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The Tumor That's Not So Tiny: Understanding the Impact of TGCT

Announcer:
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Dr. Gelderblom:
This is CE on ReachMD, and I'm Dr. Hans Gelderblom. I'm medical oncologist at the Bone and Soft Tissue Tumor Clinic at Leiden University Medical Center in the Netherlands. And I'll be talking about tenosynovial giant cell tumors, TGCT, today.

Tenosynovial giant cell tumor was previously called PVNS or giant cell tumor of tendon sheaths. It's a rare mesenchymal neoplasm arising from the synovium of joints and tendon sheaths. It's a molecular characterized, recurrent genomic aberration, often involving the CSF1 gene. And most patients affected by TGCT are young adults, and usually the disease is non-life-threatening. The disease and its treatment may impact the quality of life.

Here, you can see the skeleton, and you can see the difference between the locations of the localized—or nodular—and the diffuse form on the right-hand side. So the nodular form arises in the soft tissue near tendons or interphalangeal joints. Large joints are infrequently affected. In contrast, the diffuse form shows extensive and infiltrative involvement of the synovium of the joint and/or the tendon sheaths and extends into the extra-articular structures. It can also cause hemarthrosis, destruction of bone and cartilage with severe disability, as well as frequent local relapse after surgery.

Here, you can see some images taken from the consensus paper that was established and published in 2023. It shows an MRI imaging, which is the preferred technique for detection and characterization of TGCT.

When there's no pathology specimen available—as you can see here on the right-hand side, some examples—prior to surgery, then lab biopsy may be indicated when the radiological appearance is not typical, and an expert pathologist is then indicated.

Here, you see the burden of disease. It is a disease that recurs frequently after surgery, and you can see on the green line the diffuse form recurs in almost half of the cases, which is really important because the recovery time from surgery takes about 3 months.

So with regard to referrals, as I explained, TGCT is a new disease, and it was formerly seen as a synovitis, but it's really a benign tumor that should be treated as a tumor. Often, sports medicine physicians see these patients first, and there's a need to avoid misdiagnosis—for instance, arthritis or sports injuries—although in the patient history there might be a recent sports injury, but it could still be a TGCT.

So the primary treatment may also involve a general surgeon, an interventional radiologist, a rheumatologist, or a radiotherapist. And it's

important to acknowledge that there is not enough evidence to advise radiosynoviorthesis, cryotherapy, or standard radiotherapy. And the last one is mainly due to the fact that it may affect the function of the joint and may induce secondary tumors. So it's quite important to send these patients to a center with multidisciplinary experience with TGCT.

Awareness is therefore so much important, and I'm really happy that the patient community in this disease is very strong. You can see the TGCT support website, which is not only helpful for patients but also for physicians. And I would like to ask you to have a look at it.

So my time is up, and I hope you found this overview helpful, and thanks for listening.

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