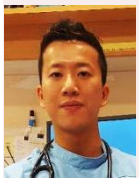
CLINICAL MEETING SUMMARIES ON 27TH SEPTEMBER 2018

Case report of pulmonary arteriovenous malformation – a not uncommon fatal disease in young fit population



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Case presentation

We are here to present a young fit lady who was diagnosed to have single large pulmonary arteriovenous malformation in right upper lobe, during her admission for difficult-to-treat brain abscess and complicated by massive pulmonary embolism.

Miss Wong was a 52 years old housewife who had no significant family, medical or surgical history. She was admitted to hospital for mild right-side weakness for 20 hours. Upon arrival to emergency department, she was fully conscious, stable vital signs, normal pulmonary, cardiovascular and abdominal findings. She was not known to have any lung lesions except the subtle increased vascular markings in right upper lobe in chest X ray during the index admission (Figure 1).

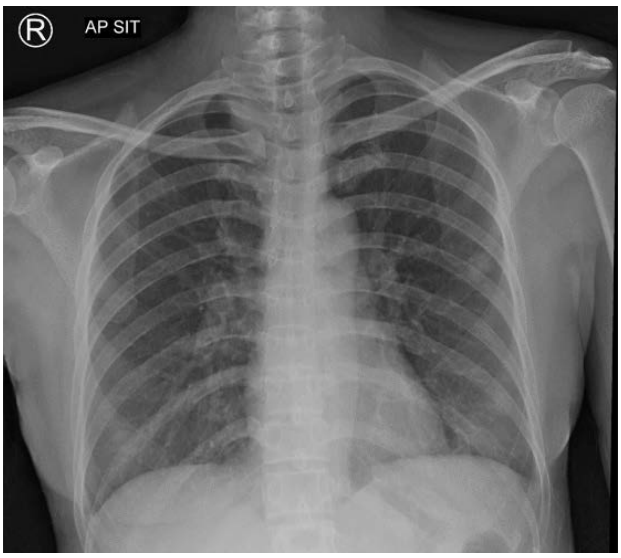

Figure 1: Initial chest X ray showing subtle increase in vascular marking in right upper lobe

This episode was first diagnosed as cerebrovascular event since CT brain on day 0 of admission showed edematous change in left parasagittal region. The weakness rapidly deteriorated to total right hemiparesis on day 2. The primary diagnosis was changed to brain abscess, as patient developed fever and serial CT brain showed more obvious 2 cm iso-dense lesion within the vasogenic edema in the left parasagittal region. It was followed by marked leukocytosis and haemoglobin drop from 8g/dL to 5.6g/dL. The diagnosis was confirmed by MRI brain on day 3 (Figure 2), and emergency operation for brain abscess drainage was done by neurosurgical unit, yielding oral flora, Streptococcus intermedius and Peptostreptococcus from the pus.

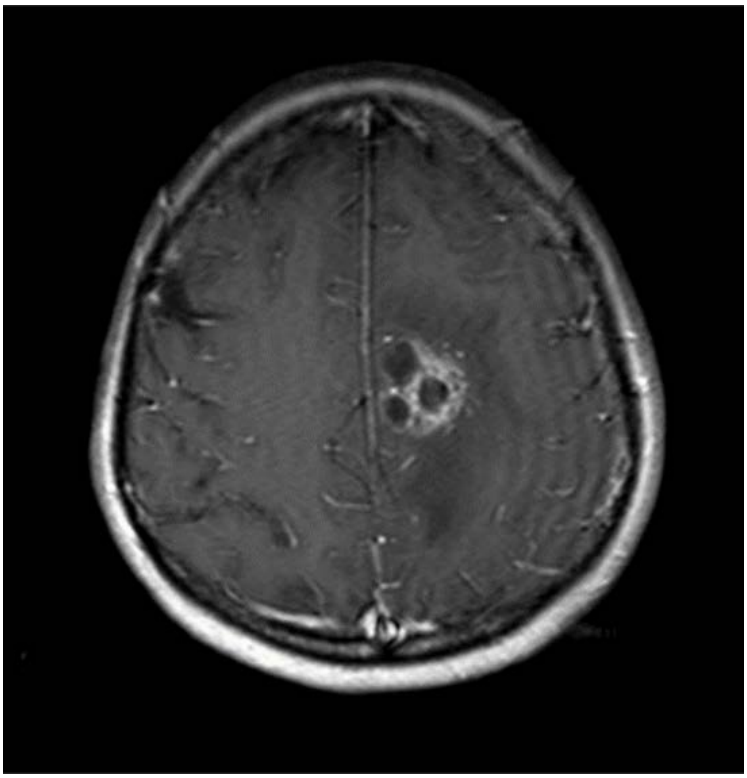

Figure 2: Left parasagittal region brain abscess on day 3 of admission

During post-operative stay in neurosurgical unit, patient had an episode of GI bleeding on day 9, which was unexplained by subsequent OGD result. Despite adequate antibiotics treatment, follow up MRI brain on day 21 showed new brain abscess formation in left high parietal lobe.

Patient developed right lower limb deep vein thrombosis on day 28, and CT thorax for pulmonary embolism incidentally found a 3cm simple pulmonary arteriovenous malformation in right upper lobe. (Figure 3)

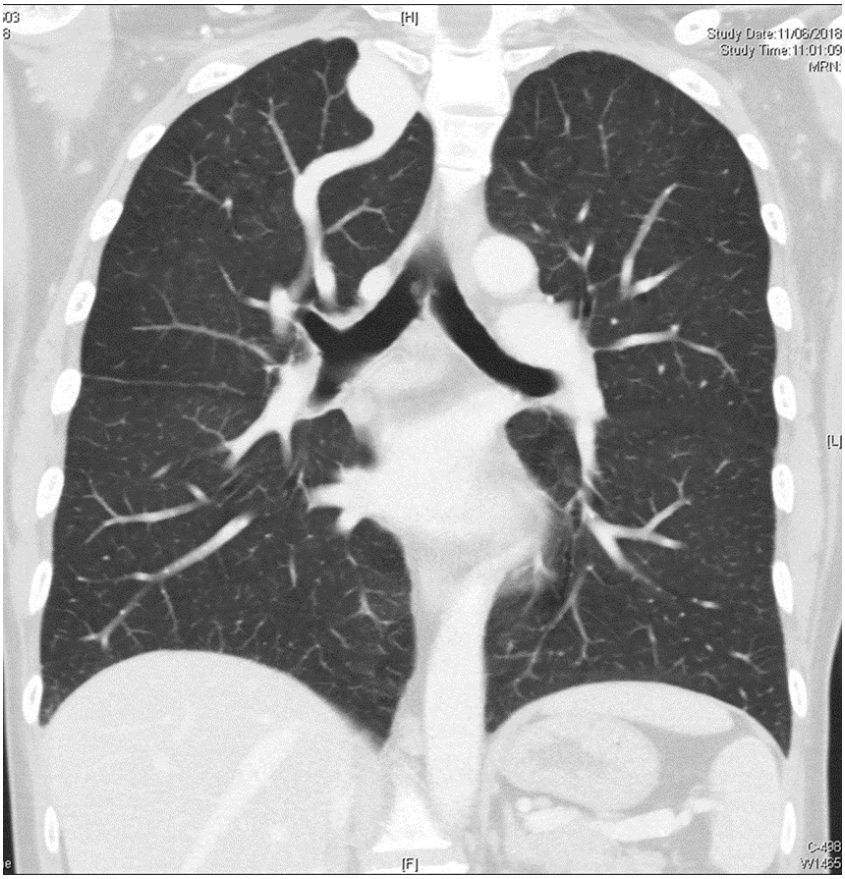

Figure 2: Incidentally found right upper lobe pulmonary AVM during investigation for pulmonary embolism

It was later found that the possible source of infection was from dental caries and dental root extraction was done on day 41 after sepsis was adequately treated. Treatment for brain abscess with combination of ceftriaxone and metronidazole was completed on day 60.

Upon further questioning, patient reviewed that she has history of recurrent epistaxis, which prompted the suspicion of hereditary hemorrhagic telangiectasia. Patient was later found to have telangiectasia over tongue and anterior nasal floor. However, USG liver did not show any co-existing hepatic AVM. Echocardiography was normal.

Patient agreed to proceed radiological embolization of the pulmonary AVM after 6 months of warfarin. Follow up CT thorax showed resolution of pulmonary embolism and showed AVM at left posterior lung base.

During embolization procedure, our radiological colleagues approached the lesion from right femoral vein to cannulate the right femoral artery. Angiogram found a right upper lobe AVM with single feeding vessel that drained to a solitary pulmonary vein via a 2.5cm vascular pouch. Ruby and POD coils were performed to the arterial flow. MVP-5Q vascular plug and Vortx-18 fibred coils were further deployed to

obliterate the residual flow. The procedure was uneventful, and minute residual flow was still detected after procedure. The small AVM in left lower lobe was left untouched.

The patient remained asymptomatic. The small AVM in left lower lobe was left for observation. The diagnosis of hereditary hemorrhagic telangiectasia was yet to be established until the genetic test result would be available.

Discussion

Pulmonary arteriovenous malformations (PAVMs) are vascular structures that provide a direct capillary-free communication between the pulmonary and systemic circulations [1]. In CT scan, it can begin as a ground glass nodule due to the enlargement of the postcapillary venule. As it grows, more small vessels would be noted in the ground glass nodule, and finally matures enough to show classical features of feeding artery connecting an enlarged draining vein via an aneurysmal sac. [2]

Though pulmonary AVM can be idiopathic, 90% of them occur as part of hereditary haemorrhagic telangiectasia (HHT). [3] HHT is an autosomal dominant genetic disease due to defect in ENG encoding endoglin in HHT type 1, ACVRL1 encoding activin receptor like kinase (ALK1) in HHT type 2, and MADH4 cause HHT in association with juvenile polyposis (JPHT). In order to establish the diagnosis, as in Curaçao's diagnostic criteria, the affected individual should have spontaneous recurrent epistaxis, telangiectasia, a visceral manifestation and an affected first degree relative. [4] Classically, telangiectasia would appear on nasal mucosa, hands, face and oral cavity. About half of them would have nasobleed by age of 10, and 80%-90% at age of 21. [5]

HHT was previously believed as a rare disease, however, lately it is estimated that it has prevalence of around 1 in 2600 in population and about 24% HHT patient would have co-existing pulmonary AVM. [6] It may be important for us to alert this diagnosis, since pulmonary AVM usually presents as consequences of right to left shunting, such as cerebral abscess, paradoxical embolism and aneurysmal sac rupture. [7] While about half of them can remain asymptomatic, about 11% of them would present as haemoptysis, 27% as cerebrovascular accident or transient ischemic attack and 12.9% as cerebral abscess. [8]

In our patient with brain abscess, pulmonary AVM was incidentally found during investigation for pulmonary embolism. There is no known association between pulmonary AVM and pulmonary embolism, except for a case report for pulmonary AVM with chronic thromboembolic pulmonary hypertension. [9] Yet there is an unexplained association between venous thrombo-embolism and brain abscess in patient with pulmonary AVM. Therefore, the

British Thoracic Society Clinical Statement on Pulmonary Arteriovenous Malformations 2017 suggest delaying non-urgent dental treatment till sepsis is settled. Our patient hence received dental root extraction on day 41 of admission.

All patients who have radiologically visible pulmonary AVM, if technically feasible, should be treated with embolization [10], as the risk of thromboembolic stroke and brain abscess is independent of feeding artery size. [7] Treatment on pulmonary AVM would tackle the problem of right to left shunting, preventing major complication, such as cerebral abscess, paradoxical embolism and sac rupture and improving oxygenation and physiological parameters. [7] [11] [12]

While traditionally endovascular coils are used for embolization [13], Amplatzer vascular plugs are becoming the preferred agent, since vascular plugs offer better occlusion to the feeding vessel to a pulmonary AVM at the neck of the venous sac, less chance of migration, especially in short and large diameter vessels. [14] [15] [16]

During pre-operative assessment, patient needs to be thoroughly assessed for any contraindications before proceeding to embolization. Co-existing hepatic AVM needs to be identified and treated first, since post capillary pulmonary hypertension (PCPH) can be reversed after hepatic AVM occlusion. [17] True pulmonary arterial hypertension is a relative contraindication to elective embolization. It modifies the complication risks in pulmonary AVM, such that the risk of paradoxical embolic stroke is substantially lower in this group of patients. [18] [19]

We should offer post-procedure clinic to patient, since 5% to 15% of them would have recanalization of previously occluded pulmonary AVM, 2% to 6% with growth of new feeding arteries to the sac, and <1% would have new or worsening pulmonary hypertension. [8] International Guidelines for the Diagnosis and Management of Hereditary Hemorrhagic Telangiectasia 2011 suggests to have CT thorax 6 – 12 months after embolization then every 3 years, and CT every 1-5 years for small untreated AVM. However, British Thoracic Society Clinical Statement on Pulmonary Arteriovenous Malformations 2017 just suggests following CXR only after embolization, and follow up untreated lesion only when symptomatic, since there is no strong evidence to support any particular protocol.

This case demonstrated the diagnostic difficulty of pulmonary AVM as the culprit of its potentially fatal complications.

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