Neuropsychiatric Masquerades: Is it a Horse or a Zebra

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Manish A. Fozdar, M.D.

- Triangle Forensic Neuropsychiatry, PLLC, Raleigh, NC
  - www.BrainInjuryExpert.com
- Consulting Assistant Professor of Psychiatry, Duke University Medical Center, Durham, NC
- Adjunct Associate Professor of Psychiatry, Campbell University School of Osteopathic Medicine

Disclosures

- Neither I nor any member of my immediate family has a financial relationship or interest with any proprietary entity producing health care goods or services related to the content of this CME activity.
- I am a non-conformist and a cynic of current medical establishment.
- I am a polar opposite of being PC. No offense intended if one taken by you.

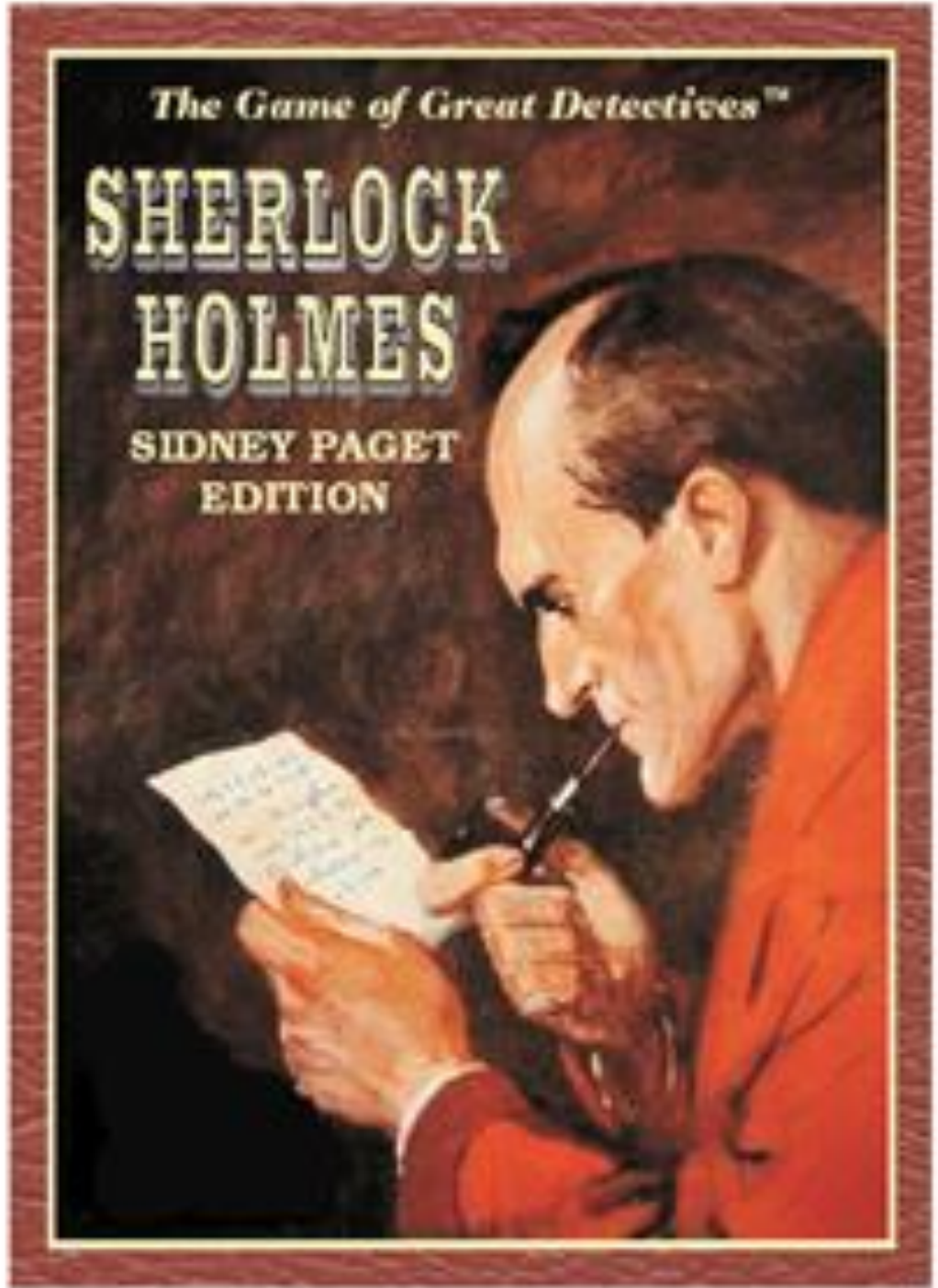
Anatomy of the talk

- Common types of diagnostic errors
- Few case examples
- Discussion of selected neuropsychiatric masquerades

When you hear the hoof beats, think horses, not zebras

- Most mental symptoms are caused by traditional psychiatric syndromes.

- Majority of patients with medical and neurological problems will not develop psychiatric symptoms.


The Game of Great Detectives™
SHERLOCK HOLMES
SIDNEY PAGET EDITION

Case

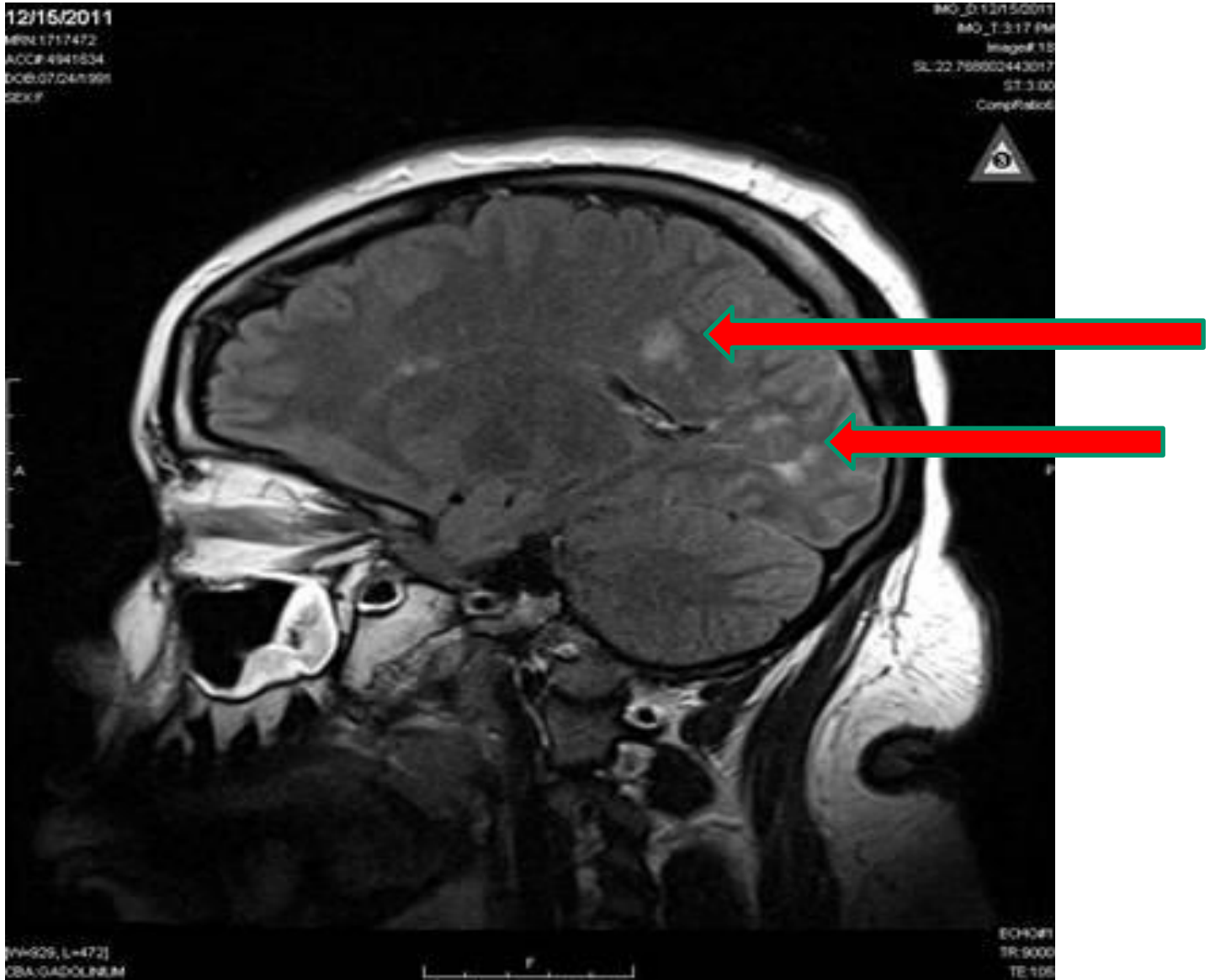
- 20 y/o AA female with h/o Bipolar disorder and several psych hospitalizations.
- Admitted a local psych hospital due to decompensation..
- While at psych hospital, she develops increasing confusion and ataxia.
- Transferred to general med-surg hospital.

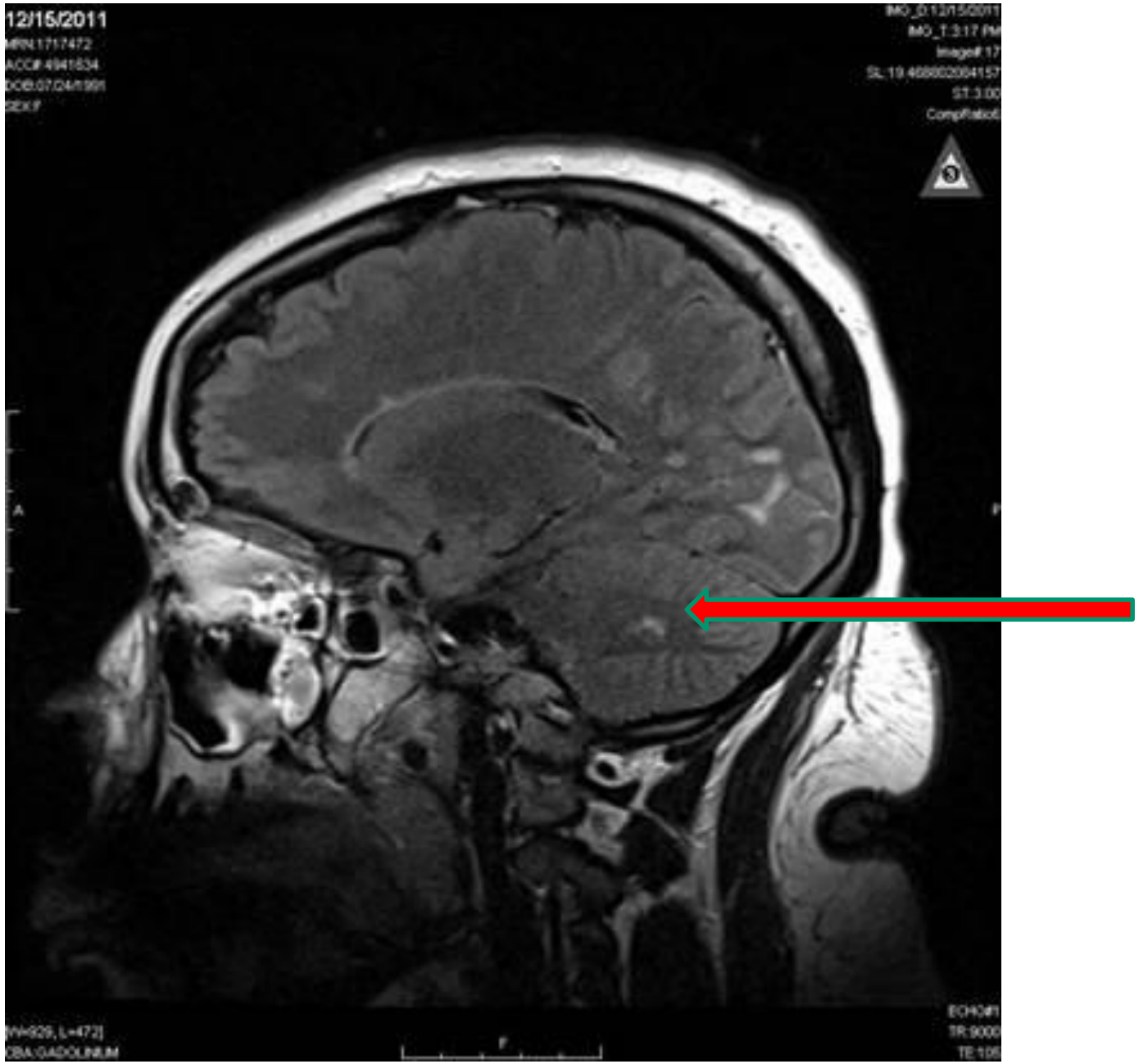
- Stayed for 2 weeks.
- Here is what happened….
- Psych C-L service consulted. We did the consult and followed her throughout the hospital stay.
- Initial work up showed Normal MRI, but was of poor quality. EEG was normal.

- She remained on the hospitalist service. 8 different hospitalists took care of her during her stay here.
- Her presentation was chalked off to “her psych disorder”, “Neuroleptic Malignant syndrome” etc.
- After some relentless persuasion, I was able to convince the (ever changing)attendings to start aggressive work up.

This is how it ended…..

Repeat MRI

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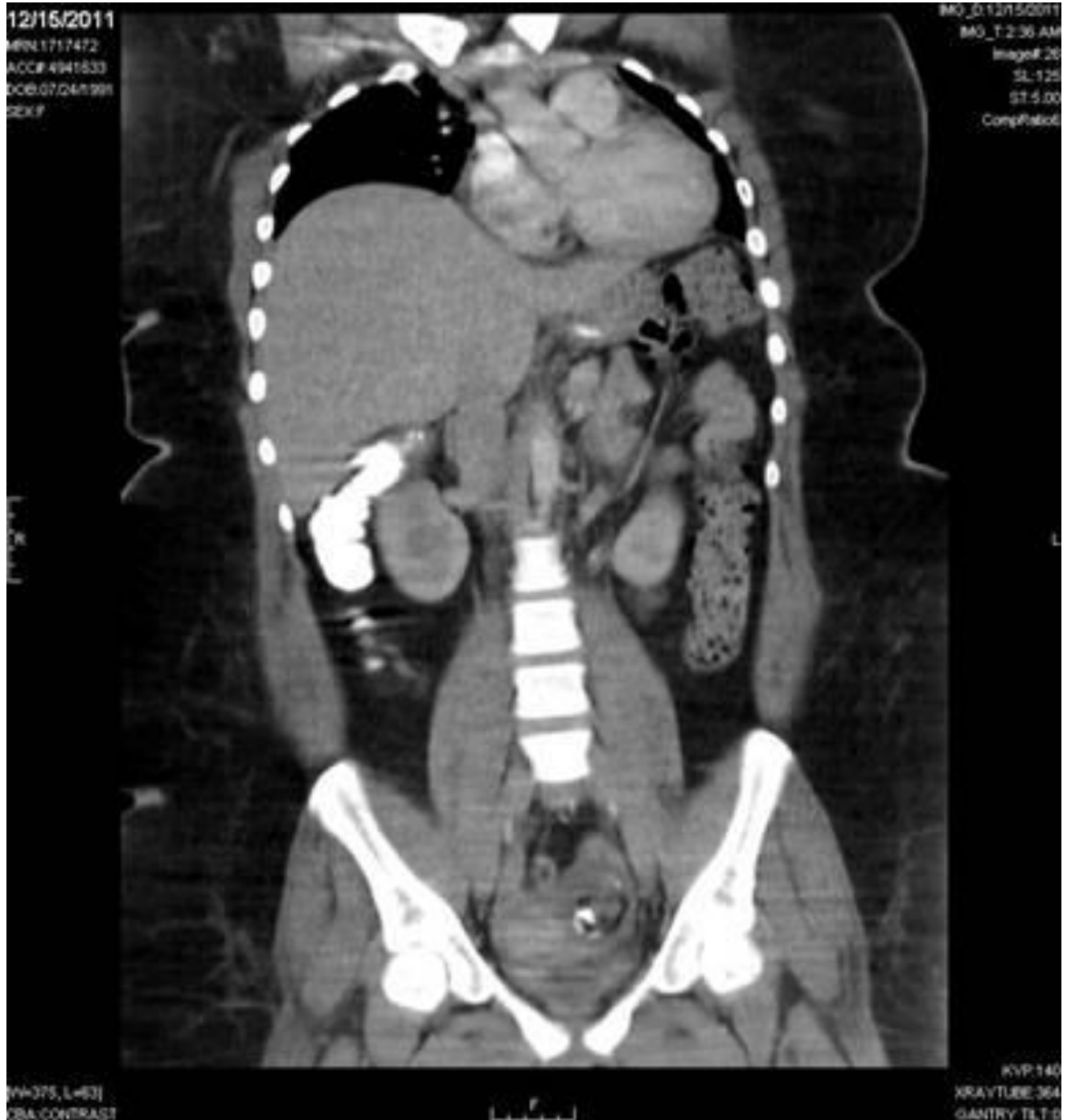
- She ended up in MICU critical care due to respiratory failure and had a seizure.
- I suspected encephalitis. Suggested whole body CT.
- CT of the abdomen revealed presence of Ovarian Teratoma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- Ovarian Teratoma was removed, received plasmapheresis and IVIG treatment.
- She got better with resolution of brain lesions after few weeks.
- She was discharged to home!
- Paraneoplastic Encephalitis was the final diagnosis.

Cognitive Errors in Diagnosis

- Cognitive biases play detrimental role when you clung to robotic models of decision making.
- *Anchoring bias*- Lock onto initial salient features of presentation and then failing to adjust your diagnosis later on when more information is available.
- *Ascertainment bias*- Stereotyping and gender bias
- *Diagnosis momentum*- Carried a diagnosis of bipolar disorder

- *Hindsight bias*- admitted to psych hospital, hence this is psychiatric.

- *Premature closure*- Closing too soon on a diagnosis. “When the diagnosis is made, the thinking stops”.

- *Psych-Out errors*- Everything and anything attributed to prior diagnosis of psych illness.

- *Yin-Yang out*- Patient has been worked up the YinYang. Nothing more to do.

- *Unpacking errors*- Failure to elicit all information.

DSM and its pitfalls

- Symptom clusters assembled under different diagnostic categories.
- Not a medical model.
- Psychiatric symptoms can be a manifestation of variety of medical/neurological/psychiatric conditions.
- Axial diagnoses are not mutually exclusive. For example, Schizophrenia on Axis I and Temporal lobe epilepsy on Axis III.

System Pitfalls

- Not enough time for evaluation- history, history, history/ location, location, location.
- Symptomatic treatment approach.
- Fragmented care, lack of communication.
- Lack of adequate training in psychiatry residency programs in behavioral neurology.

- Economic pressures of medical practice.
- 10-15 minute med checks.

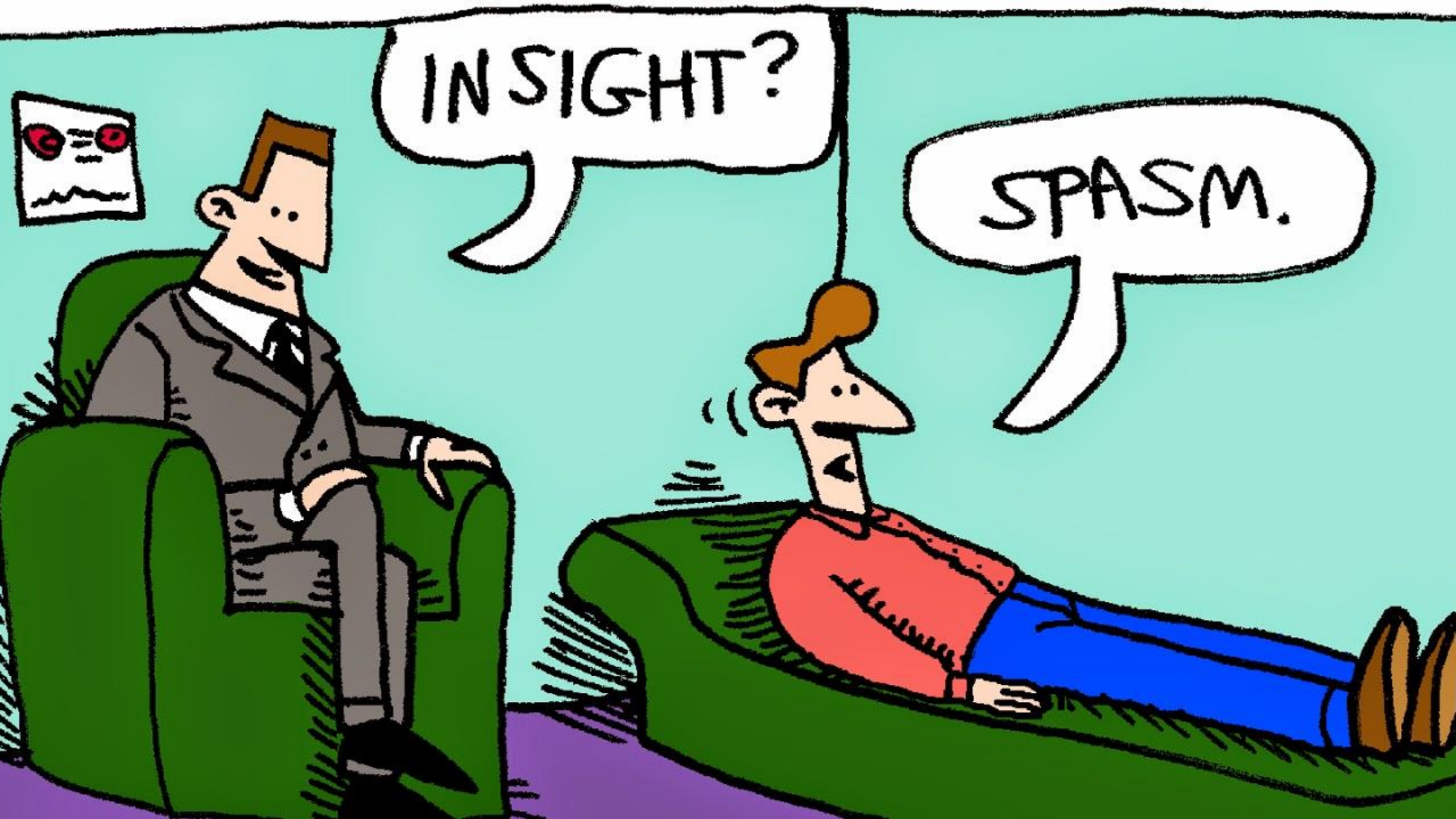
- Robert Spitzer- 2 types of errors leading to misdiagnoses as “functional” psychiatric disorders.

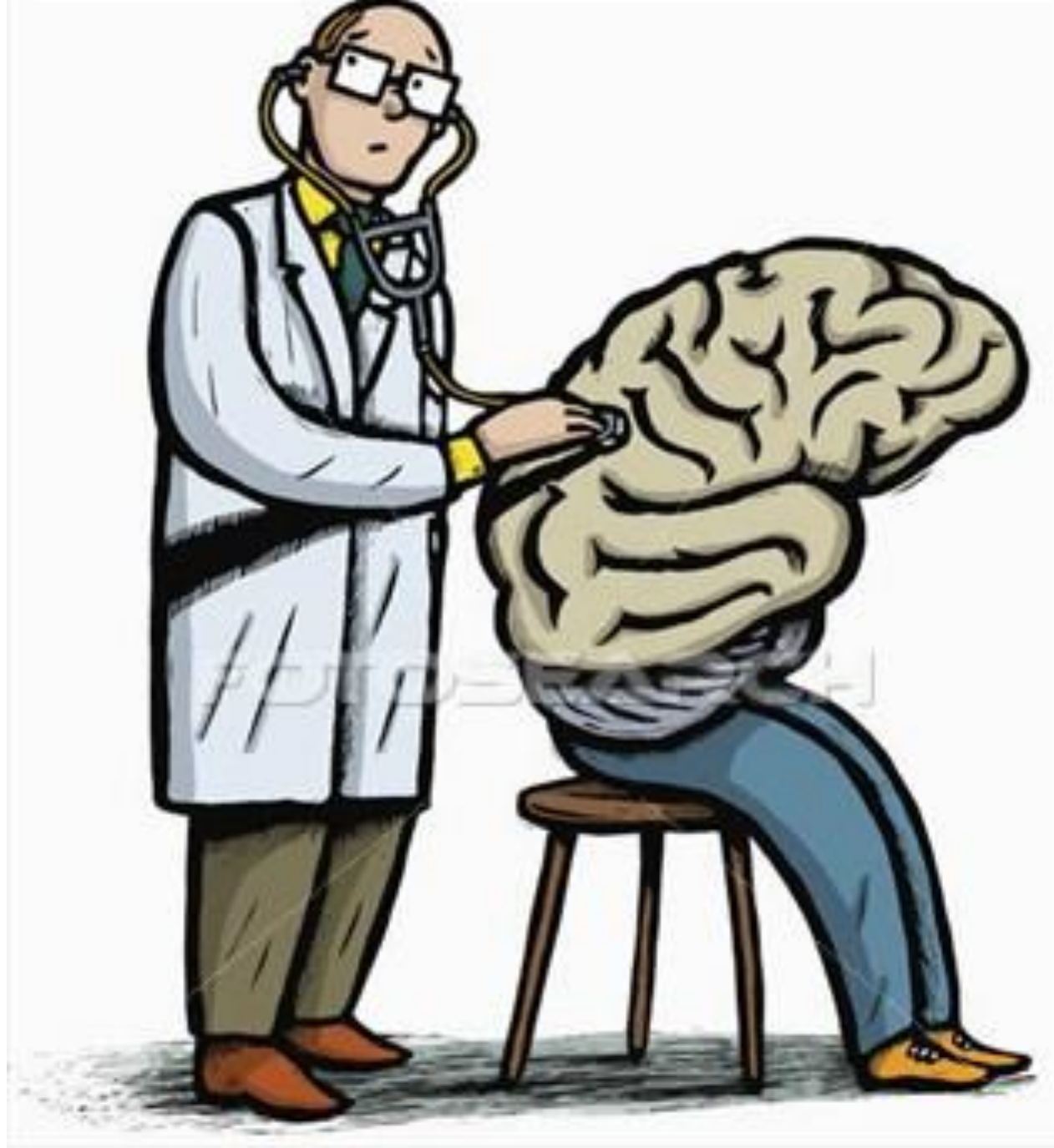
- ***Informational errors*****-** Failure to gather clinical and historical data vital to make correct diagnosis.

- ***Criterion errors*****-** Data is collected but clinician fails to recognize the illness.

- **Norman Geschwind**
- Emphasized incomplete history taking and failure to recognize regional brain syndromes leading to incorrect diagnoses.

- **Gary Tucker**
- Emphasized paying attention to clinical course of the disease.


INSIGHT?
SPASM.




Blue's Clues
Blue's Clues

Characteristics of Organicity

- Late onset presentation
- Acute onset
- Unusual presentation of known psychiatric disorders
- Treatment refractoriness
- Family history of certain medical/neurological disorders

- Absence of previous psychiatric history
- Fluctuating mental status
- Abnormal vital signs
- Neurological soft/hard signs

- 66 y/o female, healthy, with no past psych history.
- Presents with amotivation, decreased socialization, poor appetite, headaches, memory complaints, occasionally feeling confused.
- Labs normal, neuro exam normal
- Gets Dx of depression. Given few AD trials.
- No response. Worse on some meds.
- Detailed history reveals no symptoms of sadness, anhedonia, hopelessness etc.
- (++) apathy/amotivation.
- S/S fluctuated.
- Neuro exam shows frontal release signs (grasp and glabellar tap)
- MRI brain left frontal meningioma with pressure effect
- Surgery→Relief of symptoms

Acute Organic Conditions

- Fairly acute onset
- Impaired consciousness, fluctuating mental status
- Inattention, distractibility
- Little purposeful activity, apathetic
- Occasionally hyperactivity, e.g. certain deliriums
- Slow and sparse speech
- Incoherent thought process

- Ideas of reference and delusions, especially persecutory delusions.
- Lack of insight
- Memory difficulties (reduplicative paramnesia)
- Perceptual disturbances
- Hallucinations-visual more common

Chronic Organic Conditions

- Usually insidious onset
- Family, coworkers etc may notice memory impairment, general intellectual decline
- Personality problems less common at first
- Egocentricity, lack of concern for others
- Deterioration of personal hygiene
- Hoarding behavior

- Slowed thinking
- Reduplicative paramnesias- Capgras syndrome, Fregoli syndrome
- Poverty of speech
- Extreme emotional reactions- pathological laughing and crying

Frontal Lobe

- Personality changes most characteristic- disinhibition most common, apathy (pseudomania and pseudodepression)
- Lack of concern for others
- Lack of insight and judgment
- Cognitive deficits- attention, lack of planning, difficulty with multitasking
- “Silent nature” of frontal lesions- can grow large before declaring themselves

Parietal lobe

- Complex cognitive deficits, difficult to pick up on routine mental status exam or MMSE.
- Visuospatial difficulties, constructional dyspraxias
- Topographical disorientation- pt. loses himself in familiar surroundings
- Dominant parietal lobe lesions- Dysphasias, Motor apraxia, Gerstmann’s syndrome

- Non-dominant parietal lesions- left sided neglect, anosognosia, dressing dyspraxia, prosopagnosia
- Reduplicative paramnesia- Capgras, Fregoli
- Neurological exam shows cortical sensory loss, sensory extinction

Temporal lobe

- Dominant temporal lobe lesions- language difficulties
- Bilat. Medial temporal lesions- amnestic syndromes
- Personality changes along with intellectual and neurological deficits (v. Frontal lobes)
- Temporal lobe epilepsy associated symptoms

Occipital lobe

- Visual problems

Neurological Disorders

Epilepsy

- 1% of population
- Psychiatric symptoms- preictal, ictal, postictal and interictal.
- Generalized v. partial (simple or complex)
- Psychiatric symptoms common in CPS

Frontal Seizures

- Variety of clinical pictures that are often bizarre
- Automatisms- anterior and medial frontal lobe, Arm flailing, rubbing, kicking etc.
- Sexual automatisms- pelvic thrusting, genital manipulation
- Vocalizations, shouting, screaming etc.
- Disinhibition- misdiagnosis of mania, psychosis, pseudoseizures
- Difficult to detect on scalp EEG

Parietal seizures

- Sensory symptoms
- Complex hallucinations or disturbance of shape and size

Temporal Lobe Seizures

- Of most interest to psychiatrist
- Auras- often present without ensuing motor convulsion
- Epigastric sensations, depersonalization, derealization, déjà vu, jamais vu, hallucinations, intense fear and anxiety
- Confusion, disordered thinking including thought blocking

- Epileptic fugue states- less common. Patient may wander away from home. Epileptic v. psychogenic ??

- Lesser degree of dissociative episodes.

- Psychoses of epilepsy- Michael Trimble’s work.

TLE Personality
(Gastaut-Geschwind)

- Aggression
- Hypergraphia
- Hyper- religiosity
- Ethical/Moral Concerns
- Hypo/Hyper Sexuality
- Elation
- Eccentricity
- Viscosity

Humorlessness

Circumstantiality

Nonconvulsive Status Epilepticus (NCSE)

- Neuropsychiatric symptoms common and often the presenting symptoms.

- Cognitive changes, bizarre behavior, psychosis, affective disturbances, impulse control problems, speech problems, catatonic signs, autonomic disturbances.

- EEG diagnostic.

Brain Tumors

- Usually present with clear neurological s/s.
- Occasionally present with psychiatric s/s at first without overt neurological s/s. Although this is rare.
- Neuroimaging is usually diagnostic.
- Frontal lobe tumors often disguised in their presentation. Often few or no neurological s/s at first.
- May often present as dementing illness. Apathy, somnolence, akinetic mutism, irritability etc.

- Disinhibition, sexual behaviors, childishness.
- Lack of insight is striking.
- Corpus Callosum Tumors- often present with psych s/s.- especially anterior and posterior parts of CC
- Rapidly progressive cognitive decline, thought blocking, somnolence etc.

- Temporal Lobe Tumors- Highest frequency of mental s/s.
- Memory problems, speech impairment, affective disturbances.
- Occasionally psychotic illness resembling schizophrenia.

- Diencephalic Tumors- (thalamus, hypothalamus and 3rd ventricle)
- Marked amnesia, especially 3rd ventricle tumors, Confabulation resembling Korsakoff syndrome
- Somnolence and hypersomnia common.
- Psych s/s due to endocrine dysfunction in pituitary tumors

Multiple Sclerosis

- Multifaceted disease with varied manifestations
- Pts. Often diagnosed with conversion d/o, hysteria, fibromyalgia etc. until neurological deficits emerge
- Cognitive deficits present in up to 2/3 of cases. Often revealed by detailed neuropsych testing.
- Memory problems, attention deficits, problem solving

- Depression common. Reactive at times, but endogenous form may be present for some time before the disease declares itself.
- Euphoria – out of context with pt’s disability. Indicative of neurological damage.
- Bipolar disorder not as common as once thought.
- Presentation with psychosis is rare.

Parkinson’s Disease

- Classic triad- akinesia, rigidity, tremors
- Motor signs may not be evident initially and pt. may present with depression.
- Common features between depression and PD- psychomotor slowing, attention/ conc. Deficits, memory problems, flat affect etc.
- Anxiety disorder, social phobia and panic disorder in up to 25 % pts with PD.

- Hallucinations and delusions develop late in the disease. Often s/e of Antiparkinsonian meds.
- Consider PD in elderly pts presenting with depression/anxiety for the first time.

Neurosyphilis

- The Great Imitator
- Made a comeback due to HIV/AIDS.
- Neurosyphilis is a primary form of tertiary syphilis.
- Frontal lobes affected often- personality changes, irritability, decline in personal hygiene, disinhibition/mania etc.
- Dementia and depression now more common.

Herpes Simplex Encephalitis

- Prodrome of 1-7 days of URI followed by headache, fever, bizarre behavior, psychosis and fluctuating mental status.

HIV

- Think of it in a pt presenting with psych s/s without prior history, especially high risk group.
- HIV encephalopathy, AIDS-Dementia complex, opportunistic infections
- Psychiatric s/e of HIV meds.

Frontal Lobe Dementia Complex

- Not so uncommon as once thought.
- Onset usually between 45-70 yrs.
- Deterioration of personality and behavior.
- Disinhibited behavior, apathy, poor insight and judgment, neglect of personal hygiene
- Utilization behavior- tendency to touch things, use objects in sight purposelessly etc.

Pick’s Disease

- Peak onset between 50-60 yrs.
- Disinhibition, egocentricity, indulgence in foolish jokes and pranks, euphoria
- Features of Kluver Bucy syndrome- oral tendencies, overeating, touching objects in sight etc.

Huntington’s Disease

- Onset usually in 40’s & 50’s
- Psychiatric s/s could be present for yrs before neurological s/s become apparent
- Change in personality, paranoia, ideas of reference, depression, anxiety
- Family history.

Medical Disorders

Endocrine Disorders

- Thyroid disorders

➢ Hyperthyroidism- anxiety, confusion, agitation, hyperactivity, depression, hypomania, psychosis

➢ Hypothyroidism- Depression, anxiety, apathy, psychomotor slowing, memory problems

➢ Subclinical hypothyroidism

- Parathyroid disorders

➢ Hyperparathyroidism- delirium, depression, anxiety, fatigue. Associated hypomagnesemia can cause psychosis.

➢ Hypoparathyroidism- Delirium, depression, anxiety, psychosis

- Adrenal disorders
  - Addison's disease- fatigue, depression, irritability. Psychosis and confusion can occur.
  - Cushing's Syndrome- depression, memory problems, psychosis.
- Pancreatic Tumors- depression can be the sole manifestation initially. Consider it in elderly pt with back pain and new onset depression.

Systemic Lupus Erythematosus

- CNS involvement very common.
- Usually appear late in the disease, but can be present before systemic manifestations.
- Can coincide with relapses and remission of the disease itself.

Acute Intermittent Porphyria

- Autosomal dominant transmission, chromosome 11.
- Onset- anytime puberty onwards, 3rd decade most common.
- Physical s/s- acute abdominal pain, N/V, constipation, headache, seizures (20%), peripheral neuropathy
- Psych s/s- 25-75% cases. Wide range of s/s. Acute anxiety, depression, emotional lability, psychosis.
- Precipitants- inadequate nutrition, endocrine factors, ETOH, infections, medications.

Vitamin deficiencies

- B 1- Wernicke’s encephalopathy, chronic alcoholism. Triad of nystagmus, ataxia and ophthalmoplegia. May persist as Korsakoff’s syndrome with amnesia.
- B 12- Psych s/s often antedate anemia, neurological s/s. Depression, psychosis, dementia, delirium.
- Folate

Medications

- Always suspect role of meds in mental status changes.
- Requires careful history taking, collateral info etc.
- Benzodiazepines, opioids, AED, Anti HTN, Anticholinergic, Anti-Parkinsonian, HIV, steroids, chemotherapy agents etc. etc…..
- OTC and Herbal meds.(Ephedra, Triple C)

Substance Abuse

- Most common psychiatric disorder, underdiagnosed, undertreated.
- Alcohol, cocaine, marijuana, opioids, Meth, benzo., inhalants, ecstasy, hallucinogens.

Heavy Metals

- Consider it in appropriate setting.
- Occupational history, environmental exposure.
- 24 hr. urine screen.

Summary

- Think outside the box when dealing with perplexing cases.
- A & B can coexist, A can cause B, B could be due to yet unidentified C.
- History, History, History.
- Clinical course of the disease.
- Consult other physicians.

References

(1) Lishman’s Organic Psychiatry: A Textbook of Neuropsychiatry, 4th Edition. Wiley-Blackwell.

(2) The importance of cognitive errors in diagnosis and strategies to minimize them. Pat Croskerry, MD, PHD, Academic Medicine 2003, 78:775-780.