

Sarcoma

By James D. Bruckner, MD

The term sarcoma (“sar-Ko-ma”) comes from a Greek word, meaning fleshy growth. Sarcoma is defined as a cancerous tumor of the connective tissues. Normal connective tissue includes fat, blood vessels, nerves, bones, muscles, deep skin tissues, and cartilage. Sarcomas are divided into two main groups -- bone and soft tissue tumors -- and can develop in children and adults. Although rare, there are approximately 11,000 new cases of sarcoma comprised of both bone and soft tissues each year in the United States. This figure is accompanied by approximately 5,000 sarcoma-related deaths.

Bone Cancer

Bones consist of three types of tissue: compact tissue, the hard outer portion of the bone; cancellous tissue, spongy tissue inside the bone containing the bone marrow; and subchondral tissue, the smooth bone tissue of the joints. Cartilage surrounds the subchondral tissue, forming a cushion around the joints.

Bone tumors are either benign (noncancerous) or malignant (cancerous). Benign bone tumors are rarely life threatening, and they do not spread within the body; they can, however, grow and compress healthy bone tissue. Cancer that develops in the bone is called primary bone cancer. It is differentiated by secondary bone cancer, which spreads to the bone from another part of the body.

The most common type of primary bone cancer is osteosarcoma. Because it occurs in growing bones, it is most often found in children. It is very rare, with approximately 2,500 new cases diagnosed in the United States each year. The incidence is slightly higher in males than females, and no race has a higher incidence than another. Ewings sarcoma, although, is the second most common primary bone cancer, and it is even more rare among African and American blacks. Due to the rarity and severity of bone cancer, a bone cancer specialist, such as an orthopedic oncologist, should be consulted in the disease treatment. Another primary bone cancer is chondrosarcoma, which arises in cartilage and occurs more often in adults.

Symptoms of bone cancer vary, depending on the size and location of the tumor, and pain is the most common symptom. Tumors that appear in or around the joints often trigger swelling and tenderness, as well as weakening the bones, causing fractures. Systemic symptoms include weight loss, fatigue, and/or anemia.

Soft Tissue Sarcoma

Soft tissue sarcoma occurs in the muscles, fat, blood vessels, tendons, fibrous tissues, and tissues around joints. The majority of soft tissue sarcomas occur in the legs at or above the knee and less commonly in the hands, arms, and shoulders; chest; abdomen; or hips. An even smaller percentage are located in the head and neck.

Soft tissue sarcomas invade surrounding tissue and metastasize, or spread, to other organs, usually the lung, forming a secondary tumor. Secondary tumors are referred to as metastatic disease because they are part of the original cancer. Slightly more men than women develop soft tissue sarcoma, and the race distribution mirrors that of the United States population.

Early on, soft tissue sarcoma rarely causes symptoms. Because soft tissue is very elastic, these tumors can grow quite large before they are discovered, and the first symptom is usually a painless lump. As the tumor grows and presses against nearby nerves and muscles, pain or soreness and redness of the skin occurs, as well as symptoms of fever or chills. Soft tissue sarcomas are usually identified with magnetic resonance imaging (MRI) or computed tomography (CT) scans and are accurately diagnosed only by a surgical biopsy.

Diagnosis and Treatment

Primary bone and soft tissue sarcomas are treated with surgery, radiation therapy, and chemotherapy. Depending on the size of the tumor, its location, extent, and grade (severity), a combination of all or some of these treatments are used. While amputation of a limb is sometimes necessary, using chemotherapy either before or after surgery allows surgeons to save limbs in most cases. New and more effective treatments are being developed in clinical trials at many hospitals and cancer centers in the United States.

The first step in diagnosing a primary bone or soft tissue sarcoma is a complete medical history and physical examination performed by a physician. X-rays are typically used to locate a tumor first. If an x-ray suggests a tumor is present, then further testing is required such as a CT scan, MRI, or bone scan. Once the tumor is clearly identified, whether suspicious for malignancy or not, the patient is referred to an orthopedic or surgical oncologist if the tumor is located in the abdomen, spine, head, or neck. Finally, a biopsy is conducted to determine if cancer is present.

A biopsy is a procedure used to remove sample tissue from the tumor. A surgeon, usually an orthopedic oncologist, carries out the procedure using a needle or through an incision in a surgical setting. During a needle biopsy, the surgeon makes a small hole into the bone or soft tissues and removes a sample of the tumor with a small instrument. During an incisional biopsy, the surgeon cuts into the tumor, removing sample tissue. The patient is typically under general anesthesia and in a hospital operating room. A pathologist, a doctor specializing in identifying disease, then studies the cells and tissues under a microscope, determining whether the tumor is cancerous.

It is not clear why some people develop sarcoma; researchers, however, have identified some common characteristics in groups with high rates of primary bone or soft tissue sarcoma. Some studies have shown that people exposed to phenoxyacetic acid in herbicides and chlorphenols in wood preservatives have an increased risk of soft tissue sarcoma. Researchers also know that people exposed to high doses of radiation are at a

greater risk for developing soft tissue sarcoma, but they are also studying genetic abnormalities and chromosome mutations as possible causes for soft tissue sarcoma.

Additionally, adults with Paget's disease, which is a disease characterized by abnormal growth of new bone cells, have an increased risk of osteosarcoma. There are also some hereditary conditions which can increase the risk of bone cancer. People with certain inherited diseases, such as neurofibromatosis, have shown to have higher risks of soft tissue sarcoma.

James D. Bruckner, MD, recently voted one of Seattle's best adult orthopaedic surgeons, is an orthopaedic oncologist specializing in the treatment of sarcomas. He has been an orthopaedic surgeon for more than 15 years and also specializes in total joint replacement. He is a significant contributor to the innovation of joint replacement prosthetics and has created innovative implant designs for the limb salvage of numerous sarcoma patients. He obtained his medical degree at Creighton University and completed dual fellowships in adult and pediatric orthopaedic oncology at the University of Washington and Children's Hospital and Regional Medical Center.