Atypical Subependymal Giant Cell Astrocytoma

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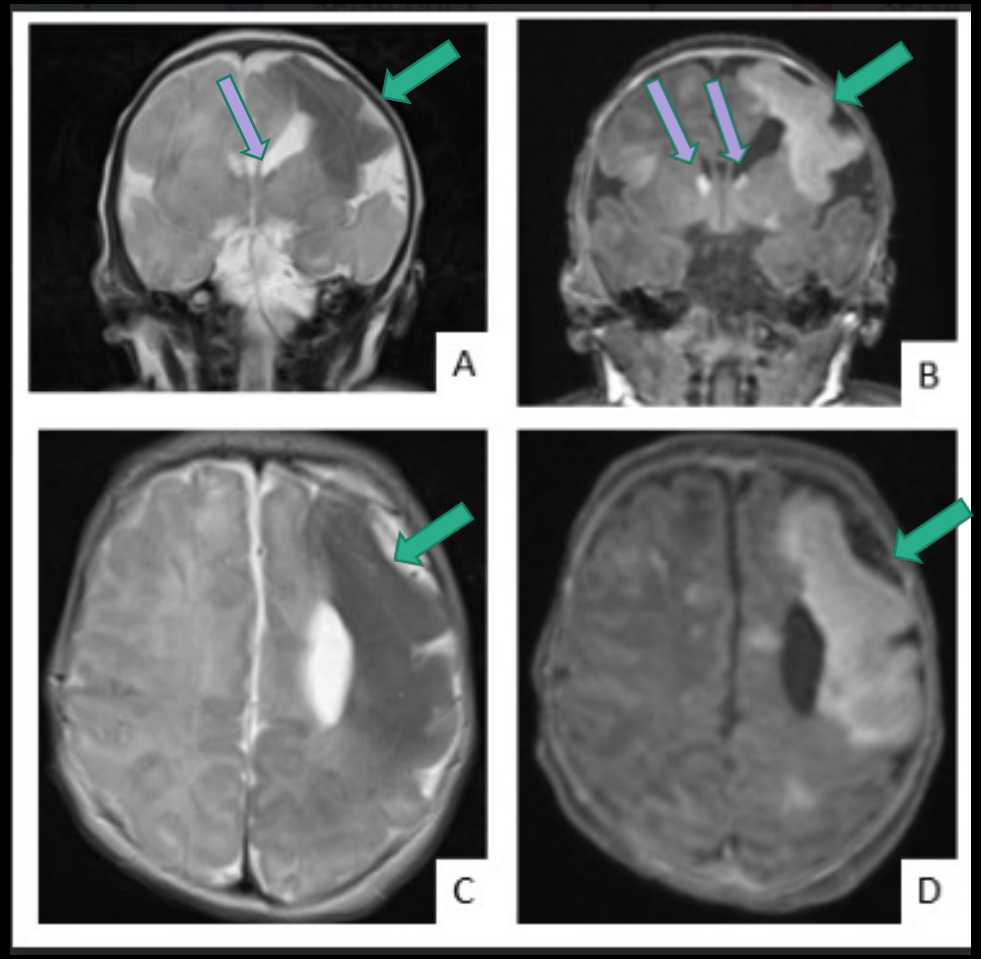
Clinical Presentation

- Term newborn presented to the ED with a chief complaint of seizure like activity.
- He had diagnosis of tuberous sclerosis complex by clinical criteria with major criteria for cortical tubers, subependymal nodules, and cardiac rhabdomyomas, as well as the minor criteria of multiple renal cysts.
- Genetic testing confirmed TSC2 mutation.

Imaging Discussion



MRI brain showed numerous areas of intraparenchymal T1 hyperintense and T2 hypointense tubers. The largest in the left frontal lobe which is measuring 6 cm (great arrows).

Multiple small subependymal nodules were also noted (purple arrow).

Management and Outcome

- Left frontal craniotomy for stereotactic resection of a large cortical tumor was performed at 13 months given daily medication refractory seizures.

- Pathology revealed the diagnosis of ‘Subependymal giant cell astrocytoma (SEGA), CNS WHO Grade 1, arising in the setting of tuber’. Patient is seizure free after surgery.

Take Home Points

- The most common location of SEGAs is the foramen of Monro, however atypical presentation could also be seen.

- SEGA is a CNS WHO grade 1 tumor and usually asymptomatic. Given the common location near the foramen of Monro, the most common presentation is obstructive hydrocephalus. However, seizure could be the initial presentation according to the location of the tumor as in this case.

References

- Almubarak AO, Abdullah J, Al Hindi H, AlShail E. Infantile atypical subependymal giant cell astrocytoma. Neurosciences (Riyadh). 2020 Jan;25(1):61-64. doi: 10.17712/nsj.2020.1.20190044. PMID: 31982898; PMCID: PMC8015630.