

Case 2: Answer

from Tokyo Teishin Hospital

Summary of Clinical history

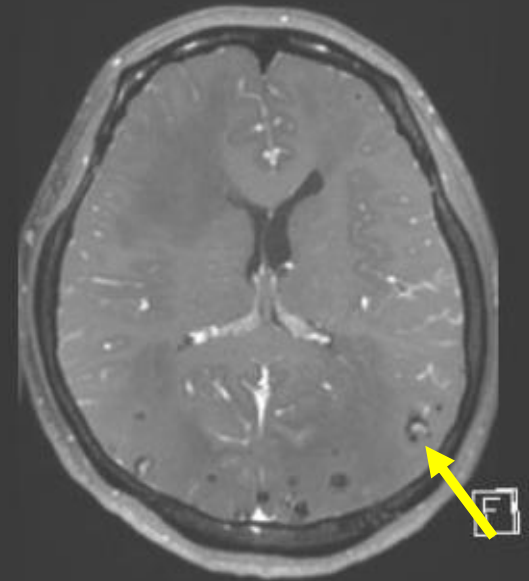
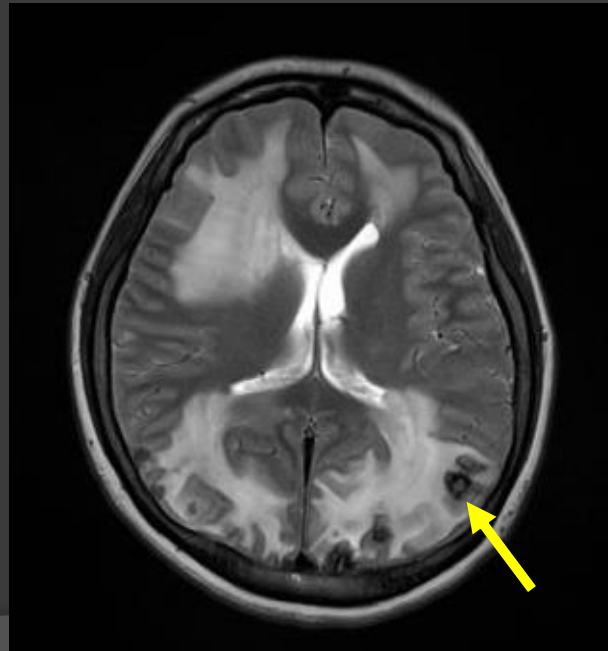
- 24 y.o. F
- cough, hemoptysis, appetite loss, BW loss of 5kg which started 7 months ago.
 - systemic manifestation
- visual disturbance of the rt. eye & severe headache 1 month ago.
 - neurologic manifestation

Rather slow progression

Summary of Imaging findings

Brain:

- **Multiple small hemorrhagic foci** throughout the brain accompanied by extensive perilesional edema.
- Most of the lesions are hypointense on T2WI, FLAIR & DWI.
- Some of them show partial contrast enhancement.
- The lesions are most evident as very hypointense nodules on SWI.



DDx: multiple hemorrhagic brain lesions

- Amyloid angiopathy
- Multiple cavernous malformations
- Angiitis
- Metastases from easily hemorrhagic lesions, such as melanoma, choriocarcinoma, angiosarcoma, epithelioid hemangioendothelioma, etc.

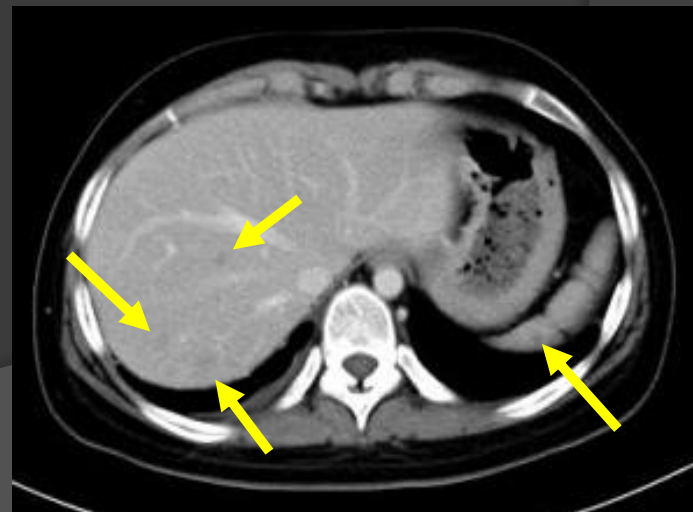
Summary of Imaging findings

Lungs:

- **Multiple small nodules** through all lung fields.
- **Multiple irregular-shaped consolidations** with air-bronchogram, **surrounding GGO** ← s/o hemorrhage.
- Interlobular septal thickenings.

Liver & Spleen:

- **Multiple small nodules**, heterogeneous densities



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Result of brain biopsy

- Sheet-like proliferation of cells w/ clear cytoplasm and round nuclei.
- Cytoplasm has some vacuoles of various sizes, some of which contain red blood cells.
- Among such cells, there are fine vascular channels associated w/ congestion, hemorrhage and hemosiderin deposition.
- Immunostaining

Positive – CD31 and Vimentin

Negative – S-100, NF, GFAP, Ibal, CD1a, CK AE1/3, EMA, LCA, CD68, and α SMA

Final Diagnosis

**Epithelioid Hemangioendothelioma
(EHE)**

involving the brain, lungs, liver & spleen.

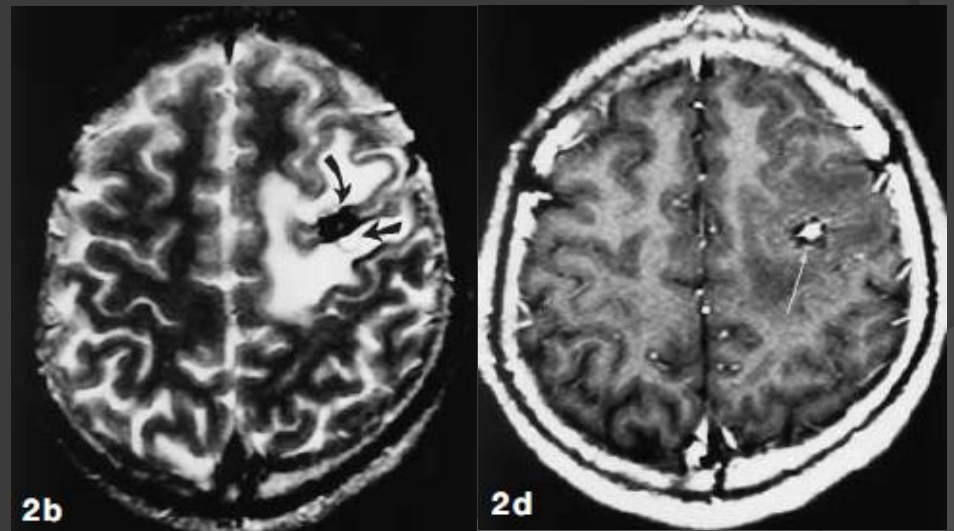
Epithelioid Hemangioendothelioma (EHE)

- Vascular neoplasm in the intima of blood vessels, described in veins of the extremities as well as in the liver and lungs.
- Clinically and histologically considered intermediate between angiosarcoma and hemangioma (low grade malign.).
- Can occur singly or in a multicentric pattern.
- Cerebral involvement is rare (0.02% of all brain tumors) but sometimes occurs as intracranial metastases.
- Recommended treatment includes excision, radiotherapy, and interferon α -2b.

Yeo SK et al. J Korean Neurosurg Soc 2007;42:129-131.
Endo T et al. J Neurooncol 2004;67:337-343.
Hamlat A et al. J Neurooncol 2004;67:361-366.

Cerebral imaging findings of EHE

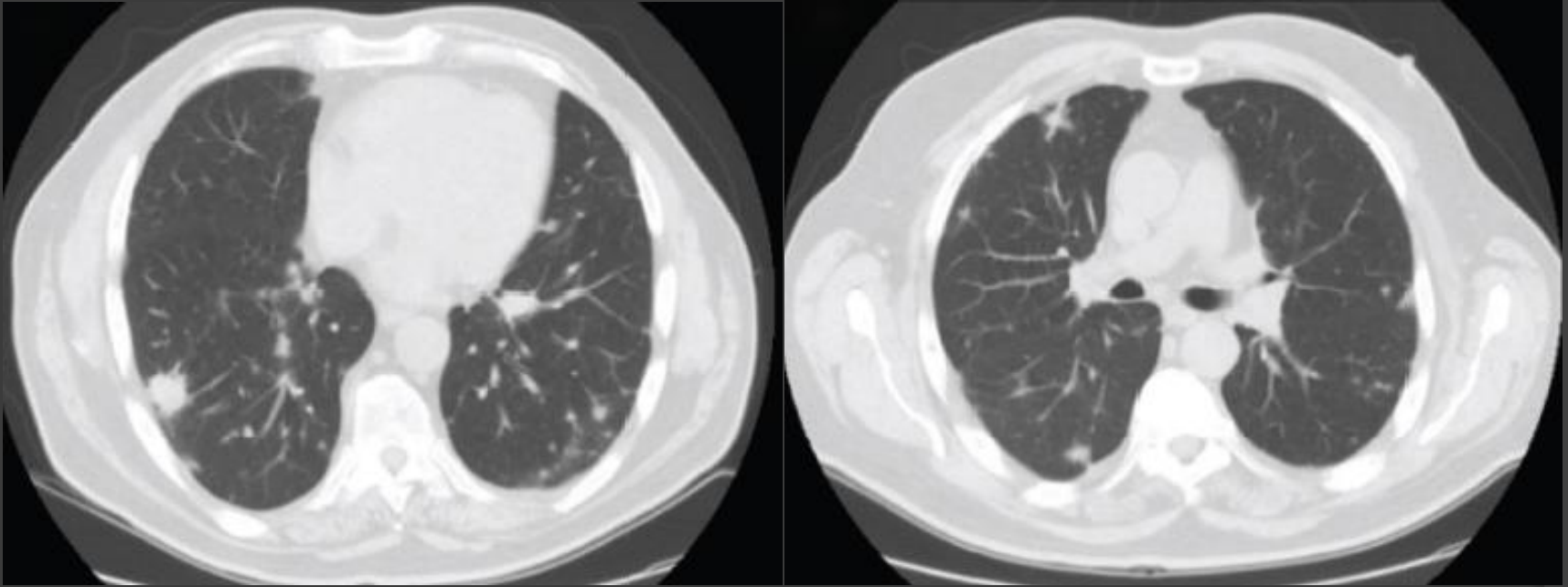
- Solid appearance with variable intensities on T1WI and hyperintensity/heterogeneous intensity on T2WI.
- Typically shows diffuse and strong enhancement on CE-T1WI, with or without peripheral hypointensity on T2WI.
- Solid-cystic variant and a fatty component have also been reported.



Chan YL et al. Neuroradiology 2001;43:848-850.

Rocha Oliveira PC et al. Arq Neuropsiquiatr. 2012;70:637-638.

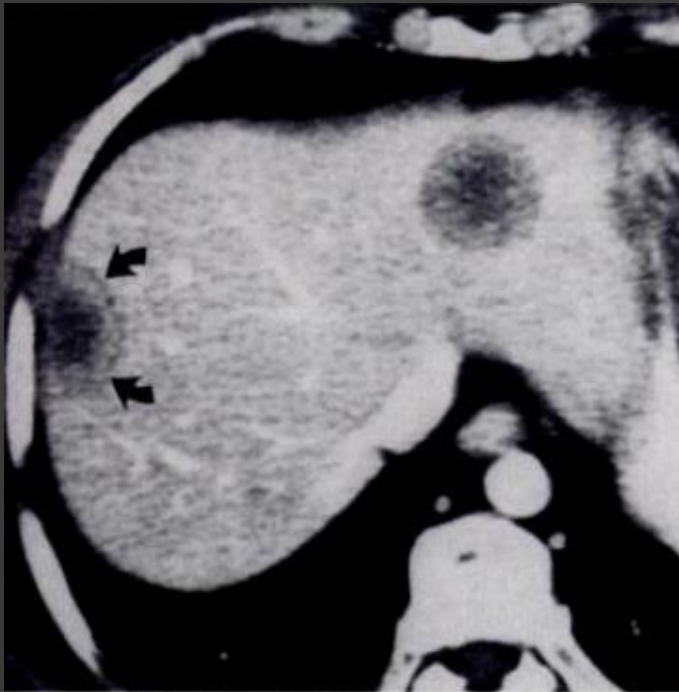
Pulmonary imaging findings of EHE



Cazzuffi R et al. Case Rep Med. 2011;2011:262674

- Multiple pulmonary nodules (IVBAT: Intravascular bronchioloalveolar tumor)
- Can be modified by tumoral hemorrhage

Hepatic imaging findings of EHE



Miller WJ et al. Am J Roentgenol. 1992;159(1):53-7.

- Often laminated, conglomerated in the liver, in the peripheral location predominantly, capsular retraction

Take Home Points

- A single or multiple extensively hemorrhagic lesion(s) in the brain could be EHE, especially in a slowly progressive presentation.
- Search for involvement of other organs is recommended after suggesting an intracranial EHE.

References

1. Yeo SK, Kim JH, et al: Intracranial epithelioid hemangioendothelioma. J Korean Neurosurg Soc 2007;42:129-131.
2. Endo T, Su CC, et al: Malignant intracranial epithelioid hemangioendothelioma presumably originating from the lung: case report. J Neurooncol 2004;67:337-343.
3. Hamlat A, Casallo-Quilliano C, et al: Epithelioid hemangioendothelioma of the infundibular-hypothalamic region: case report and literature review. J Neurooncol 2004;67:361-366.
4. Chan YL, Ng HK, et al: Epithelioid haemangioendothelioma of the brain: a case report. Neuroradiology 2001;43:848-850.
5. Rocha Oliveira PC, Alcantara FP, et al: Cerebral epithelioid hemangioendothelioma with thoracic simultaneous involvement: advanced MRI features. Arq Neuropsiquiatr. 2012;70:637-638.