Acute Chorea and Hemiballism-Hemichorea

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Greek word *khoreia*: choral dance

“Saint Vitus Dance”

- involuntary
- irregular
- seemingly random
- semi-directed
- “restlessness” to the observer
Greek word *ballismos*: jumping about or dancing

“violent form of chorea”

- sudden
- vigorous
- forceful movements
- usually whole limb
Gradual onset and progression

Most inherited causes of chorea

Rapid onset

Acquired causes: vascular, drug-induced chorea...
Etiologies of acute-onset chorea

Vascular
- Ischemic stroke
- Hemorrhagic stroke
- Cavernous angioma
- Mitochondrial encephalopathy with lactic acidosis and stroke (MELAS)

Metabolic
- Nonketotic hyperglycemia
- Hypoglycemia
- Hyperthyroidism

Pregnancy

Iatrogenic
- Anticonvulsants
- Oral contraceptives

Inflammatory
- Multiple sclerosis
- Sarcoidosis
- Infectious
  - Cryptococcal granuloma
  - Toxoplasmosis
  - Mycoplasma
  - Tuberculoma
  - Human immunodeficiency virus encephalitis

Autoimmune/paraneoplastic
- Systemic lupus erythematosus
- Antiphospholipid antibody syndrome
- Scleroderma
- Behcet disease
- CRMP5 antibodies
- NMDA antibodies

Group A beta-hemolytic streptococcus (Sydenham chorea)
Past medical history

High cardiovascular risk
Rash and arthralgia → systemic lupus erythematosus
Symptoms of thyrotoxicosis
Risk factors for HIV transmission.
Recent streptococcal infection or history of rheumatic fever → Sydenham’s chorea
Drug exposure
Hemodialysis → uremia
Diabetes
Pregnancy test → Chorea gravidarum in first trimester
HIV test → AIDS
Erythrocyte sedimentation rate and antinuclear antibody → Systemic lupus erythematosus
Anti-dsDNA → Relatively specific to systemic lupus erythematosus
Anticardiolipin antibodies and lupus anticoagulant → Antiphospholipid syndrome
Thyroid function tests → Thyrotoxicosis
Anti-streptolysin O titre (recent streptococcal infection) → Sydenham’s chorea/PANDAS
Antibasal ganglia antibodies → Associated with post-infective chorea, chorea gravidarum and oral contraceptive-induced chorea
Full blood count → ↑ Haematocrit in polycythaemia
Cell mass → Sensitive test for polycythaemia rubra vera
Cerebrospinal fluid analysis → inflammatory/neoplastic causes

CT / MRI hemichorea / hemiballismus = structural lesion
Easy and straightforward in ...
... acute onset hemichorea in patients with lacunar stroke

Multiple anatomical locations
Stroke
Paroxysmal Hemiballism/Hemichorea Resulting from Transient Ischemic Attacks

Carmen Gasca-Salas and Anthony E. Lang

*Movement Disorders-Clinical Practice; 3:3, 2016.*
Hemichorea-Hemiballism in a Patient with Temporal-Parietal Lobe Infarction Appearing After Reperfusion with rtPA

Takenobu Murakami, Tomohiro Wada, Itaru Sasaki, Kenji Yoshida, Mari Segawa, Suguru Kadowaki, Akioh Yoshihara, Shunsuke Kobayashi, Akihiko Hoshi, Yoshihiro Sugiura and Yoshikazu Ugawa

Vascular lesions

- < 1% of vascular stroke
- Ischaemic or haemorrhagic lesions of the basal ganglia and adjacent white matter (territory of the middle or posterior cerebral artery)
- Spontaneous remission (surgery)
- Uncommon causes:
  - Moyamoya disease (intracranial vasculopathy that presents with ischaemic lesions or, less commonly, haemorrhagic stroke of the basal ganglia)
  - ‘post-pump chorea’: complication of extracorporeal circulation
Easy and straightforward in ... cases of Sydenham chorea in “endemic” areas

• Onset 8-9; mostly girls
• 4-8 weeks after group A β-hemolytic streptococcal pharyngitis
• Clinical features
  • Chorea (20% hemichorea)
  • Motor impersistence, hypotonia, tics, abnl EOMs
  • Behavioural abnormalities

Video F Cardoso courtesy
Drug-induced

• Oestrogen-containing oral contraceptives
• Anticonvulsants: phenytoin, carbamazepine, valproate, gabapentin
• Antiparkinsonian drugs: LD, DA
• Thyroid replacement
• CNS stimulants: cocaine, amphetamines, methylphenidate
• Lithium
• Tricyclic antidepressants
• Neuroleptics
  • Withdrawal-emergent
  • Tardive
Drug-induced
Chorea caused by lithium intoxication: A case report and literature review

Gerald D. Podskalny, Stewart A. Factor

Movement Disorders; 11:6, 1996. 733-737
Immune-mediated

• Sydenham’s chorea
• Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections (PANDAS)
• Post-infectious chorea
• Systemic lupus erythematous
• Anti-phospholipid antibody syndrome
• Henoch-Schonlein purpura
• Behcet’s disease
• Polyarteritis nodosa
• Paraneoplastic chorea
• Multiple sclerosis
• Autoimmune encephalitis
Paraneoplastic chorea associated with breast cancer

Jana Martinková, Peter Valković, Ján Benetin

*Movement Disorders; 24:15, 2009. 2296-2297*
Clinical case

45 yrs onset of abnormal movements

46 yrs first falls

Uncle with a diagnosis of dementia

No previous psychiatric disease

No previous intake of neuroleptics

First seen in a urgent care department due to a fall
Clinical case

- Brain and spinal lesions
- Abnormal visual evoked potentials
- Positive IgG oligoclonal bands
Metabolic
- Pregnancy (Chorea gravidarum)
- Hyperthyroidism
- Hyperglycemia
- Hyponatremia

Infectious
- HIV
  - primary infection
  - opportunistic infection
- Borreliosis
- Polycitaemia rubra vera
Identification: 68 year-old woman

Present history: acute left arm and leg involuntary movements of 15 days` duration

Past history: 15 year-long, uncontrolled Diabetes mellitus type 2; arterial hypertension

Observation: generalized chorea predominant on left hemibody, left hemifacial grimacing

Laboratorial analysis: hyperglycemia (serum glucose 450 mg/dL), HbA1c 15.8%
Clinical case
Putaminal petechial haemorrhage as the cause of non-ketotic hyperglycaemic chorea: a neuropathological case correlated with MRI findings

Tiago A Mestre, Joaquim J Ferreira and José Pimentel

Clinical case

56 yrs

Acute-onset involuntary limb and facial movements occurring in the setting of uremia

Previous medical history: diabetes mellitus, end-stage renal disease secondary to diabetic nephropathy (undergoing hemodialysis for 15 years)

Observation: limb chorea and dystonia (facial grimacing)

Laboratorial analysis: Creatinine 5.9 mg/dL, Urea 88 mg/dL; glucose 108 mg/dL; Na+ 137 mEq/L; K+ 4.2 mEq/L
Clinical case
Management:
- Adjustment of dialysis program
- Treatment with sulpiride (50 mg/d)

Evolution:
- Resolution of involuntary movements after 10 days
- Resolution of Cranial MRI basal ganglia lesions at 3 months of follow-up
Chorea gravidarum

Associated

rheumatic fever antiphospholipid antibody syndrome
systemic lupus erythematosus
encephalitis
syphilis
Remission of cause

Autoimmune disorders - immunosuppression

Neuroleptic
- Haloperidol
- Risperidone

Dopamine-depleting agent
- Tetrabenazine

Benzodiazepine
- Clonazepam

Anticonvulsant
- Valproic acid
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