MD2B - Pediatric Oncology: Osteosarcoma

Overview

Osteosarcoma is the most common primary bone cancer in children. It arises from cells that produce new bone and is characterized by the presence of immature bone tissue. This cancer has a bimodal age distribution, most commonly affecting adolescents and young adults, suggesting a link between rapid bone growth and tumor formation. Approximately 1000 new cases of osteosarcoma are diagnosed annually in the United States accounting for about 3% of childhood tumors. Around 500 of these cases occur in children under 20 years of age. Boys are affected more often than girls, and African American children are affected more frequently than Caucasian children. While osteosarcoma can occur in any bone, it most often occurs in large bones with high growth rates, such as the distal femur (thigh bone), proximal tibia (shin bone), and proximal humerus (arm bone). Osteosarcoma frequently metastasizes to the lungs and other bones.

Risks

Certain genetic conditions and environmental factors have been associated with the development of osteosarcoma. Hereditary retinoblastoma, Li-Fraumeni syndrome, Rothmund-Thomson syndrome, Ollier's disease, osteogenesis imperfecta, polyostotic fibrous dysplasia and Paget's disease have all been found to increase the likelihood of developing osteosarcoma. In addition, a history of high dose radiation treatment has been found to increase the risk of developing osteosarcoma.

Symptoms

Osteosarcoma often presents with pain, swelling, redness, or limitation of movement at the site of the tumor. When the tumor is in the leg, children may develop a limp. When the tumor is in the arm, children may experience arm weakness and difficulty lifting heavy objects. The cancer can also weaken the bone, making it prone to fractures even with minimal trauma. Since these tumors occur frequently in active adolescents, initial symptoms are often misdiagnosed as sports-related injuries. Any bone or joint pain that does not improve with conservative therapies such as rest and icing, should be investigated further to rule out serious conditions such as infection or cancer.

Diagnosis

A physician will perform a physical examination, schedule imaging tests, and order laboratory tests to help distinguish osteosarcoma from other bone disorders.

Imaging studies

- X-rays: A diagnostic test using energy beams to produce images of internal tissues, bones, and organs
    - High yield: In osteosarcoma, a sclerotic lesion is observed that is often described as a "sunburst" or "onion skin" pattern. Commonly, elevation of the periosteum may result in the appearance of "Codman's triangle"
- Bone Scans: A diagnostic test using nuclear imaging to visualize bone degeneration, inflammation, or tumor
- Magnetic Resonance Imaging (MRI): A diagnostic imaging procedure that uses large magnets, radio frequencies, and a computer to generate detailed images of internal structures including the spinal cord

and nerve fibers

- Computed Tomography (CT or CAT) Scan: A diagnostic imaging procedure that combines X-rays and computer technology to produce cross-sectional images of the body, often used to visualize soft tissues. If osteosarcoma is suspected, a chest CT will be performed to ensure the cancer has not spread to the lungs

Tissue samples

Samples from the primary tumor and from the bone marrow will be obtained by inserting a needle into the tumor and into the hip bones. These samples will then be examined under the microscope to identify any abnormal or cancerous cells. Obtaining a tissue sample through biopsy is crucial for distinguishing osteosarcoma from other bone conditions, such as Ewing's sarcoma and osteomyelitis.

Treatment

Before the use of chemotherapy, which began in the 1970s, osteosarcoma was treated with surgical resection, usually requiring limb amputation. Unfortunately, over 80% of patients developed metastatic disease, often in the lungs. The introduction of chemotherapy to treatment protocols has significantly increased survival rates for osteosarcoma. Unlike Ewing's sarcoma, however, osteosarcoma is not very responsive to radiation therapy.

Surgery

Surgery is used to obtain local control of the tumor. This may involve a total limb amputation, or a limb-salvage procedure, where only the tumor and some surrounding healthy tissues are removed. If a limb salvage procedure is performed, the removed bone or joint can be replaced with allografts or prosthetic devices. The choice between limb amputation and limb salvage surgery depends on factors such as the tumor’s location, size, and spread, as well as the patient’s age, bone development, and lifestyle. For patients with osteosarcoma that has not spread beyond the bone, studies have shown no difference in overall survival between limb-sparing surgery and total amputation.

Chemotherapy

Chemotherapy is a systemic treatment and treats cancer cells throughout the body. Chemotherapy can be used prior to surgery to shrink the tumor. It can also be utilized after surgery to eliminate any remaining cells.

Prognosis

The addition of chemotherapy to treatment regimens has greatly increased the survival rates of children with osteosarcoma. Over 70% of patients who present with localized disease achieve long term cure. Patients who have pelvic tumors tend to have poorer outcomes, and those with metastases at the time of diagnosis have survival rates under 30%.