MYXOID MESENCHYMAL TUMOR with EWSR-1 FUSION

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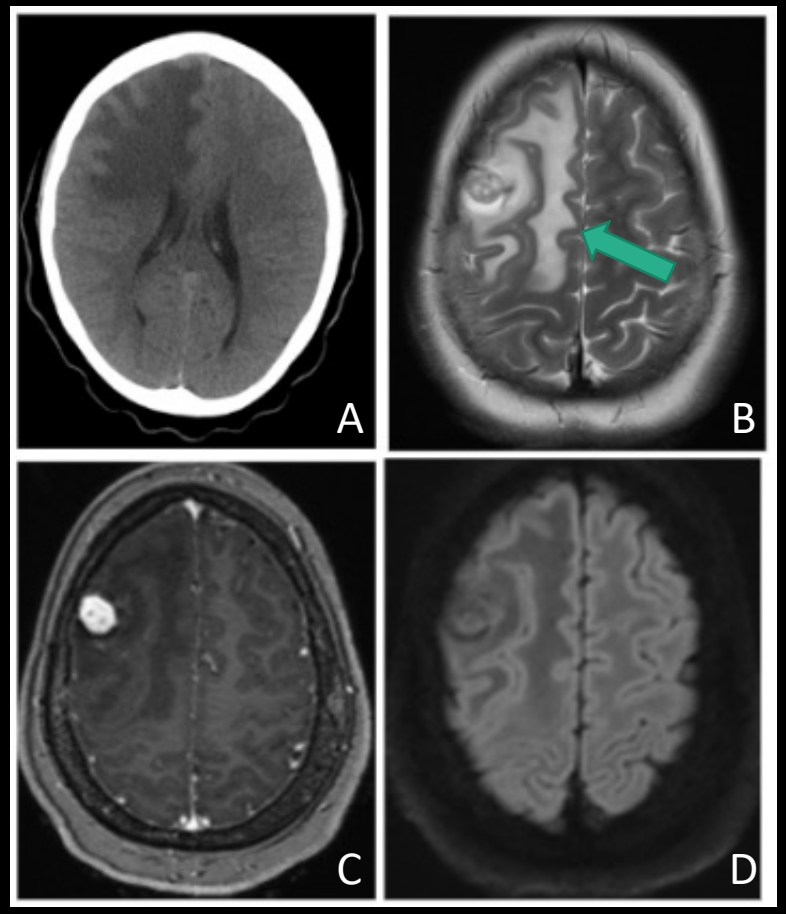
Clinical Presentation

- 13 year old female presents to the ED with reported seizure-like activity.

Imaging Discussion



- A. Non-contrast CT revealed vasogenic edema in the right frontal lobe without discrete mass.

- MRI B. T2WI, C. T1WI post contrast and D. DWI showed avidly enhancing circumscribed mass in the right frontal cortex with mild diffusion restriction and significant vasogenic edema (green arrow).

Management and Outcome

- Patient underwent right frontal craniotomy for gross total resection of tumor and pathology revealed ‘Myxoid Mesenchymal Tumor with EWSR-1 fusion’.

Take Home Points

- Intracranial myxoid mesenchymal tumor (IMMT) is a rare neoplasm occurs most frequently in adolescents and young adults. Case reports show that adolescents and young adults with intracranial myxoid mesenchymal tumors are positive for EWSR1 rearrangement.

References

- Sloan EA, Chiang J, Villanueva-Meyer JE, Alexandrescu S, Eschbacher JM, Wang W, Mafra M, Ud Din N, Carr-Boyd E, Watson M, Punsoni M, Oviedo A, Gilani A, Kleinschmidt-DeMasters BK, Coss DJ, Lopes MB, Raffel C, Berger MS, Chang SM, Reddy A, Ramani B, Ferris SP, Lee JC, Hofmann JW, Cho SJ, Horvai AE, Pekmezci M, Tihan T, Bollen AW, Rodriguez FJ, Ellison DW, Perry A, Solomon DA. Intracranial mesenchymal tumor with FET-CREB fusion-A unifying diagnosis for the spectrum of intracranial myxoid mesenchymal tumors and angiomatoid fibrous histiocytoma-like neoplasms. Brain Pathol. 2021 Jul;31(4):e12918. doi: 10.1111/bpa.12918. Epub 2021 Jan 28. PMID: 33141488; PMCID: PMC8089120.