

TREATMENT UPDATE:

Tenosynovial Giant Cell Tumor (TGCT)

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Treatment Update: Tenosynovial Giant Cell Tumor (TGCT)

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Tenosynovial Giant Cell Tumor (TGCT) is a rare, benign tumor that grows in the lining of joints or tendon sheaths.

While TGCT is non-cancerous, the tumor can grow and cause damage to the surrounding tissue and structures of the affected area. Symptoms include pain, swelling, warmth or tenderness, reduced range of motion and instability of the affected joint.

There are two types of TGCT:

- **Localized:** The tumor is slow-growing and typically confined to a small area, most commonly the fingers. Wrists and hands can also be affected.
- **Diffuse:** It is more aggressive than the localized type and the tumor can spread outside the joint. Diffuse TGCT primarily affects large joints such as the knee, hip, ankle or shoulder. There may be multiple tumors at the same time and it has a higher rate of recurrence than the localized type.

TGCT is diagnosed through a combination of physical exam, imaging tests (e.g., X-rays, MRI) and biopsy. A sample of synovial fluid (fluid found in the joints) may be taken to look for blood, which is a sign of TGCT.

Treatment Options

Although TGCT is not cancerous, it is most often treated by an oncologist, as they understand how tumors grow and can guide treatment decisions.

Treatment for TGCT is individualized depending on the type of TGCT, its location, its risk of recurring and the symptoms being experienced. Active surveillance (close monitoring) may be an approach if there are no symptoms, the tumor is slow-growing or if surgery poses a significant risk.

Surgery

Surgery is the main treatment for TGCT. The tumor (or tumors) is excised (removed) along with any inflamed joint tissue. The surgery may be “open” (cutting the skin and tissues) or arthroscopic, a less invasive procedure in which a few cuts are made around the joint and a small camera is inserted to guide the surgery. Additional surgery may be needed if the tumor (or tumors) returns.

Total joint replacement (arthroplasty) may be needed in advanced cases where chronic inflammation has caused severe joint degeneration. Arthroplasty improves function and reduces pain by replacing the damaged joint with plastic, metal or ceramic parts.



Radiation

Radiation therapy is primarily used after surgery for diffuse or recurrent TGCT.

- **External beam radiation** uses a machine, called a linear accelerator, to direct multiple beams of radiation to the tumor. The use of CT (computerized tomography) and PET (positron emission tomography) scans allows radiation oncologists to accurately target tumors, shaping the radiation beams to the size and dimensions of the tumor to help spare healthy tissue.
- **Image-Guided Intensity-Modulated Radiotherapy (IG-IMRT)** is a newer, more precise technique that further minimizes damage to surrounding healthy tissue.

Targeted Therapy

Targeted therapies are designed to target the specific cell mechanisms that are important for the growth and survival of tumor cells.

If surgery is not possible or effective, there are two oral therapies approved by the U.S. Food and Drug Administration (FDA) designed to inhibit the Colony-Stimulating Factor 1 Receptor (CSF1R): pexidartinib (Turalio), approved in 2019, and vimseltinib (Romvimza), approved in 2025. The overexpression of CSF1R drives the tumor formation in TGCT.

The Importance of Clinical Trials

Clinical trials are the standard by which we measure the worth of new treatments and the quality of life of patients as they receive those treatments. For this reason, doctors and researchers urge people with certain medical conditions to take part in clinical trials.

Your doctor can guide you in making a decision about whether a clinical trial is right for you. Here are a few things that you should know:

- Often, people who take part in clinical trials gain access to and benefit from new valuable treatments.
- Before you participate in a clinical trial, you will be fully informed as to the risks and benefits of the trial, including any possible side effects.
- Most clinical trials are designed to test a new treatment against a standard treatment to find out whether the new treatment has any added benefit.
- You can stop taking part in a clinical trial at any time for any reason.

Treatment Side Effects

All treatments can cause side effects. It's important that you report any side effects that you experience to your health care team so they can help you manage them. Report them right away—don't wait for your next appointment. Doing so will improve your quality of life and allow you to maintain your treatment plan. It's important to remember that not all patients experience all side effects, and patients may experience side effects not listed here.

Radiation

The side effects of radiation therapy can include dryness, swelling, peeling, redness and (rarely) blistering. If a reaction occurs, contact your health care team so the appropriate treatment can be prescribed. It's especially important to contact your health care team if there is any open skin or painful area, as this could indicate an infection. Infections can be treated with an oral antibiotic or topical antibiotic cream.

Side Effects of Targeted Therapy

Pexidartinib is associated with serious liver-related side effects, and the medication is only available through the FDA-mandated Turalio REMS (Risk Evaluation and Mitigation Strategy) program. There is no evidence of liver injury with vimseltinib; the most common side effects include swelling, rash, headaches and fatigue.

General Side Effects

Some side effects may occur across treatment approaches. This section provides tips and guidance on how to manage these side effects should they occur.

Managing Digestive Tract Symptoms

Nausea and vomiting

- Avoid food with strong odors, as well as overly sweet, greasy, fried or highly seasoned food.
- Eat meals that are chilled, which often makes food more easily tolerated.
- Nibble on dry crackers or toast. These bland foods are easy on the stomach.
- Having something in your stomach when you take medication may help ease nausea.

Diarrhea

- Drink plenty of water. Ask your doctor about using drinks such as Gatorade which provide electrolytes. Electrolytes are body salts that must stay in balance for cells to work properly.
- Over-the-counter medicines such as loperamide (Imodium A-D and others) and prescription drugs are available for diarrhea but should be used only if necessary. If the diarrhea is bad enough that you need medicine, contact a member of your health care team.

- The BRAT diet (bananas, rice, applesauce, toast) and soluble fiber such as oats, bran and barley can help with diarrhea. Foods high in insoluble fiber, such as leafy greens and most fruits should be avoided as they can worsen diarrhea. Oily foods, caffeine and alcohol should also be avoided.
- Avoid food high in refined sugar and those sweetened with sugar alcohols such as sorbitol and mannitol.

Constipation

- As hydration is important to avoid constipation, make sure to drink plenty of fluids. Also, limit your intake of caffeine and alcoholic beverages, as they can cause dehydration.
- Include foods high in fiber in your daily diet, such as fruit (especially pears and prunes), vegetables and cereals. If your health care team approves, you may want to add synthetic fiber to your diet, such as Metamucil, Citrucel or FiberCon.
- Be as physically active as you can, after checking with your health care team on the level of physical activity that is right for you.
- If your doctor has prescribed a “bowel regimen,” make sure to follow it exactly.



Loss of appetite

- Eating small meals throughout the day is an easy way to take in more protein and calories, which will help maintain your weight. Try to include protein in every meal.
- To keep from feeling full early, avoid liquids with meals or take only small sips (unless you need liquids to help swallow). Drink most of your liquids between meals.
- Keep high-calorie, high-protein snacks on hand such as hard-boiled eggs, peanut butter, cheese, ice cream, granola bars, liquid nutritional supplements, puddings, nuts, canned tuna or trail mix.
- If you are struggling to maintain your appetite, talk to your health care team about whether appetite-building medication could be right for you.

Managing Fatigue

Fatigue (extreme tiredness not helped by sleep) is one of the most common side effects of many treatments. If you are taking a medication, your doctor may lower the dose of the drug, as long as it does not make the treatment less effective. If you are experiencing fatigue, talk to your doctor about whether taking a smaller dose is right for you.

There are a number of other tips for reducing fatigue:

- To be able to sleep well at night, avoid excessive sleep during the day.
- Take short walks or do some light exercise, if possible.
- Try easier or shorter versions of the activities you enjoy.
- Ask your family or friends to help you with tasks you find difficult or tiring.

There are also prescription medications that may help, such as modafinil. Your health care team can provide guidance on whether medication is the right approach for your individual circumstances.

Fatigue can be a symptom of other illnesses, such as anemia, diabetes, thyroid problems, heart disease, rheumatoid arthritis and depression. So be sure to ask your doctor if they think any of these conditions may be contributing to your fatigue.

Managing Pain

To help your doctor prescribe the best medication, it's useful to give an accurate report of your pain. Keep a journal that includes information on:

- Where the pain occurs
- When the pain occurs
- How long it lasts
- How strong it is on a scale of 1 to 10, with 1 being the least amount of pain and 10 the most intense
- What makes the pain feel better and what makes it feel more intense

There are a number of options for pain relief, including prescription and over-the-counter medications. It's important to talk to a member of your health care team before taking any over-the-counter medication, to determine if they are safe and will not interfere with your treatments.

Physical therapy, acupuncture and massage may also be of help in managing your pain. Consult with a member of your health care team before beginning any of these activities.

Communicating With Your Health Care Team

As you manage your TGCT, it's important to remember that you are a consumer of health care. The best way to make decisions about health care is to educate yourself about your diagnosis and get to know the members of your health care team, including doctors, nurse practitioners, physician assistants, nurses, dietitians, social workers and patient navigators.

Here are some tips for improving communication with your health care team:

Start a health care journal. Having a health care journal or notebook (either on paper or in a digital format) will allow you to keep all of your health information in one place. You may want to write down the names and contact information of the members of your health care team, as well as any questions for your doctor.

Prepare a list of questions. Before your next medical appointment, write down your questions and concerns. Because your doctor may have limited time, ask your most important questions first and be as specific as possible.

Bring someone with you to your appointments or have them be present during telehealth sessions. Even if you have a journal and a prepared list of questions or concerns, it's always helpful to have support when you go to your appointments. The person you bring may also think of questions to ask your doctor or remember details about your symptoms or treatment that you may have forgotten.

Write down your doctor's answers. Taking notes will help you remember your doctor's responses, advice and instructions. You can also ask the person who accompanies you to take notes for you, either in your journal or on a tablet or smartphone.

Record your visit if your doctor allows it. Recording the conversation with your doctor gives you a chance to hear specific information again or share it with family members or friends.

Incorporate other health care professionals into your team.

Your oncologist is an essential member of your health care team, but there are other health care professionals who can help you manage your diagnosis and treatment:

- Your primary care physician should be kept updated about your TGCT treatment and any test results.
- Your local pharmacist is a great source of knowledge about the medications you are taking. Have all of your prescriptions filled at the same pharmacy to avoid the possibility of harmful drug interactions.
- Make sure your oncologist knows of any other medical conditions you have or any pain you are experiencing so that they can consult with your primary care physician or specialists as needed.

Remember, there is no such thing as over-communication.

CancerCare's Free Support Services and Programs

It can be very difficult to receive a diagnosis of TGCT, and adjusting to the necessary changes in your life can be challenging.

CancerCare can help. We are a national nonprofit organization providing free, professional services to anyone affected by cancer. Our licensed oncology social workers can provide support and education, help in navigating the complicated health care system and offer information on support groups and other resources.

To learn more about how CancerCare helps, call us at 800-813-HOPE (4673) or visit www.cancercares.org.

You will likely also build your own personal support network composed of family and friends. In doing so, it's best to take some time to think about the people in your life and how they are best suited to help. Match the task to their strengths—ask a family member who loves to shop to pick up something for you at the store, or ask a friend who's a good listener to come over for a chat.



Frequently Asked Questions

Q: How rare is TGCT?

A: TGCT is extremely rare, with only 11-50 cases per million people each year. Although it can affect people of any age, it is most commonly diagnosed between the ages of 30 and 50, and is slightly more common in women than in men.

Q: What is the recurrence risk of TGCT? Are there any dietary or lifestyle changes that help reduce recurrence?

A: The lifetime recurrence risk in localized TGCT is as high as 15%; in diffuse TGCT it can be as high or higher than 70%. No specific diet or lifestyle modifications have been identified as reducing the risk of TGCT or its recurrence.

Q: Is TGCT ever treated with chemotherapy or immunotherapy?

A: TGCT is not treated with chemotherapy or immunotherapy. The only medicines approved by the FDA to treat TGCT are targeted therapies that inhibit (block) the Colony-Stimulating Factor 1 Receptor (CSF1R); the overexpression of which leads to tumor growth.

Q: Are there any clinical trials in progress for TGCT?

A: The phase III MANEUVER trial is evaluating the investigational therapy pimicotinib for the treatment of TGCT. Like the two FDA-approved therapies, pimicotinib inhibits CSF1R.

Resources

CancerCare®

800-813-HOPE (800-813-4673)

www.cancercare.org

Information from EMD Serono

www.tgctwrecks.com

TGCT Support

www.tgctsupport.org

CLINICAL TRIAL WEBSITES**Clinicaltrials.gov**

www.clinicaltrials.gov

National Cancer Institute

www.cancer.gov

Carebox (formerly EmergingMed)

www.careboxhealth.com

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