



ESPR

European Society of
Paediatric Radiology



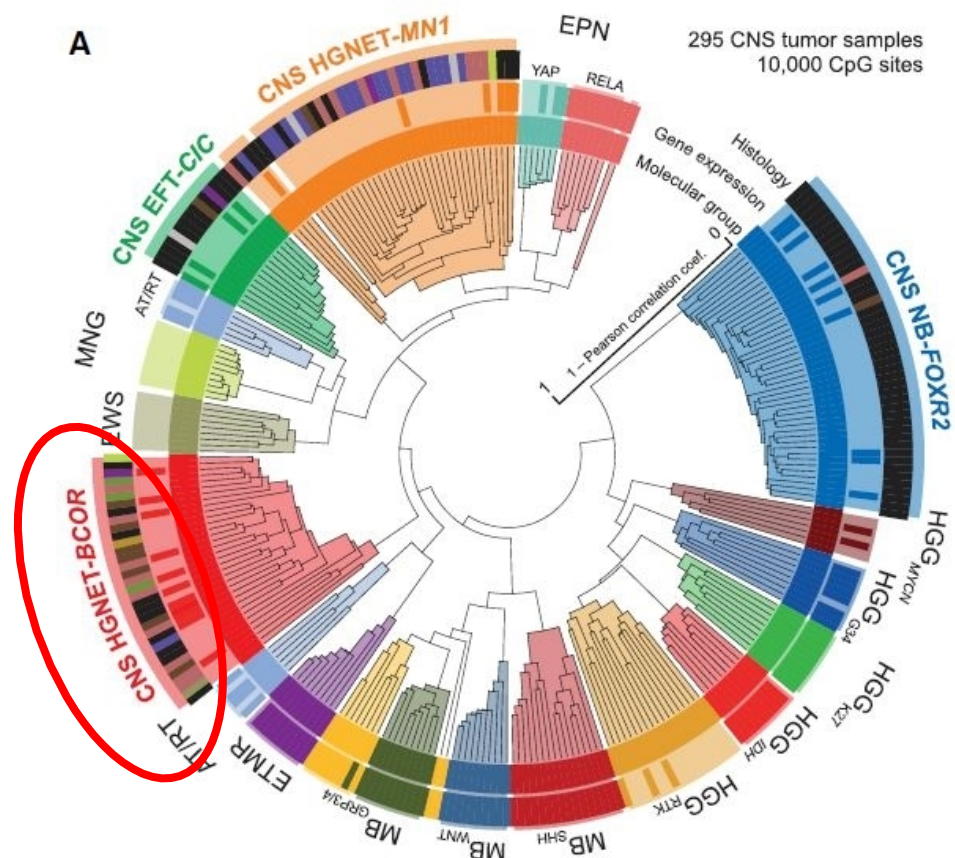
Imaging features with histopathologic correlation of CNS high-grade neuroepithelial tumors with BCOR internal tandem duplication

L Cardoen, A Tauziède-Espariat, V Dangouloff-Ros, S Moalla, N Nicolas, CJ Roux, Y Bouchoucha, F Bourdeaut, K Beccaria, S Bolle, G Pierron, C Dufour, F Doz, N Boddaert, HJ Brisse



New Brain Tumor Entities Emerge from Molecular Classification of CNS-PNETs

Dominik Sturm,^{1,2,3,89} Brent A. Orr,^{4,89} Umut H. Toprak,^{2,5,89} Volker Hovestadt,^{2,6,89} David T.W. Jones,^{1,2}



The 2021 WHO Classification of Tumors of the Central Nervous System: a summary

Embryonal tumors

Medulloblastoma

Medulloblastomas, molecularly defined

Medulloblastoma, WNT-activated

Medulloblastoma, SHH-activated and *TP53*-wildtype

Medulloblastoma, SHH-activated and *TP53*-mutant

Medulloblastoma, non-WNT/non-SHH

Medulloblastomas, histologically defined

Other CNS embryonal tumors

Atypical teratoid/rhabdoid tumor

Cribriform neuroepithelial tumor

Embryonal tumor with multilayered rosettes

CNS neuroblastoma, *FOXR2*-activated

CNS tumor with *BCOR* internal tandem duplication

CNS embryonal tumor

- Sturm D, Orr BA, Toprak UH et al. New brain tumor entities emerge from molecular classification of CNS-PNETs. *Cell* 2016;164(5):1060-1072
- Louis DN, Perry A, Wesseling P et al. The 2021 WHO Classification of Tumors of the Central Nervous System: a summary. *Neuro Oncol* 2021 Aug 2;23(8):1231-1251

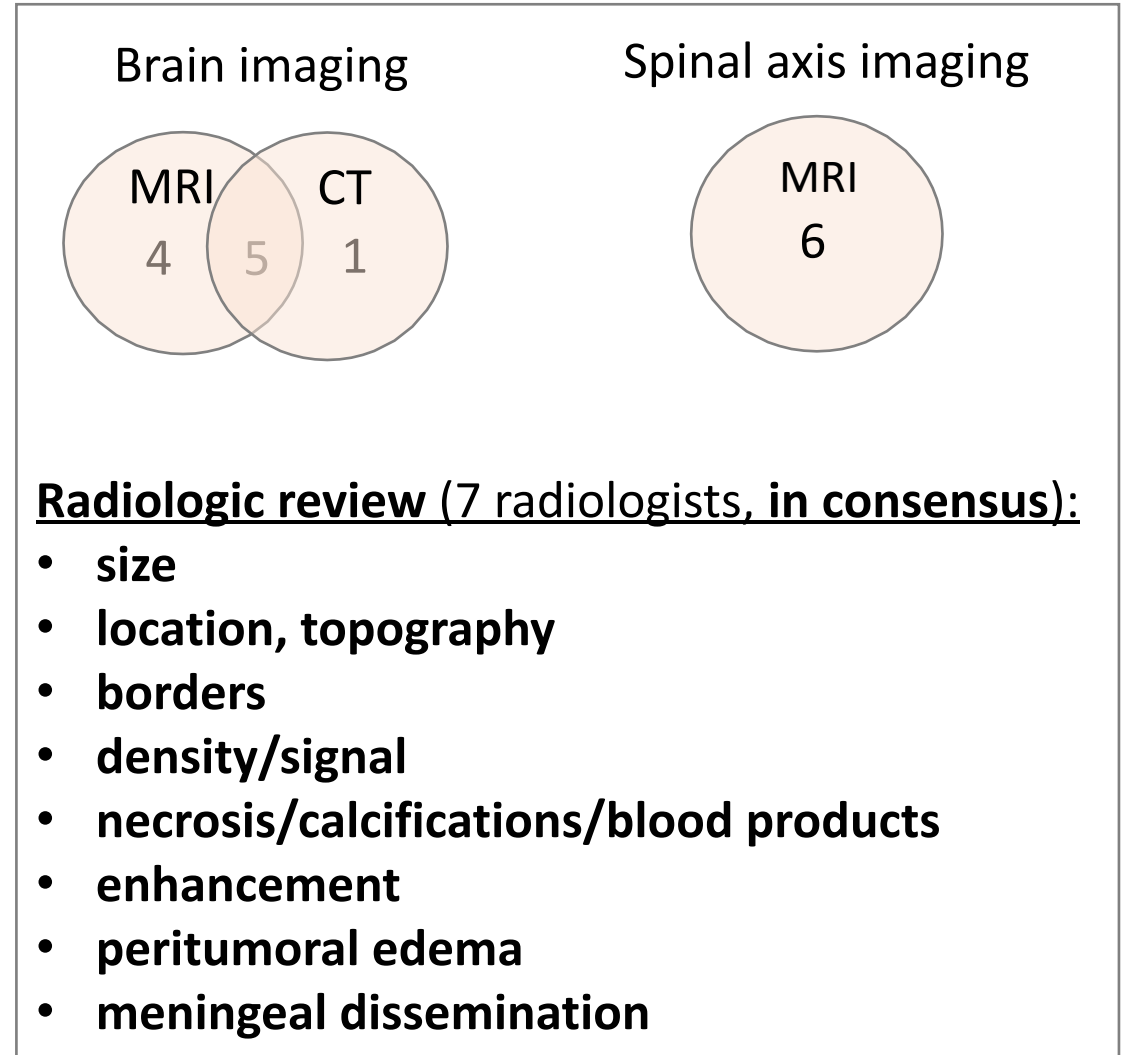
- **Retrospective**, non interventional study
- **10 children**
- RNA-sequencing or DNA-methylation analysis

Clinical data

- gender
- age

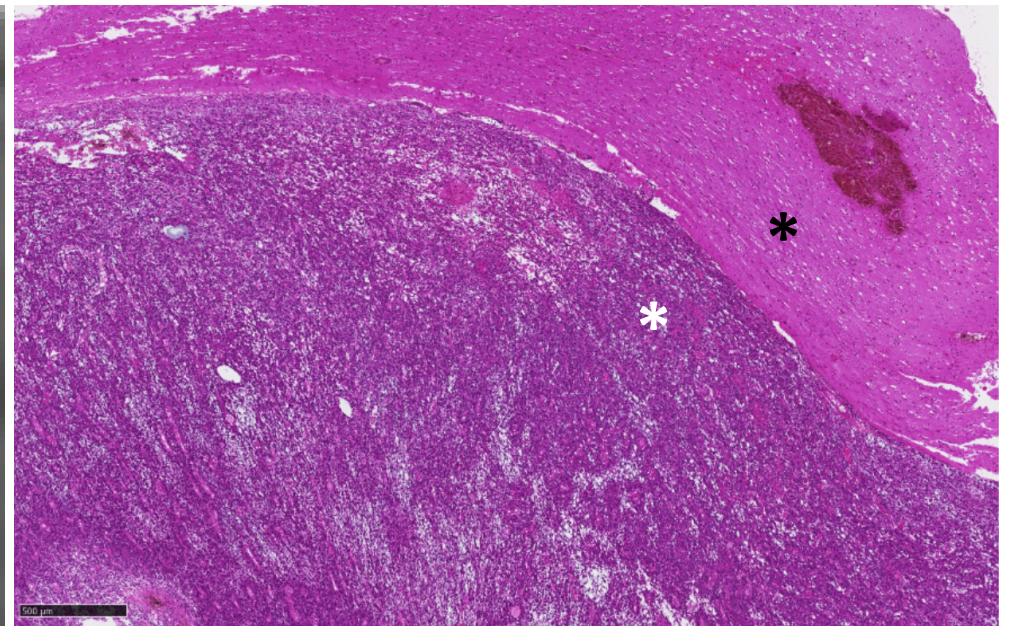
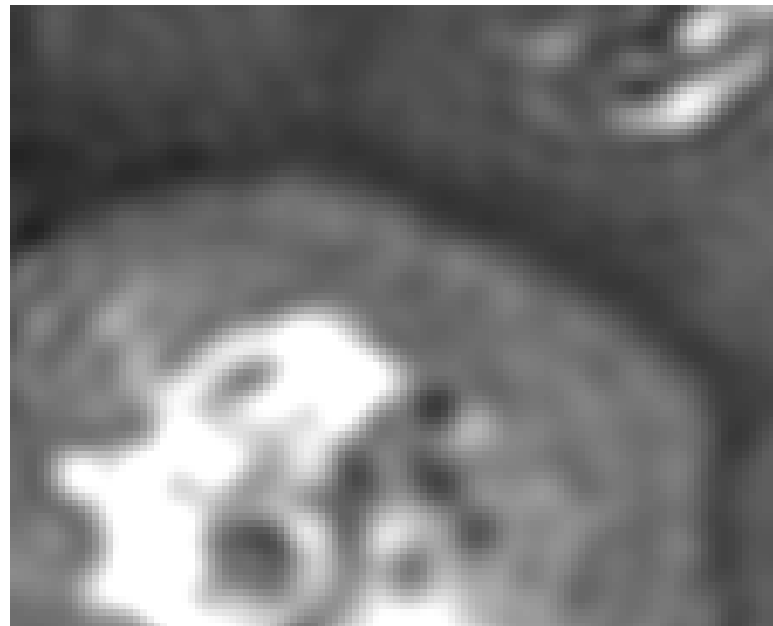
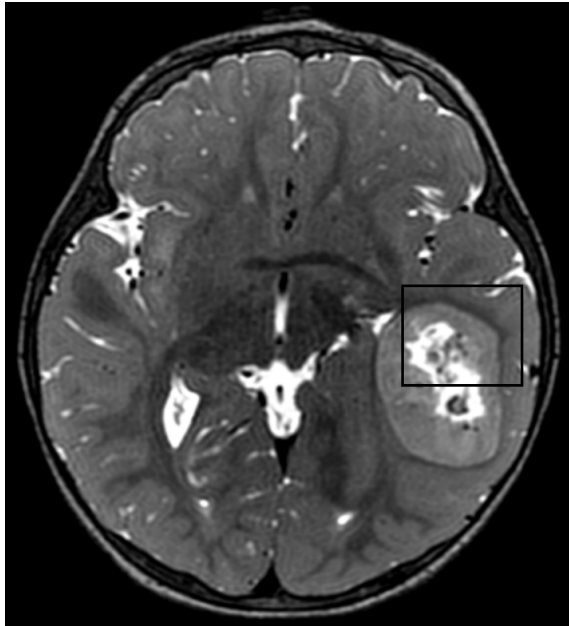
Histopathologic review (1 neuropathologist):

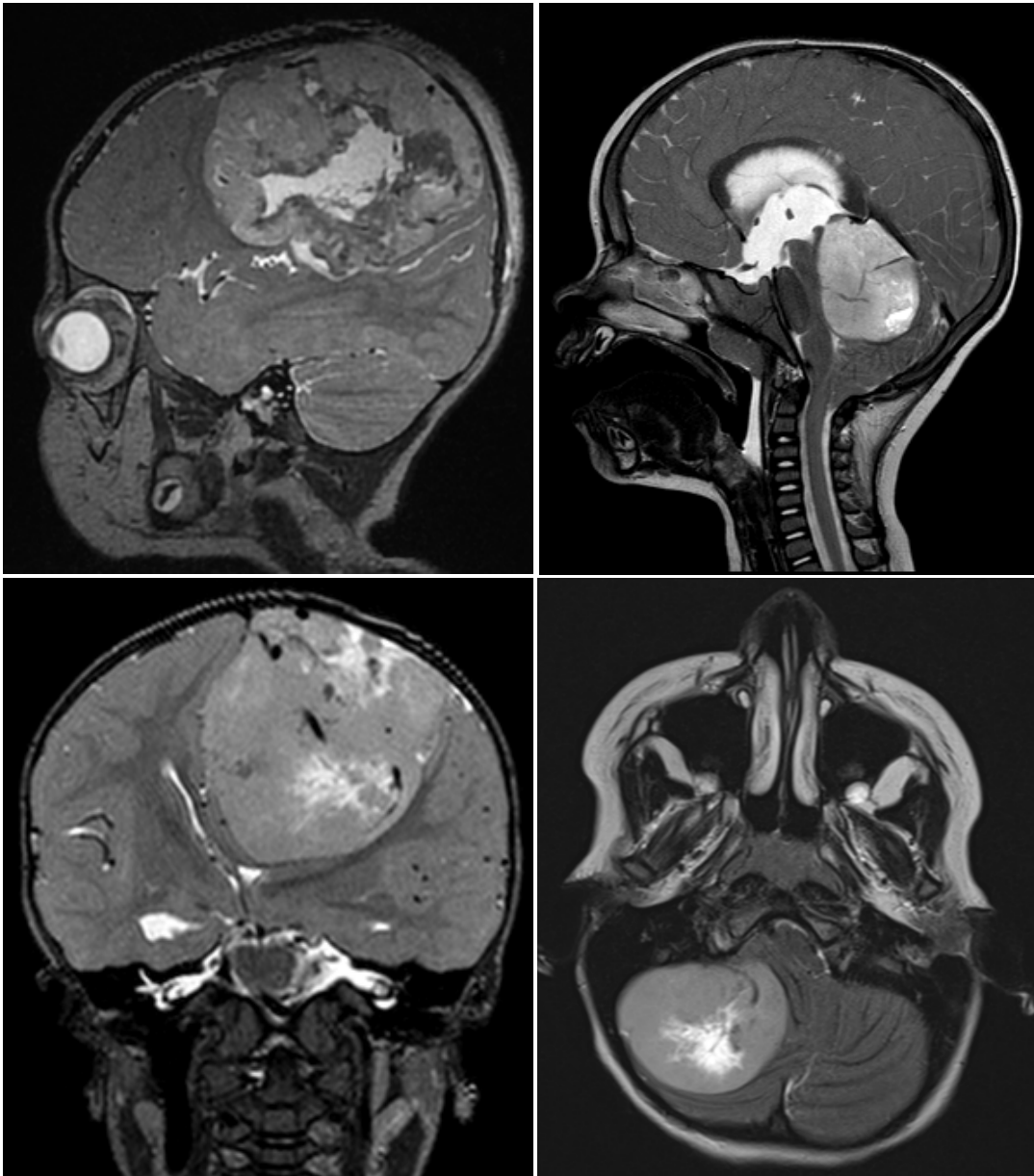
- cellular density
- necrosis
- calcifications
- hemorrhagic modifications
- density of the vascular network
- microvascular proliferation



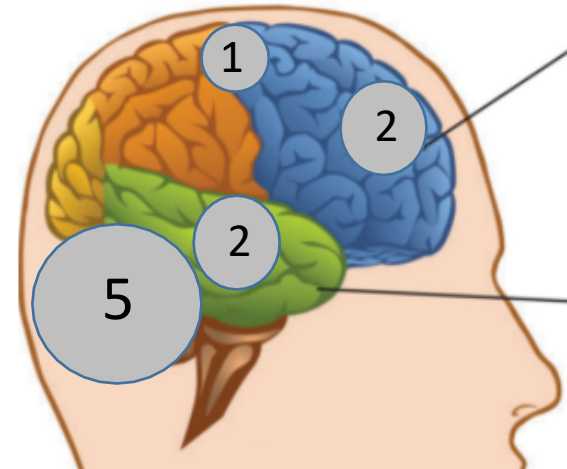
- 7 ♀ – 3 ♂
- 1,8 years

- **Large mass (max diameter 6 cm (4.7-9.2 cm))**
- **Well-defined borders**

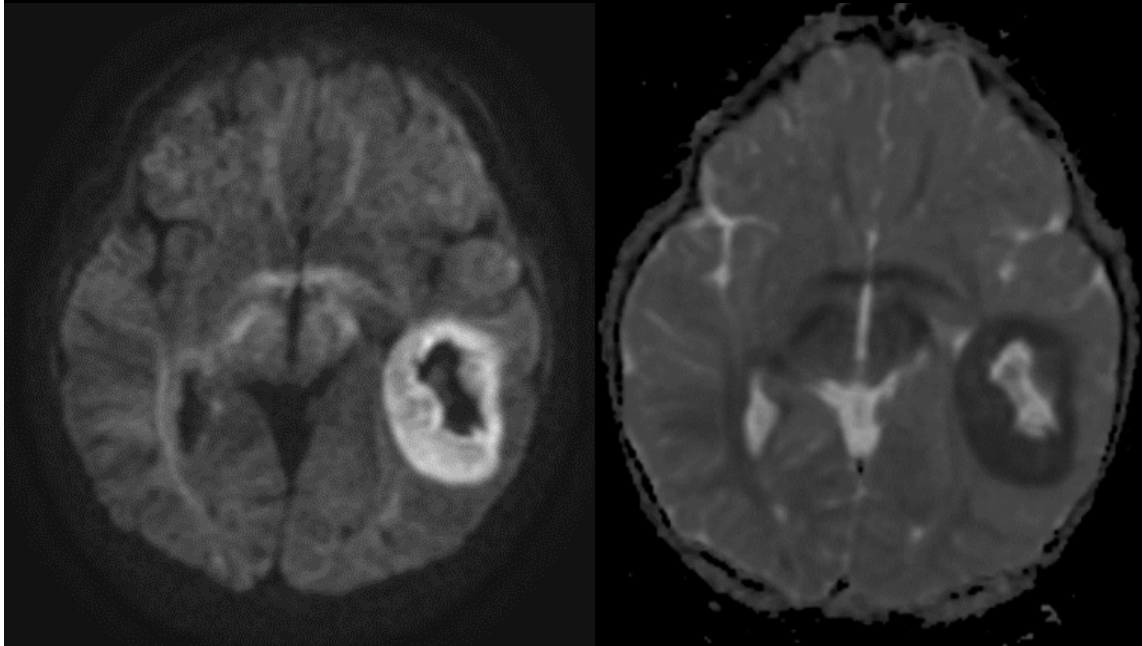
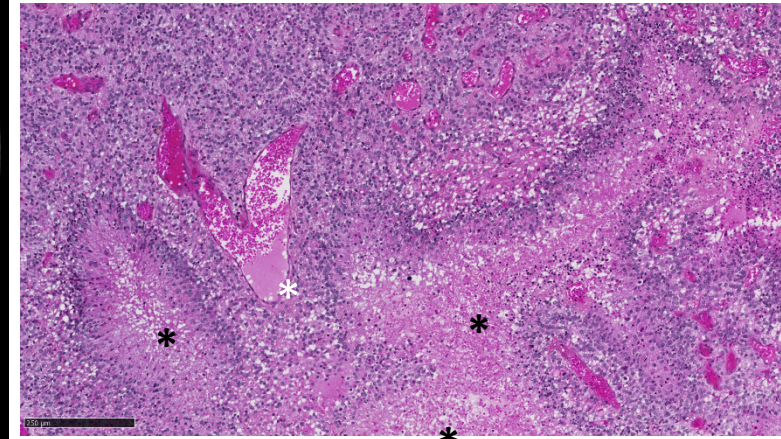
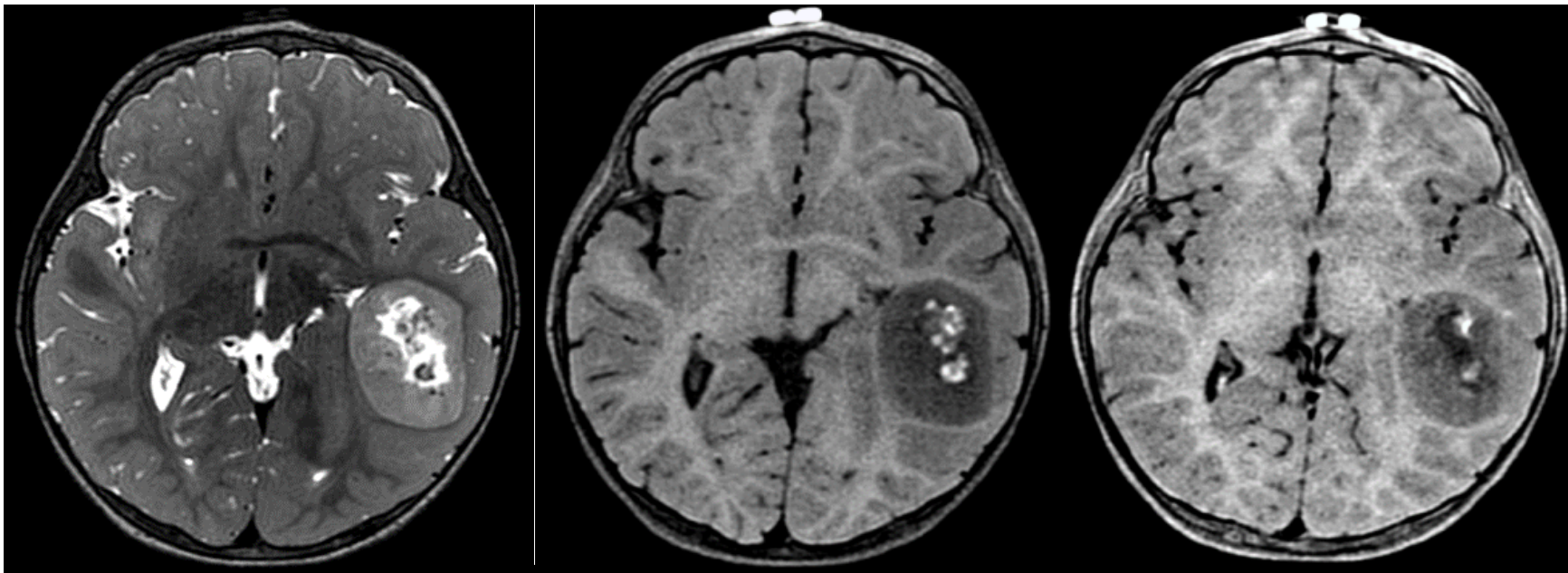




- Topography
 - **supra- and infratentorial**

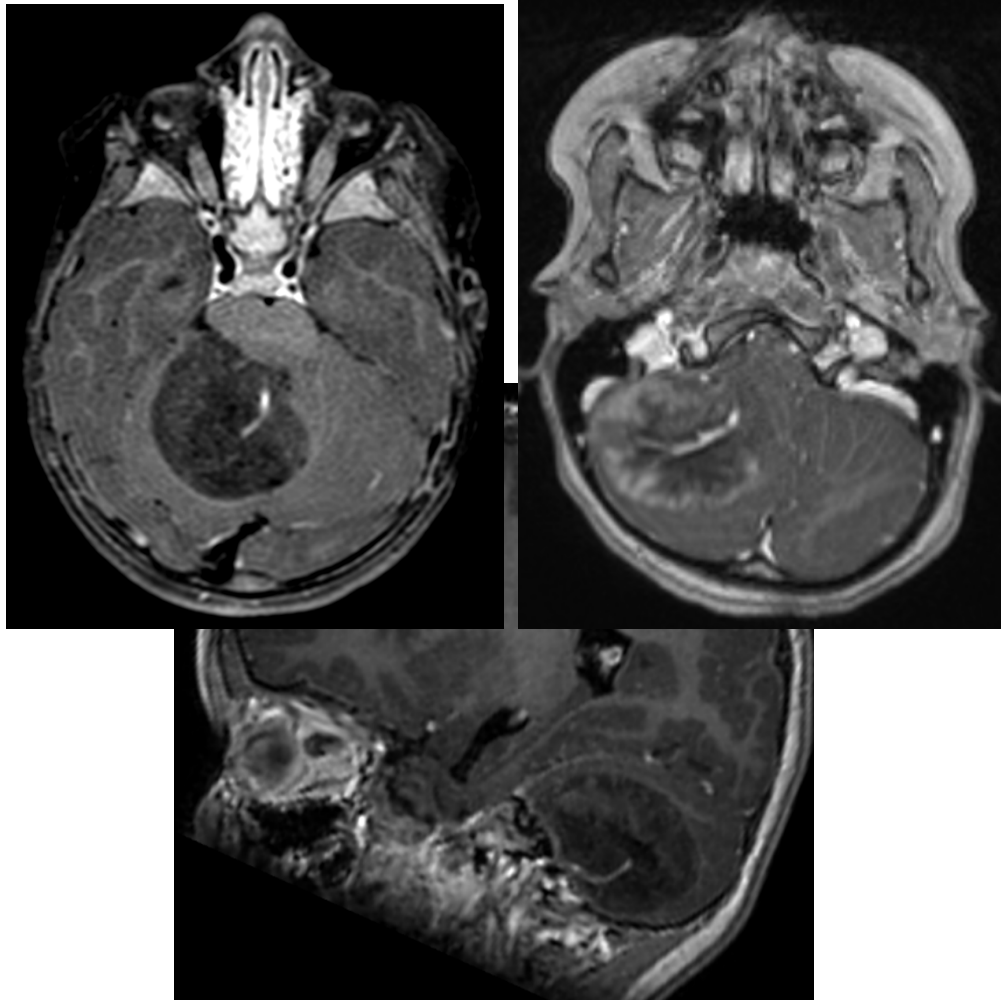


- **intra-axial**
 - **peripheral** (contact with dura mater)
-
- **No peritumoral edema**

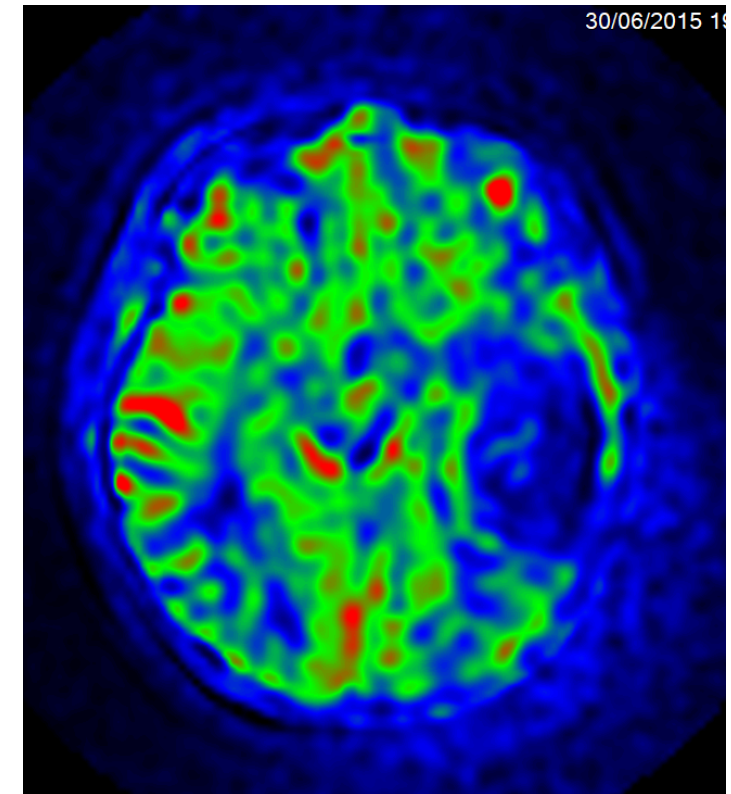


- Slightly hypersignal T2
- Hyposignal T1
- **Restricted diffusion**
- **Weak contrast** enhancement
- **Central necrosis**
- **Blood** products

- Weak contrast enhancement
- Central vessel



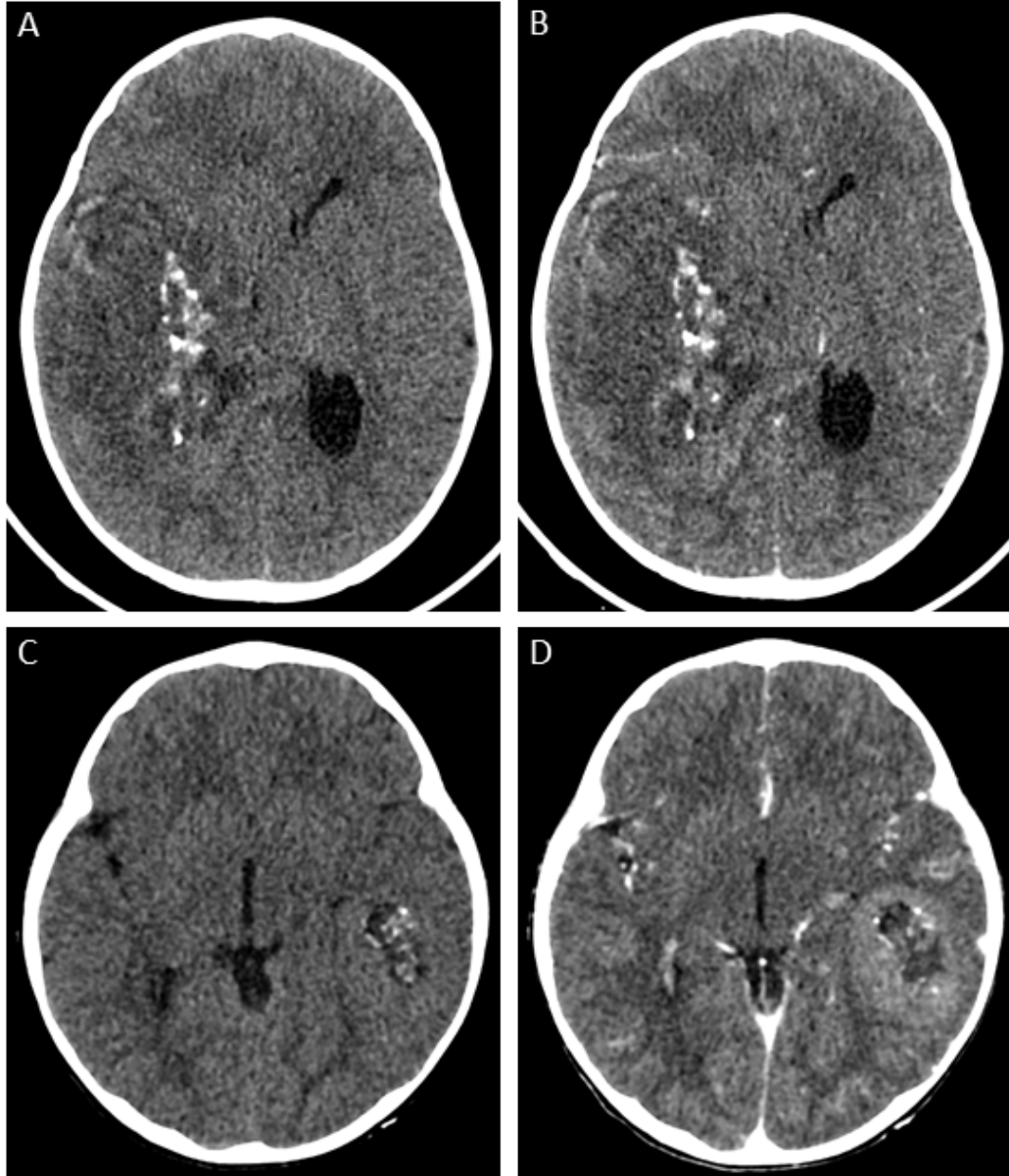
- Low CBF
- High rCBV



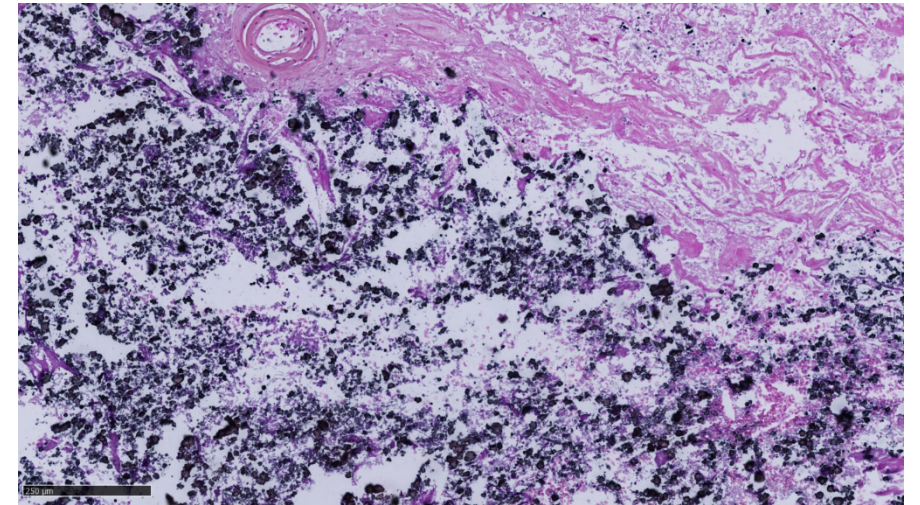
TDM

IV-

IV+



- Iso/hypodense (wrt GM)
- **Calcifications:** border of the central necrotic area and the solid peripheral portion of the tumor



Conclusion

CNS tumor with BCOR internal tandem duplication

large peripheral intra axial masses

well-defined borders

no peritumoral edema

restricted diffusion

weak contrast enhancement

frequent central necrosis, hemorrhage and calcifications

intratumoral veins

no leptomeningeal dissemination at the time of diagnosis.

L Cardoen, A Tauziède-Espariat, V Dangouloff-Ros et al. (2022) Imaging features with Histopathologic Correlation of CNS High-Grade Neuro-epithelial Tumors with a BCOR Internal Tandem Duplication. Am J Neuroradiol (43)1:151-156