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A rare case of primary CNS angiosarcoma presenting as an incomplete Gerstmann syndrome: A case report and review of literature

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Introduction: Primary central nervous system angiosarcoma is a very rare malignancy, with only 22 cases reported in the literature in the past 26 years.

Clinical presentation: A 60-year old Filipino woman, housewife, presented with 1 week history of difficulty performing daily mathematical equations, difficulty reading words, and difficulty writing. On neurologic exam, patient was found to be oriented to person, place, and time. There was note of dyscalculia, agraphia, and right-left disorientation. There was no finger agnosia, agraphesthesia, astereognosia, nor apraxia. Cranial MRI showed a 4x4x4.1 cm mass at the left parietal lobe. Total excision of the mass done and biopsy revealed angiosarcoma of the brain. Thorough search for another systemic lesion failed to show any other angiosarcomatous lesion. The patient refused any further radiotherapy or chemotherapy.

Discussion: Soft tissue sarcomas comprise a small percent 1% of the malignant neoplasms, and of which only 0.5% comprise the primary brain OR spine sarcoma. In the review of literature that we did, of the 22 reported cases, the tumor was commonly located supratentorially (9/22), affecting middle to late ages with 3:1 male to female ratio. It is an aggressive tumor with a <2 year (range of survival is 4 months - 3½ years) survival independent of the age, location, or whether chemotherapy or radiotherapy was given. As with the best of our knowledge, there has been no recorded case of angiosarcoma which presented with Gerstmann syndrome in the literature.

Conclusion: The study described the first reported case of primary CNS angiosarcoma in the Philippines and the 23rd worldwide. It was also described the first case of angiosarcoma which presented with Gerstmann syndrome. Due to the rarity of this disease, much is still yet to be known in its risk factors, definitive treatment, and prognosis.

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