33 Question Exam

Nutritional Diseases

In his words: Important to know about mineral excesses - wilson's disease, hemochromatosis, and does disease resolve from a micronutrient excess or deficiency?

- Wilson's Disease: results from inherited genetic mutation of gene related to copper storage → excess copper is toxic to brain and liver
    - Unique sign of Wilson's disease is brown ring around iris
- Hemochromatosis: excess iron (iron overload) → it is NOT usually caused by excess DIETARY iron; instead it is caused by inherited genetic mutations of genes related to iron absorption and storage OR too much iron from frequent blood transfusions
- Do these diseases resolve from a micronutrient excess or deficiency?: micronutrient deficiency

In his words: Question on scurvy definitely a question → his interesting case → oral manifestations

- Cause of scurvy: vitamin C deficiency → leads to impaired collagen formation → leads to bleeding tendency due to poor vessel support (gums, skin, periosteum and joints)
    - Bleeding in joints results in joint pain and swelling
    - Other effects are inadequate synthesis of osteoid and impaired wound healing
- Symptoms of scurvy:
    - Gingival
        - Swelling, ecchymoses, bleeding, loosening of teeth → scorbutic hyperplastic gingivitis
    - Bone
        - Joint swellings, and arthralgia
    - Advanced
        - Poor wound healing
- Why do a biopsy for scurvy? → to rule out very serious diseases like leukemia
- What did the biopsy show for scurvy? → hemorrhage and lack of cohesive CT (suggestive of scurvy) AND lack of neoplasia (no other leukemia or other cancerous conditions)
- Treatment protocol:
    - Vitamin C supplementation for one month or until symptoms resolve → children: 100-300 mg/day; adults: 500-1000 mg/day + additional iron and vitamin supplements

In his words: Slide that has table of vitamins (nicknames and deficiency causes) → 2 questions

Nutrient	also known as	Deficiency causes
Vitamin A	Retinol	Varying levels of vision impairment
Vitamin B1	Thiamin	Beriberi: cardiovascular and neuronal impairment
Vitamin B2	Riboflavin	
Vitamin B3	Niacin	Pellagra: dermatitis (roughened skin), dementia, diarrhea
Vitamin B6	Pyridoxine	
Vitamin B12	Cobalamin	Pernicious anemia
Vitamin C	Ascorbic Acid	Scurvy (see subsequent slides)
Vitamin D		Rickets (children) Osteomalacia (adults) Poor mineralization of skeleton - bone fragility and weakness
Vitamin E	α-tocopherol	
Vitamin K		Inadequate blood clotting
Iron		Iron deficiency anemia

Bone Pathology

In his words: Question about odontogenic cysts/tumors. What's the bony lesion that surrounds an impacted tooth?

- Dentigerous cyst: common cyst always associated with an impacted tooth

In his words: Question about gout

- Gout: a particular type or arthritis in which uric acid crystals deposit in joints
    - Produce episodic, severe pain
        - Redness, swelling and pain most often on big toe
        - Joint discomfort may last a few days to a few weeks
    - Dietary factors and systemic diseases contribute to propensity of gout

In his words: Know slide about arthritis - joint pain (heavy slide)

- Arthritis definition: inflammation of the joints and subsequent destruction of cartilage
- Symptoms: pain, reduced range of motion, and stiffness
- Types of arthritis:
    - Osteoarthritis - overwhelmingly the most common
        - Aging, abnormal loading of joints, crystal deposition, constant wear & tear
        - pain/dysfunction usually constant
    - Rheumatoid arthritis
        - Cause by autoantibodies against antigens within the joint spaces
        - Serological testing needed for making diagnosis
        - May be seen concurrently with lupus, scleroderma, rheumatic fever
    - Psoriatic arthritis
        - An arthritis found in patients with psoriasis
        - Thought to be autoimmune, but Dx based on concurrent skin lesions

In his words: Another question about what is pathologic fracture?

- Pathological fracture is when a fracture occurs either spontaneously or with very minor trauma to bones already weakened by disease → most of the diseases talked about in this lecture lead to weakening of bones and therefore, affected patients are more susceptible to pathological fracture

In his words: What kind of cancers often metastasize to the jaw?

- BLT, with a Cold, Kosher, Pickle
    - Breast, Lung, Thyroid, Colon, Kidney, Prostate

In his words: Slide on all tumors - Ewing sarcoma, osteosarcoma, osteoblastoma → Primary bone cancers i.e. osteosarcoma, are not the most common cancers affecting bone → it may be the most common PRIMARY, but not the most common cancer overall; Primary bone tumors - where do they often present?

- Ewing Sarcoma:
    - Rare, aggressive, malignant tumor of mesenchymal stem cell origin
    - Mostly in children and adolescents
- Osteosarcoma:
    - Malignant neoplasm of osteoblastic origin
- Osteoblastoma AKA osteoid osteoma:
    - Benign neoplasm of osteoblasts; may be destructive, but still benign
    - Radiolucent or mixed radiopaque/radiolucent
- What are the most common cancers affecting bone? → primary bone cancers (i.e. osteosarcoma) are NOT the most common cancers affecting bone; it may be the most common PRIMARY, but not the most common cancer overall
    - Cancer inside bone is most commonly caused by metastatic disease (just like cancer seen in the brain)
- Where do primary bone tumors often present?: often affect the long bones (femur, tibia) because the long bones grow for the longest period of time and therefore cells in are mitotically active for a longer duration of time (the more mitotically active a cell type is, the greater chance there is of developing a neoplasm)

In his words: Osteogenesis imperfecta question

- Osteogenesis imperfecta: diverse group of genetic diseases all characterized (in general) by osteopenia and bone fragility
    - Numerous different mutations in many genes; most cases exhibit mutations in type 1 collagen genes, COL1A1, COL1A2
- Clinical features of osteogenesis imperfecta:
    - Depending on severity… multiple bone fractures, growth impairment, bone deformities, hyperextensive joints (may see facial deformities)

- Some cases: blue sclera and hearing deficits
- Dental alterations: will resemble what is seen in dentinogenesis imperfecta → opalescent teeth, pulpal obliteration

In his words: Question on paget's disease

- Paget Disease: abnormal, haphazard deposition of bone with unknown cause→ results in skeletal distortion and weakening
- Who is affected?: primarily people of anglo-saxon descent (high rates in UK, Australia, New Zealand) AND disease is adult onset, usually over 40
- Clinical and radiographic features:
  - Cotton wool appearance of bone radiographically
  - Bone pain
  - Bones become thickened, enlarged, weakened and susceptible to fracture
  - Pelvis, femur, lumbar, vertebrae and skull are most affected
  - Lab testing: elevated serum alkaline phosphatase levels

In his words: Question on osteopetrosis

- Osteopetrosis: rare group of hereditary skeletal disorders characterized by markedly increased bone density.
  - Increased bone density does NOT lead to stronger bones, it leads to bones more susceptible to pathologic fracture. Why? → A: normally, the stronger bone cortex and spongy medulla allow for greater resistance to traumatic force. In osteopetrosis, marrow spaces fill with bone and makes the bone weaker and more susceptible to fracture
- General characteristics:
  - Numerous types of disease with spectrum
  - Some clinical features:
    - Reduced hematopoiesis because bone grows into marrow spaces
    - Increased fractures and osteomyelitis
  - Mutations in one of several genes cause disease

In his words: Question on hypophosphatasia

- Rare, metabolic bone disease characterized by deficiency of alkaline phosphatase
- One of the first presenting signs of the disease is premature loss of the primary teeth
  - Tooth loss is caused by lack of cementum on root surfaces
- Clinical manifestations similar to rickets
  - Growth impairment, bowing of bones, weak bones due to poor mineralization of skeleton

In his words: Question on osteomalacia

- Osteomalacia: Vitamin D deficiency in adults (rickets in children)
    - Weak overall bone structure - susceptible to pathological fracture
    - No tooth abnormalities, dentition has already been developed

In his words: Question on 10 year old patient comes with swelling of left maxilla, radiograph shows glass ground appearance of the affected bones, no other signs and symptoms of disease are noted. What is the best diagnosis?

- Fibrous dysplasia

In his words: Question about medication related osteonecrosis of the jaw

- A side effect of bisphosphonate drugs is MRONJ
- You can see exposed, necrotic bone in the jaw following medication
- The exposed, necrotic bone may get infected and result in osteomyelitis
- Greater severity in cancer patients vs. osteoporosis patients

Infectious Immunological Disease

In his words: Bunch of diseases - which one of these is genetically mediated cause of immune deficiency?

- Bruton's agammaglobulinemia: X-linked B cells are not produced which leads to a lack of antibodies (specifically gamma globulins)
- SCID (Severe combined immunodeficiency disease): Bubble boy
    - Many different mutations (some on X, some autosomal), presentations and inheritance patterns
    - Usually a deficiency of B and T cells
- Chediak-Higashi Syndrome: Decrease in phagocytosis
    - Specifically - reduced neutrophil function
    - Leads to high susceptibility to bacterial infections
        - Other manifestations include albinism and early onset periodontal disease
- Wiskott-Aldrich syndrome: X-linked
    - Eczema, thrombocytopenia, immune deficiency
- Transient hypogammaglobulinemia of infancy: **NOT Genetic!**
    - Infant is slow to start making its own immunoglobulins leading to immune compromise in infancy

In his words: Another question: 43 year old male presents with fatigue and shortness of breath, numerous nodules in lung, they were biopsied and histopathology analysis showed noncaseating granulomas and no fungal organisms. What is the best diagnosis?

- Sarcoidosis

In his words: 55 year old female shows with hardening of skin manifested by microstomia, as well as shortening of fingers in claw-like position, fingers turn blue when cold. What is the best diagnosis?

- Scleroderma (systemic sclerosis)

In his words: An immunocompromised patient with an aggressive infection produces black, necrotic tissue on his palate. What is the diagnosis?

- Mucormycosis - deep fungal

In his words: Which is the most common cause of meningitis in HIV AIDS patients?

- Cryptococcal meningitis - can be fatal

In his words: Which is a common infection obtained at a hospital setting?

- Nosocomial infection

In his words: Slide in infectious disease lecture that goes over terminology - which one is a fungal infection or bacterial infection → histoplasmosis, actinomycosis, mycosis fungoides??

- Mycosis usually means fungal infection, but not always
- Fungal infections:
    - Histoplasmosis, blastomycosis, coccidiomycosis, cryptomycosis, aspergillosis
- Mycosis fungoides: not a fungal infection; it's a T cell lymphoma found on skin
- Actinomycosis: bacterial infection
- Basically know that mycosis fungoides and actinomycosis are the only ones that are not fungal

In his words: What are the oral and skin manifestations of HIV?

- Kaposi's Sarcoma - purple nodules papules and macules
- Necrotizing Gingivostomatitis - punched out papilla similar to leukemia, neutropenia, and agranulocytosis
- Oral hairy leukoplakia (EBV mediated)
- Deep fungal infections - oral blisters, ulcers, or swellings similar to oral cancer

In his words: Question on etiology of oral hairy leukoplakia and Kaposi's sarcoma? The etiologic agent comes from which kind of viruses?

- Oral hairy leukoplakia: EBV mediated
- Kaposi's sarcoma: unusual vascular neoplasm
    - Purple nodules, papules, macules
    - HHV8 associated
    - Associated with immune compromise
        - HIV/AIDS
        - Organ transplant patients
        - Immunosenescence

Hematopathology

In his words: Question on aplastic anemia

- Aplastic anemia: rare and life threatening
  - Causes: environmental toxins and antibiotic (chloramphenicol)
  - Pancytopenia: failure of bone marrow to produce all of the blood cells (RBCs, WBCs, platelets)
  - Oral manifestations: petechial hemorrhages, gingival swelling and spontaneous bleeding, ulceration, pallor and severe periodontal disease.

In his words: Question: 18 year old female presents with spontaneous bleeding on gingiva, present since birth. What is the most likely cause? → know who gets hemophilia, thrombocytopenia, who gets Von Willebrand's disease

- Most likely cause: Von Willebrand
- Hemophilia A and B: males only
- Von Willebrand's disease: females and males
- Thrombocytopenia: marked decrease in circulating platelets → acquired so can't be this

In his words: Question on anemia in general → all of the following are true except one, which one is the exception?

- WHO Definition of anemia: low blood concentration of hemoglobin (<13.0 g/dL)
  - Most often a sign/symptom of another underlying disease
  - Often seen in elderly (but not due to old age)
  - Main symptoms are weakness and fatigue, due to less oxygen
- Most anemias are acquired (NOT inherited)
  - Nutritional deficiencies most common cause worldwide (iron, B12, folate)
  - Inflammatory diseases, chronic infections, liver or kidney disease
  - Diseases of spleen (splenomegaly)
  - Blood cancers (leukemia), cancer treatment (chemotherapeutic drugs)
  - Internal bleeding
- Inherited anemias: Thalassemia and sickle cell disease
  - Defects in genes that produce Hb, hence called hemoglobinopathies

In his words: Which of the following diseases are not likely to produce necrotizing ulcerative gingivostomatitis? Question would be which of the entities produce some level of immunocompromised?

- Diseases that cause NUG:
  - Cyclic neutropenia
  - Generalized neutropenia
  - HIV/AIDS

In his words: Question on leukemia - know which one is seen in young kids

- Types of leukemia:
    - Acute lymphocytic → this one is seen mostly in kids; one of the most common childhood malignancies
    - Acute myelogenous
    - Chronic lymphocytic
    - Chronic myelogenous

In his words: What disease is often seen in young children?

- Diseases usually seen in children:
    - Burkitt lymphoma
    - Acute lymphocytic leukemia
    - Cyclic neutropenia

In his words: Question about what you might see on biopsy in a patient with multiple myeloma?

- Multiple myeloma:
    - Malignancy of plasma cell origin
    - Presents in bone
    - Multifocal
    - Almost always in adults
    - Punched out lesions radiographically
    - Characterized by presence of Bence Jones proteins in urine or serum
        - These are immunoglobulin light chains (plasma cells produce antibodies, so a neoplasm of plasma cells will produce a lot of circulating antibodies

In his words: Question on Hodgkin's disease

- Hodgkin's Disease (AKA Hodgkin's Lymphoma):
    - Malignancy of lymph nodes and spleen
    - Malignant cells are Reed-Sternberg cells
    - Cervical and supraclavicular nodes are most common sites
    - Bimodal peak age (peak #1 between 15-35; peak #2, older than 50) .
    - Many subtypes → all carry different prognosis, treatment regimen

In his words: What is the first line treatment for leukemia?

- Chemotherapy

In his words: Where do we see (what tissues) hyperplastic lymphoid tissue?

- Lymphoid hyperplasia: proliferation of lymphoid tissue in response to insult (usually infection)
- Where do we see lymphoid hyperplasia? → anywhere there is a collection of lymphoid tissue… tonsils, spleen, lymph nodes, and MALT (mucosa associated lymphoid tissue)

- <u>Where do we see lymphoid hyperplasia intraorally?</u>
    - Walderyer's ring - lymphoid tissue in oropharynx
        - Tonsils - filled with lymphoid tissue
            - Pharyngeal, palatine, lingual
        - Soft palate
    - Where else? Floor of mouth, posterolateral tongue