“Heartbeat in the head”–An atypical presentation of dural arteriovenous malformation

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ABSTRACT

Pulsatile tinnitus is an uncommon presenting symptom and is perceived to be repetitively synchronous with the patient’s heartbeat. Prompt recognition of this rare symptom and subsequent early identification of the underlying etiology is essential for timely management to prevent potentially major complications. Here we highlight a case of a middle-aged female who presented with symptom of “heartbeat in the head” and was later found to have left dural arteriovenous fistula.

Key Words: Pulsatile tinnitus, Dural arteriovenous fistula

1. INTRODUCTION

Pulsatile tinnitus can be best defined as an auditory perception which is repetitively synchronous to the patient’s own heartbeat.[1] There are multiple vascular and non-vascular causes of pulsatile tinnitus. Dural arterial venous fistula (DAVF), arteriovenous malformation and internal carotid stenosis or dissection are important vascular causes while non-vascular causes include glomus tumor, benign intracranial hypertension and anemia.[2, 3] We present a case of DAVF in a middle-aged female who had unique symptom of “heart beat in the head”. She was extensively investigated and diagnosed with DAVF (Borden type I) which was managed conservatively.

2. CASE PRESENTATION

A 54-year-old Chinese female presented with report of “heartbeat in the head” for the last three months. It started around the left temporal region and left ear, which was exacerbated by brisk walking and neck flexion or extension movements. Her symptom was progressive, often severe, and did not respond to simple analgesia. There was no prior history of upper respiratory tract infection, trauma, surgery, anemia or oral contraceptive pill usage. On auscultation, a left sided carotid bruit was heard. Her motor power, cerebellar, sensory examination and deep tendon reflexes were normal. Her otoscopy examination findings were unremarkable.

Carotid ultrasound imaging demonstrated turbulent high flow over the left External Carotid Artery (ECA), with accompanying loud pulsatile bruit over the left upper neck to the mastoid region. Further computed tomography angiography (CTA) of the cerebral vessels (see Figure 1) showed larger left ECA caliber compared to the right. In addition, there was asymmetric contrast opacification of abnormally enlarged left tentorial veins, left transverse sinus, left sigmoid sinus, and left internal jugular vein. These findings collectively suggested an underlying dural arteriovenous fistula (DAVF). Digital subtraction angiography (DSA) later demonstrated...
a left tentorial and left transverse sinus DA VF draining into
the left sigmoid and transverse sinus supplied by multiple
arterial feeders, predominantly from the occipital and middle
meningeal branches of the left ECA (see Figure 2). Other
feeders arising from the right ECA, meningeal branches of
the left posterior cerebral and left vertebral arteries were also
observed. There was retrograde venous flow with no cortical
venous reflux or drainage.

Her imaging findings were consistent with a Borden type I fistula which did not warrant emergent surgical interven-
tion. She was given adequate reassurance and was regularly
followed up in the outpatient clinic. Repeated CTA three
months later showed reduction in the size of her DAVF.

Figure 1. CTA showing larger left ECA caliber (blue arrow)
as compared to the right ECA (white arrow).

3. DISCUSSION

One of the vascular cause for pulsatile tinnitus is DAVF. It is
a vascular anomaly in which there is an abnormal connection
within the dura between meningeal arteries and dural venous
sinuses. It accounts for 10%-15% of all intracranial vascular
(arteriovenous) malformations. There is no clear hypothe-
sis behind the formation of DAVF, but prior trauma, tumors,
dural venous sinus thrombosis or surgery such as previous
craniotomy as well as hormonal alteration during pregnancy
or menopause have been known to result in fistula forma-
tion. Detection rate of DAVF as per a Scottish population
based study was 0.16 per 10,000 people per year. [6, 7]

DAVF, when symptomatic, can present in many ways. Symp-
toms usually depend on the location of fistula, size and pat-
tern of venous drainage. [8–11] Commonly reported symptoms
are pulsatile tinnitus (if the fistula is located just behind the
ear, e.g., sigmoid-transverse sinus fistula); chemosis, blur-
ing of vison or even blindness (if fistula is behind the eyes,
e.g., cavernous sinus fistula); seizures, paralysis or ataxia
(if fistula is large and causing cerebral venous congestion)
and myelopathy or tetraplegia (if DAVF draining into per-
imedullary veins). [12, 13]

DAVF is often an incidental finding during a routine Magn-
etic resonance angiogram. In symptomatic cases, initial
screening can be done by MRI and MR angiography (MRA)
which is recommended with good diagnostic accuracy. [14]
Non-invasive CTA of the head and neck region can be used as
an alternative imaging modality for the evaluation of vascular
pathology. Despite advancement in imaging modalities digi-
tal subtraction angiography (DSA) remains the gold standard
modality in diagnosing DAVF. [15–17] In DSA study, a catheter
is introduced through a groin puncture and is directed under
x-ray guidance into different arteries to reach the cerebral
circulation. DAVF is diagnosed when contrast dye, admin-
istered through the catheter, appears in the affected vein via
the fistula connection.

Borden and Cognard categorized DAVF into various sub-
types on the basis of venous drainage pattern. [19, 18] Cognard’s
classification correlates venous drainage patterns with in-
creasingly aggressive neurological clinical course. It divides
DAVF into 5 subtypes according to the following features:
location of fistula, presence of cortical venous drainage, direction of flow and presence of venous ectasia (see Table 1).

| Table 1. Cognard’s classification of DAVF |
|------------------------------------------|
| Type I | Confined to sinus, antegrade flow, no cortical venous drainage / reflux |
| IIa | Confined to sinus, retrograde flow (reflux) into sinus, no cortical venous drainage / reflux |
| IIb | Drains into sinus with reflux into cortical veins, antegrade flow |
| Ia + b | Drains into sinus with reflux into cortical veins, retrograde flow |
| Type III | Drains directly into cortical veins (not into sinus) drainage (40% haemorrhage) |
| Type IV | Drains directly into cortical veins (not into sinus) drainage with venous ectasia (65% haemorrhage) |
| Type V | Spinal perimedullary venous drainage, associated with progressive myelopathy |

According to the site of location, presence/absence of cortical venous drainage, Borden divided DAVF into three types (see Table 2).

| Table 2. Borden’s classification of DAVF |
|----------------------------------------|
| Type I | Anterograde sinus flow and no cortical venous reflux. |
| Type II | Retrograde sinus flow and cortical venous reflux. |
| Type III | No flow inside the sinus and exclusive cortical venous reflux. |

Cortical venous reflux or drainage pattern is considered to be associated with higher complications such as intracranial haemorrhage. Hence DAVF with no cortical drainage (Borden type I, Cognard types I, IIa) has relatively benign course and risk of haemorrhage is considered low.[19,20]

When considering treatment options, endovascular fistula closure is the mainstay of DAVF treatment. It is offered for high grade fistulas (Borden II, III or Cognard IIb, III, IV, V) or low grade fistula with debilitating symptoms.[20,21] Low grade fistulas (Borden I, Cognard I, IIa) with minimal symptoms are managed conservatively.[25,26] Fistula closure can be achieved by catheter embolization. In this procedure, agents such as the metal coils, glue-like substances, stents or other devices are used to close the fistula.[22] If catheter embolization is contraindicated or not feasible, radiation treatment using gamma knife can be used to close the fistula. However, this is a slow treatment and can take months to years to be effective.[23] Surgery is considered as last option in cases when endovascular approaches have failed or are not feasible. Various surgical options available include direct intraoperative embolization of meningeal vessels, resection of abnormal dura and disconnection of the retrograde leptomeningeal venous drainage.[24] Close outpatient clinic follow up is advised. Appropriate management approach, should be discussed among a multidisciplinary team which includes neurologist, neurosurgeon and intervention radiologist. The risk and benefits of intervention should be weighed against the expected clinical course of the lesion.

This case illustrates DAVF, an important etiology of pulsatile tinnitus and its unique presentation as heartbeat in the head. It emphasizes on the importance of carotid vessel auscultation to exclude bruit which may signify an underlying anatomical pathology of carotid vasculature. DAVF should be considered as possible etiology for pulsatile tinnitus when no other obvious cause was found after extensive investigations. Subsequent management depends on location and presence or absence of cortical venous drainage of the fistula. DAVF with no cortical drainage (Borden type I, Cognard types I, IIa) has relatively benign course and can be managed conservatively with a close follow up to monitor progression of fistula.

**CONFLICTS OF INTEREST DISCLOSURE**
The authors have declared no conflicts of interest.

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