomas since hypo-attenuation of the lesion relative to the grey matter supports a diagnosis of TMS.² Magnetic resonance spectroscopy and as well as cerebrospinal fluid oligoclonal band where available, may also be useful for diagnosis.^{10,11}

Neurosurgeons need to understand and appreciate these clinical and neuroimaging findings so as to hasten appropriate diagnosis and reduce unwarranted surgical or medical interventions which the index case was subjected to. In some cases, biopsy of the lesion is inevitable in order to clinch the diagnosis but could be misleading in some other cases with poor index of suspicion like the index case.^{3,4,10,12}

TMS is rare in children and is without any clear gender predilection.^{2,4} Also some of these children with TMS are more at risk of multiple sclerosis in the future.⁷ Solitary lesions are common but multiple lesions have been documented in up to 70% of patients especially among children.^{6,7} Clinical presentations are usually polysymptomatic and depend largely on the location of the lesion. However, motor and cognitive symptoms predominate.^{2,6,7} The location of the lesion in the dominant hemisphere of the index patient contributed greatly to the morbidity. Quadriparesis as noted in the index patient can be attributed to the cervical cord involvement.

The common sites of predilection for TMS are parietal, fronto-parietal and infratentorial.⁸ The involvement of spinal cord and optic nerves in the index patient underscores the importance of evaluating these areas for demyelination.⁵ However, the cervical spinal cord is the most common part of the spinal cord involved though there has been report of thoracic cord involvement.⁹

TMS is primarily an inflammatory lesion and the mainstay of treatment is steroids and other anti-inflammatory drugs including immunosuppressants which may reduce the frequency and severity of relapses. The role of surgery is limited and probably necessary when biopsy is needed to distinguish TMS from other differential diagnosis like gliomas. Response to methylprednisolone in the index case was remarkable. Some patients may recover without neurological deficit and with disappearance of lesions radiologically.¹⁰

TMS has variable outcomes depending on the extent of the demyelination. Although TMS is mostly monophasic, some cases may recur. However longer follow-up period is needed to remark on the tendency of recurrence in the index case. The index patient at six months follow-up visit to the outpatient had mild residual hemiparesis. However, she could walk unaided with significant improvement in speech and quality of life.

In conclusion, this rare disease can easily be misdiagnosed when the index of suspicion is low. It poses a serious diagnostic dilemma for the neurosurgeons, radiologists and pathologists. In resource poor areas, the finding of open ring enhancement in post contrast MRI scan is a reliable diagnostic guide and may prevent unnecessary extensive neurosurgical interventions.

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