Sarcoma – the Forgotten Cancer

July is Sarcoma Awareness Month. Deemed a “forgotten cancer” because of its rarity, sarcomas are cancers that start in bone, muscle, connective tissue, blood vessels or fat, and can be found anywhere in the body. In general, there are two types of sarcoma: soft tissue sarcoma and bone sarcoma with more than 50 different subtypes.

There are two categories of sarcomas:

Soft tissue sarcomas

The term soft tissue refers to tissues that connect, support, or surround other structures and organs of the body. Soft tissue includes muscles, tendons (bands of fiber that connect muscles to bones), fibrous tissues, fat, blood vessels, nerves, and synovial tissues (tissues around joints).

Malignant (cancerous) tumors that develop in soft tissue are called sarcomas, a term that comes from a Greek word meaning “fleshy growth.” There are many different kinds of soft tissue sarcomas. They are grouped together because they share certain microscopic characteristics, produce similar symptoms, and are generally treated in similar ways. (Bone tumors [osteosarcomas] are also called sarcomas, but are in a separate category because they have different clinical and microscopic characteristics and are treated differently.)

Non-soft tissue sarcomas

Non-Soft Tissue Sarcomas – The most common type of bone cancer is osteosarcoma, which develops in new tissue in growing bones. Another type of cancer, chondrosarcoma, arises in cartilage. Evidence suggests that Ewing’s sarcoma, another form of bone cancer, begins in immature nerve tissue in bone marrow. Osteosarcoma and Ewing’s sarcoma tend to occur more frequently in children and adolescents, while chondrosarcoma occurs more often in adults.

In 2019, over 12,750 cases of soft tissue sarcoma and 3,500 cases of bone sarcoma are expected to be diagnosed in the United States, according to data from the National Cancer Institute’s Surveillance, Epidemiology, and End Results Program (SEER). Approximately 5,270 and 1,660 people are expected to die from soft tissue and bone sarcomas, respectively. The five-year survival rate for soft tissue sarcomas is 64 percent, while the survival rate is 66 percent for bone sarcomas.

Here’s a list of five facts to know about sarcoma:

1. Sarcomas are rare.

Sarcomas are rare in adults and make up approximately 1% of all adult cancer diagnoses. They are relatively more common among children. Between 1,500 and
1,700 children are diagnosed with a bone or soft tissue sarcoma in the U.S. each year. This makes up about 15% of cancers in children under the age of 20.

2. **Sarcoma can develop nearly anywhere in the body.**
   Common areas sarcoma tumors grow in include the legs, hands, arms, head, neck, chest, shoulders, abdomen and hips. About 50%-60% of soft tissue sarcomas occur in extremities (arms and legs).

3. **For most sarcomas, the cause is not known.**
   Although there are no known risk factors for sarcoma, certain genetic conditions, radiation exposure and some chemicals may increase the risk of developing sarcoma in some individuals.

   Genetic conditions such as Neurofibromatosis, Gardner syndrome, Li-Fraumeni syndrome, Retinoblastoma, Werner syndrome, Gorlin syndrome, tuberous sclerosis and damage to the lymph system can increase the risk of developing sarcoma.

   Patients might also develop sarcomas from radiation given to treat other cancers like breast cancer, cervical cancer, lymphoma and retinoblastoma. The sarcoma often starts in the area of the body that had been treated with radiation, and on average, can take about 10 years to develop. Radiation exposure accounts for less than 5% of sarcomas.

   Exposure to vinyl chloride, arsenic, dioxin and herbicides that contain high doses of phenoxyacetic acid may also increase risk of sarcoma.

   Additionally, researchers are now studying genetic abnormalities and chromosome mutations as possible causes as well.

4. **Sarcomas are difficult to detect and diagnose.**
   Because they are so rare and can take multiple forms in multiple locations, sarcomas are difficult to detect and often misdiagnosed. In their early stages, soft tissue sarcomas rarely display any symptoms other than a painless lump. As the tumor grows, pain may occur depending on where the tumor is located, or if it presses on nearby nerves.

5. **If you’ve been diagnosed with sarcoma, get a second opinion.**
   With a rare and complex cancer, such as sarcoma, it is important to seek the best possible care to improve your chance of survival. Many doctors have never seen or treated a patient with sarcoma. Get a second opinion from a doctor that specializes in sarcoma about the initial diagnosis and the treatment plan, even if you are being treated at a major cancer center. Using advanced genomic and molecular technologies, NFCR-funded research has made significant discoveries in the development of and treatment for sarcoma.
SARCOMA
SOFT TISSUE & BONE

Sarcomas are cancers of the bone and connective tissue. It is made up of many "subtypes" because it can arise from a variety of tissue structures - nerves, muscles, joints, bone, fat, blood vessels.

If not caught early enough, sarcomas can invade surrounding tissue and metastasize to other tissues and organs of the body. The most frequent location are the limbs since this is where the majority of the body's connective tissue resides. They are commonly hidden deep in the body.

Researchers still don't know exactly what causes most cases of soft tissue and bone sarcoma.

SIGNS + SYMPTOMS

In early stages, there may be no symptoms. As the tumor grows, it may cause other symptoms, such as pain or soreness as it presses against nearby nerves and muscles. Some other common symptoms are:

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PAIN OFF AND ON IN THE AFFECTED AREA, WHICH MAY BE WORSE AT NIGHT

SWELLING, WHICH MAY START WEEKS AFTER THE PAIN

A LIMP (IF SARCOMA IS IN LEG)

TREATMENT

- Surgical removal of cancer tissue
- Chemotherapy to kill cancer cells
- Radiation therapy to kill cancer cells

TYPES OF SARCOMA

SOFT TISSUE

- There are about 50 different types of soft tissue sarcomas
- They can be found in any soft tissue in any part of the body
- Most of them develop in the arms or legs

- The most common types in adults are: malignant fibrous histiocytoma, liposarcoma, and leiomyosarcoma
- The most common type in children is rhabdomyosarcoma

BONE

- Osteosarcoma is the most common type
- It can occur at any age but mostly in children/young adults
- Ewing tumors are the second most common in children
- Most tumors develop in the bones around the knee or the proximal humerus

DON'T IGNORE YOUR LUMPS AND BUMPS!