Sarcomas of the Bone: Staging and Treatment

What is staging for cancer?

Staging is a way to find out how much cancer is in your body, how far it has spread, and what treatment can be used. Staging looks at the size of the tumor, where it is, and if it has spread to other organs. Tests like CT, bone scan, MRI,PET scan, and biopsies may be done to help stage your cancer. Your providers need to know about your cancer and your health so that they can plan the best treatment for you.

Staging looks at the size of the tumor, where it is, and if it has spread to other organs. This article focuses on primary bone cancers (cancers that start in bones) that are most often seen in adults, such as:

- Chondrosarcoma.
- Chordoma.
- Ewing sarcoma.
- Giant cell tumor of bone.

For more information about other types of bone tumors, see our osteosarcoma and bone metastasis articles.

There are two systems used to stage sarcoma of the bone:

1. The Musculoskeletal Tumor Society (MSTS) staging system, also called the Enneking system. It has three parts:

- **G**- Describes the grade of the tumor. Grade is based on what the cancer looks like under a microscope. You may have either of these grades:
  - Low grade (G1) which look more like normal cells.
  - High grade (G2) which look more abnormal.
- **T**- Describes the extent of the “primary” tumor. The tumor can be kept to the bone, called “intracompartmental (T1)” or the tumor can spread beyond the bone to nearby tissue or organs, called “extracompartmental (T2).”
- **M**- Describes if the cancer has spread to other organs (called metastases). Tumors that have not spread are considered M0, while tumors that have spread are M1.

2. The “TNM system,” as described by the American Joint Committee on Cancer has three parts:

- **T**- Describes the size/location/extent of the "primary" tumor in the bone.
- **N**- Describes if the cancer has spread to the lymph nodes.
- **M**-describes if the cancer has spread to other organs (called metastases).

How is sarcoma of the bone staged?

Staging for sarcoma of the bone is based on:

- The location and size of your tumor on imaging tests and what is found after surgery.
- If there is spread to other organs (metastasis).

- If there is cancer in your lymph nodes.

The staging systems can be very complex. Below is a summary. Talk to your provider about which system is used and the stage of your cancer.

Musculoskeletal Tumor Society (MSTS) System

Stage IA (G1, T1, M0): The tumor is low grade (G1), intracompartmental (T1), and has not spread to the lymph nodes or other organs (M0).

Stage IB (G1, T2, M0): The tumor is low grade (G1), extracompartmental (T2), and has not spread to the lymph nodes or other organs (M0).

Stage IIA (G2, T1, M0): The tumor is high grade (G2), intracompartmental (T1), and has not spread to the lymph nodes or other organs (M0).

Stage IIB (G2, T2, M0): The tumor is high grade (G2), extracompartmental (T2), and has not spread to the lymph nodes or other organs (M0).

Stage III (G1 or G2, T1 or T2, M1): The tumor is low grade (G1) or high grade (G2), intracompartmental (T1) or extracompartmental (T2), and has spread to lymph nodes or other organs (M1).

The TNM System

The TNM system below is used mostly for sarcoma of the bone tumors of the arms, legs, trunk, skull, or facial bones. It is not often used for cancer that starts in the pelvis or spine. If you have cancer that started in the pelvis or spine, talk with your provider about how your cancer is staged.

Stage IA (T1, N0, M0, G1 or GX): The main tumor is no more than 8 centimeters (cm), or about 3 inches wide. The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0). The cancer is low grade (G1), or the grade cannot be determined (GX).

Stage IB (T2-T3, N0, M0, G1 or GX): The main tumor is more than 8 cm wide (T2) or there is more than one tumor in the same bone (T3). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0). The cancer is low grade (G1), or the grade cannot be determined (GX).

Stage IIA (T1, N0, M0, G2 or G3): The main tumor is no more than 8 cm wide (T1). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0). The cancer is high grade (G2 or G3).

Stage IIB (T2, N0, M0, G2 or G3): The main tumor is more than 8 cm wide (T2). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0). The cancer is high grade (G2 or G3).

Stage III (T3, N0, M0, G2 or G3): There is more than one tumor in the same bone (T3). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0). The cancer is high grade (G2 or G3).

Stage IVA (Any T, N0, M1a, Any G): The main tumor can be any size, and there may be more than one in the bone (Any T). The cancer has not spread to nearby lymph nodes (N0). It has spread only to the lungs (M1a). The cancer can be any grade (Any G).

Stage IVB (Any T, N1 or Any N, Any M or M1b, Any G): The main tumor can be any size, and there may be more than one in the bone (Any T). The cancer has spread to nearby lymph nodes (N1) or might not have spread to nearby lymph nodes (Any N). It may or may not have spread to distant organs like the lungs or other bones (Any M) or it has spread to distant parts of the body, such as other bones, the liver, or the brain (M1b). The cancer can be any grade (Any G).

How is sarcoma of the bone treated?

Treatment for sarcoma of the bone depends on many things, like your cancer stage, age, overall health, and

testing results. Your treatment may include some or all of the following:

- Surgery.
- Radiation Therapy.
- Chemotherapy.
- Targeted Therapy.
- Immunotherapy.
- Other medications.
- Clinical Trials.

Surgery

You will need to see an orthopedic surgeon (a doctor who specializes in bone and joint surgery) who has worked with sarcoma of the bone tumors. Whenever possible, a biopsy and surgery to remove the tumor should be planned together. The goal of surgery is to remove all of the cancer. The tumor plus some normal tissue around the tumor are removed to lessen the chance of the cancer coming back after surgery.

Two main types of surgery for sarcoma of the bone in the limbs are:

- **Limb-salvage surgery:** Also called limb-sparing. The tumor must be in the arm or leg. The surgeon tries to remove all of the tumor while saving the nearby ligaments, tendons, nerves, and blood vessels so that you can still use your limb as before. If the tumor cannot be removed without affecting the rest of the limb, amputation may be needed.
- **Amputation:** If your tumor is very big or if it reaches into blood vessels or nerves, the limb may need to be removed. Your surgeon will look at MRI scans to plan how much of the limb should be removed. Most times, your surgeon will round off where the amputation is made so that it will fit into a prosthetic (artificial) limb.

Surgery for tumors in other parts of the body may include:

- **Pelvic (hip):** It can be hard to remove the whole tumor with surgery. Chemotherapy is often given before surgery to shrink the tumor. Radiation therapy may also be given after surgery to get rid of all of the cancer.
- **Lower jawbone:** The whole lower part of the jaw may be removed. The jaw can later be replaced with bones from other parts of the body. Radiation therapy may also be used if the whole tumor can’t be removed.
- **Spine or skull:** These are most often treatedwith a mix of chemotherapy, radiation, and surgery.
- **Joint fusion:** Also called “arthrodesis.” Some tumors in joints are hard to remove while keeping the joint in working order. Surgery can be done to fuse the two bones of the joint together.

The most important part of surgery for sarcoma of the bone is the physical therapy and rehabilitation that happens after surgery. Your care team will help set you up with physical, occupational, and other therapies. These will help you with any changes to your mobility or movement after surgery.

Radiation Therapy

Radiation therapy uses high-energy rays kill cancer cells. Radiation therapy is often used in combination with surgery and/or chemotherapy. Radiation may be given before or after surgery. The most common type of radiation used for sarcomas of the bone is external beam radiation therapy. A different kind of radiation therapy used for bone tumors uses radioactive medications, like radium-233. This type of medication is injected into a vein and collects in the bones. Once the medication is in the bones, it starts to kill off cancer cells with its radiation.

Chemotherapy

Chemotherapy is the use of anti-cancer medicines that go through your whole body to treat cancer cells. These medicines may be given through a vein (IV, intravenously) or by mouth. Chemotherapy may be given before surgery (called neoadjuvant treatment), with radiation (called chemoradiation), after surgery (called adjuvant treatment), or as the main treatment if your cancer has spread to other parts of the body or if you can't have surgery.

Chemotherapy may be used to treat Ewing sarcoma and osteosarcoma but is not often used to treat other types of bone cancer, like giant cell tumors, chordomas, and chondrosarcomas.

Some common chemotherapy medications used to treat sarcomas of the bone are doxorubicin, cyclophosphamide, ifosfamide , etoposide, topotecan, irinotecan, temozolomide, cabozantinib, and gemcitabine.

Targeted Therapy

Targeted therapies are medications that target something specific to the cancer cells, stopping them from growing and dividing. Most targeted therapy medications for sarcomas of the bone work on proteins called kinases. These medications are used to treat chordomas that have spread or have come back after treatment (recurrence). Some may also be used to treat advanced chondrosarcomas.

Targeted therapies used to treat sarcomas of the bone are dasatinib, pazopanib, sunitinib, imatinib, erlotinib, lapatinib, sorafenib, and regorafenib.

Immunotherapy

Immunotherapy medications work with the immune system to kill cancer cells. Pembrolizumab is an immunotherapy medication that may be used to treat some sarcomas of the bone. Pembrolizumab binds to the "programmed death receptor" (PD1) protein found on T-cells to stimulate (rev up) your immune system to find and kill cancer cells.

Other Medications

Denosumabis a medication that blocks a protein called RANKL. The RANKL protein tells cells called osteoclasts to break down bone. By blocking RANKL, bones are less likely to break down. This can be helpful with certain types of giant cell tumors of bone that have either come back after surgery or cannot be removed with surgery.This medication is given as a subcutaneous (under the skin, "subq") injection.

Clinical Trials

You may be offered a clinical trial as part of your treatment plan. To find out more about current clinical trials, visit the OncoLink Clinical Trials Matching Services.

Making Treatment Decisions

Your care team will make sure you are included in choosing your treatment plan. This can be overwhelming as you may be given a few options to choose from. It feels like an emergency, but you can take a few weeks to meet with different providers and think about your options and what is best for you. This is a personal decision. Friends and family can help you talk through the options and the pros and cons of each, but they cannot make the decision for you. You need to be comfortable with your decision – this will help you move on to the next steps. If you ever have any questions or concerns, be sure to call your team.

You can learn more about bone cancer at OncoLink.org.

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