Babinski-Nageotte Syndrome Secondary to Hemorrhagic Medullary Cavernoma in a District Hospital: A Variant of Wallernberg Syndrome

Ganesh Kasinathan*, Rudra Dewi Thavarajan
Department of Internal Medicine, Segamat Hospital, KM 6 Jalan Genuang, Segamat, Johor, Malaysia
*Corresponding author: concorde842000@yahoo.com

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Abstract
Babinski-Nageotte syndrome is a rare clinical entity in a young healthy individual. This syndrome includes a group of neurological symptoms resulting from injury to both medial and lateral medulla of the brain. It is a variant of Wallernberg syndrome. This case report describes a 27 year old healthy Malay gentleman who presented with a three day history of right facial numbness and weakness associated with left sided body hemiparesis and hemiparethesia. He had obvious right hypoglossal nerve palsy. He also complained of vertigo, dysphagia, dysarthria and vomiting. He denied having any hiccups. Physical examination revealed an obvious right Horner’s syndrome with upper motor neuron right facial nerve palsy. There was no facial tenderness. Motor power of the left upper and lower limbs were graded as 2/5. His gag reflex on the right side was absent with abundant oral secretions. Jerky nystagmus of the right eye at all gazes was seen along with diplopia. A plain computed tomography scan of the brain showed a hyperdense lesion suggestive of hemorrhagic medullary cavernoma. He subsequently developed respiratory distress due to pneumonia in which he required mechanical ventilation. However, he succumbed to his illness due to Extended Spectrum Beta Lactamase (ESBL) Klebsiella pneumonia on day 6 of admission. This crossed finding is diagnostic of Babinski-Nageotte syndrome. The unavailability of Magnetic Resonance Imaging/Angiography facility may provide a diagnostic challenge to elucidate the exact etiology for this syndrome in a district hospital.

Keywords: Babinski-Nageotte, hemiparethesia, nystagmus, cavernoma, pneumonia


1. Introduction
Babinski-Nageotte syndrome is also called an alternating brainstem syndrome. It is a variant of Wallernberg syndrome. It was first described in 1902, and later named after the neurologists who initially investigated it, Joseph Babinski and Jean Nageotte. [1] Babinski-Nageotte syndrome is a lateral "Wallenbergian" medullary lesion with the spreading of the lesion to the more basal localized pyramidal tract. [2] This syndrome has mainly been reported to result from atherosclerotic occlusion of the vertebral artery. [3] Typical symptoms consists of prominent dysphagia, hiccups, ataxia, vertigo, dysarthria, ipsilateral Horner’s syndrome and contralateral limb weakness with sensory loss.

2. Case Presentation:
A previously healthy 27 year-old male presented with sudden onset right facial numbness for the past three days. His facial numbness and weakness were associated with left sided body hemiparesis and hemiparethesia. He also complained of nausea, vomiting, dysarthria and dysphagia. His dysphagia was significant as he complained of abundant secretions in the oral cavity. He denied having fever, headache, neck stiffness, hiccups or seizures. He complained of diplopia of the right eye at all gazes. He was a non smoker and a teetotaler. Besides that, he also denied taking any recreational drugs or traditional medications prior to this. He worked as a mechanic at a car workshop. He had no significant family history. Physical examination showed that he was alert with a full Glasgow Coma Scale score. He was seen restless. He had a blood pressure of 147/92 and a regular heart rate of 94 beats per minute. He had a right Bernard-Horner’s syndrome associated with jerky nystagmus and diplopia at all gazes. The motor power of the left upper and lower limbs was graded as 2/5 with normal muscle tone and no clonus. Reflexes were not brisk. Extensor plantar response was equivocal bilaterally. There was obvious loss of pinprick sensation over the left upper and lower limbs. Proprioception and vibration sense were normal bilaterally. Cardiovascular and respiratory examinations were unremarkable. No carotid artery bruit audible. The complete blood count showed a normal hemoglobin of 14g/dL and a platelet count of 220000/uL.
His total white count was 13.7 x 10^9/L. He had normal renal and liver function tests with no derangement in coagulation profile. The fasting blood glucose was 5.5 mmol/L. His fasting lipid parameters were all within normal ranges. Anti-nuclear antibodies and rheumatoid factors were negative. He had normal inflammatory markers. The chest radiograph showed a normal sized heart with no evidence of Pancoast tumor. A 12-lead electrocardiogram showed he was in sinus rhythm. 2D transthoracic echocardiography revealed normal sized heart chambers with good left ventricular function. No evidence of atrial myxoma or intracardiac thrombus seen as a source of embolism. Plain computed tomography scan of the brain (Figure 1) supported the diagnosis of hemorrhagic medullary cavernoma. A subsequent plain computed tomography scan of the brain (Figure 2) repeated 48 hours later did not show any signs of evolution. In a district hospital setting like this, there were no Magnetic Resonance Imaging /Angiography facility or cerebral angiography available for diagnostic imaging. Consultation with a neurosurgeon of a tertiary hospital was immediately arranged. However, a conservative management plan was opted as the family and patient refused consent for any neurosurgical intervention. Subsequently, the patient developed severe respiratory distress whilst in ward which necessitated mechanical ventilation in the intensive care unit. However, on day 6 of admission, he succumbed to his illness due to Extended Spectrum Beta Lactamase (ESBL) Klebsiella pneumonia.

3. Discussion

Babinski-Nageotte syndrome is regarded as a combination of the medial and lateral medullary syndrome. [4] Because the arterial blood supply to the dorsolateral and medial parts of the medulla comes from different blood vessels, this syndrome is characterized by the simultaneous occurrence of lesions of both areas. [4] In this patient, the probable etiology to this syndrome was the hemorrhagic medullary cavernoma. Cavernomas are uncommon congenital vascular anomalies of the brain characterized by sinusoidal vascular spaces adjacent to each other with no intervening brain parenchyma and occur in any part of the central nervous system (CNS).[5] The cause for these cavernomas are not fully understood. Brainstem cavernomas account for 18-35% of CNS cavernomas. [6] The risk of hemorrhage is much higher with brainstem cavernomas compared to cavernomas located at other sites of the neurological system. Many reports have demonstrated a worse prognosis for patients whose cavernomas were subtotally removed than for those whose cavernomas were not surgically treated. [7] Conservative therapy may be one of the treatment options for the symptomatic brainstem cavernoma. [7] Conservative approach may consists of periodic monitoring of Magnetic Resonance Imaging (MRI) to assess the size of the lesion. In this patient, the neurosurgical team decided for a conservative approach as they thought it would be too risky to operate as surgical intervention may result in permanent neurological deficits and the family refused consent for surgery.

Surgical removal (“resection”) may be considered if at least one of the following is true [8]:

- The lesion abuts the surface of the brainstem that abuts the pia mater, the outer covering of the brain. This type of lesion is called “exophytic”.
- Repeated hemorrhages result in progressively worse deficits.
- Acute hemorrhage is external to the “capsule” of the lesion. In other words, blood from a hemorrhage is entering brain tissue surrounding the cavernous angioma.
- The cavernous angioma has grown to a point where it is pressing upon surrounding brain tissue in a way that causes a visible compression of the surrounding tissue.

Expert opinion varies regarding the indication and timing of surgery, but if haemorrhage appears associated with worsening of the neurological deficit surgical evacuation of the lesion and hematoma is recommended [9]. Complete removal of the lesion is essential to prevent re-haemorrhage, which may occur in up to 43% of surgical cases [10].

4. Conclusion

Babinski-Nageotte syndrome is a rare condition which should be considered for a patient who presents with a crossed finding. The unavailability of Magnetic Resonance Imaging/Angiography/Cerebral Angiography facility may provide a diagnostic challenge to elucidate the exact etiology for this syndrome in a district hospital.
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Statement of Competing Interests

The authors declare that there is no conflict of interest regarding the publication of this paper.

List of Abbreviations

None.

Informed Consent

Informed consent was obtained from the patient for the publication of the study.

References


