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

This information is about peripheral T-cell lymphoma Not Otherwise Specified (PTCL-NOS). PTCL-NOS is when a T-cell lymphoma doesn't fit into any particular category of T-cell lymphoma.

On this page

We have separate information about the topics in **bold font**. Please get in touch if you'd like to request copies or if you would like further information about any aspect of lymphoma. Phone 0808 808 5555 or email information@lymphoma-action.org.uk.

What is PTCL-NOS?

T-cell lymphomas are very rare. PTCL-NOS is a type of **T-cell lymphoma** – a **non-Hodgkin lymphoma** that develops from white blood cells called **T cells**. PTCL-NOS includes all T-cell lymphomas that don't fit into any other category. It is one of the more common types of these rare T-cell lymphoma.

The lymphomas within this group can be quite different from each other. However, they might share some common symptoms. As more research is done in the future, it is likely that PTCL-NOS will be split into several subtypes of T-cell lymphoma rather than being discussed as a single type of lymphoma.

Who gets PTCL-NOS?

T-cell lymphoma is a very rare type of lymphoma. PTCL-NOS is the most common T-cell lymphoma, accounting for around 1 in 3 cases. It can occur at any age, but it is more common in people in their 60s. It tends to affect more men than women. Most people have **advanced stage** lymphoma when they are diagnosed.

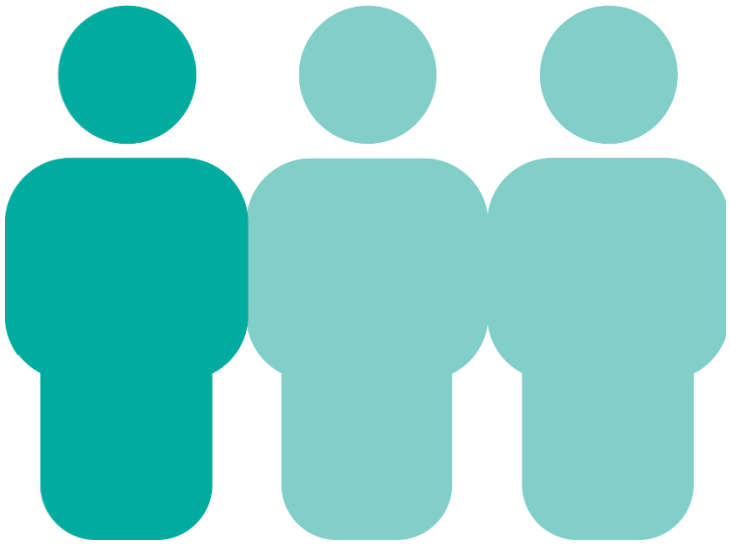

Figure: PTCL-NOS accounts for around 1 in 3 cases of T-cell lymphomas.

Symptoms of PTCL-NOS

The most common symptom people with PTCL-NOS might have is a **swollen lymph node or nodes**. These can develop anywhere in the body but the most common places are the neck, armpit or groin. PTCL-NOS is also commonly found outside of the lymph nodes (extranodal areas).

- 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 platelet levels).
- The liver and spleen might be enlarged, causing **bloating or tummy (abdominal) pain**. Lymphoma elsewhere in your gut might cause pain, sickness, diarrhoea or vomiting.
- The skin can be affected by itchy red patches.
- If you have lymphoma in your chest, you might feel short of breath, develop a cough or have pain or a feeling of pressure in your chest.

Fevers, night sweats and unexplained weight loss (known as '**B symptoms**') are also common in people with PTCL-NOS.

Looking back, I only recognised my symptoms in hindsight. I was getting more infections than usual and losing more weight than expected. Then my stomach started to swell and I started having night sweats and lower itchy limbs. I sought medical advice after I developed a high temperature and hand cramps. A PET scan revealed I had PTCL-NOS
Clare, affected by PTCL-NOS

Treatment of PTCL-NOS

PTCL-NOS is the most common type of T-cell lymphoma, but like all T-cell lymphomas, it is still rare. This makes it difficult for doctors to determine which treatment will give you the best outcome.

There are no standard treatments for PTCL-NOS. Several **targeted drugs** are being tested for PTCL-NOS and different **chemotherapy regimens** (combinations of drugs) are being compared in **clinical trials**. Your doctor might ask you if you would like to take part in a clinical trial to test new treatments and help find out what the best treatment is for PTCL-NOS.

If you don't want to take part in a clinical trial, or if there isn't one that is suitable for you, you are likely to be treated with **chemotherapy**. The most common chemotherapy regimen used for PTCL-NOS is **CHOP**:

- **c**yclophosphamide
- doxorubicin (or **h**ydroxydaunorubicin)
- vincristine (also known as **O**ncovin®)
- **p**rednisolone (a steroid).

Your lymphoma specialist might also recommend a chemotherapy drug called **e**toposide in addition to CHOP (CHEOP), or you might have a different chemotherapy regimen altogether. If you are not fit enough to have CHOP or CHEOP, you might be treated with less intensive chemotherapy, such as gemcitabine or bendamustine.

If you respond to chemotherapy and you are well enough, your doctor is likely to recommend that you have a **self (autologous) stem cell transplant**. This gives you a better chance of staying in **remission** (no evidence of lymphoma) after treatment.

I had chemotherapy which removed the cancerous cells, but I then had an allogeneic (donor) stem cell transplant. It involved spending a lot of time in isolation so I decided to make it like a retreat! I took in my own duvet instead of using hospital sheets and blankets. I put posters up all over the walls – like I was at university again! I brought my Playstation and a load of books and a diary.
Clare, affected by PTCL-NOS

Relapsed or refractory PTCL-NOS

PTCL-NOS might respond well to initial treatment, but it is common for it to come back (**relapse**) after either an initial response or having achieved a complete **remission**. However sometimes, PTCL-NOS fails to respond to treatment (refractory lymphoma). In these cases, your doctor might consider:

- a different chemotherapy regimen, such as GDP (Gemcitabine, Dexamethason, Platinum), GemOx (Gemcitabine, Oxaliplatin), DHAP (**d**examethasone, **h**igh-dose cytarabine [also known as **A**ra-C] and cisplatin [also known as **P**latinol®]) or
  ICE (**i**fosfamide, **c**arboplatin and **e**toposide)
- a **donor (allogeneic) stem cell transplant** if your lymphoma responds to more 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. Drugs that have been or are being tested in PTCL-NOS include:

- **antibody–drug conjugates** such as **brentuximab vedotin**
- **immune checkpoint inhibitors** such as avelumab and pembrolizumab
- **proteasome inhibitors** such as bortezomib or carfilzomib
- **HDAC inhibitors** such as vorinostat, romidepsin or belinostat
- a chemotherapy drug called **pralatrexate**
- **antibody treatments** such as alemtuzumab, which binds to a protein called CD52 on T cells

Some of these 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. To find out more about clinical trials or search for a trial that might be suitable for you, visit **Lymphoma TrialsLink**.

References

The full list of references for this page is available on our website. Alternatively, email **publications@lymphoma-action.org.uk** or call 01296 619409 if you would like a copy.

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