Case Report

Gerstmann Syndrome; A Frequent Pathology Presenting With A Rare Symptomatology

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Summary

We report a case of glioblastoma multiforme (GBM) of the left parietal lobe presenting with Gerstmann Syndrome (GS). A 54-year-old woman presented dyscalculia. The computed tomography depicted a cystic mass in the left parietal lobe, thus she was referred to our department. Her detailed neurological examination revealed left-right disorientation, finger agnosia and agraphia with dyscalculia. Pathological examination of the tumor revealed GBM. This most common primary malignancy of the central nerve system rarely presents with GS. Detailed neurological examination may locate the pathology of the brain prior to the radiological examinations and is still precious despite the rapid progression of the radiological technologies.

Key words: Gerstmann Syndrome, Glioblastoma Multiforme, dyscalculia, finger agnosia

INTRODUCTION

Gerstmann Syndrome (GS) consists of acalculia, agraphia, finger agnosia and left-right disorientation\(^{(2,4,5,6,9)}\). It is commonly associated with the focal lesions of the left angular gyrus, although a few cases are reported due to diffuse pathologies of the brain including lead intoxication, carbon monoxide poisoning, alcoholism and atrophy\(^{(2,4)}\). It has been discussed from the early times of definition of the syndrome, whether four cardinal symptoms of GS have a common base or only appear by chance association\(^{(6)}\). Even some authors like Benton regarded the syndrome as a fiction or an artifact of false observation\(^{(1)}\). However, there have also been some other
authors who stress that the existence of this rarely seen syndrome strongly suggests pathology in the posterior parietal lobe of the dominant hemisphere\(^8\).

Symptoms of the central nervous system (CNS) tumors vary in a great spectrum depending on the location of the pathology. We describe a patient with glioblastoma multiforme (GBM) in the left parietal lobe presented with an unusual symptomatology “Gerstmann Syndrome” for intracranial mass lesions.

**CASE PRESENTATION**

Fifty four years old right handed woman, suffering from moderate headache, applied to a physician when she developed dyscalculia. Patient running a family owned grocer in rural Turkey, who had no prior neurological or psychiatric illnesses, was immediately noticed by her husband when she started to miscalculate the bills of the shoppers. She was referred to our department when a left parietal cystic mass had been detected on cranial CT scan (Figure 1).

At the admission, the patient's score according to Glasgow Coma Scale (GCS) was found to be 15. She had no motor deficits and pathological reflexes. Her fundoscopic examination was bilaterally normal. Neuropsychological examination was also normal. On the neurological examination, the four cardinal symptoms of GS (acaleulia-dyscalculia, agraphia, finger agnosia and left-right disorientation) were found. Dyscalculia was the alarming sign for the family to refer to a physician at the beginning of all. Cerebral MRI depleted the cystic mass, with a mural like nodule, enhanced by gadolinium administered (Figure 2). Patient was operated via a left parieto-occipital craniotomy and the mass was removed. Her symptoms were immediately lost after the operation as well as the radiological appearance (Figure 3). Pathological examination revealed GBM. She was discharged 4 days after the operation with an un-eventful follow-up period. Patient is under therapy regarding the protocol of our own institution's neuro-oncology council.

![Figure 1: Pre-operative, pre- and post-contrast (A&B) axial CT scan images revealing the cystic mass of the left parietal lobe.](image-url)
GBM is known to be the most frequent primary malign tumor of the CNS\(^3\). It accounts for 12-15% of all intracranial neoplasms and 60-75% of astrocytic tumors. The incidence is about 3-4 new cases per 100,000 population per year in most North American and European countries. It may be diagnosed at any age but preferentially affects adults, with a peak incidence at between 45 and 75 years of age\(^{10}\). In a series of 987 GBM's from the University Hospital of Zurich, the most frequently affected sites were the temporal (31%), parietal (24%), frontal (23%), and occipital lobes (16%).

Location of our case presented is not exceptional, however, presenting symptoms are unusual and rare for intracranial neoplastic lesions. As known, clinical history of the disease is usually short. In our case nearly a sudden onset of acalculia is experienced. Nausea and vomiting like signs due to increased intracranial pressure were absent, while only mild headache was disturbing.

There are authors adding some other symptoms related to GS. Tucha et al., suggest toe agnosia along with defined

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**DISCUSSION**

**Figure 2:** Pre-operative axial and sagittal MRI images of the tumor of the left parietal before (A) and after (B&C) the administration of gadolinium. Figure 2B and 2C depict the mural like nodule of the cystic mass, enhanced by gadolinium.

**Figure 3:** Early post-operative axial CT scan image depicting the resolution of the cystic mass.
four cardinal symptoms\(^{(11)}\). They reported a patient suffering from GBM in left parietal lobe that they observed toe agnosia with symptoms of GS. Our patient was not examined for toe agnosia as the operation was performed prior to our awareness of this report. In post-operative period her examination for toes was normal with the improvement of other symptoms as we mentioned above.

Rusconi et al.\(^{(7)}\) pursued an alternative hypothesis according to which the selective association of the four symptoms that has been reported in pure cases might rather be due to a circumscribed subcortical lesion yielding a disconnection syndrome. They examined the functional neuroanatomy that could account for pure Gerstmann syndrome, which is the selective association of acalculia, finger agnosia, left-right disorientation, and agraphia by means of functional MRG images in 5 healthy subjects. In every subject, the parietal activation patterns across all four domains consistently connected to a small region of subcortical parietal white matter at a location that is congruent with the lesion in a well-documented case of pure Gerstmann syndrome. The authors concluded that their functional neuroimaging findings are not in agreement with Gerstmann's postulate of damage to a common cognitive function underpinning clinical semiology. They interpreted that pure forms of Gerstmann's tetrad do not arise from lesion to a shared cortical substrate but from intraparietal disconnection after damage to a focal region of subcortical white matter\(^{(7)}\). GBM manifests mostly in the subcortical white matter of the cerebral hemispheres\(^{(10)}\). In our case, the symptomatology of GS is probably due to intraparietal disconnection after damage to a focal region of subcortical white matter, as stated by Rusconi et al.

Various neurosurgical pathologies may manifest with pure or related GS due to their locations, which are expected to disturb the angular gyrus of the dominant, often left, parietal lobe. Chronic subdural hematoma and GBM are among reported pathologies\(^{(5,11)}\).

Briefly we conclude that a detailed neuro-examination is still precious for localization of any pathology of the nervous system. GS is a clinical entity despite all the debate among the neuro-scientists. Patients with any kind of pathology vascular, tumoral or traumatic should be inspected more detailed for any component of this syndrome; Acalculia, left-right disorientation, finger agnosia and agraphia. Presence of any of these symptoms should suspect the clinician for the other symptoms of the tetrad. GS has the high value in localization and the lesion is mainly localized to angular gyrus of the dominant hemisphere.

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