Case 14890

Eurorad

Parietal anaplastic ganglioglioma in a 3-year-old girl

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Section: Paediatric radiology
Area of Interest: Neuroradiology brain Paediatric
Procedure: Diagnostic procedure
Imaging Technique: MR
Imaging Technique: MR-Diffusion/Perfusion
Imaging Technique: MR-Spectroscopy
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 3 years, female

Clinical History:

A 3-year-old-girl presented with headaches and partial complex seizures for a few months. There were findings suggestive of increased intracranial pressure.

Imaging Findings:

An extensive mass located in the left parietal region with ill-defined borders was demonstrated, with involvement of the left thalamus. The mass had heterogeneous signal intensity, predominantly isointense on T1WI and T2WI, with cystic/necrotic zones and bleeding in the posterior portion of the tumour. It demonstrated mild / moderate perilesional oedema predominantly in the upper parietal region, which demonstrated restriction of diffusion. The lesion impinged on the body of the left lateral ventricle, causing moderate dilation of the left occipital and temporal horns. On T1FS+Gd the lesion demonstrated moderate heterogeneous enhancement, mainly posteriorly. Diffusion-weighted sequences showed restricted diffusion in the perilesional oedema. On spectroscopy, there is a marked increase in the choline peak, with a decrease in the N-acetylaspartate and Cr peaks (tumoral pattern).

Discussion:

Gangliogliomas are tumours in which nerve and glial cells, usually astrocytes, participate in the neoplastic process; in this sense, they differ from most primary CNS neoplasms in which only the glial cells show neoplasia. Gangliogliomas are uncommon tumours, constituting approximately 3% of brain tumours in children and approximately 6% of supratentorial paediatric brain tumours; they are found in older children and young adults more frequently than younger children or infants [1].
The best diagnostic imaging clue is a partially cystic, enhancing, cortically-based mass in child/young adult with temporal/parietal lobe epilepsy. A cyst with an enhancing mural nodule is classic, but nonspecific for ganglioglioma. There are 3 morphologic patterns [1, 2]:
- The most common: Circumscribed cyst + mural nodule
- Solid tumour (often thickens, expands gyri)
- Uncommon: Infiltrating, poorly-delineated mass (our case)

CT findings are of a mass which is often non-specific. General features include an iso- or hypodense nodule-mass

which is frequently calcified (35%). Bony remodelling or thinning can indicate the slow growing nature of the tumour. Enhancement is seen in approximately 50% of cases (involving the solid non-calcified component) [1, 3, 4].

On MRI, the ganglioglioma may have sharply or poorly defined margins. The tumour may be solid, cystic, cystic with a mural nodule, or have the appearance of many small cysts. Signal intensity on T1-weighted sequences is variable (and often mixed); T2-weighted sequences generally reveal variable signal intensity in the cystic component depending on the amount of proteinaceous material or presence of blood products. Regions of slight T1 hyperintensity, probably representing calcification, may be helpful in identifying these neoplasms. Solid portions of the tumour enhance variably. On T2* calcified areas may show blooming. Solid portions of the tumour enhance variably. Usually there is not restriction on DWI images, but reduced diffusion in solid components can be shown. Spectroscopy demonstrates a choline peak with decreased n-acetyl-aspartate, a pattern suggestive of tumour. The peripheral location within the hemispheres and the erosion of the adjacent calvarium can be helpful in making the diagnosis of ganglioglioma.

Symptoms are site-specific, but are typically long-standing. Ganglioglioma is the most common tumour associated with chronic temporal lobe epilepsy. [1, 4].
Surgical resection is treatment of choice. Radiation therapy and/or chemotherapy may be helpful for aggressive or unresectable tumours [4].
Differential Diagnosis List: Parietal anaplastic ganglioglioma, Dysembryoplastic neuroepithelial tumour (DNET), Pleomorphic xanthoastrocytoma (PXA), Oligodendroglioma, Desmoplastic infantile astrocytoma

Final Diagnosis: Parietal anaplastic ganglioglioma

References:

Barkovich AJ (2012) Pediatric Neuroimaging Fifth Edition. Lippincott Williams & Wilkins Chapter 7: Intracranial, orbital, and neck masses of childhood
Wolf LüdemannEmail authorRouzbeh BananChristian HartmannHelmut BertalanffyConcezio Di Rocco (2017) Pediatric intracranial primary anaplastic ganglioglioma. Child's Nervous System 33(2): 227–231 (PMID:27921213)
Bianchi F, Tamburrini G, Massimi L, Caldarelli M (2016) Supratentorial tumors typical of the infantile age: desmoplastic infantile ganglioglioma (DIG) and astrocytoma (DIA). A review. Child's Nervous System Childs Nerv Syst. 2016 Oct;32(10):1833-8. doi: 10.1007/s00381-016-3149-4 (PMID:27659826)
Zanello M, Pagès M, Roux A, Peeters S, Dezamis E, Puget S, Devaux B, Sainte-Rose C, Zerah M, Louvel G, Dumont SN, Meder JF, Grill J, Huberfeld G, Chrétien F, Parraga E, Sauvageon X, Varlet P, Pallud J. (2017) Epileptic seizures in anaplastic gangliogliomas. Br J Neurosurg 31(2):227-233. doi: 10.1080/02688697.2016.1220506 (PMID: 27550627)

Figure 1

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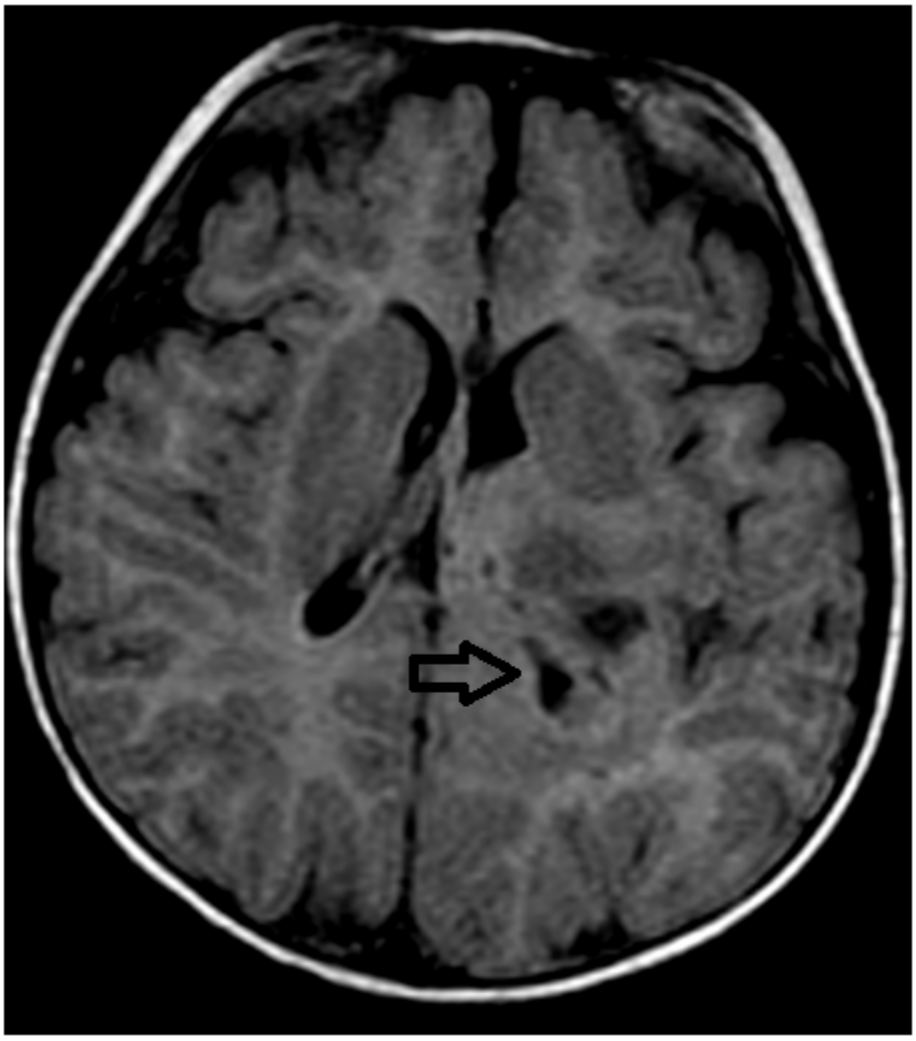

Description: An isointense lesion partially defined with hypointense cystic-necrotic areas is shown in the left posterior parietal region (arrow), and extending to the left thalamic region. **Origin:** Department of radiology. Hospital Universitario Virgen de la Arrixaca, Murcia (Spain).

Figure 2

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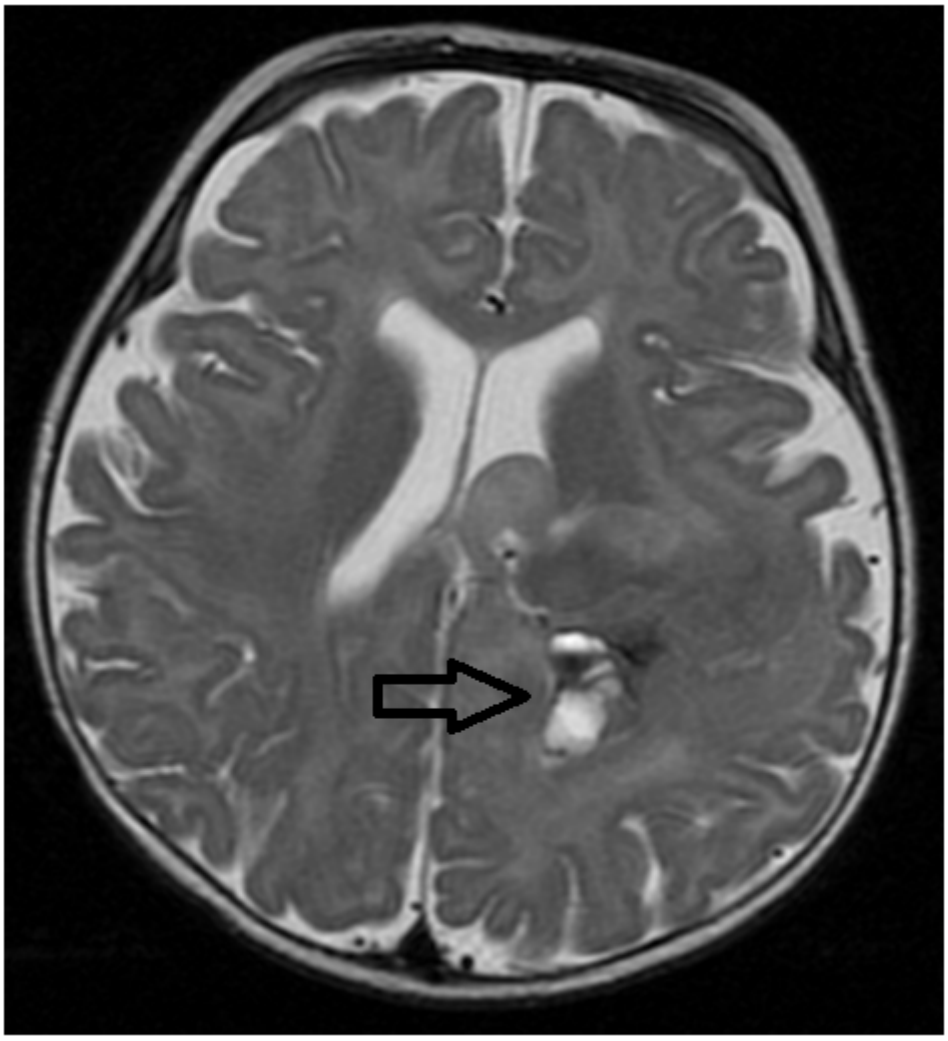

Description: An isointense lesion partially defined with hyperintense cystic-necrotic areas is shown in the left posterior parietal region (arrow), and extending to the left thalamic region. T2 hypointensities corresponding to calcifications/blood (arrow). **Origin:** Department of radiology. Hospital Universitario Virgen de la Arrixaca, Murcia (Spain).

Figure 3

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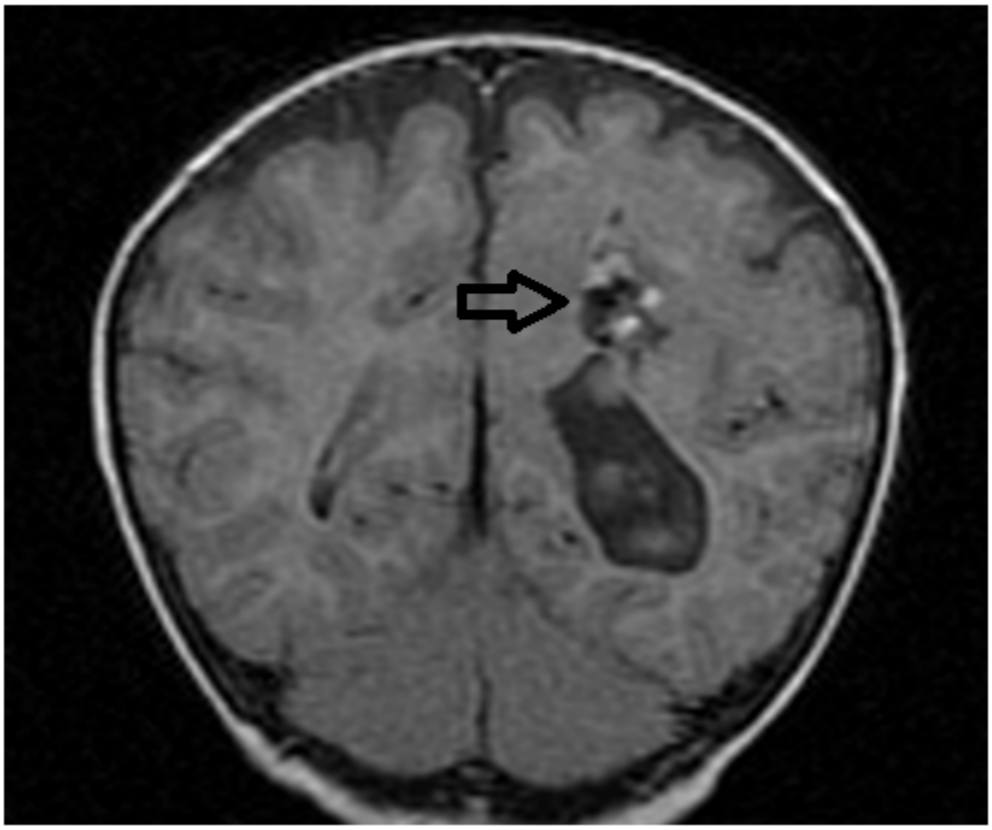

Description: Coronal T1 shows small areas of heterogeneous intensity that make up a mass of ill-defined limits (arrow). **Origin:** Department of radiology. Hospital Universitario Virgen de la Arrixaca, Murcia (Spain).

Figure 4

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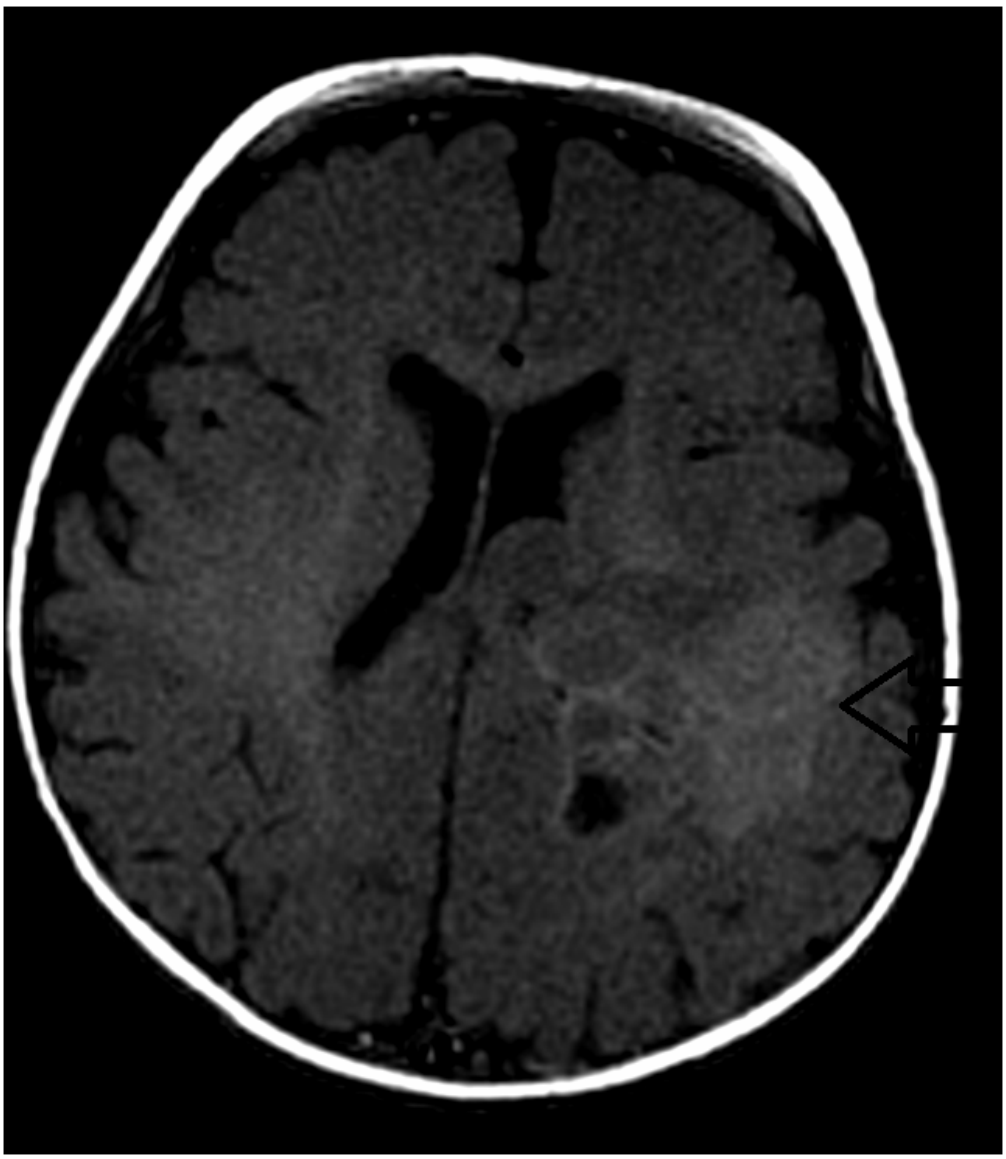

Description: FLAIR sequence demonstrates mild / moderate perilesional oedema predominantly in the upper parietal region (arrow), which demonstrates restriction of diffusion. **Origin:** Department of radiology. Hospital Universitario Virgen de la Arrixaca, Murcia (Spain).

Figure 5

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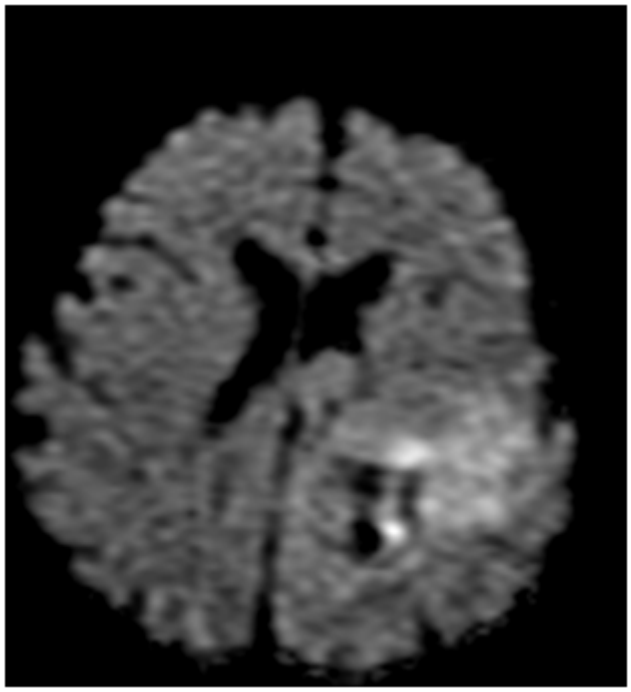

Description: On diffusion-weighted sequences, hyperintensity of the perilesional oedema is demonstrated. On the ADC map the same areas are hypointense. **Origin:** Department of radiology. Hospital Universitario Virgen de la Arrixaca, Murcia (Spain).

Figure 6

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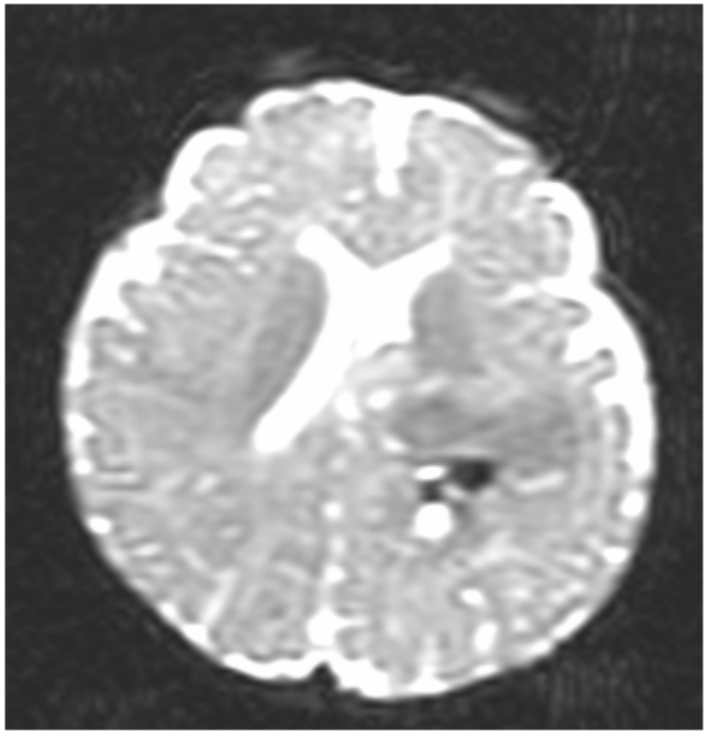

Description: On diffusion-weighted sequences, hyperintensity of the perilesional oedema is demonstrated. On the ADC map the same areas are hypointense. **Origin:** Department of radiology. Hospital Universitario Virgen de la Arrixaca, Murcia (Spain).

Figure 7

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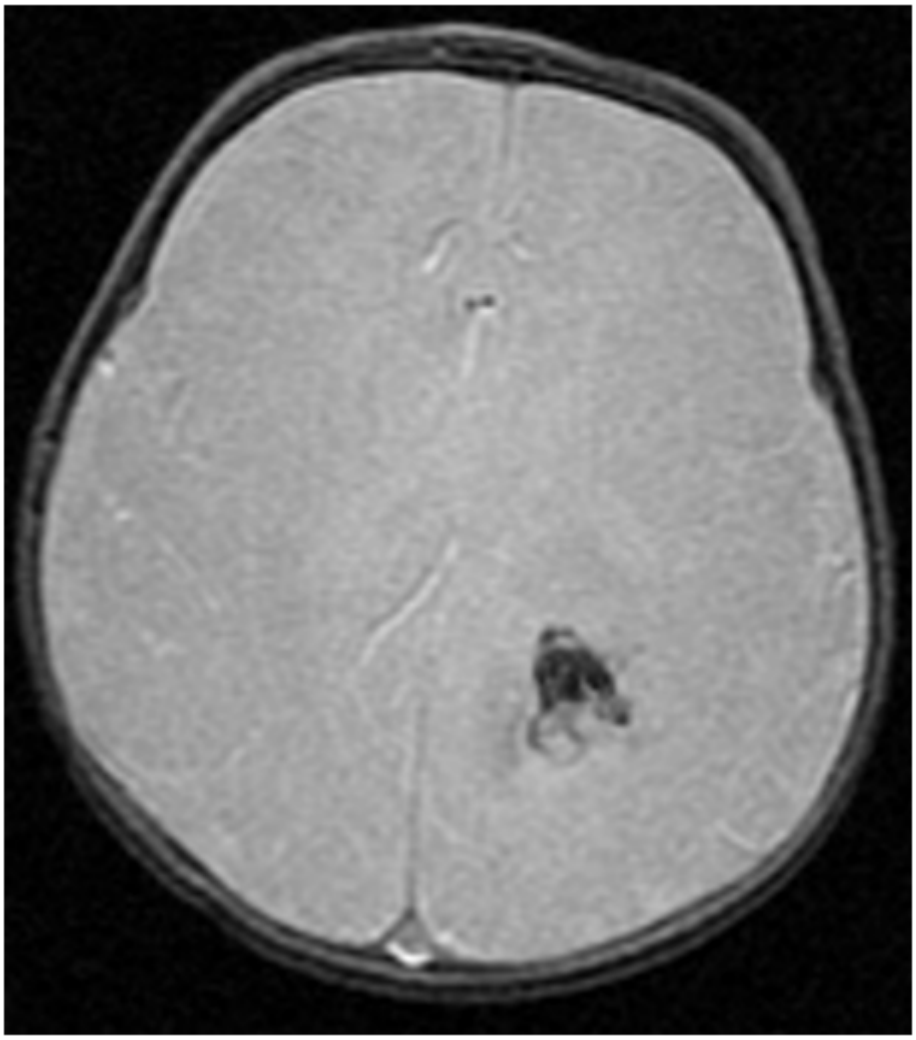

Description: In the posterior area of the tumour in this gradient echo sequence, there is loss of signal corresponding to areas of bleeding / calcification. **Origin:** Department of radiology. Hospital Universitario Virgen de la Arrixaca, Murcia (Spain).

Figure 8

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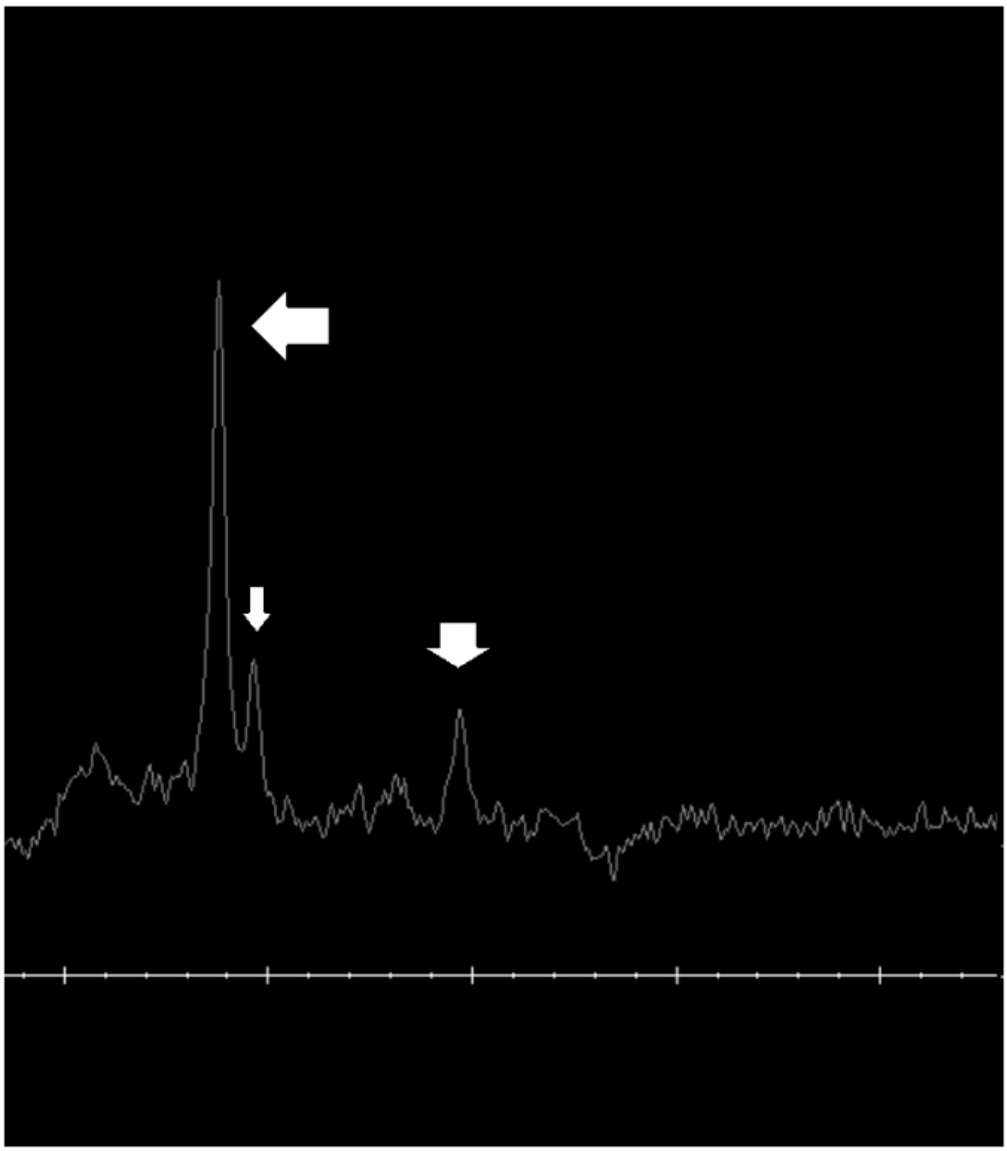

Description: Spectroscopy at TE=144ms of the tumour area shows a marked increase in the choline peak (superior arrow), with a decrease in the N-acetylaspartate peak and Cr peak (inferior arrows), (tumoral biochemical pattern). **Origin:** Department of radiology. Hospital Universitario Virgen de la Arrixaca, Murcia (Spain).

Figure 9

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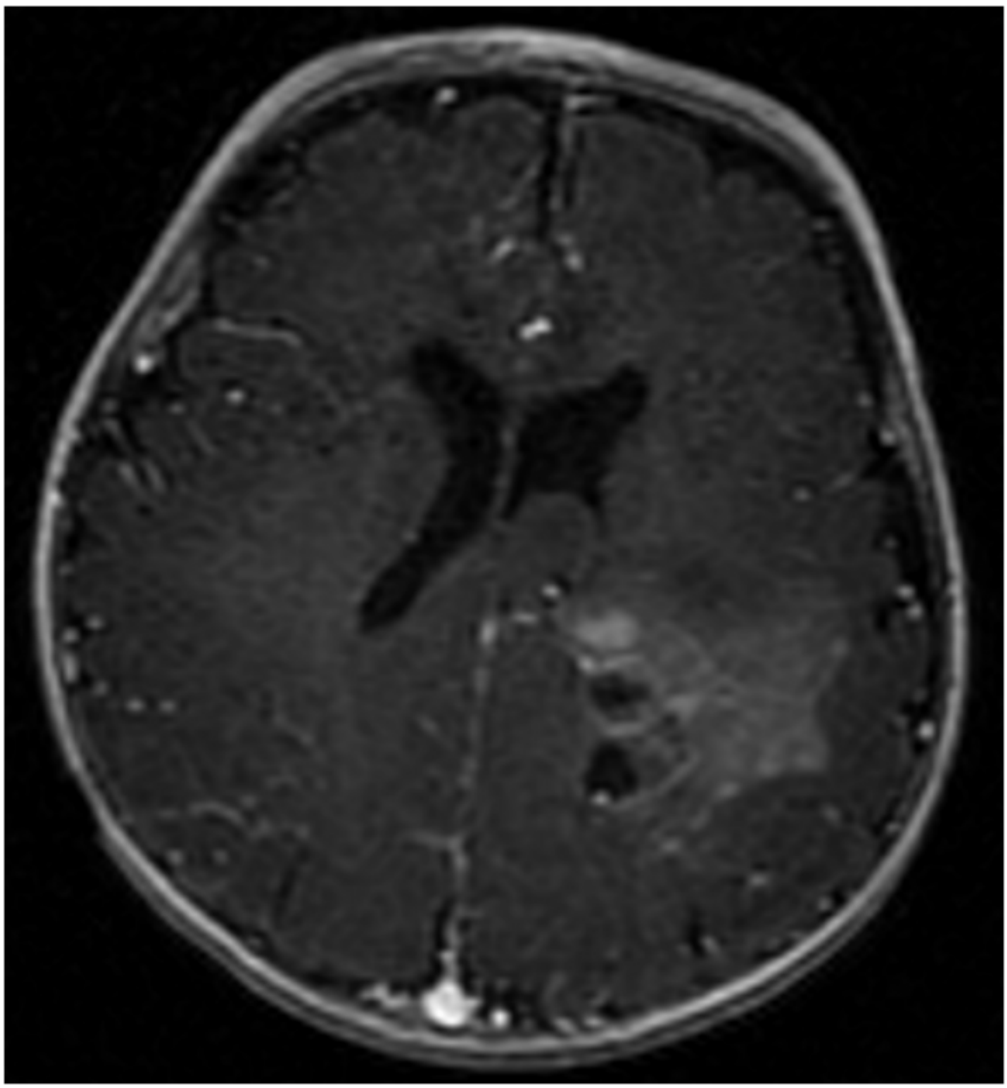

Description: The lesion demonstrates moderate heterogeneous enhancement, mainly posteriorly.
Origin: Department of radiology. Hospital Virgen de la Arrixaca, Murcia, Spain.