Extranodal NK/T-cell lymphoma (nasal type)

This information page is about extranodal NK/T-cell lymphoma, nasal type – a T-cell lymphoma that usually starts around the nose.

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What is extranodal NK/T-cell lymphoma and who gets it?

Extranodal NK/T-cell lymphoma is a rare type of non-Hodgkin lymphoma (NHL) that develops from natural killer (NK) cells or T lymphocytes (T cells). NK and T cells are both types of lymphocyte. This lymphoma develops at extranodal sites (outside the lymph nodes, also known as glands) most often the nose, which is why it is called ‘nasal-type’.

We have an overview page that explains more about what T-cell lymphoma is.
This type of lymphoma is most common in people from Asia and Central and South America. It is rare in people from other parts of the world, including the UK. It usually occurs in adults, at an average age of around 50. It affects more men than women.

There is a link between extranodal NK/T-cell lymphoma and infection with a virus called the Epstein–Barr virus (EBV), which also causes glandular fever. Around 9 in 10 people in the general population have been infected with EBV by adulthood and the vast majority do not go on to develop lymphoma. It is not known why a few people who have EBV also develop extranodal NK/T-cell lymphoma.

## Symptoms

Extranodal NK/T-cell lymphoma is usually found only in the area around the nose (‘nasal’) when it is diagnosed. This means it is most often at stage 1E or stage 2E, where ‘E’ stands for ‘extranodal’. A few people have more widespread lymphoma (stage 3 or 4) when they are diagnosed.

A destructive mass most often forms inside the nose or in the sinuses (air-filled spaces) around the nose.

You might have other symptoms that affect your nose, eyes, mouth or face, like:

- a blocked nose
- discharge or bleeding from your nose
- weepy eyes
- swelling of your face
- problems swallowing
- problems with your teeth.

The lymphoma can spread to areas around your nose, like your upper airway, eyes and mouth.

Sometimes, the lymphoma starts at other sites, for example the bowel or skin. Rarely, it is found in the bone marrow (the spongy tissue in the centre of
your bones where blood cells are made). What symptoms you have depends on where the lymphoma is growing, for example if it is in your skin, you might have a rash.

‘B symptoms’ (fevers, night sweats and weight loss) are common in people with extranodal NK/T-cell lymphoma. These symptoms often appear together.

Treatment

T-cell lymphomas are rare, so it is difficult to determine which treatment gives the best outcome. You might be asked if you would like to take part in a clinical trial to help test new and current treatments.

Most people with extranodal NK/T-cell lymphoma have radiotherapy to the areas affected by lymphoma, particularly if the lymphoma is localised to the area around the nose. If your lymphoma is widespread (stage 3 or 4), you are likely to have chemotherapy alone.

Chemotherapy is used together with radiotherapy to reduce the risk of relapse (lymphoma coming back). You usually have your course of chemotherapy either at the same time as you are having radiotherapy or after your radiotherapy.

Chemotherapy regimens (combinations of drugs) that include the drug L-asparaginase are usually recommended, for example the SMILE regimen, which includes the drugs:

- dexamethasone (a steroid)
- methotrexate
- ifosfamide
- L-asparaginase
- etoposide.

Depending on your health and the extent of your lymphoma, your doctor might suggest a different treatment, for example:
Chemotherapy regimen containing the drug gemcitabine, possibly with L-asparaginase

a targeted drug, usually through a clinical trial

the CHOP chemotherapy regimen.

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.

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 extranodal NK/T-cell lymphoma

It is common for extranodal NK/T-cell lymphoma to relapse (come back) after being in remission. Sometimes the lymphoma is refractory (doesn’t respond) to treatment. Should this be the case for you, your doctor might consider:

- a different chemotherapy regimen
- 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

There are several targeted drugs that can be effective in treating extranodal NK/T cell lymphoma, either on their own or together with chemotherapy.
These are usually only available if you are taking part in a clinical trial because they are still being investigated. You can find out more about clinical trials and search for a trial that might be suitable for you at Lymphoma TrialsLink.

Drugs being tested in extranodal NK/T cell lymphoma, but not yet approved in the UK, include:

- **combined treatments** that use antibodies to deliver chemotherapy to lymphoma cells, for example brentuximab vedotin (Adcetris®).
- **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 carfilzomib. 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. You are usually seen every 1–3 months at first, then every 6 months after a year or so. Your follow-up usually stops after around 3 years if you remain in remission.
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
- Stem cell transplants
- Glossary

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