



## Case Report

# Cerebral Abscess as a first Presentation for Hereditary Haemorrhagic Telangiectasia: A Case Report

**Shareefa Abdulghaffar\*, Noor AlBadrawi, Ahmed Bedair Mohamed, Ebrahim Ahmed Mohamed**

<sup>1</sup>Rashid Hospital, Department of Radiology, Dubai Health Authority, PO Box 4545 Dubai, United Arab Emirates

**\*Corresponding author:** Shareefa Abdulghaffar, Rashid Hospital, Department of Radiology, Dubai Health Authority, PO Box 4545 Dubai, United Arab Emirates

**Citation:** Abdulghaffar S, AlBadrawi N, Mohamed AB, Mohamed EA. (2021) Cerebral Abscess as a first Presentation for Hereditary Haemorrhagic Telangiectasia: A Case Report. Ann Case Report 6: 715. DOI: 10.29011/2574-7754.100715

**Received Date:** 13 December 2021; **Accepted Date:** 17 December, 2021; **Published Date:** 22 December 2021

### Abstract

Hereditary Haemorrhagic Telangiectasia (HHT) is a rare autosomal dominant disease with multisystem and multi-organ involvement. Patients usually present with recurrent epistaxis, mucocutaneous telangiectasia and arteriovenous malformations (AVMs) in multiple organs in the body including the lungs and brain. Hereditary Haemorrhagic Telangiectasia is a clinical diagnosis based on the presence of the previously mentioned disease manifestations or family history of the disease. Presence of pulmonary arteriovenous malformations in these patients increases the risk of septic or non-septic embolization to the systemic circulation. We present a case of a 32-year-old Asian male with acute onset of confusion and fever. Imaging with contrast enhanced computed tomography of the brain and chest showed multiple cerebral abscesses and pulmonary AVMs. The cerebral abscesses were most likely sequelae of paradoxical septic embolization from the multiple pulmonary AVMs.

**Keywords:** Hereditary Haemorrhagic Telangiectasia; Cerebral abscess; pulmonary arteriovenous malformations; Osler-Weber-Rendu

### Case presentation

A 32-year-old Asian male, previously healthy, presented to the Accident & Emergency Department with a history of sudden onset of acute confusional state associated with fever, chills, and headache for the last 3 days. No history of loss of consciousness, seizures, sensory or motor loss, night sweats, abdominal pain, nausea or vomiting. He denied any history of previous similar episodes. There was no significant past medical, surgical, or family history.

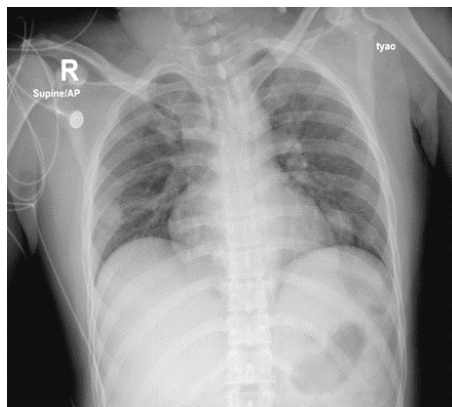
On physical examination, the patient was febrile (low-grade fever with temperature of 38°C), yet hemodynamically stable. GCS was 10 out of 15. He was confused and not oriented to person, place, time and unable to obey one-step commands. Neurological examination showed reactive pupils bilaterally, positive neck

stiffness, up-going plantar reflex on right side and proptosis of both eyes. Otherwise, rest of cranial nerves were intact with preserved gait, sensory & motor power in four limbs.

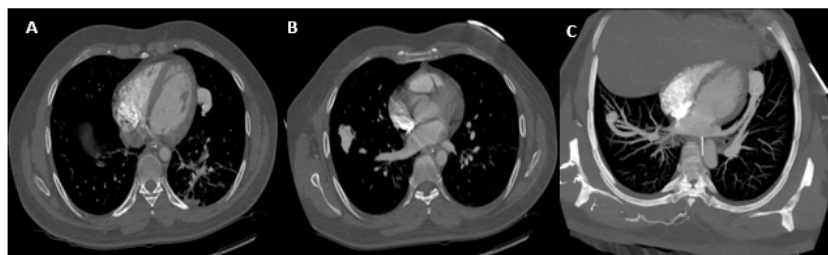
### Investigations & Imaging findings

Basic laboratory and haematological findings revealed elevated white blood cell count (WBC = 24.3 x 10<sup>3</sup> u/L), raised erythrocyte sedimentation rate (ESR = 41 mm/hr) and C-reactive protein (CRP = 114.4 mg/L). Lumbar puncture was done and Cerebrospinal fluid (CSF) culture showed growth of *Citrobacter Koseri*, *Pseudomonas Aeruginosa* and *Klebsiella Pneumoniae*. Acid fast bacilli test was negative for *Mycobacterium Tuberculosis* infection.

Chest radiograph showed multiple lung nodules in lower zones of both lungs with prominent vessels shadows seen merging with them (Figure 1). Further evaluation with contrast-enhanced computerized tomography (CT) of the chest (Figure 2) revealed bilateral pulmonary AVMs.

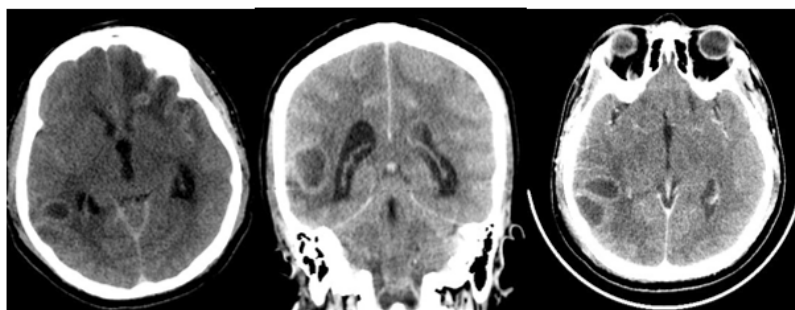


**Figure 1:** Antero-posterior supine chest radiograph showing two well-defined nodules in lower zones of both lungs with prominent vascular shadows merging with them.



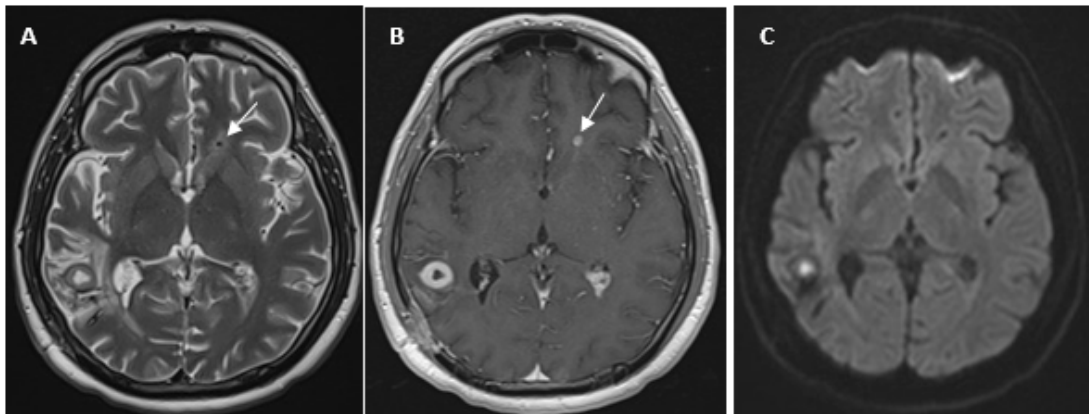
**Figure 2:** Contrast-enhanced CT scan of the chest. Image (A) Axial section of the chest showing a lobulated pulmonary nodule in the inferior lingular segment of the left upper lobe supplied by an enlarged segmental pulmonary artery branch and drained by an enlarged left inferior pulmonary vein (red arrow). Image (B) showing another right lower lobe lobulated enhancing nodule supplied by a segmental pulmonary artery branch and drained by an enlarged right inferior pulmonary vein (white arrow). Image (C) Axial MIP images of the chest showing both bilateral lower lobe nodules drained by the bilateral corresponding inferior pulmonary veins.

In view of the patient's neurological symptoms, contrast-enhanced CT scan of the brain was also performed and it revealed multiple, small, supra-tentorial, cerebral abscesses in the left frontal and right parieto-occipital lobes, associated with mild hydrocephalus. Abnormal leptomeningeal and ventricular wall enhancement was noted, consistent with meningitis and ventriculitis (Figure 3).



**Figure 3:** (A,B) Axial and coronal Contrast enhanced CT scan of the brain showing multiple ring enhancing cerebral lesions with surrounding vasogenic oedema noted at the left frontal and right parieto-occipital regions (white arrows) . Image (c) Axial images of the brain showing abnormal enhancement of the leptomeninges and the ventricular ependymal lining in keeping with meningitis and ventriculitis.

Throughout admission, the patient's clinical condition deteriorated and a follow up contrast enhanced MRI study of the brain was done and it confirmed the CT brain findings (Figure 4).



**Figure 4:** Axial MR images of the brain (A) Axial T2-weighted image showing a right parieto-occipital region well-defined hyperintense cystic lesion with hypo-intense rim and surrounding oedema. A similar but smaller lesion noted at the left frontal lobe (white arrow). (B) Both lesions show thick rim of peripheral enhancement on axial gadolinium-enhanced T1-weighted image. (C) The right parieto-occipital lesion shows central area of restricted diffusion on DW image.

### Treatment & Outcomes

In view of the patient clinical presentation, radiological and laboratory findings, the patient was admitted under the care of neurosurgical team and underwent insertion of external ventricular drainage through a burr hole. Patient was kept in intensive care unit and under air-borne precautions. He completed a 6 weeks course of IV therapy of meropenem and vancomycin. Unfortunately, the patient continued to deteriorate clinically with persistent low GCS throughout admission and passed away.

### Discussion

Hereditary Haemorrhagic Telangiectasia (HHT), or Osler-Weber-Rendu disease, is a rare autosomal dominant multi-organ disorder. While it is a multi-system disease, it is mainly characterized by multiple arteriovenous malformations (AVMs) involving nasal cavity (90%), skin and mucosal membranes (90%), liver (30%), gastrointestinal tract (20%), pulmonary vasculature (50%) and CNS (5-10%) [1,2].

Healthy patients suddenly presenting with multiple cerebral abscesses and no previous history of an underlying cause should raise the suspicion of pulmonary AVMs. The abnormal pulmonary vasculature is known to cause cerebral abscesses which are serious neurological complications seen in up to 10% of these cases [3]. It is thought to be a result of paradoxical septic embolization passing through the abnormal arteriovenous fistulous communication and bypassing the pulmonary capillary bed and lodged in the grey-white matter junction of the brain parenchyma with subsequent cerebral abscess formation [1,4]. Therefore, brain abscess should

be suspected in HHT patients presenting with neurological symptoms. Patient with HHT can also have other central nervous system involvement including cerebral ischemic infarction, cerebrovascular malformation (CVM), and metabolic disorders [2,5]. Trans-arterial vaso-occlusion is indicated in cases of PAVMs particularly those with feeding arteries exceeding 3mm in diameter as they are more likely to shower emboli [3].

Cross-sectional imaging with contrast-enhanced CT scans of the brain, chest and abdomen is the modality of choice in assessment of possible HHT to identify the vascular malformations. They would reveal the presence of visceral AVMs commonly involving the brain, lungs and liver [5]. The gold standard for diagnosis of PAVMs is by thin sections contrast enhanced CT of the chest which is superior to digital subtraction angiography due to its higher sensitivity and non-invasive nature [6]. CT of the chest also allows assessment of the diameter of the feeding arteries of these AVMs to correctly direct the clinicians to proper treatment [3]. Trans-thoracic contrast echocardiography is the screening test of choice for PAVMs; however, definite diagnosis of HHT will eventually require genetic testing for the responsible mutation [3,6].

Treatment of cerebral abscess is planned through multidisciplinary approach, involving both medical and surgical approach. It is generally recommended for the patient to undergo surgical drainage if the abscess measures >2.5 cm in diameter, 6–8 weeks of intravenous antibiotic treatment, and to have serial imaging through CT or MRI every 15–20 days [7,8]. In our case, the patient's diagnosis was delayed and was found to be complex with the presence of the AVMs. In addition to that, the patient's

poor clinical status did not allow for surgical drainage of the brain abscesses or to undergo pulmonary digital subtraction angiography of the PAVMs for diagnostic and therapeutic purposes to the possible underlying cause of the multiple abscesses.

In conclusion, HHT can cause radiological and treatment challenges, and knowledge of the pathology allows prompt diagnosis and management of its complications.

### Learning points

1. HHT manifests as arteriovenous malformations in different parts of the body.
2. Radiology plays an important role in the diagnosis and management of HHT through image-guided endovascular therapy.
3. Undiagnosed cases of HHT may lead to high mortality and poor outcomes due to increased risk of embolic stroke and brain abscess, as presented in this case.

### References

1. Jaskolka J, Wu L, Chan RP, Faughnan ME. (2004) Imaging of hereditary hemorrhagic telangiectasia. *AJR Am J Roentgenol.* 183: 307-314.
2. Carette MF, Nedelcu C, Tassart M, Grange JD, Wislez M, et al. (2009) Imaging of hereditary hemorrhagic telangiectasia. *Cardiovasc Intervent Radiol.* 32: 745-757.

3. Olitsky SE. (2010) Hereditary hemorrhagic telangiectasia: diagnosis and management. *Am Fam Physician.* 82: 785-790.
4. Todo K, Moriwaki H, Higashi M, Kimura K, Naritomi H. (2004) A small pulmonary arteriovenous malformation as a cause of recurrent brain embolism. *AJNR Am J Neuroradiol.* 25: 428-430.
5. Moradi M, Adeli M. (2014) Brain abscess as the first manifestation of pulmonary arteriovenous malformation: A case report. *Adv Biomed Res.* 3: 28.
6. Brouwer MC, Coutinho JM, van de Beek D. (2014) Clinical characteristics and outcome of brain abscess: systematic review and meta-analysis. *Neurology.* 82: 806-813.
7. Saboo SS, Chamarthy M, Bhalla S, Park H, Sutphin P, et al. (2018) Pulmonary arteriovenous malformations: diagnosis. *Cardiovascular diagnosis and therapy,* 8: 325-337.
8. Sell B, Evans J, Horn D. (2008) Brain abscess and hereditary hemorrhagic telangiectasia. *South Med J.* 101: 618-625.