

## Case Submission Guide 3.7.25 with sample case images

### **Selecting a case:**

The ACR Case-in-Point is a case-of-the-day CME program with the goals of teaching diagnostic and interventional radiology. Our target audience includes medical trainees and practicing radiologists. This program provides an opportunity for radiology trainees to generate scholarly work. Please use the lists below as a guide for selecting an appropriate case, submitting your case idea for review by our editors, and completing the case submission if your idea is accepted.

### **We are looking for cases that include:**

- Common imaging presentations of both common and uncommon diseases.
- Uncommon imaging presentations of common diseases.
- Pathognomonic imaging features (i.e., Aunt Minnies).
- Imaging findings that carry a classic differential diagnosis.
- Common Interventional Radiology (IR) procedures for common diseases with clear teaching points in imaging findings, techniques, or outcomes.
- Common IR procedures for uncommon diseases.

### **Cases to avoid:**

- Case with uncommon imaging presentations of uncommon diseases.
- Cases that leave doubt about the actual diagnosis.
- Cases with nonspecific imaging findings, poor image quality, or where the key imaging feature is missing.
- Cases with uncommon IR procedures for uncommon diseases.
- Cases with uncommon IR procedures not widely accepted as standard of care.

### **Case Suggestion Form:**

**Name, contact info, etc.** All authors should be in medical school and above.

**Diagnosis.**

**Differential Diagnosis Title.**

- This can be based upon a location (e.g., cerebellopontine angle cistern masses), an imaging feature (e.g., tree-in-bud opacities), or clinical feature (e.g., pediatric right lower quadrant pain). See 'Differential Diagnosis' section below for details.
- For IR procedures, this can be a disease or clinical situation requiring treatment (e.g., acute splenic laceration, or uterine arteriovenous fistula) or the name of the interventional radiology procedure. See below for details.

**Differential Diagnosis entities and description.**

- Provide 3-5 entities/diseases to include in the differential diagnosis. For each entity, in bullet format, list the most common imaging findings of the entity and how it can be distinguished from the disease you are presenting in the case. See below for details.

### **Questions section:**

- This should include 1-2 image-based questions from your case. Submit all your case images.
- Create 3-5 questions. These questions should be useful to those preparing for ACR In-Training exams (DXIT, TXIT), maintenance of certification exams, or test practical knowledge for the practicing radiologist.

**Options for question format:**

- Image-based point-and-click questions where the reader is asked to look at an image and click on something specific in the image, such as an abnormal finding or an anatomic structure.
- Multiple choice questions with a single correct response.
- Multiple choice questions with multiple correct responses.

**Options for question content:**

- Tests knowledge of the key imaging feature(s) of the disease.
- Tests knowledge of the differential diagnosis for an imaging finding or location-based differential diagnosis.
- Tests knowledge of imaging anatomy.
- Test knowledge of optimal treatment options of the disease and indications in IR.
- Test knowledge of key steps, imaging features, outcomes, and complications associated with IR procedures.
- Tests non-interpretive skills in diagnostic radiology include radiation safety, MRI safety, use of contrast media, contrast reactions, practical medical physics, imaging artifact identification, ethical considerations, giving bad news, etc.
- Questions about surgical/medical management, clinical diagnostic features, laboratory, or pathology findings are less important than imaging features but can be included only if the knowledge is useful to a practicing radiologist.

**What makes a good question?**

- No ambiguity: The correct answer(s) are clearly correct. Incorrect answer choices are clearly incorrect. If an answer choice could be disputed, then consider a different answer choice or different question.
- Be careful in your question construction that you are testing the reader's knowledge and not the reader's test-taking skill.
- Information is current: The optimal imaging, procedures, disease, and therapies can change over time.
- Limit question stems to 1-3 sentences. If medical history or the presenting complaint is included in the question stem, provide only the history that is directly relevant. This brevity is meant to simulate actual clinical radiology practice where the patient information provided with the imaging order is often minimal.

**Question explanations:**

- For each answer choice, describe why the response is either correct or incorrect. Treat this as a teaching opportunity. Limit the explanation to 1-3 sentences.

See sample case questions and answers in the published case format below:

### History

A 40-year old male presents with status epilepticus three weeks following stem cell transplantation.

### Question

1.1 Where is the pertinent abnormality located?

- A Cerebral cortex
- B Hippocampi
- C Thalami
- D White matter



### Explanation

#### Question

1.1 Where is the pertinent abnormality located?

You Selected **B: Hippocampi**

#### Explanation

✓ **The is abnormal hypodensity of the hippocampi. Normally, the hippocampi are hyperdense relative to white matter.**

#### Other explanations:

✗ **Cerebral cortex**

The grey-white matter differentiation of the cerebral cortex is preserved. There is normal relative hyperdensity of the cortex

✗ **Thalami**

The grey-white matter differentiation of the thalami is preserved. There is normal relative hyperdensity of the thalami.

✗ **White matter**

There is homogenous normal density of the white matter.

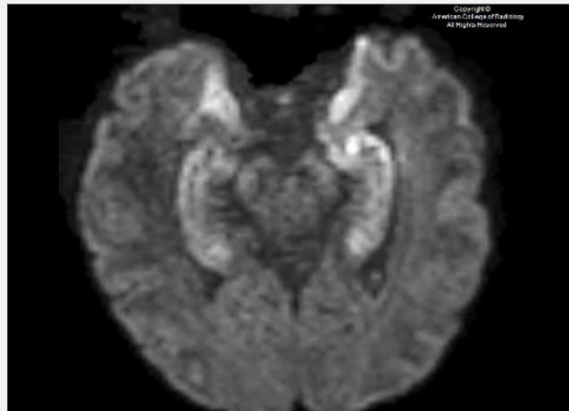
[Check your score](#)

### Question

2.1 Diffusion weighted imaging and ADC map (not shown) revealed abnormal diffusion restriction in the bilateral hippocampi. What should be included in the differential diagnosis for bilateral hippocampal diffusion restriction? (Check all that apply and click the Submit button.)

- Hypoxic-ischemic injury
- Diffuse axonal injury
- Human herpesvirus-6 infection
- Status epilepticus
- Herpes simplex encephalitis

Submit



### Question

2.1 Diffusion weighted imaging and ADC map (not shown) revealed abnormal diffusion restriction in the bilateral hippocampi. What should be included in the differential diagnosis for bilateral hippocampal diffusion restriction? (Check all that apply and click the Submit button.) (You have already attempted this question. The CORRECT RESPONSES are shown below.)

There were 5 items presented to you.

### Explanation

You got 40% (2 of 5) correct.

The following choices SHOULD have also been checked:

- Status epilepticus
- Herpes simplex encephalitis

The following choices SHOULD NOT have been checked:

- Diffuse axonal injury

Hypoxic-ischemic injury has variable patterns depending on the age of the patient and duration and degree of hypoxia. Bilateral hippocampal diffusion restriction is one such manifestation. The typical pattern of diffuse axonal injury are punctate foci of susceptibility and diffusion restriction in the peripheral white matter of the cerebrum, corpus callosum, deep grey nuclei and brainstem. Human herpesvirus-6 encephalitis most commonly manifests as bilateral hippocampal diffusion restriction and FLAIR hyperintensity. A common manifestation of status epilepticus is FLAIR hyperintensity with or without diffusion restriction in one or both hippocampi. Herpes simplex encephalitis commonly manifests with unilateral or bilateral hippocampal diffusion restriction.

[Check your score](#)

### Imaging Findings of [disease or IR procedure]:

- This section begins with the name of your case diagnosis. It follows with a list specifying the most common and important imaging findings of the disease, *and not necessarily the imaging findings encountered in your specific case*. Be sure to mention the appearance of the disease on different imaging modalities, if relevant. Also mention if one modality is most important for the diagnosis. Include 1 or 2 annotated images with captions **from your case** on this page and reference the annotations in your caption of imaging finding.
- **For IR procedures**, state the procedure name and list key steps of the technique, equipment, key imaging findings, and outcomes associated with the procedure.

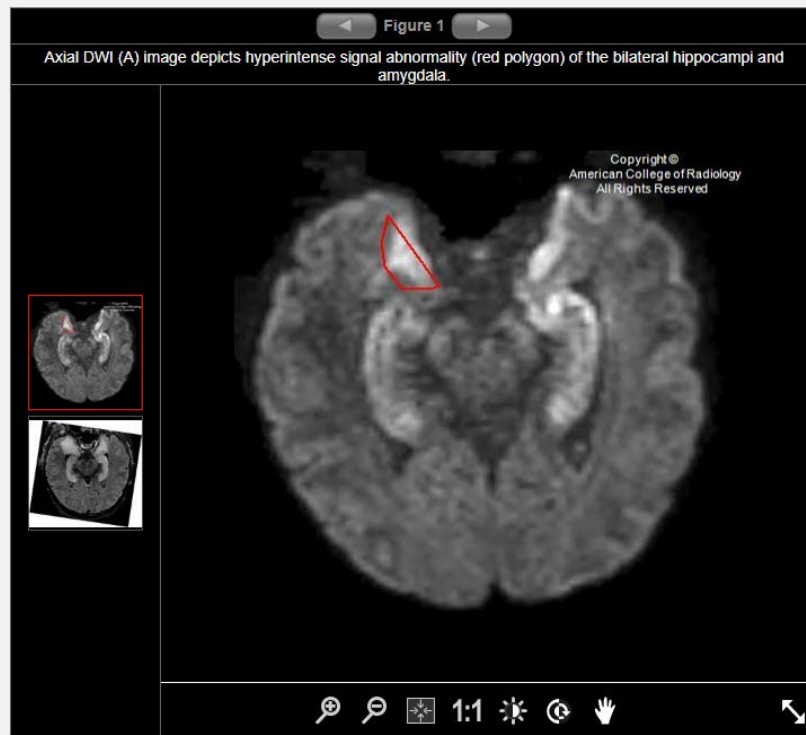
### See a sample of the Imaging findings page of a case (also includes your case diagnosis):

#### Diagnosis

Human herpes virus 6 encephalitis

#### Imaging Findings of Human herpesvirus 6 Encephalitis

- T2/FLAIR hyperintensity and diffusion restriction of the bilateral hippocampi and amygdala is most common.
- There is no abnormal contrast enhancement.
- Swelling is uncommon – a feature useful for distinguishing HHV6 from herpes simplex encephalitis1.
- On CT, hypoattenuation is uncommonly seen. Atrophy is uniformly seen in the chronic phase, after 30 days.



### **Differential Diagnosis of [lesion location, imaging feature, clinical feature]:**

- Please select a feature from your case to create a list of differential diagnoses. The differential diagnosis should be based on one of the three options below. You must specify what your list of differential diagnoses is in reference to (examples below). *The name of the disease you are presenting is not an acceptable title for your differential diagnosis list.*
  1. **Lesion location:** For example, you would specify cerebellopontine angle cistern masses, lesions of the appendix, terminal tuft masses, etc.
  2. **Imaging feature:** For example, you would specify sunburst periosteal reaction, ring-enhancing brain lesion, tree-in-bud opacities, bilateral basal ganglia T2 hyperintense signal abnormality, etc.
  3. **Clinical feature:** For example, you specify pediatric sensorineural hearing loss, hemoptysis, fever in immunocompromised patients, right lower quadrant pain, etc.
- Choose 3-5 entities/diseases to include in your differential diagnosis. For each disease, create a list of 4-6 concise bullet points that detail the most common imaging findings of the disease and the best way to differentiate the alternative diagnosis from the disease you are presenting in your case. Generating and narrowing differential diagnosis lists are some of the most important functions of the radiologist. Accordingly, this section is valuable to our readers.
- For each entity you include in the differential diagnosis section, submit one image depicting typical imaging findings of the disease. Preferably the image comes from your own case file/institution and must not have been published previously. If that is not available, then search the Case-in-Point case archive for the disease and take a screen capture of a representative image depicting the disease. Annotate each image and provide a brief caption that starts with the disease name. For example, "Status Epilepticus: Axial DWI images depict low-level diffusion restriction in the bilateral hippocampi."
- Additionally, for each entity you include in the differential diagnosis section, search the Case-in-Point archive for that disease and provide the Case ID number of that case. Our editors will use this to create a hyperlink within your case.
- **For IR procedures**, this section can be used for:
  - Differential diagnoses of imaging finding
  - Describing related or alternative IR procedure options
  - Presenting different techniques or equipment in the same procedure
  - Demonstrating potential different outcomes and complications associated with the procedure.

## See a sample of the Differential Diagnosis page below.

### Differential Diagnosis of Bilateral hippocampal T2/FLAIR hyperintensity

#### Status epilepticus:

- Most commonly manifests as T2/FLAIR hyperintensity and swelling of cerebral cortex, medial temporal lobes and hippocampi, pulvinar of the thalamus, and subcortical white matter. Signal abnormality confined to just the bilateral hippocampi and amygdala is uncommon.
- Can be unilateral or bilateral.
- Can be associated with diffusion restriction.

#### Hypoxic-ischemic injury :

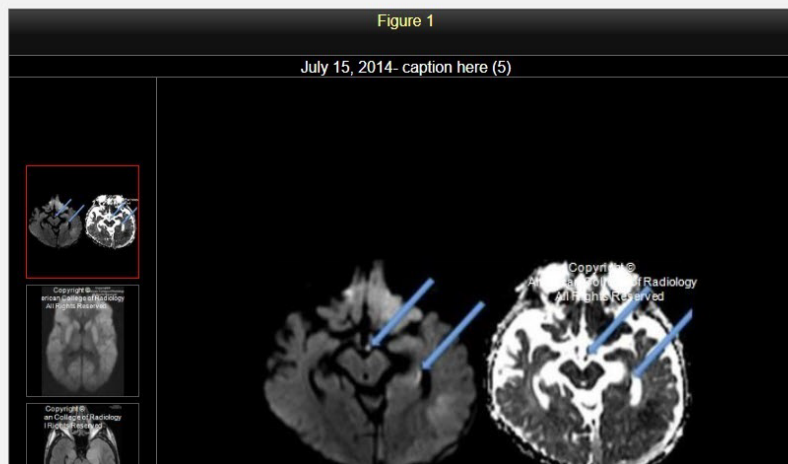
- Patterns of brain injury depend on patient age along with severity and duration of hypoxia-ischemia.
- In adults with severe HII, diffusion restriction and FLAIR hyperintensity of grey matter is most common (deep grey nuclei > cerebral cortex > hippocampi).
- In adults with mild or moderate HII, watershed pattern of infarction is most common.

#### Herpes simplex encephalitis :

- In addition to signal FLAIR and DWI abnormality of the hippocampi (unilateral or bilateral), HSE will almost always involve other parts of the brain.
- Some degree of asymmetric involvement of the insula, inferior frontal lobes, deep grey nuclei and other parts of the temporal lobes is almost always present.
- Intraparenchymal hemorrhage is also common.

#### Autoimmune encephalitis:

- This category includes a wide variety of paraneoplastic and non-paraneoplastic causes with variable imaging appearances.
- Bilateral mesial temporal lobe FLAIR hyperintensity and diffusion restriction is a common pattern.
- Can be indistinguishable from HHV6 encephalitis on imaging.



### Clinical Issues of diagnosis:

- In a paragraph of 150 words or less, provide an overview of the key clinical features of the disease you are presenting. This is a generic, general discussion of the disease and should not reference your specific case. It can include some or all the following: typical clinical presentation of the disease, disease prevalence, demographics, diagnosis, natural history, prognosis, pathology findings, treatment, or prognosis. Please describe what role imaging plays in the diagnosis; however, do not rehash all the imaging findings of the disease, as already seen in "Imaging Findings".
- **For IR procedures**, this section also includes a discussion of general indications, key technical aspects, expected outcomes, complications, and efficacy of the procedure.

### Case Discussion:

- In a paragraph of 150 words or less, provide an overview of your patient's specific case. It should include a concise summary of your patient's presentation, diagnosis, imaging findings, clinical course, specific IR procedure or treatment and outcome. Also, it can address the following:
  1. Were any of your case's clinical features or imaging findings atypical for the disease?



2. Did your patient's images pose a diagnostic challenge or leave some uncertainty?
3. Was there an imaging finding or clinical feature that clinched the diagnosis?
4. In reading the images or preparing this case, did you learn something that others may benefit from?
5. For IR procedures, were there any unique or challenging aspects or steps in the technique?
6. For IR procedures, were there any complications or negative outcomes that could have been avoided?

**See a sample of the Clinical Issues & Case Discussion page below.**

#### **Clinical Issues of human herpes virus 6 encephalitis**

Human herpes virus-6 (HHV-6) is ubiquitous with up to 90% seropositive rate by two years of age<sup>2</sup>. The primary infection presents in childhood as a self-limited disease characterized by fever, rash (roseola) and pharyngitis. The virus can remain latent in a variety of tissues and organs. The most common presentation of HHV-6 in adulthood is a reactivation of the virus in the few weeks following stem cell or solid organ transplantation. Clinically, patients present with fever, behavior changes, antegrade amnesia and seizure. Only two-thirds of patients with HHV-6 encephalitis will have findings on brain MRI<sup>3</sup>, typically signal abnormalities of the bilateral medial temporal lobes. Analysis of blood and CSF for DNA evidence of HHV-6 is confirmatory. Ganciclovir and foscarnet are commonly used in treatment<sup>4</sup>; HHV6 is resistant acyclovir. Long-term sequelae include seizure and antegrade amnesia.

#### **Case Discussion**

Our patient's case of HHV-6 encephalitis followed a typical clinical course. He presented 3 weeks following hematopoietic stem cell transplantation for acute lymphocytic leukemia. His presenting symptoms included fever and seizure and his initial work-up included lumbar puncture and brain MRI, among other tests. The MRI depicted typical imaging features of HHV-6 encephalitis – T2/FLAIR hyperintensity and diffusion restriction of the bilateral hippocampi and amygdale. Based on the presenting symptoms and MRI, there was some diagnostic concern for herpes simplex encephalitis (HSE). However, the symmetric distribution of signal abnormality, lack of swelling/mass effect, and lack of intraparenchymal hemorrhage on MRI all favored HHV-6 rather than HSE. Following treatment with intravenous ganciclovir, the patient's encephalitis resolved and he has been seizure-free although he reports cognitive slowing since th

#### **References:**

- Cite any references you used in writing this case. Citations are required when discussing numbers or findings from the peer-reviewed literature. Common radiological or medical knowledge does not require citation. Use AMA citation format as seen below.

#### **References**

1. Noguchi T, Yoshiura T, Hiwatashi A, et al. CT and MRI findings of human herpesvirus 6-associated encephalopathy: Comparison with findings of herpes simplex virus encephalitis. *AJR*. 2010; 194:754–760. DOI:10.2214/AJR.09.2548
2. Zerr D, Meier A, Selke S, et al. A population-based study of primary human herpesvirus 6 infection. *N Engl J Med* 2005; 352:768-776. DOI: 10.1056/NEJMoa042207
3. Berzero G, Campanini G, Vegezzi E et al. Human herpesvirus 6 encephalitis in immunocompetent and immunocompromised hosts. *Neuro Immunol Neuroinflamm*. 2021;8(2):e942; DOI: 10.1212/NXI.0000000000000942
4. Ward K, Hill J, Hubacek P, et al. Guidelines from the 2017 European Conference on Infections in Leukaemia for management of HHV-6 infection in patients with hematologic malignancies and after hematopoietic stem cell transplantation. *Haematologica*. 2019; 104(11):2155-2163. doi:10.3324/haematol.2019.223073

#### **Authors:**

- Cases should include at least 1 radiologist at the attending level. All authors should be at the level of medical school and above. For any pathology images please include the pathologist as an author, especially if they have written captions.



**CME questions:**

- Submit five true-or-false type questions that relate to the content provided in the case. Please ensure there is no ambiguity about the correct answer.

**CME Posttest**

**Not yet published**

**Question**

**1.1** evaluate the following (Please respond to the following with TRUE or FALSE.)

- T  F The most common MRI findings of HHV-6 are FLAIR hyperintensity and diffusion restriction in an asymmetric destruction the bilateral medial and anterior temporal lobes, insulae and frontal lobes.
- T  F The primary medical therapy for HHV-6 encephalitis acyclovir.
- T  F Intraparenchymal hemorrhage and swelling/mass effect can both help in distinguishing HHV-6 encephalitis from HSE.
- T  F Status epilepticus, autoimmune encephalitis, and hypoxic ischemic injury can all result in T2/FLAIR signal abnormality of the hippocampi.
- T  F Upward gaze palsy (parinaud syndrome) is a common presentation of HHV-6 encephalitis.