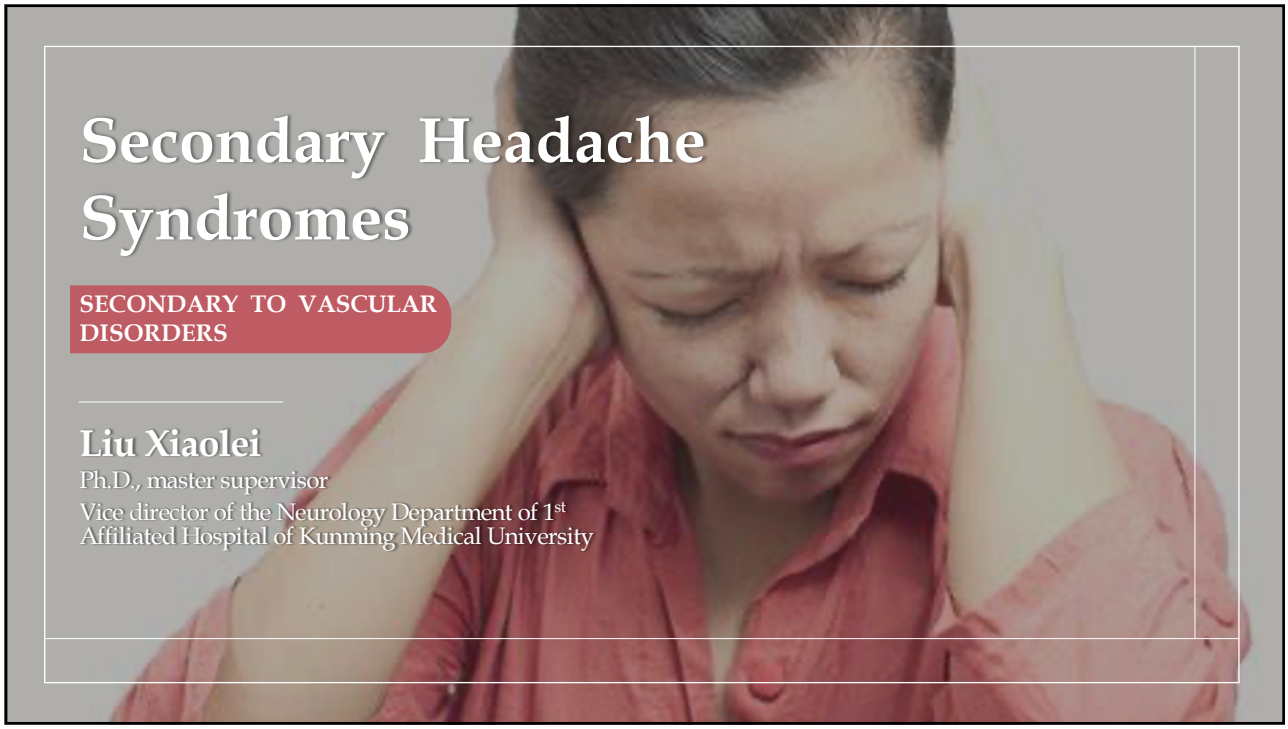

Secondary Headache Syndromes

SECONDARY TO VASCULAR DISORDERS

Liu Xiaolei
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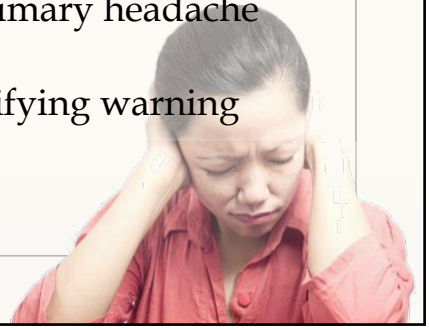
Epidemiology

- Worldwide, almost 3 billion people have a headache disorder
- Of those, approximately 1.89 billion have tension-type headache and 1.04 billion have migraine
- In addition, serious secondary causes of headache invariably present with clinical features that are consistent with or indistinguishable from the most common primary headache disorders
- Therefore, a standardized approach to identifying warning signals in all patients is necessary



Lancet Neurol 2018;17(11):954-976.

2

Red Flags for a Potentially Life-Threatening Headache

SNOOP4

Red Flags	Description/Examples
Systemic symptoms/signs/disease	Fever, chills, rash, myalgia, night sweats, weight loss, comorbid systemic disease (eg, human immunodeficiency virus [HIV], immunocompromised state, malignancy), pregnancy or postpartum
Neurologic symptoms/signs	Change in mental status or level of consciousness, diplopia, abnormal cranial nerve function, pulsatile tinnitus, loss of sensation, weakness, ataxia, history of seizure/collapse/loss of consciousness
Onset sudden	Onset sudden or first ever, severe or "worst" headache of life, thunderclap headache (pain reaches maximal intensity instantly after onset)
Older onset	Onset after 50 years of age
Pattern change	**P1: P**rogressive headache (eg, to daily, continuous pattern), **P2: P**recipitated by Valsalva maneuver **P3: P**ostural aggravation **P4: P**apilledema

Modified with permission from Dodick DW, Semin Neurol.

3

International Classification of Headache Disorders, Third Edition (ICHD-3)

Secondary Headache Disorders:

- Headache attributed to trauma or injury to the head and/or neck
- Headache attributed to cranial and/or cervical vascular disorder
- Headache attributed to nonvascular intracranial disorder
- Headache attributed to a substance or its withdrawal
- Headache attributed to infection
- Headache attributed to disorder of homoeostasis
- Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth, or other facial or cervical structure
- Headache attributed to psychiatric disorder

Cephalalgia 2018;38(1): 1–211.

4

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Cephalalgia 2018;38(1): 1–211.

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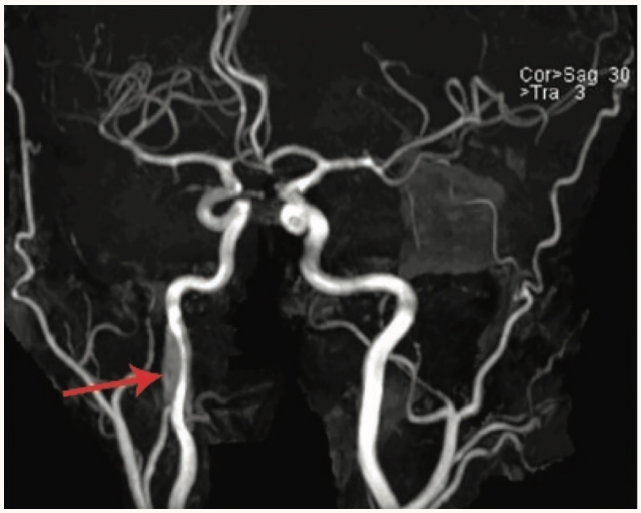
A case

A 41-year-old man presented for evaluation of right-sided neck pain that began 2 weeks prior while weight lifting. He reported a prior history of episodic migraine with visual aura since adolescence that had significantly improved over the last few years. Shortly before the onset of his neck pain, he experienced transient visual scintillations (which he attributed at the time to a visual aura), followed by a right posterior temporal headache. His headache and neck pain persisted, despite treatment with naproxen and sumatriptan, which he had used for his migraines.

On examination, he was noted to have mild right-sided ptosis and miosis. MRI of the brain was normal. Urgent magnetic resonance angiography (MRA) of the head and neck was performed and revealed an acute dissection of the right internal carotid artery with an intramural hematoma. He was started on antiplatelet therapy and fortunately avoided any further neurovascular sequelae.

6

A case



Magnetic resonance angiogram (MRA) of the head

an acute dissection of the right internal carotid artery with an intramural hematoma (arrow)

7

A case

- Patients with a history of a primary headache disorder can also develop secondary headache conditions with symptoms that may overlap with their primary syndrome
- A detailed neurologic examination is essential even in patients with a known primary headache disorder, particularly when a change in headache pattern occurs

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Headache Secondary To Vascular Disorders

1. Arterial dissection
2. Intracranial hemorrhage
3. Acute ischemic stroke
4. Cerebral venous sinus thrombosis
5. Reversible cerebral vasoconstriction syndrome (RCVS)
6. Severe arterial hypertension
7. Cardiac cephalalgia

9

Arterial dissection

- Headache occurs in 60% to 95% of cases of carotid artery dissections, 70% of cases of vertebral artery dissections
- usually unilateral with face/neck pain on the same side
- may be accompanied by ipsilateral Horner syndrome or amaurosis fugax
- Lower cranial neuropathies, cerebellar signs, and visual field defects can also accompany the headache

1. Neurology 1995; 45(8):1517–1522.
2. Cephalalgia 1994;14(1):33–36.

10

Intracranial hemorrhage

Subarachnoid Hemorrhage (SAH)

- The "worst headache of life"
- Accompanied by neck stiffness without fever
- 25% of cases of thunderclap headache are secondary to subarachnoid hemorrhage
- Up to 50% of patients with subarachnoid hemorrhage may present with transient or milder headache (sentinel bleed) and therefore are at risk for delayed diagnosis with subsequent morbidity
- Headache features: occipital location, a "stabbing" quality, a rapid peak of intensity (within 1 second of onset), and associated meningismus
- Focal neurologic signs may be present

1. Cephalalgia. 2002;22(5):354–360.
2. N Engl J Med 2000;342(1):29–36.
3. Headache 2018;58(3):364–370

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Intracranial hemorrhage

Subdural Hematoma

- A more insidious onset of headache
- Similar to those of a brain tumor (as a result of mass effect)
- Mental status changes can also be present
- The elderly are at higher risk for developing subdural hemorrhages (frequently from unwitnessed falls)

1. Neurosurg Rev 2018;41(2):549–556.

12

Acute Ischemic Stroke

- Headache has been reported to occur in 27% of cases of acute stroke
- Factors that have been independently associated with headache at ictus include female sex, younger age, prior history of migraine, and cerebellar as well as right hemispheric location of stroke
- Migraine with aura is also associated with an increased risk of stroke

Stroke 2005;36(2):e1–e3.

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Cerebral Venous Sinus Thrombosis

- Headache is the most common but least specific feature
- Present in approximately 75% to 90% of cases
- Other signs include focal neurologic deficits, altered mental status, seizure, and papilledema
- Risk factors for cerebral venous thrombosis include female sex (4:1 female-to-male ratio), pregnancy or postpartum state, and use of estrogen-containing hormonal contraceptives

N Engl J Med 2005;352(17):1791–1798.

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Reversible Cerebral Vasoconstriction Syndrome

- RCVS, or Call-Fleming syndrome, is characterized by recurrent severe headache attacks in combination with the radiologic finding of diffuse segmental vasoconstriction of intracranial arteries that resolves over a 3-month period
- Reported triggers include exposure to certain substances or medications (such as marijuana, tacrolimus, cyclophosphamide, pseudoephedrine, selective serotonin reuptake inhibitors [SSRIs]), carcinoid tumor, and the puerperium period
- RCVS headaches are often bilateral, brief in duration (1 to 3 hours), recurrent over a span of days to weeks, and are sudden in onset, rapidly reaching a maximal severe intensity (thunderclap)

Lancet Neurol 2012;11(10):906–917.

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Arterial Hypertension

- Headache may arise when systolic blood pressure of 180 mmHg or more and/or diastolic blood pressure of 120 mmHg or more
- May occur with or without symptoms of encephalopathy (eg, lethargy, confusion, visual disturbances, or seizure)
- Typically characterized by bilateral or diffuse, pulsating, and aggravated by physical activity

Arch Neurol 1976;33(4):281–288.

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Cardiac Cephalalgia

- Refers to a headache that occurs in temporal relation to the onset of acute myocardial ischemia
- Headache features: location →bifrontal, bitemporal, or occipital, intensity→ mild to severe, and duration →minutes to hours)
- Nausea may also accompany the headache, which may sometimes resemble migraine
- Cardiovascular risk factors are frequently present
- Ischemic changes can be seen on ECG
- Distinguishing cardiac cephalalgia from migraine is essential to avoid the inappropriate administration of triptan or ergot medications, which are contraindicated in coronary syndromes because of their vasoconstrictive effects

Curr Pain Headache Rep 2015;19(4):14.

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Take Home Message

- Red Flags for a Potentially Life-Threatening Headache **(SNOOP4)**
- Classification of Headache Disorders **(ICHD-3)**
- Headache attributed to vascular disorders
  1. Arterial dissection
  2. Intracranial hemorrhage
  3. Acute ischemic stroke
  4. Cerebral venous sinus thrombosis
  5. Reversible cerebral vasoconstriction syndrome (RCVS)
  6. Severe arterial hypertension
  7. Cardiac cephalalgia

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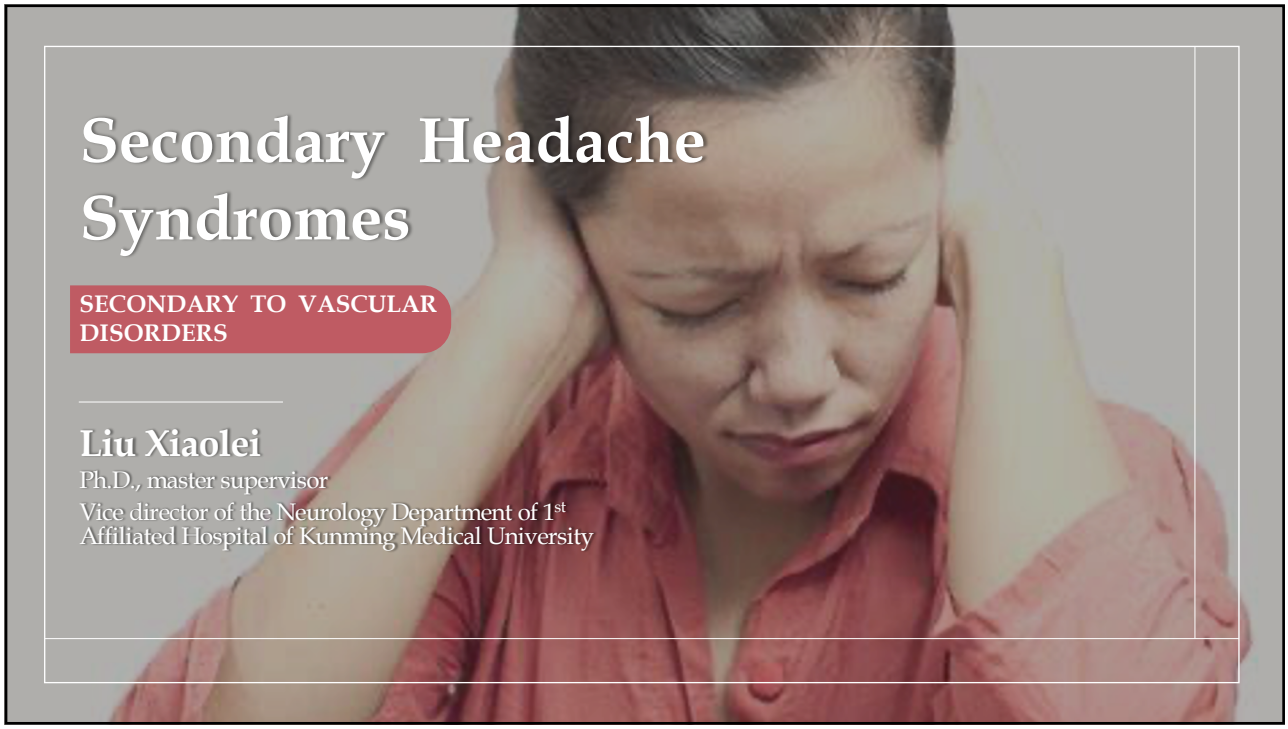

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