Peripheral T-cell lymphoma not otherwise specified (PTCL-NOS)

When a T-cell lymphoma doesn’t fit into any of the other types of T-cell lymphoma, it is called peripheral T-cell lymphoma not otherwise specified (PTCL-NOS). This page is about PTCL-NOS.

On this page

What is PTCL-NOS and who gets it?

Symptoms

Treatment

Relapsed or refractory PTCL-NOS

Research and targeted treatments

Follow-up

What is PTCL-NOS and who gets it?

PTCL-NOS is the most common type of mature T-cell lymphoma, accounting for around a third of all cases of T-cell lymphomas diagnosed. The lymphomas within this group can be quite different to each other. However, they share some similarities, such as the symptoms they cause. When more evidence becomes available in the future, PTCL-NOS is likely to be split into several subtypes.
PTCL-NOS can occur at any age, but it is more common to be first diagnosed in people around the age of 60. It is more common in men – for every 3 men diagnosed with PTCL-NOS, 2 women are diagnosed. Most people have advanced stage disease when they are diagnosed.

Symptoms

People with PTCL-NOS often have enlarged (swollen) lymph nodes (glands) in their neck, armpit or groin – often in more than 1 of these places. PTCL-NOS is also often found in extranodal areas (outside of the lymph nodes):

- the bone marrow (the spongy tissue in the centre of bones where blood cells are made) is often affected, which can lead to anaemia (low red blood cells) and thrombocytopenia (low platelets)
- the liver and spleen might be enlarged
- the skin can be affected by itchy red patches.

‘B symptoms’ (fevers, night sweats and weight loss) are common in people with PTCL-NOS; these symptoms often appear together.

Treatment

Most people with PTCL-NOS are treated with chemotherapy. Because T-cell lymphomas are rare, it is difficult to determine which treatment gives the best outcome. New, targeted drugs are being tested for PTCL-NOS and different chemotherapy regimens (combinations of drugs) are being compared in clinical trials. You might be asked if you would like to take part in a clinical trial to help test new treatments and to find out what the best treatment is for PTCL-NOS.

The most common chemotherapy regimen used for PTCL-NOS is CHOP. This is made up of:

- cyclophosphamide
- hydroxydaunorubicin
Chemotherapy is given in cycles. The drugs are given on certain days, which are followed by a break in treatment for your body to recover before the next cycle begins.

Other options might include CHOEP (CHOP with the drug etoposide) or regimens that include the drug gemcitabine.

If you are well enough, and your lymphoma has been reduced by chemotherapy, your doctor is likely to recommend you have more chemotherapy together with a stem cell transplant. This is to give you a better chance of staying in remission (no evidence of lymphoma) for a long time. ICE chemotherapy (ifosfamide, carboplatin and etoposide) is often used before a stem cell transplant, but your doctor might suggest a different regimen.

Relapsed or refractory PTCL-NOS

It is common for PTCL-NOS to relapse (come back) after being in remission. Sometimes, the lymphoma is refractory (doesn’t respond) to treatment. In these cases, your doctor might consider:

- a different chemotherapy regimen, eg GEM-P (gemcitabine, cisplatin and methylprednisolone)
- an allogeneic (donor) stem cell transplant if your lymphoma is reduced by chemotherapy and you are well enough
- a targeted drug, usually through a clinical trial.

Research and targeted treatments

Many targeted drugs are being tested to see if they can help people with T-cell lymphoma.
Some of the targeted drugs described here might be available to you through a clinical trial. If you are interested in taking part in a clinical trial, ask your doctor if there is a trial that might be suitable for you. You can find out more about clinical trials and search for trials at Lymphoma TrialsLink.

Drugs being tested in PTCL-NOS but not yet approved in the UK include:

- **Cell signal blockers** that disrupt signals that keep cancer cells alive or allow them to grow. They act on different pathways in cells:
  - **HDAC inhibitors** like belinostat (Beleodaq®) or romidepsin (Istodax®), both of which are approved in the US for use in people with relapsed or refractory T-cell lymphoma.
  - PI3-K inhibitors like duvelisib
  - **PD1 or PD-L1 inhibitors** like nivolumab and avelumab
- **Immunomodulatory drugs** like lenalidomide (Revlimid®) and thalidomide. These affect the activity of the immune system in several different ways. They can help the immune system to attack the lymphoma cells and prevent the lymphoma from growing.
- **Proteasome inhibitors**, like bortezomib (Bortezomib Accord or Velcade®) and carfilozomib. These disrupt the balance of proteins in cells, causing them to die.

New drugs are being developed and tested all the time. This is not intended to be a comprehensive list of drugs in development.

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**Follow-up**

When your treatment is finished, you have regular **follow-up appointments** at the hospital. People who stay in remission after treatment for PTCL-NOS are usually followed up for around 3 years after the end of their treatment.

You should contact your medical team or GP if you develop any symptoms of lymphoma or have other concerns between your appointments. Your specialist might bring your appointment forward if they think they need to see you sooner.
References

These are some of the sources we used to prepare this information. The full list of sources is available on request. Please contact us by email at publications@lymphoma-action.org.uk or phone on 01296 619409 if you would like a copy.


Further reading

- Overview of T-cell lymphomas
- Chemotherapy
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