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Case Report

Atypical Presentation of Gerstmann Syndrome in a Patient with Encapsulated Right Chronic Subdural Hematoma

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Abstract

Gerstmann syndrome, occasionally dubbed the angular gyrus syndrome is a constellation of clinical features that depict a potential inherent problem affecting the supramarginal and angular gyrus of the dominant left parietal cortex resulting in a partial or full complement of a tetrad of symptoms. It is a syndrome that depicts a derangement in key functions of the parietal lobe of the dominant hemisphere which is the left cerebral cortex in most people. Its cause can arise from a myriad of traumatic, vascular, tumoral, metabolic or infectious cerebral aetiologies. Chronic subdural hematoma is a common condition prevalent in the elderly population and managed by neurosurgeons worldwide. Its pathophysiology involves elaboration of fibrin degradation products, liquefaction of blood clots and new membrane formation due to fibroblast activity on a subdural hematoma over a period of time which warrants surgical treatment to ameliorate clinical features attributable to it. This article highlights the management of a relatively rare case in which features of Gerstmann syndrome were noted in a patient who had radiologic evidence of chronic subdural hematoma in the right non dominant hemisphere, bringing to the fore the importance of detailed neurologic evaluation of patients with parietal lobe lesions in either hemispheres for features of Gerstmann syndrome in order to avoid incidence of missed diagnosis which may influence holistic management of patients presenting symptom and reflect as suboptimal outcome following treatment.

Keywords

Atypical, Presentation, Gerstmann Syndrome, Chronic Subdural

1. Introduction

Chronic subdural hematoma is a collection of blood between the outer cortical surface of the brain and the meningeal layer of the dura. It is usually a result of bleeding from veins bridging the aforementioned surfaces, Dural venous sinuses and emissary veins. [1] It is a relatively common condition managed worldwide by neurologic surgeons. Initially named pachymeningitis hemorrhagica interna, it is fairly common in the elderly population with 63 yrs being the average age of incidence. Risk factors for its occurrence include seizures, head trauma, cerebrospinal fluid shunts, alcohol abuse, use of anticoagulants as well as in hemiplegic patients who are at risk of head trauma from frequent falls. [2]

Over the course of time, fibroblast usually invade the blood to form membranes. This is succeeded by the proliferation of new

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capillaries, elaboration of fibrin degradation products and consequent liquefaction of the blood clot with the later inhibiting coagulation thereby keeping the blood in a liquefied form. [2]

Gerstmann syndrome is a relatively rare condition that occurs as a result of a lesion to the angular and supramarginal gyrus of the left (dominant) hemisphere of the affected individuals. Famously named after Austrian born American neurologist Josef Gerstmann, its clinical manifestation include inability to identify ones fingers (finger agnosia), difficulty with calculation (acalculia), ability to read but inability to write (agraphia without alexia), left-right disorientation, inability to carry out verbal commands for activities that could be have been otherwise performed with ease (ideomotor apraxia) and bilateral astereognosis. [3]

Its Myriad of etiologies include but not limited to brain tumors, abscess, multiple sclerosis, middle cerebral artery aneurysm, cortical atrophy, progressive multifocal leukoencephalopathy and chronic subdural hematoma. It is usually seen in left parietal lobe lesions. [4]

There is paucity of literature reporting the occurrence of this dominant parietal lobe syndrome in association with chronic subdural hematoma. We report a case of Gerstmann syndrome in an elderly male with chronic subdural hematoma.

2. Case Report

A 67-year-old right-handed male patient presented with a 3-week history of difficulty differentiating the left from the right side of his body, inability to identify his fingers by name, difficulty with calculations and inability to carry out verbal commands on tasks which he could easily do prior to onset of symptoms. There was associated generalized throbbing headache, left sided body weakness and 4 episodes of generalized tonic clonic seizures He is a known hypertensive of 6 years duration regular on daily dose of 10 mg Amlodipine and 75 mg Clopidogrel, there was no prior history of head trauma, transient ischemic attack nor alcohol abuse.

General physical examination was essentially normal, neurologic examination revealed him to be confused with a Glasgow coma scale score of 14 (Eye opening - 4, Best Verbal response - 4, Best motor response -6). He had a pronator drift of his left upper limb with increased tone and reflexes in the left limbs. He had decreased sensation in the dermatomes of the left side of his body. He had significant difficulty differentiating his left from right body parts, identifying his fingers by name neither could he properly name familiar objects placed on his palms with his eyes closed.

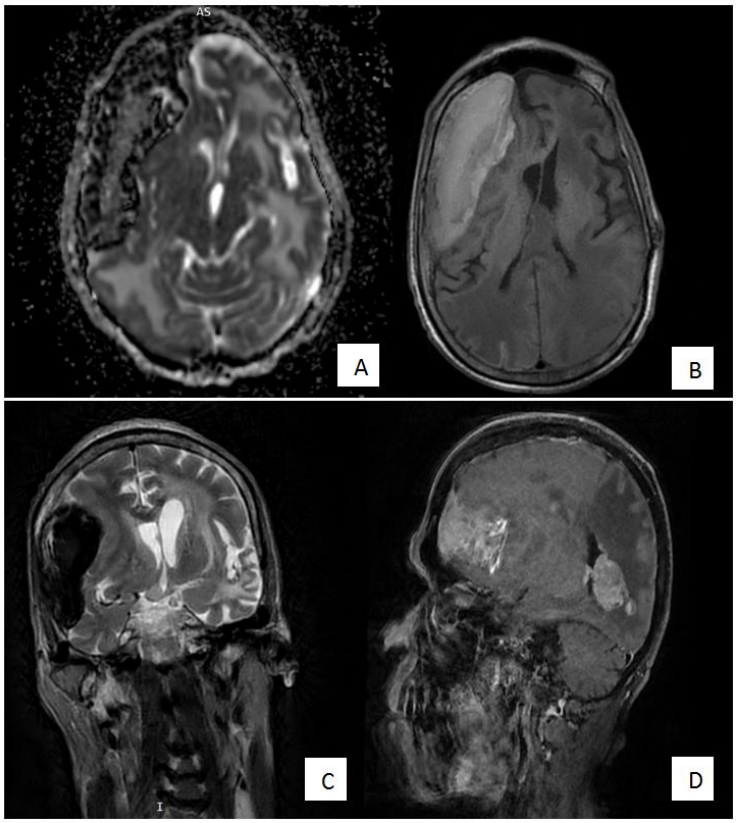

Figure 1. *Cranial MRI images of the right fronto-parietotemporal chronic subdural hematoma (A) ADC image showing hypointense region with necrotic debris within the capsule of the hematoma. (B) Axial T1 Image showing hyperintensity of the lesion with midline shift and right ventricular effacement. (C) Coronal T2 image showing a hypo intensity of the lesion (D) Sagittal contrast image showing contrast enhancement.*

He had dysmetria and could not perform the heel to shin test nor the tandem walk. He walked with a left hemiplegic gait. His Folstein mini mental state examination score was 13/30 with deficits in the orientation, registration, attention, calculation and language components of the scale.

He had a Cranial computerized tomography scan which revealed an extra-axial bilobular/pear shaped extra-axial lesion in the right fronto- parietotemporal region which exerts a mass effect with a midline shift and obliteration of the right lateral ventricle.

A clinical diagnosis of Gerstmann syndrome in an elderly patient with chronic subdural hematoma was made. His pre-operative laboratory investigations were essentially normal and he subsequently had a right fronto- parietotemporal craniotomy and evacuation of the hematoma with excision of pyogenic membranes Histology analysis of the resected specimen confirmed it to be the pyogenic membrane of a chronic subdural hematoma.

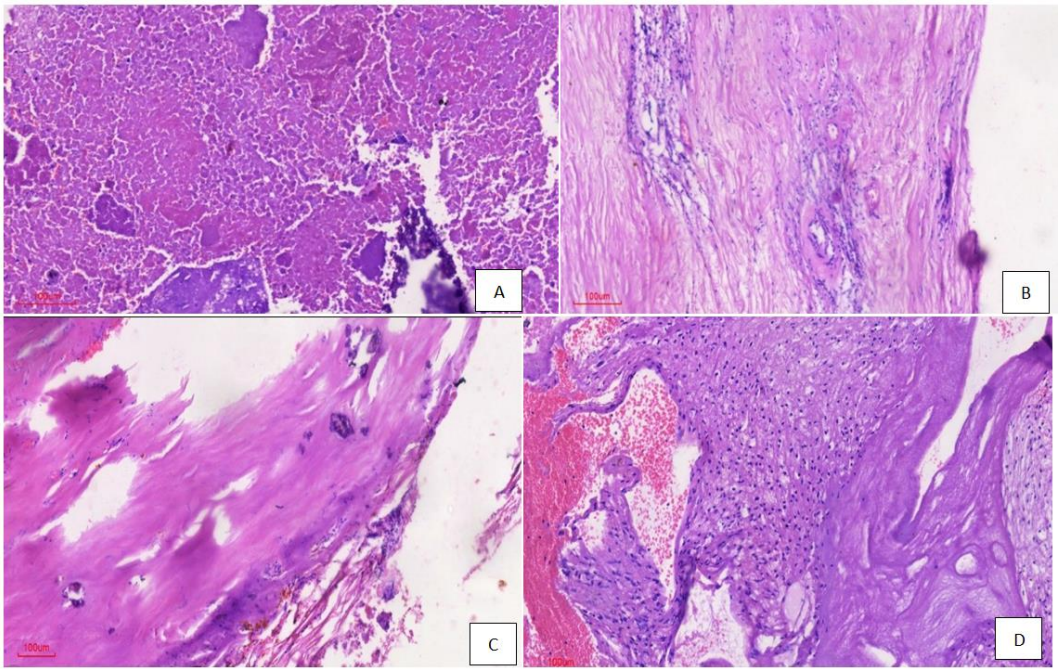

Figure 2. *Histopathologic slides of the membrane of the chronic subdural hematoma (A) Necrotic tissue with calcification (B) Oedema, fibrosis, thickened blood vessels and inflammatory cells (C) Fibrous tissue with calcifications (D) Brain tissue with focus of fibrous tissue and hemorrhage.*

Post operatively the patient was placed on anticonvulsants, analgesics and neurophysiotherapy. He showed remarkable improvement in neurology evidenced by an optimal Glasgow coma scale score of 15 and resolution of some of his presenting symptoms such as ideomotor apraxia, astereognosis and left-right disorientation. Finger agnosia resolved at one-month post-surgery, however acalculia persisted. He was discharged to clinic follow up on post-operative day 4 during which he had no fresh complaints and was noted to be ambulant on both lower limbs with no associated hemiparesis. He is currently on regular outpatient neurophysiotherpy sessions and has sustained his neurologic improvement since after the surgery.

3. Discussion

Gerstmann syndrome is a neurologic disorder in which affected patients present with symptoms such as acalculia, left-right disorientation, finger agnosia and agraphia. [5] Our patient presented with all the cardinal features of the syndrome in addition to having ideomotor apraxia and astereognosis.

These features are usually due to a lesion in the Brodmann areas 39 and 40 (angular and supramarginal gyrus) respectively of the dominant (left) parietal lobe. In a unique contrast, our patient who was right-handed, had a right sided lesion and presented with the features of the syndrome. Plausible explanations for this could be a possible anomalous functional and language dominance in the cerebral cortex as well as a possible reversal of lateralization due to an atypical brain functional anatomy. [6-8] However physiologic imaging studies could not be done in our patient to establish cerebral dominance due to unavailability of a functional magnetic resonance image.

Examination findings in our patient revealed varying severity of symptoms as agraphia, acalculia and left-right disorientation were markedly affected in comparison to other features. This presentation was similar to findings in both

right vascular [9] and left [10] sided lesions. An explanation for the co-existence of these clinical features could be an anatomical proximity of the different neuronal circuits involved in the manifestation of these symptoms. [11] However, diffuse tensor imaging (DTI) have been able to elucidate an integral subcortical white matter region close to the intraparietal sulcus whose fibres are connected to the four regions implicated in Gerstmann syndrome. [12] In consonance with different rates in neurologic improvement in affected patients, [13] our patient showed variable timelines in resolution of his symptoms with finger agnosia resolving last and acalculia persisting.

The integral role of brain cortical mapping has been elucidated with its importance underscored in delineation of the functional anatomy of regions that may be implicated in Gerstmann syndrome, hence optimizing management outcomes in affected patients. [14]

Disconnection between association fibers linking the frontal and parietal lobes due to ischemia has been inferred as one of the pathophysiologic mechanisms of gerstmann syndrome [15] and this could be the ase in our patient who have had a long standing increased intracranial pressure from the chronic subdural hematoma.

Similar to findings in 34.62% of cases of chronic subdural hematoma, [16] the histologic analysis of the pyogenic membrane of the chronic subdural hematoma showed hemorrhagic inflammatory features, thickened blood vessels and fibrous tissues with calcification.

4. Conclusion

This documentation brings to light that though Gerstmann syndrome is commonly reported in left (dominant) hemispheric lesions, it can occur also in right sided lesions and hence, a high index of suspicion should be entertained when either posterior parietal lobes are involved in a lesion. We also advocate for advanced neuroimaging such as SPECT, DTI and Functional MRI to establish cerebral dominance and monitor neurophysiologic functions after treating the etiology that precipitated the symptoms.

Abbreviations

SPECT	Single Photon Emission Tomography Scan
DTI	Diffusion Tensor Imaging
MRI	Magnetic Resonance Imaging

Author Contributions

Kelechi Michael Azode: Conceptualization, Data curation, Methodology, Resources, Supervision, Writing - original draft, Writing - review & editing

Ese-Enaorho Ewoye: Formal Analysis, Investigation, Software, Validation, Visualization

Conflicts of Interest

The authors declare no conflicts of interest.

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Research Field

Kelechi Michael Azode: Neurosurgery, Neuroscience, Neuro-radiology, Neuro-oncology

Ese-Enaorho Ewoye: Morbid anatomy, Forensic medicine