Pediatric Oncolgy

Signs and Symptoms

The symptoms and signs of cancer are variable and nonspecific in pediatric Unlike cancers in adults, childhood cancers usually dissemination of disease at diagnosis is common and presenting symptoms or signs are often caused by systemic involvement.

Relevant point in the history

Most of the symptoms and signs are not specific and might represent other possibilities in a differential diagnosis.

Pain was one of the initial presenting symptoms in >50% of children
Pallor fatigue, exercise intolerance
Fever
Bleeding : skin or mucosal bleeding
Weight loss
Anorexia, fatigue, malaise, irritability
Organ infiltration can cause lymphadenopathy
Testicular enlargement (leukemia)
Central nervous system (CNS) involvement (cranial neuropathies, headache, seizures).
Respiratory distress may be caused by severe anemia or mediastinal node compression of the airways

Mass effects: Solid tumors may produce mass effects that are nonspecific, such as compression of the thoracic airways or superior vena cava (lymphoma), the optic chiasm and hypothalamic-pituitary region (craniopharyngioma), and the 4th ventricle (cerebellar astrocytoma) , abdominal mass in wilms tumour

Physical Examination

Findings in a child with malignancy are dependent on whether the cancer is systemic or localized.

The cancers most common in children involve the **lymphohematopoietic system** . When the bone marrow is compromised by malignancy (e.g., leukemia, disseminated neuroblastoma), typical findings include
pallor from Anemia
Bleeding, petechiae, or purpura from thrombocytopenia or coagulopathy; Cellulitis or other localized infection from leukopenia; and skin nodules (especially in infants)
hepatosplenomegaly

Lymphadenopathy cervical lymph nodes is common in children
Superior vena cava syndrome from an anterior mediastinal **mass** including respiratory distress, and facial and neck plethora and edema.
Abnormalities of the central nervous system (CNS) that can indicate cancer include : decreased level of consciousness, cranial nerve palsies, ataxia, afebrile seizures, ptosis, decreased visual activity
Neuroendocrine deficits, and increased intracranial pressure (papilledema).
Any focal neurologic deficit in the motor or sensory system,especially a decrease in cranial nerve function, should prompt furtherinvestigation for malignancy.

Abdominal masses
Upper abdominal mass : Wilms tumor, neuroblastoma, hepatoblastoma, germ cell tumors, and sarcomas.
Enlargement of the liver
Mid abdominal masses include non-Hodgkin lymphoma, neuroblastoma, germ cell tumors, and sarcomas.
Lower abdominal masses include ovarian tumors, germ cell tumors, and sarcomas.
Rhabdomyosarcoma usually appears as an extremity mass, particularly in adolescents..
Sacrococcygeal masses in neonates are usually **teratomas**
Ophthalmologic presentation of malignancy includes a **white pupillary reflex, Proptosis** can be produced by rhabdomyosarcoma, neuroblastoma, lymphoma, and Langerhans cell histiocytosis. Horner syndrome, iris heterochromia, and opsoclonus-myoclonus all suggest a diagnosis of neuroblastoma.

Leukemia :
On physical examination, findings of pallor, listlessness, purpuric and petechial skin lesions, or mucous membrane hemorrhage can reflect bone marrow failure
Lymphadenopathy, splenomegaly,
Hepatomegaly less often,.
Patient with Bone or joint pain : may have tenderness over the bone or joint swelling and effusion.
Increased intracranial pressure that indicate leukemic involvement of the CNS. These include papilledema ,retinal hemorrhages, and cranial nerve palsies.
Respiratory distress usually is related to anemia but can occur in patients as the result of a large anterior mediastinal mass (e.g., in the thymus or nodes). This problem is most frequently seen in adolescent boys with T-cell ALL.
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Early Detection

The prognosis of malignancy in children depends primarily on tumor type, extent of disease at diagnosis, and rapidity of response to treatment. Early diagnosis helps to ensure that appropriate therapy is given in a timely manner and thus optimizes the chances of cure.
Delays in diagnosis are particularly likely in certain clinical situations. The
Lymphoma , especially during adolescence, often manifests as an anterior mediastinal mass. Symptoms such as chronic cough, unexplained shortness of breath, or "new-onset asthma" are typical with this presentation and are often overlooked. Tumors of the nasopharynx or middle ear can mimic infection. Prolonged, unexplained ear pain, nasal discharge, retropharyngeal swelling, and trismus should be investigated as possible signs of malignancy.
Early symptoms of **leukemia** may be limited to prolonged or unexplained low-grade fever or bone and joint pain. Blood counts with abnormalities in ≥2 cell lines might indicate the need for bone marrow examination, even when leukemic blast cells are not seen in the blood smear (see Table 520.1).
Select children with certain chromosome abnormalities, such as Down syndrome, Klinefelter syndrome, and WAGR syndrome (Wilms tumor, *a* niridia, *g* enital abnormalities, mental *r* etardation); children with overgrowth syndromes, such as Beckwith- Wiedemann syndrome and hemihypertrophy; and children with certain inherited single-gene disorders, including retinoblastoma, *P53* mutations (Li-Fraumeni

syndrome), familial adenomatous polyposis, and neurofibromatosis (see Table

Ensuring the Diagnosis

When a malignant neoplasm is suspected, the immediate goal is to confirm the diagnosis. A diagnosis can often be established on the basis of the patient's age, symptoms, and location of masses. Selected imaging techniques and tumor markers .Especially when a solid tumor is present, the pediatric oncologist, surgeon, and pathologist should work as a team to determine the site of biopsy, amount of tissue required, and whether fine-needle aspiration, percutaneous image-guided biopsy, incisional biopsy, or excisional biopsy and tumor resection are indicated.

For select situations, at the time of the initial diagnostic procedure, plans for bone marrow aspiration and biopsy

It is important that fresh tissue not be placed in formalin. Besides routine light microscopy, pathologic evaluation may include immunochemistry, flow cytometry, cytogenetics, and molecular genetic studies (e.g., fluorescence in situ hybridization and reverse-transcriptase polymerase chain reaction). Emerging technologies include DNA microarray analysis and cancer genome sequencing that can identify specific gene expression patterns and sequences in tumors. In time, these technologies might ensure more accurate classification and treatment.

Staging

Once a specific diagnosis is confirmed, studies to define the extent of the malignancy are necessary to determine prognosis and treatment.

outlines the minimum evaluation required for common pediatric malignancies.

In addition, for many tumors (e.g., Wilms tumor, neuroblastoma, rhabdomyosarcoma) a surgical staging system is used. Surgical stage can be determined at the time of the initial diagnostic procedure or subsequently. For example, a patient who has abdominal surgery for possible Wilms tumor or neuroblastoma should have careful evaluation and biopsy of all adjacent lymph nodes.

Diagnosis and Staging

Accurate diagnosis and staging of the extent of disease are imperative, especially for childhood cancers that have high cure rates, because the

nature of therapy depends strongly on the type of cancer. In addition, **prognostic subgroups** based on the stage of disease have been established for most cancers that occur in
children.

Diagnostic imaging is a critical phase of evaluation in most children with solid tumors (i.e., cancers other than leukemia). MRI, CT, ultrasonography, scintigraphy (nuclear medicine scans), positron emission tomography (PET), and spectroscopy, as appropriate, all serve a clear purpose in the evaluation of children with cancer, not only before treatment to determine the extent of disease and the appropriate therapy but also during follow-up to determine whether the
therapy was effective.

methods of obtaining tumor tissue (e.g., fine-needle aspiration, percutaneous image-guided biopsy) can be performed in pediatric centers with appropriate expertise in diagnostic imaging, interventional radiology, cytology, and anesthesia support. *Sentinel node mapping* is helpful in the staging of some children's cancers.

T-ALL also usually has a higher leukocyte count.

B-lymphoblastic leukemia is the most common immunophenotype, with onset at 1-10 yr of age. The median leukocyte count at presentation is 33,000/μL, although 75% of patients have counts <20,000/μL; thrombocytopenia is seen in 75% of patients and hepatosplenomegaly in 30–40% of patients. In all types of leukemia, CNS symptoms are seen at presentation in 5% of patients (10–15% have blasts in cerebrospinal fluid [CSF]). Testicular involvement is rarely evident at diagnosis, but prior studies indicate occult involvement in 25% of boys. There is no indication for testicular biopsy

Treatment option of oncology

More than 2 of the primary modalities are often used together, with chemotherapy being the most widely used, followed, in order of use, by surgery, radiation therapy, and biologic agent therapy and Palliative Care