AMSER Case of the Month July 2023

HPI: 2-month-old previously healthy female presenting with inconsolability and episodes of emesis

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Patient Presentation

HPI: 2-month-old presenting to an outside hospital for inconsolability and non-bloody non-bilious emesis that began the evening prior to presentation. Mother noted mild cough and a “bump” in the location of the anterior fontanelle. History was otherwise negative for additional symptoms.

Past Medical History: None. Born at 39 weeks via planned cesarean due to gestational diabetes mellitus. No birth-related complications.

Vitals: BP 91/74, pulse 179, temp 37.5, resp 33, spO2 100%

Physical Exam: Irritable, not opening eyes spontaneously, firm bulging anterior fontanelle, left pupil 3mm minimally reactive, right pupil 4mm minimally reactive, tachycardic, pulmonary and abdominal exams unremarkable, hypotonia of left upper extremity, decerebrate movements of right upper extremity

Pertinent Labs: WBC 9.4, Hemoglobin 7.5, Platelets 489, Lumbar puncture (at OSH prior to imaging) showing RBCs in CSF

What Imaging Should We Order?

ACR Appropriateness Criteria:

Acute mental status change, suspect increased intracranial pressure

Variant 1: **Acute mental status change. Increased risk for intracranial bleeding (ie, anticoagulant use, coagulopathy), hypertensive emergency, or clinical suspicion for intracranial infection, mass, or elevated intracranial pressure. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
CT head without IV contrast	Usually Appropriate	☢ ☢ ☢
MRI head without IV contrast	Usually Appropriate	O
MRI head without and with IV contrast	May Be Appropriate	O
CT head without and with IV contrast	May Be Appropriate	☢ ☢ ☢
CT head with IV contrast	Usually Not Appropriate	☢ ☢ ☢

This imaging modality was ordered by the ER physician at the OSH prior to transfer.

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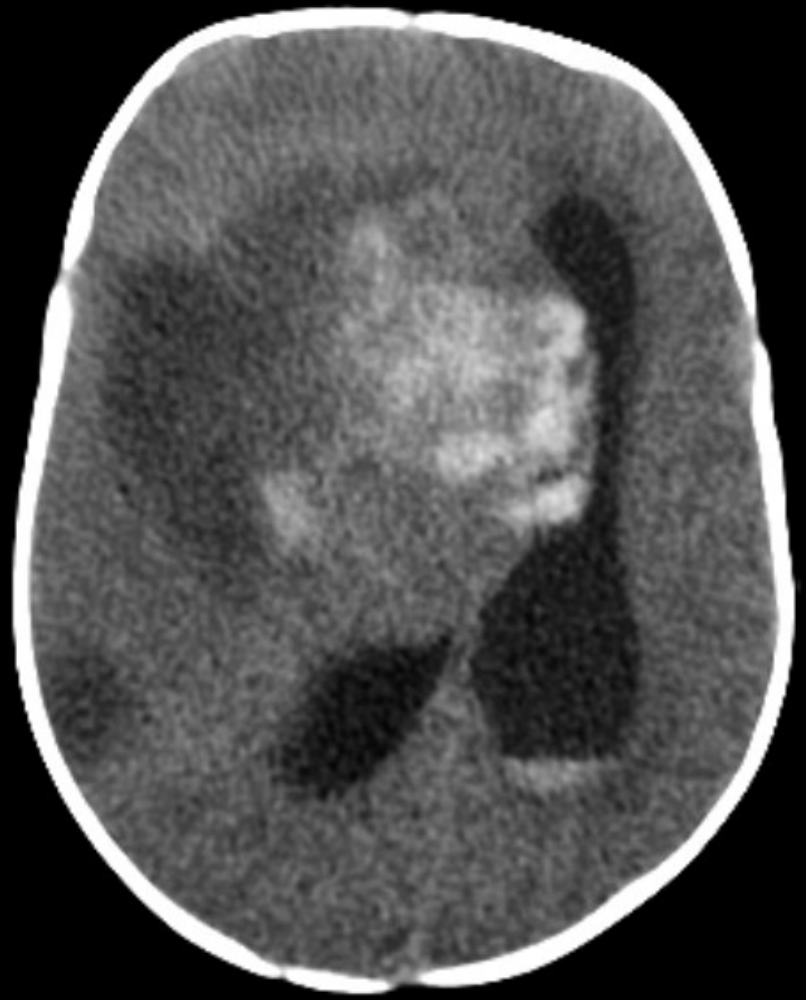
Findings (unlabeled)



Findings (labeled)

Large complex cystic and solid mass in the right frontoparietal region with internal hemorrhage

Ventricular dilation, indicating obstructive hydrocephalus

Leftward midline shift

Layering intraventricular blood products

What Additional Imaging Should We Order?

ACR Appropriateness Criteria:

Acute mental status change, known intracranial mass

Variant 2: **Acute or progressively worsening mental status change in patient with a known intracranial process (mass, recent hemorrhage, recent infarct, central nervous system infection, etc). Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
CT head without IV contrast	Usually Appropriate	☢☢☢
MRI head without and with IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
CT head without and with IV contrast	May Be Appropriate	☢☢☢
CT head with IV contrast	May Be Appropriate	☢☢☢

This imaging modality was ordered by the PICU team after facility transfer.

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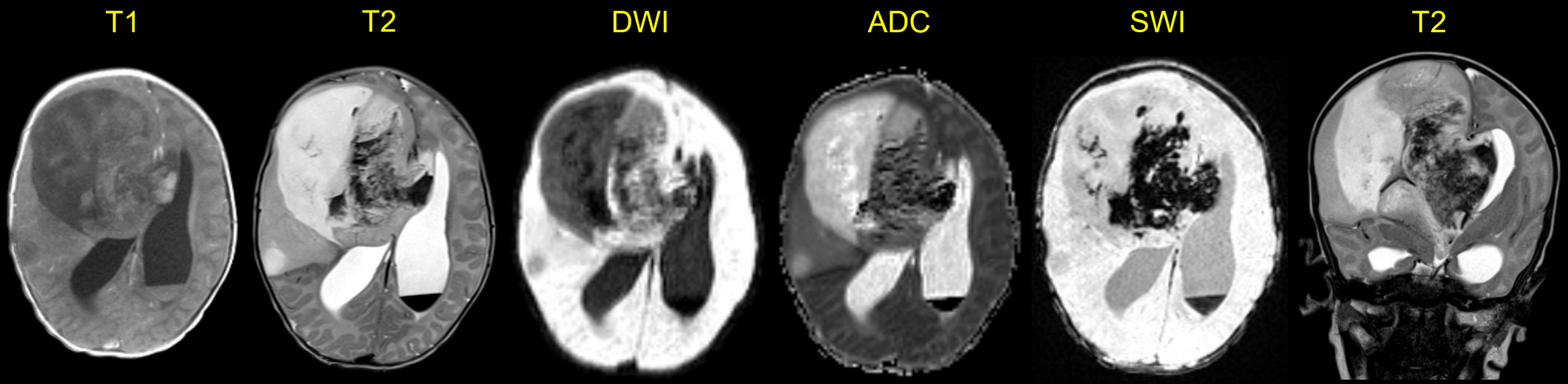
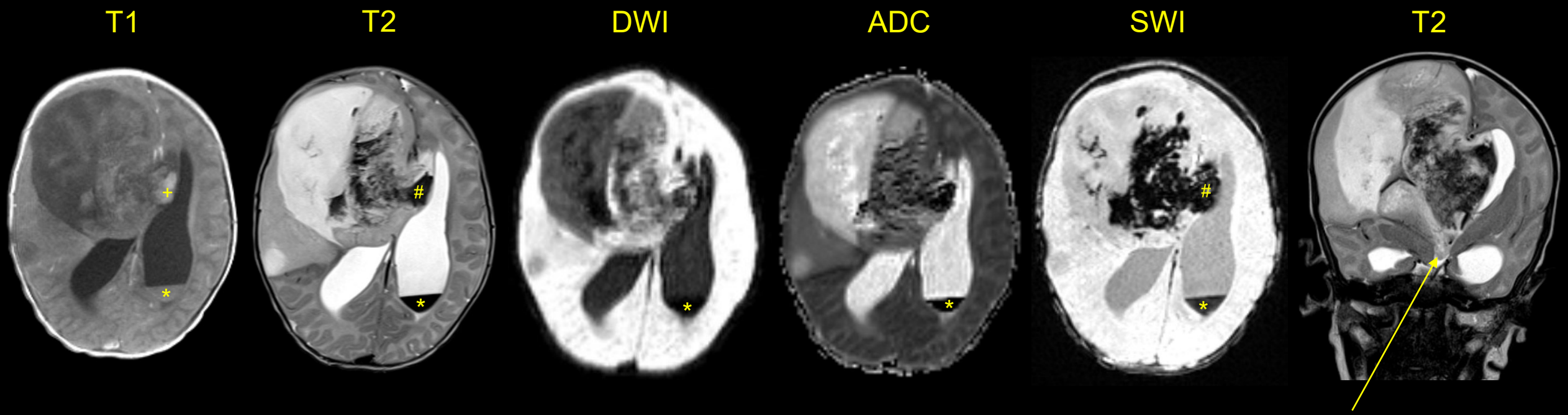
Findings (unlabeled)



Findings (labeled)



A 10.5 x 7 x 8.5 cm mixed cystic and solid mass is seen in the right frontoparietal region crossing midline with ~1.5 cm of leftward midline shift.

Large internal area of low signal intensity on T2/SWI images (#), with scattered high T1 intensity areas (+), most likely reflects intratumoral hemorrhage. Layering hemorrhage is seen in the left lateral ventricle(*).

The majority of the tumor demonstrates facilitated diffusion relative to gray matter on DWI and ADC images.

Tumor extension into the third ventricle with obstruction of the foramina of Monro results in obstructive hydrocephalus.

Final Dx:

Surgical Pathology: **Diffuse pediatric-type high-grade glioma**, CNS WHO grade 4, not otherwise represented in pathologic reference cohort

Case Discussion

- A recently recategorized tumor in the 5th edition of the WHO Brain tumor classification (2021), formerly pediatric glioblastoma
- Usually, supratentorial in location
- Although named “diffuse,” infant-type tumors can appear well-circumscribed as in this case
- Additional radiographic features may include contrast enhancement, intratumoral bleeding, and perilesional edema
- Differential diagnosis may include ependymomas, embryonal tumors, desmoplastic infantile tumors, or other types of gliomas

Case Discussion

- Treatment
  - Generally, gross total vs. subtotal resection followed by radiotherapy and chemotherapy
  - This patient underwent gross total resection with subsequent ventriculoperitoneal shunt placement
  - The patient developed post-operative seizures, now well-controlled medically, and is doing well
- Prognosis
  - Poor prognosis overall
    - May differ by molecular subtype
    - Studies reporting 14 – 44 months survival among subtypes

References:

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