Chapter 6
Hematology/Oncology

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Questions

1. Clinical manifestations of sickle cell anemia include:
   1. Hand-foot syndrome, painful often symmetrical swelling of the hands and feet
   2. Painful, vaso-occlusive crises
   3. Acute chest syndrome
   4. More frequent bacterial infections
      A. 1, 2, 3
      B. 2, 4
      C. 4 only
      D. All of the above

2. Sickle trait:
   1. Is found in approximately 8% of the African-American population in America
   2. Is found in approximately 3% of the Hispanic population in America
   3. Is found in <1% of racial groups in America other than Hispanic and African-American
   4. Is not associated with hemolytic anemia
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

3. The acute chest syndrome:
   1. Is the leading cause of death in sickle cell patients after the age of 10 years
   2. Is only seen in infants with SS disease
   3. Is a syndrome of hypoxemia, CXR infiltrates, and pulmonary infection/infarction
   4. Is best treated with nebulized bronchodilators and vigorous hydration
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

Answers

1. E. All of the above

Sickle hemoglobin differs from normal adult hemoglobin by one amino acid substitution, glutamic acid for valine, at position 6 on the beta chain. Individuals heterozygous for HbS are resistant to falciparum malaria. Affected homozygous SS individuals have severe hemolytic anemia since the sickled cells are poorly deformable and brittle. Clinical manifestations are rarely seen before 6 months of age with the hand-foot syndrome often seen in 1–2-year-old children.
SS disease is a chronic hemolytic anemia with associated crises such as splenic sequestration crises, aplastic crises, and vaso-occlusive crises. Pain crises are the most common type of vaso-occlusive crisis. Below is a list of common clinical manifestations seen in SS disease:

Cerebrovascular accidents, acute chest syndrome, priapism gallbladder disease, hematuria
Renal concentrating defect, cardiomyopathy, infections
A variety of psychological problems including school failure and depression

2. E. All of the above

Individuals heterozygous for HbS typically have no signs or symptoms of sickle cell disease. Rarely, these individuals have painless hematuria. The diagnosis of sickle trait is made with hemoglobin electrophoresis. The RBCs in people with sickle cell trait contain 30–40% HbS; thus sickling does not occur under normal circumstances. In unusual conditions such as shock, very high altitude, or extremely demanding exercise, a vaso-occlusive crisis may occur.

3. B. 1, 3

This clinical syndrome may occur as a complication of postoperative atelectasis. Initially, the child may not appear severely ill, but the condition can progress rapidly. Early detection of any pulmonary compromise in a child with sickle cell disease, followed by vigorous treatment (CPT, incentive spirometry, etc.), is essential given the high mortality of children who develop the syndrome.

4. Regarding infection in children with sickle cell disease:
   1. Osteomyelitis is relatively common, particularly with salmonella.
   2. Encapsulated organisms such as pneumococcus and *Haemophilus influenzae* type b are common etiologic agents.
   3. Serious infection is particularly common in the first 5–6 years of life.
   4. With the newer vaccines, infections are no longer a problem for these children.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

5. Therapy for vaso-occlusive crises includes:
   1. Adequate analgesia
   2. Antibiotics
   3. Adequate hydration
   4. Immobility
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

6. Acute lymphoblastic leukemia (ALL) :
   1. Is the most common leukemia in childhood
   2. Has its peak incidence in children at 10 years of age
   3. May relapse in the bone marrow or CNS
   4. Is treated with total body irradiation
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

7. Chemotherapeutic agents used in the treatment of low-risk ALL include:
   1. Prednisone
   2. Vincristine
   3. Intrathecal methotrexate (MTX)
   4. Bleomycin
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

4. A. 1, 2, 3

The polyvalent pneumococcal vaccines currently available are poorly immunogenic in children under the age of 5 years. Prophylactic penicillin is effective in preventing serious pneumococcal infections in these younger children. Full immunization status is especially important in children with sickle cell disease.

5. B. 1, 3

Children with frequent pain crises are difficult to assess. Children afflicted with this chronic condition often have a flat affect making observer assessment of their degree of discomfort unreliable. Analgesics should not be withheld if the child reports pain, however. Treatment is directed toward preventing complications such as the acute chest syndrome, uncovering etiologies such as infections (osteomyelitis, pneumonia), and providing adequate hydration, nutrition, and comfort.

6. B. 1, 3

ALL occurs with slightly greater frequency in boys than girls. It is subclassified on the basis of immunologic, cytogenetic, and molecular genetic markers. The median ages for the various types of ALL range from <1 year to 7 years. Presenting signs and symptoms are usually nonspecific and include anorexia, lethargy, and irritability. Pallor, bleeding and fever, and signs of bone marrow failure prompt medical attention. There are approximately 200 new cases of ALL/year. Based on survival, ALL is characterized into standard and high risk. Standard risk characteristics in addition to cytogenetic and immunologic factors include age 2–9, female gender, white race, absence of adenopathy, WBC count $<10 \times 10^9$, and absence of CNS disease.

7. A. 1, 2, 3

Without treatment of sanctuaries, relapses in the CNS and testicles were common. Induction generally consists of vincristine, prednisone, and asparaginase, accompanied by intrathecal methotrexate, hydrocortisone, and Ara-C. CNS irradiation is effective in minimizing CNS disease, but it also produces late neuropsychiatric effects.

8. Hodgkin's disease:
   1. Has a bimodal age distribution with peak incidences in the second and fifth decades of life
   2. Commonly presents with painless enlarged cervical lymph nodes
   3. Often causes enlarged mediastinal lymph nodes which may cause cough or other respiratory symptoms
   4. Is sensitive to both chemotherapy and radiation
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

9. Acute cervical adenitis, inflammation of one or more lymph nodes in the neck, is caused by:
   1. *Staphylococcus aureus*
   2. Atypical mycobacteria
   3. Group A streptococcus
   4. Adenoviruses
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

10. Relatively common noninfectious causes of cervical adenitis include:
    1. Hodgkin's disease
    2. Non-Hodgkin's lymphoma
    3. Neuroblastoma
    4. Hemangiomas
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

8. E. All of the above

The lymphadenopathy seen is usually in the cervical area, but axillary and inguinal nodes are sometimes part of the presentation. Hepatosplenomegaly is rare. Fever, weight loss, and night sweats are seen. Staging is important in prognosis and in determining treatment. Disease-free survival rates from 60% to 90% are achieved, based on the staging at diagnosis.

9. A. 1, 2, 3

With chronic infection, signs such as erythema, warmth, and fluctuance are absent. Nodes associated with malignancy are firm and may be fixed to underlying structures or overlying skin. Another infectious cause of cervical adenitis is Kawasaki's disease.

10. B. 1, 3

The most common presentation of Hodgkin's disease is painless, firm adenopathy. Cervical or supraclavicular nodes are commonly involved. Significant enlargement of the nodes in the anterior mediastinum leads to cough, respiratory distress, and cardiovascular embarrassment. This mediastinal involvement is seen the most in older children with Hodgkin's disease. Hodgkin's disease is staged using the Ann Arbor system:

Stage I disease involves a single LN area or a single extralymphatic site.
Stage II is more extensive but on one side of the diaphragm.
Stage III disease is seen on both sides of the diaphragm.
In stage IV, the malignancy is disseminated, involving greater than 1 extralymphatic site.

Neuroblastoma has a very varied presentation. Abdominal pain and mass are a common presentation, but in children with localized disease, adenopathy is also seen.

11. Neuroblastoma, the most common extracranial tumor of childhood and the most frequently diagnosed cancer in infants, presents in a variety of ways including:
    1. With a hard painless mass in the neck
    2. As an abdominal or thoracic mass
    3. With bone pain from skeletal and bone marrow metastases
    4. With seizures from CNS metastases
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

12. Wilms' tumor of the kidney:
    1. Commonly presents with an asymptomatic flank mass
    2. Is often diagnosed in children at 2–4 years of age
    3. Is associated with hypertension in up to 60% of cases
    4. Often metastasizes to the lungs
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

13. Manifestations of graft versus host disease (GVHD) in children include:
    1. Maculopapular rash
    2. Generalized erythroderma with bullae and desquamation
    3. Liver dysfunction manifested by elevated bilirubin
    4. GI disturbances manifested primarily by diarrhea
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

14. Brain tumors:
    1. Are the most common solid tumor in children
    2. In children between the ages of 2 and 12 years are most often located in the posterior fossa
    3. May present with signs of increased intracranial pressure
    4. May present with focal neurological signs
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

11. A. 1, 2, 3

This tumor has a variable presentation since it may develop at any site of the sympathetic nervous system. It is generally discovered as a mass or masses on exam or on a radiologic scan. Treatment varies depending on the stage at diagnosis, as does the chance for survival. Overall cure rate, with all stages included, is approximately 50%.

12. E. All of the above

This tumor accounts for most renal cancer in children. The asymptomatic mass is often discovered by a parent. Surgical removal is indicated even in cases where pulmonary metastases have occurred. With chemotherapy following surgery, survival ranges from 50% to <90% depending on histology and stage.

13. E. All of the above

GVHD occurs when there is a disparity of histocompatibility antigens between the recipient of a bone marrow transplant and the donor marrow. Donor T lymphocytes damage various tissues in the host especially the skin, GI tract (mucositis), and liver. There are acute and chronic forms of the disease with chronic having a worse prognosis.

14. E. All of the above

Brain tumors are the second most common reported malignancy in children and adolescents. Surgery and radiation are the mainstays of treatment. CNS tumors may present with headache, worse in the morning. These tumors are classified by location (infratentorial, supratentorial) and histology.

15. Regarding posterior fossa tumors:
    (a) They tend to present with symptoms of raised intracranial pressure.
    (b) The most common histology, cerebellar astrocytoma, also has the best prognosis.
    (c) Morning headache with associated vomiting may be part of the presentation.
    (d) Medulloblastoma is the second most common posterior fossa tumor.
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

15. E. All of the above

Infratentorial (posterior fossa) tumors predominate in children 4–14 years of age. Nearly one-half of pediatric brain tumors arise in the cerebellum, frequently astrocytomas and medulloblastomas. In another classification system, medulloblastomas, which are poorly differentiated, very malignant tumors, are termed primitive neuroectodermal tumors (PNET).