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Case Report

Delayed Recovery of Bilateral Supplementary Motor Area Syndrome After the Resection of a Central Neurocytoma

Alasdair FitzGerald[1], Irrum Aneela[1,*], Paula Choszczewski[1], David Summers[2], Chandrasekaran Kaliaperumal[3]

[1]Department of Neurological Rehabilitation, Astley Ainslie Hospital, Edinburgh, SCOTLAND.
[2]Department of Neuroradiology, Royal Infirmary of Edinburgh, Edinburgh, SCOTLAND.
[3]Department of Neurosurgery, Royal Infirmary of Edinburgh, Edinburgh, SCOTLAND.

ABSTRACT

Supplementary Motor Area (SMA) syndrome is a rare complication following brain tumor (localized in the supplementary motor area) removal and is characterized by temporary paralysis and mutism. These symptoms typically resolve within hours to days. This case report challenges the typical presentation of SMA Syndrome. We report a unique case where a patient developed right dense hemiplegia (severe weakness on one side of the body) and mutism after resection of an intraventricular central neurocytoma (a benign tumor within the brain's fluid-filled cavities). This deviates from the usual transient nature of SMA syndrome. This case report delves into the extended duration of these post-surgical symptoms in this patient. It compares the case with existing literature on SMA syndrome to understand the reasons behind this atypical presentation. By analysing similar cases, we aim to shed light on potential factors that might influence the severity and duration of recovery in SMA syndrome. This case highlights the importance of considering variations in SMA syndrome presentation. It emphasizes the need for further research to improve our understanding of the factors influencing recovery patterns after surgery in this region of the brain.

Keywords: Central neurocytoma, Supplementary motor area syndrome, Frontal aslant tract, Hemiplegia, Mutism.

Correspondence:
Dr. Irrum Aneela (MBBS and MRCP)
Department of Neurological Rehabilitation, Astley Ainslie Hospital, Edinburgh, SCOTLAND.
Email: irrum.aneela@nhs.scot

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INTRODUCTION

The SMA lies within the interhemispheric fissure, in the caudal region of the frontal lobe, just anterior to the primary motor cortex and the precentral sulcus, and superior to the cingulate sulcus.[1-3] Its role is not clearly defined but believed to be involved in learning and sequencing coordinated intentional movement including speech and bimanual dexterity or inhibiting or switching between sequences.[4,5] Surgery in the supplementary motor area can be complicated by contralateral limb paresis and mutism known as SMA syndrome. It is estimated that 10% of glioblastomas and 27% of gliomas arise in this region.[6]

Typically, SMA syndrome symptoms are temporary. In the most recent comprehensive systemic review of 236 cases, complete recovery was noted to occur on average within 45 days, or in 79.7% of cases within a year, with partial recovery in 19.5%[1] with complete persistence of hemiplegia and mutism in only 0.8% or 2 cases.[5] Functional MRI evidence three months following surgery suggests sufficient plasticity of the SMA to restore interhemispheric connectivity to levels identified pre-operatively.[7]

CASE PRESENTATION

A male patient in his 30s presented with a twenty-one-month history of headache followed by transient episodes of photophobia, anosmia, visual aura, intermittent hemiplegia and two episodes of aphasia. Neurological examination at presentation was normal. His symptoms were attributed to possible migraines following neurologist assessment. He was commenced on amitriptyline 10mg at night providing symptomatic relief.

He subsequently experienced a new onset of flashes and white outs of his vision prompting assessment by an optician who diagnosed likely optic neuritis. He was referred for an ophthalmology assessment which revealed bilateral papilledema with end organ damage and a bilateral nasal field defect (left > right).

Urgent Computer Tomography (CT) and subsequent Magnetic Resonance Imaging (MRI) (Figure 1) showed a large, irregular lobulated and coarsely calcified mass lying predominantly within the left lateral ventricle and extending across the foramen of Monro, measuring approximately 45 mm x 66 mm x 33 mm. There was associated hydrocephalus.

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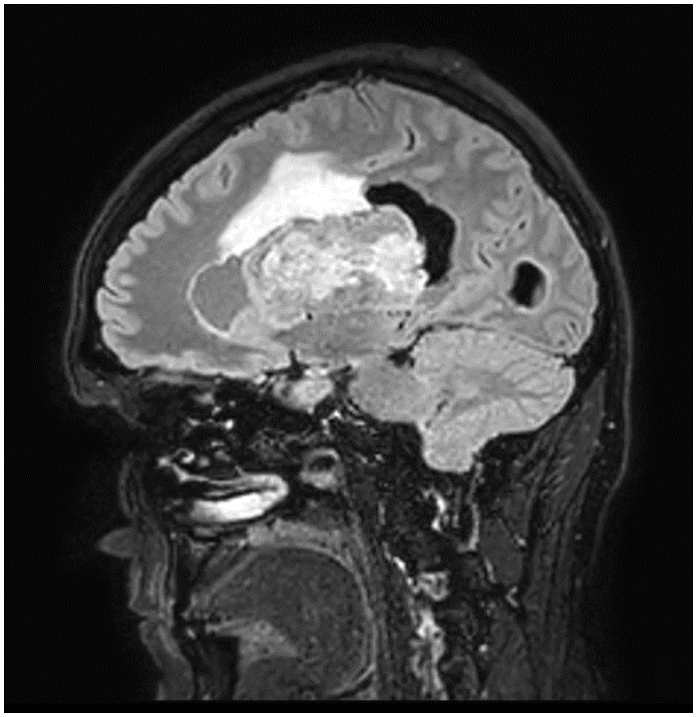

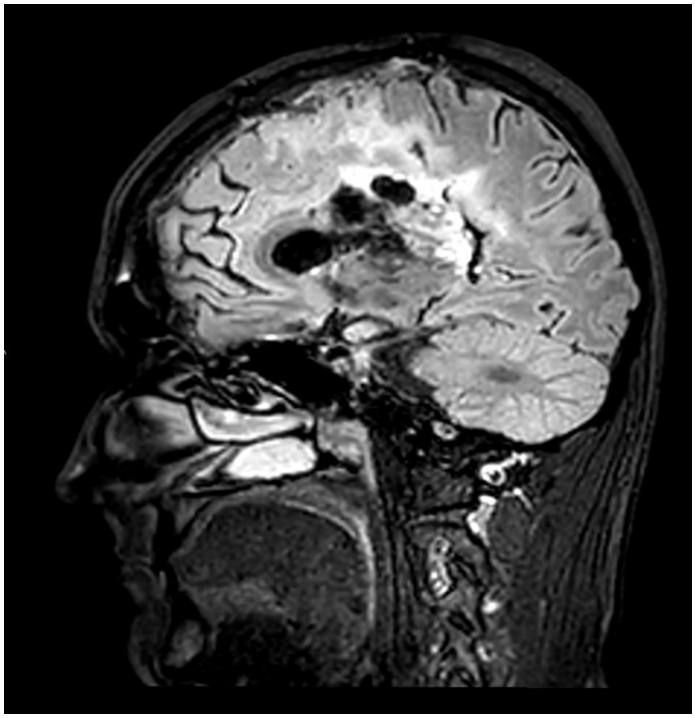

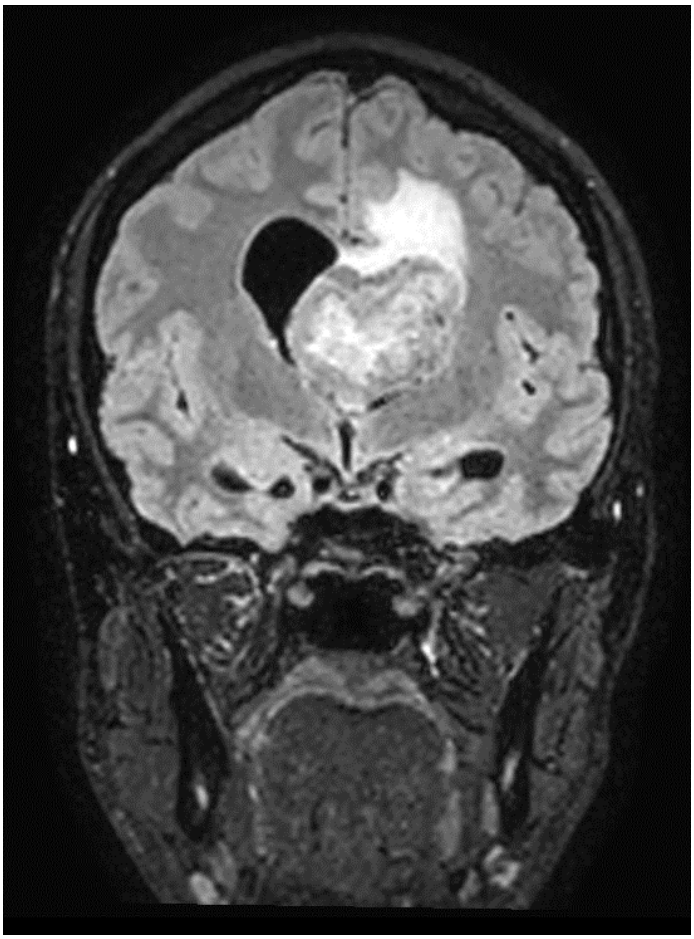

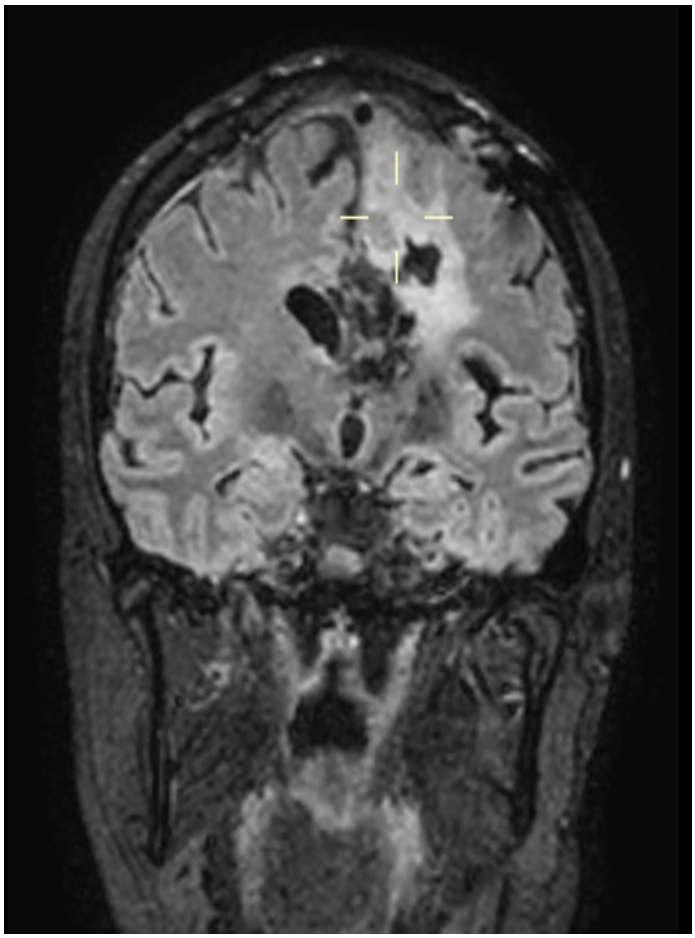

Figures 1: (a) Preoperative sagittal view of tumour (FLAIR) and (b) Preoperative coronal view of tumour (FLAIR) demonstrating 7.0 x 5.0 x 4.3 cm mass within the left lateral ventricle causing local subependymal and adjacent superior white matter oedema, compression of the cerebral peduncles and midbrain with a lateral ventricular hydrocephalus.

Figures 2: (a) (Post-operative Sagittal view of tumour (FLAIR)) & (b) (Post-operative Coronal view of tumour (FLAIR) 12 months post-operation demonstrating left SMA encephalomalacia and callosal injury in region that correlates to crossed frontal aslant tract.

Figures 1a (Preoperative sagittal view of tumour (FLAIR)) & 1b (Preoperative coronal view of tumour (FLAIR)).

Craniotomy and debulking of the tumour was performed. Following neuro-oncology multidisciplinary planning, interhemispheric, transcallosal debulking was performed (post operative imaging suggests 80% of the tumour was removed) and a right frontal External Ventricular Drain (EVD) was inserted to prevent hydrocephalus. Immediately following surgery, he developed SMA syndrome manifesting as dense right hemiplegia, mutism and left deviating gaze palsy. Histopathology of the tumour confirmed the diagnosis of central neurocytoma, WHO grade 2.

One week following surgery, he developed seizures progressing to status epilepticus in association with increased size of left ventricles with significant mass effect and tonsillar descent attributable to blockage of his EVD. The EVD was replaced, and he was commenced on antiepileptic medication.

Outcome and follow up

He was provided with intense neurorehabilitation post operatively. His left gaze palsy resolved completely after a few weeks. At seven months postoperatively, his dysphasia has improved to a negligible extent in that he remains non-verbal and can use gesture but inconsistently. He continues to have complete paralysis of his right upper limb and has spastic paresis of his right lower limb limiting mobility to short distance indoor walking using a tripod stick. In this regard his persisting functional loss is comparable to that experienced in the two cases uniquely described in literature to date.[1,5]

Table 1: Comparison of Reported Cases and Index Case.

	Baker *et al.* – Case 1	Baker *et al.* – Case 2	Index Case
Demographics	62-year-old female	77-year-old male	Caucasian male in mid 30s
Clinical picture	Presented with refractory seizures.	Presented with a two-week history of progressive right-sided weakness.	Twenty-one-month history of headache, photophobia, transient episodes of hemiplegia and aphasia.
Post-operative development of SMA syndrome symptoms.	Immediately postoperatively, the patient was found to have mutism and was unable to move the right side of her body.	Postoperatively, the patient demonstrated worsening right-sided hemiparesis as well as mutism.	Immediately developed aphasia, left deviating gaze palsy and right dense hemiplegia.
Past medical history	Nil described	Nil described	Nil
Pre-op MRI findings	Low-grade glioma of the left frontoparietal region, hyperintensities seen on coronal T2 and on sagittal T2 FLAIR imaging.	Hyperintensities seen on T2 sagittal and coronal imaging suggestive of glioblastoma.	Large tumour in the left intraventricular region with surrounding oedema in the cingulate gyrus and hydrocephalus.
Tumour pathology	Low grade glioma.	Glioblastoma.	Grade II Central Neurocytoma.
Treatment	Awake craniotomy for tumour resection The ventricle was widely entered in both cases suggesting that the corpus callosum was cut.	Nonawake craniotomy for tumour resection The ventricle was widely entered in both cases suggesting that the corpus callosum was cut.	Inter hemispheric trans callosal tumour de bulking with 80% tumour resection.
Post-op MRI findings	Post resection of tumour revealing dissection through the SMA into the corpus callosum.	Postoperative images of T1 with contrast enhancement show dissection into the corpus callosum.	Partial resection of large left intraventricular tumour Persisting ventricular dilation particularly on the left with sulcal effacement.
Outcome	The patient sustained these deficits permanently with no improvement at three years.	The weakness has not improved on long-term follow-up He also remains largely averbal.	Gaze palsy improved after a few weeks Limited improvement in motor function and speech after seven months.

A repeat MRI brain three months post-surgery revealed a reduced tumour size and improvement of his hydrocephalus. He will require radiotherapy for the central neurocytoma, and this has been deferred due to his current clinical condition. There has been minimal improvement after seven months which is quite unusual for SMA syndrome.

Figures 2a (Sagittal view of tumour site one year post operation (FLAIR)) & 2b (Coronal view of tumour site one year post operation (FLAIR)).

DISCUSSION

It's unclear as to what is the cause of SMA syndrome. We speculated that the long pre-presentation history, large tumour volume, involvement of the cingulate gyrus, hydrocephalus and the required neurosurgical interventions were the added contributors to the persistence of symptoms in this case. However there does not appear to be any universal contributory factor in terms of gender, age, tumour pathology or grade, laterality, specific location in terms of SMA versus cingulate gyrus versus superior frontal gyrus, extent of resection or use of intraoperative monitoring[1] nor are there any identified factors that predict severity or persistence. Local ischaemia, retraction injury as a necessary operative process or cerebral oedema are factors.[7] There was no evidence of any ischaemia or haemorrhage noted in the patient's post operative neuroimaging.

A "crossed frontal aslant tract"[5] has been described based on MRI imaging of two patients with permanent SMA syndrome, identifying a pattern of nonhomologous connections through the corpus callosum connecting the premotor area to the contralateral premotor and SMAs, with projections to a previously described frontal aslant tract.[8] Functional MRI studies suggest that recovery appears to be linked to increase in connectivity between the ipsilateral primary sensorimotor cortex and contralateral SMA and premotor areas compared to preoperative connectivity.[7]

While it has already been suggested in a case series of two, that persistence of SMA syndrome appears to be linked to connections through the corpus callosum connecting the ipsilateral primary sensorimotor cortex and the contralateral SMA and premotor areas via a previously described "frontal aslant tract", there is a need to identify further similar cases. We compare (Table 1) our index case to previously reported similar cases.[5] MRI images in our case demonstrate injury in this frontal aslant region, evidence that supports this previous postulation.

CONCLUSION

Although the persistence of SMA syndrome is rare, the possibility should be highlighted as a post operative risk in the consenting process in relevant neurosurgical cases. There is still a need to define what contributes to this persistence. Further case reports or series should provide evidence to support the view that it relates to corpus callosum connections or the frontal aslant tract. Aside from tumour location, there are no identified clinical or demographic risk factors that predict severity or persistence of SMA syndrome.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

AUTHORS' CONTRIBUTIONS

Dr. Irrum Aneela is the corresponding author, drafted the text and obtained the informed consent. Dr. Paula Choszczewski is responsible for drafting of the text. Mr. Chandrasekaran Kaliaperumal is the clinician in charge of the clinical care of the patient and is responsible for critical revision of intellectual content, accuracy and integrity. Dr. Alasdair FitzGerald is responsible for literature search and critical revision of drafts; collating tables and sourcing/editing of images. Dr. David Summers provided and reported on the images.

REFERENCES

1. Palmisciano P, Haider AS, Balasubramanian K, Dadario NB, Robertson FC, Silverstein JW, *et al.* Supplementary Motor Area Syndrome After Brain Tumour Surgery: A Systematic Review. World Neurosurgery 2022;165:160-71.e2.
2. Vergani, F., Lacerda, L., Martino, J., Attems, J., Morris, C., Mitchell, P., *et al.* White matter connections of the supplementary motor area in humans. Journal of Neurology, Neurosurgery, and Psychiatry, 2014;85(12):1377-85. https://doi.org/10.1136/jnnp-2013-307492
3. Ribas, G. C. The cerebral sulci and gyri. Neurosurgical Focus, 2010;28(2):E2. https://doi.org/10.3171/2009.11.FOCUS09245
4. Nachev, P., Kennard, C. & Husain, M. Functional role of the supplementary and pre-supplementary motor areas. Nat Rev Neurosci, 2008;9:856-69. https://doi.org/10.1038/nrn2478
5. Baker, CM, Burks, JD, Briggs, RG, *et al.* The crossed frontal aslant tract: A possible pathway involved in the recovery of supplementary motor area syndrome. Brain Behav. 2018;8:e00926. https://doi.org/10.1002/brb3.926
6. Duffau, H., & Capelle, L. Preferential brain locations of low-grade gliomas. Cancer, 2004;100(12):2622-6. https://doi.org/10.1002/ (ISSN)1097-0142
7. Vassal M, Charroud C, Deverdun J, Le Bars E, Molino F, Bonnetblanc F, *et al.* Recovery of functional connectivity of the sensorimotor network after surgery for diffuse low-grade gliomas involving the supplementary motor area. J Neurosurg. 2017;126(4):1181-90. doi: 10.3171/2016.4. JNS152484. Epub 2016 Jun 17. PMID: 27315027.
8. Catani, M., Dell'acqua, F., Vergani, F. Short frontal lobe connections of the human brain. Cortex, 2012;48(2):273-91. https://doi.org/10.1016/ j.cortex.2011.12.001.

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