PRACTICE AID

Tools for Characterizing and Diagnosing TGCT

PeerView Oncology

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Tenosynovial Giant Cell Tumor (TGCT): a rare, locally aggressive, mesenchymal neoplasm that most often arises in the synovium of joints, bursae, or tendon sheaths

General Diagnostic Factors for TGCT[1-5]

*Presentation can be **localized** or **diffuse***

Localized TGCT[6-10]

- Mostly monoarticular and commonly seen in small joints
- Majority occur in the digits
- Lesions are usually nodular, well-circumscribed, at least partially encapsulated, and small
- Common presenting symptom is painless swelling
- Pain may develop, especially with tumor growth

Diffuse TGCT[6-10]

- Mostly monoarticular and commonly seen in large joints
- Most often occurs in the knee
- Tumors are usually large (>5 cm) and have poorly defined borders
- Common symptoms are pain, swelling, limited joint motion; locking/catching may also occur
- Pain may be insidious, intermittent, or sustained

In localized and diffuse TGCT, tumors can interfere with joint function

Differential Diagnosis of TGCT

Localized TGCT	Diffuse TGCT
• Localized trauma/ joint degeneration • Ganglion cyst • Xanthomas • Hemangiomas • Giant cell tumor of the bone	• Meniscal tear or sports injuries • Synovial proliferative disorders, such as rheumatoid arthritis and synovial chondromatosis • Fibromas • Xanthomas • Soft tissue sarcomas

Other Important Considerations

- Diagnosis is most commonly confirmed via MRI
- Diagnosis can also be determined by presurgical biopsy or during surgery
- Delays in diagnosis can result in treatment planning at a more advanced stage when there is tumor involvement in bones, muscles, or tendons, particularly in diffuse disease

1. Gouin F et al. *Orthop Traumatol Surg Res.* 2017;103(1s):s91-s97. 2. Bisbinas I et al. *J Foot Ankle Surg.* 2004;43:407-411. 3. Illian C et al. *J Med Case Rep.* 2009;3:9331. 4. Akinci O et al. *Acta Orthop Traumatol Turc.* 2011;45:149-155. 5. Verspoor FGM et al. *Rheumatology (Oxford).* 2014;53:2063-2070. 6. Lucas DR. *Arch Pathol Lab Med.* 2012;136:901-906. 7. de Saint Aubain Somerhausen N, van de Rijn M. In: Fletcher CDM eds. WHO Classification of Tumours of Soft Tissue and Bone. Vol 5. 4th ed. Lyon, France: IARC Press;2013:100-101. 8. Mastboom MJL et al. *Acta Orthop.* 2017;88:688-694. 9. Rao AS et al. *J Bone Joint Surg Am.* 1984;66:76-94. 10. Gelhorn HL et al. *Clin Ther.* 2016;38:778-793.

Access the activity, "Putting Tenosynovial Giant Cell Tumor on Notice: Multidisciplinary Treatment and Implications of New Science," at **PeerView.com/TGCT.**

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Understanding the TGCT Management Landscape

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The NCCN recommends that patients with TGCT are evaluated and managed by a multidisciplinary team with experience and expertise in sarcoma[1]

Multidisciplinary Management

- A team-based approach to managing TCGT is recommended
- Specialists collaborating on TGCT care typically include surgeons, medical oncologists, pathologists, radiologists, orthopedists/orthopedic oncologists, oncology nurses, and physical/rehabilitation therapists

Surgery

- Localized and diffuse TGCT are typically treated with surgery to remove or reduce the size of the tumor
- Surgery is the most common way to treat TGCT; techniques include open and arthroscopic surgery
- Surgery can be curative; but in some cases, particularly in diffuse disease, surgery may not prevent recurrence

Targeted Therapy (Kinase Inhibitors)[2]

- Pexidartinib is FDA approved for the treatment of adults with symptomatic TGCT associated with severe morbidity or functional limitations and not amenable to improvement with surgery
  - Dosing: 400 mg orally twice daily on an empty stomach, at least 1 hour before or 2 hours after a meal or snack
  - Category 1 recommended treatment option (NCCN)[1]
  - The Marketing Authorization Application is validated by the European Medicines Agency (EMA)
- Different types of patients can be considered candidates for pexidartinib, including individuals with recurrent TGCT for whom additional surgery would worsen functional limitation, or those who are not surgical candidates after evaluation by a multidisciplinary team

Palliative therapy options for patients with TGCT may include: pain medications, anti-inflammatory drugs, or steroids

Imatinib mesylate is recommended as a category 2A treatment option (NCCN)[1]

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Recurrent TGCT is a challenging condition that requires the expertise of many disciplines to ensure effective management

Guide to Recurrent Disease[3-7]

- Up to 15% of localized TGCT can recur when the tumor is completely resected
- 20%-50% of diffuse TGCT may recur post resection
- Patients with diffuse TGCT may require multiple surgeries, which can lead to substantial morbidity, possible future joint arthroplasty, and/or, in extremely rare cases, amputation
- Long-term course of recurrent disease may be associated with loss of joint function or early secondary osteoarthritis

Multidisciplinary management of TGCT, including recurrent disease, should take place at specialized sarcoma centers

Many resources are available to assist your patients in finding specialized care centers

https://sarcomaalliance.org/resources/sarcoma-center/list/#

http://sarcomahelp.org/sarcoma-centers.html

1. NCCN Clinical Practice Guidelines in Oncology: Soft Tissue Sarcoma. Version 4.2019. https://www.nccn.org/professionals/physician_gls/PDF/sarcoma.pdf. Accessed October 21, 2019. 2. Turalio (pexidartinib) Prescribing Information. https://dsi.com/prescribing-information-portlet/getPIContent?productName=Turalio&inline=true. Accessed October 21, 2019. 3. Ravi V et al. *Curr Opin Oncol.* 2011;23:361-366. 4. Xie G-P et al. *PLOS One.* 2015;10:e0121451. 5. Gouin F et al. *Orthop Traumatol Surg Res.* 2017;103(1s):s91-s97. 6. Dines JS et al. *Arthroscopy.* 2007;23:930-937. 7. Ogilvie-Harris DJ et al. *J Bone Joint Surg Am.* 1992;74:119-123.

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