

CASE REPORT

Complicated Ruptured Mycotic Intracerebral Emboli from Infective Endocarditis Presenting Like Meningoencephalitis

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ABSTRAK

Jangkitan endokarditis merupakan sejenis penyakit yang berisiko tinggi dan boleh membawa maut. Namun, untuk mendiagnosa penyakit ini merupakan satu cabaran buat doktor di Jabatan Kecemasan. Ini adalah kerana spektrum dan manifestasi penyakit ini yang begitu luas dan tidak spesifik. Ia boleh berlaku secara akut tetapi tanpa tanda-tanda imunologi salur darah yang klasikal atau secara kronik dengan tanda-tanda yang tidak spesifik dan boleh menyerupai manifestasi penyakit lain. Tanda-tanda penyakit ini juga boleh disebabkan oleh embolisasi sistemik, satu komplikasi yang serius terutama sekali jika terjadi di salur darah otak. Aneurisma mikosis otak merupakan komplikasi yang paling digeruni dan boleh disalah diagnosa sebagai jangkitan otak primer seperti keradangan otak dan meninges. Kami ingin melaporkan satu kes sistemik embolisasi dari jangkitan endokarditis di mana pesakit mengalami tanda-tanda neurologi disebabkan oleh aneurisma mikosis otak. Pesakit ini mengalami demam yang berpanjangan, sakit kepala dan defisit neurologi fokal di mana di peringkat awal telah disalah diagnosa sebagai keradangan otak dan meninges. Pesakit yang mempunyai tanda-tanda sebegini perlu disiasat dengan lebih teliti terutama di peringkat jabatan kecemasan. Ini bagi membolehkan diagnosa yang tepat diperolehi dan rawatan awal dan agresif dapat dimulakan bagi menjamin prognosis yang lebih baik buat pesakit.

Kata kunci: endokarditis, infektif, kronik, meningoensefalitis, sakit kepala

ABSTRACT

Infective endocarditis (IE) is a potentially life-threatening disease which carries high risk of morbidity and mortality. The variability of clinical presentation of

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infective IE remains a diagnostic challenge for the Emergency Physicians. The clinical manifestation may present as an acute, rapidly progressive infection with the absence of classical immunological vascular phenomenon or as subacute or chronic disease with vague constitutional symptoms that may mislead initial assessment and mimic other conditions. Symptoms may also manifest as a result of systemic embolization which can be catastrophic and life-threatening especially if it ends up in the cerebral circulation. IE complicated by cerebral mycotic aneurysm (CMA) is the worst neurological sequelae and can be misdiagnosed as a primary intracranial infection such as meningoencephalitis. Here, we report a case of neurological emergency secondary to systemic embolization of IE with a devastating diagnosis of CMA. History of prolonged fever associated with headache and focal neurological deficit led to the initial diagnosis of meningoencephalitis due to a low clinical suspicion of the disease in the Emergency Department. In emergency setting, a combination of high degree of clinical suspicion together with thorough history, physical examination and diagnostic imaging are crucial in order to guide and establish the diagnosis of this potentially devastating disease. Early recognition and initiation of aggressive treatment is crucial to provide better prognosis and higher survival rate for patients with CMA.

Keywords: chronic, endocarditis, headache, infective, meningoencephalitis

INTRODUCTION

Infective endocarditis (IE) is defined as an infection of a native or prosthetic heart valve, the endocardial surface, or an indwelling cardiac device (Cahill & Prendergast 2016). The incidence is uncommon and has been estimated between 3-10 cases per 100,000 populations per year. However, recent studies reported a rising trend in the incidence (Cahill et al. 2017). Despite advances in diagnostic and treatment modality of IE, the mortality remains high with 20-30% in hospital mortality rate, essentially unchanged over the past two decades (Bin Abdulhak et al. 2014).

Vascular embolic event is one of the minor criteria in Modified

Duke Criteria to diagnose IE. The pathognomonic signs which are Roth's spot, splinter haemorrhages, Janeway lesions, or Osler nodes are uncommon (Murdoch et al. 2009). Cerebral mycotic aneurysm (CMA) is even more uncommon where it occurs in only 3-10% of all cases of native IE (Sonneville et al. 2011; Silverman & Upshaw 2007). Therefore, one tends to forget that the neurological symptoms demonstrated is due to a septic emboli phenomenon. Hence, it can be misdiagnosed as a primary intracranial infection.

The variability of clinical presentation of infective endocarditis and its complications poses a diagnostic challenge for the Emergency Physicians. Identification of patient at

risk and familiarity with various clinical manifestation of IE is crucial to facilitate prompt diagnosis as well as implement effective treatment and management.

CASE REPORT

A 21-year-old lady was brought to the Emergency Department (ED) with history of decrease level of consciousness and lateralizing gaze of both eyes for one day. This was also associated with photophobia, vomiting and diarrhoea. There was history of fever for the past 2 months associated with one month history of non-productive cough and generalised headache for 2 weeks. There was no history of recent trauma.

Upon presentation, her vitals were temperature 39°C, heart rate 120/minutes, regularly regular, blood pressure 80/50 mmHg, respiratory rate 22/minutes and oxygen saturation under room air was 95%. Her Glasgow coma scale (GCS) was 10 with E3, V2 and M5. Her pupils were unequal and neurological examination revealed hyperreflexia of the left upper and lower limb. The Babinski reflex was positive bilaterally. There were no skin rashes or peripheral stigmata of infective endocarditis such as Osler's nodes or Janeway lesion. Cardiovascular and respiratory examinations were normal. Other systemic examinations were unremarkable.

The laboratory tests revealed white cell count $10.2 \times 10^9/L$, haemoglobin 9.2 g/dL, and platelet $203 \times 10^9/L$. Arterial blood gas under high flow mask showed pH: 7.49, pCO_2 : 22.2 mmHg, pO_2 : 174 mmHg and HCO_3 :



Figure 1: Non contrast Computed Tomography shows hypodensity at the right thalamus shown in white arrow

16.9 mmol/L with glucometer of 11.6 mmol/L. Chest X-ray at first presentation showed right perihilar opacities. An initial non contrast CT brain in the ED only showed hypodensity at the right thalamus (Figure 1).

She developed 2 episodes of generalised tonic-clonic seizure which was aborted with intravenous diazepam. In view of her poor GCS recovery and the inability for her to protect her airway, a rapid sequence induction was performed and she was successfully intubated. She was subsequently admitted to the intensive care unit for continuation of care as a case of meningoencephalitis.

An urgent bedside echocardiography was performed once the patient was admitted to the intensive care unit revealing a large vegetation measuring 1.5cm x 0.5cm at the mitral valve with severe mitral regurgitation and minimal pericardial effusion. Cardiac chambers size was normal with no obvious hypokinesia and ejection

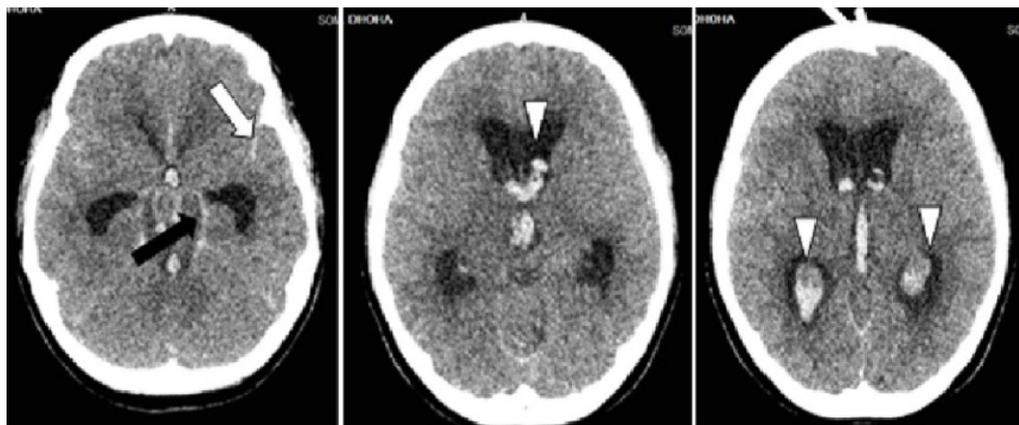


Figure 2: Computed Tomography Angiography of brain presence of mycotic aneurysm (black arrow) with subarachnoid haemorrhage (white arrow). There is also presence of hydrocephalus and intraventricular haemorrhage shown by white arrow head

fraction was 64%. Patient was started on intravenous ampicillin, gentamicin and cloxacillin with inotropic and ventilator support continued. The blood culture and sensitivity result was only available after 2 days and was positive for *Streptococcus viridans*.

A referral was made to a cardiothoracic hospital nearby for possible mitral valve replacement and removal of vegetation. However, patient continued to deteriorate and became more haemodynamically unstable. Hence, an urgent CT angiography of brain was done which showed presence of mycotic aneurysm in the posterior cerebral artery with subarachnoid bleed. There was diffuse cerebral oedema with hydrocephalus and intraventricular bleed (Figure 2). She was then referred to the neurosurgical team who then decided for conservative management in view of presence of brain death. Her condition deteriorated further whereby she developed three episodes of pulseless electrical activity and

subsequently succumbed to death on day 5 of admission.

DISCUSSION

IE can either be acute or subacute where the presentation may differ and the diagnosis criteria using Duke's classification may not be easily applicable. The complexity and variability of clinical manifestation poses a challenge in establishing the diagnosis of IE in the emergency setting (Habib et al. 2015). This potential deadly disease should be suspected in a variety of clinical situations such as pyrexia of unknown origin, unexplained embolic phenomena, or symptoms that raise concern for connective tissue disease or multisystem organ involvement (Schauer et al. 2014).

In the acute course of IE, the classical peripheral stigmata such as Osler's nodes, Janeway lesions, and Roth spots may be few or absent. These immunological vascular phenomena are more characteristic of the later

stages of subacute form of untreated IE and much more common in left sided valvular involvement (Cahill & Prendergast 2016; Bin Abdulhak et al. 2014). Most of the patients with subacute form of IE may manifest with low grade fever and malaise which do not seem to correspond to a serious disease (Pilar 2016). The most common clinical manifestation in IE is fever (90%) and murmur (85%) (Cahill & Prendergast 2016).

IE can also present with complications, particularly heart failure or metastatic infection secondary to systemic embolization. They may manifest with sign and symptoms of heart failure such as shortness of breath or orthopnoea, focal neurological complaint due to a cerebral embolic, back pain associated with vertebral osteomyelitis or cough, and pleuritic chest pain due to pulmonary embolization (Schauer et al. 2014). Neurological complication occurs in 20-40% of patients with IE as a result of embolization from endocardial vegetation and associated with high mortality (Wojda et al. 2016). A study done by Garcia-Cabrera et al. 2013, found that the mortality in IE patient with neurological complication was 45%, compared to 24% in patients who did not experience neurologic sequelae (Garcia-Cabrera et al. 2013).

CMA accounts to 10% of all neurological complications and carries the worst outcome. The mortality is as high as 16-30% in non-ruptured cases and 49-80% in ruptured ones (Sonnevile et al. 2011; Wojda et al. 2016). The clinical presentation of CMA varies from having focal

neurological deficit, headache, altered mental status, seizure or may remain asymptomatic in some patients (Sonnevile et al. 2011). However, when CMA ruptures, the presentation acutely changes into reduced level of consciousness indicating an intracranial haemorrhage.

The management of CMA depends on the presence of intracranial haemorrhage, the anatomic location and the clinical course of the disease. CMA can be managed surgically in addition to standard treatment of IE which is high dose intravenous antibiotics. Endovascular techniques are preferred in patients with high surgical risk, patients with multiple CMA or inaccessible to open surgery. However, a ruptured aneurysm warrants an urgent open brain surgery. Surgical intervention was shown to be safe, effective and increased the survival rate compared to the conservative management alone. Therefore, prompt diagnosis and early aggressive treatment is crucial as these will determine the outcome of the patient (Yuan & Wang 2017).

In the present case, the patient presented with meningoencephalitis like symptoms in combination with evidence of IE. However, CT angiography of the brain was only done when her condition worsened. Studies have shown that 65% of patients with CMA presented initially with IE. Therefore, it is highly recommended that patients who were diagnosed with IE who also had neurological symptoms should undergo neuroimaging studies to rule out CMA (Carmelli et al. 2018). An early diagnosis of intracranial

mycotic aneurysm without bleeding would allow either an endovascular or surgical intervention to be performed where the survival rate was shown to be higher. A progressive symptoms and deterioration of neurological status would indicate a ruptured aneurysm which carries a graver diagnosis and prognosis. At this point of time, the risk would be higher for the patients to undergo any surgical intervention.

CONCLUSION

Diagnosis of infective endocarditis necessitates integration of clinical findings, microbiological analysis, and imaging results. Patients with history of prolonged fever and neurological symptoms with addition of a newly found cardiac murmur should be promptly investigated and treated empirically as a case of IE. They should also undergo neuroimaging study preferably CT angiography of the brain to look for CMA which is the deadliest and most dreaded complication of IE. Early recognition and initiation of aggressive treatment is crucial to provide better prognosis and higher survival rate for patients with CMA.

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