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Angioimmunoblastic T-cell lymphoma (AITL) and follicular T-cell lymphoma (FTCL)

This information is about angioimmunoblastic T-cell lymphoma (AITL) and follicular T-cell lymphoma (FTCL). These are two types of T-cell lymphoma that develop from the same type of white blood cell, and are treated in the same way.

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 are AITL and FTCL?

Angioimmunoblastic T-cell lymphoma (AITL) and follicular T-cell lymphoma (FTCL) are types of **T-cell lymphoma** – **non-Hodgkin lymphomas** that develop from white blood cells called **T cells**. Many T-cell lymphomas have complicated names based on the type of cell they develop from or particular proteins they make.

AITL and FTCL are officially classed as **slow-growing (low-grade)** lymphomas but they often behave like **fast-growing (high-grade)** types. They develop from a particular type of white blood cell called a 'follicular helper T cell'. They are sometimes grouped together under the name 'nodal T-cell lymphomas with T-follicular helper (TFH) phenotype'.

Although they develop from the same type of cell, **lymph node biopsies** from people with AITL and FTCL look different under a microscope. In AITL, affected **lymph nodes** contain abnormal immune cells and lots of new, small blood vessels. In FTCL, abnormal immune cells are mainly found in parts of the lymph nodes called the 'follicles'.

About 1 in 3 people with AITL or FTCL have abnormal **B cells** as well as abnormal T cells. This can make it difficult to diagnose AITL and FTCL accurately.

Who gets AITL or FTCL?

AITL and FTCL account for about 1 in every 5 cases of T-cell lymphoma. Only around 140 people are diagnosed with AITL or FTCL in the UK each year. These types of lymphoma usually affect older people, typically around the age of 70. They affect men and women equally.

Scientists don't know what causes AITL or FTCL. These types of T-cell lymphoma have been linked to infections including viruses called Epstein-Barr virus (EBV) and human herpesvirus 6 (HHV-6). However, the potential relationship between having these infections and AITL or FTCL remains unclear and further research is ongoing.

Symptoms of AITL and FTCL

Most people with AITL present to their doctor with **advanced stage** lymphoma (stage 3 or 4) . AITL often causes 'systemic' symptoms that affect your whole body. FTCL is usually less widespread when it is diagnosed and the most common symptoms of AITL and FTCL are:

- fever, night sweats and unexplained weight loss ('B symptoms')
- **swollen lymph nodes**, usually in several places
- **a swollen liver and spleen**, which might make you feel bloated or cause tummy (abdominal) pain
- **itching**
- **skin rash**.

I developed, what felt like, a bad throat infection. The lymph nodes in my neck became very enlarged, and I thought I must have picked up a strange infection on a business trip.
Owen, affected by angioimmunoblastic T-cell lymphoma

AITL and FTCL often affect your **bone marrow** (the spongy tissue in the centre of your bones where blood cells are made), which can lead to low levels of red blood cells (**anaemia**) and platelets (**thrombocytopenia**). Occasionally there might be a build-up of fluid around the lungs (pleural effusion), which can cause breathing difficulties.

The lymphoma can also affect how well your **immune system** works so you might find it **harder than usual to fight off infections**.

In some people with AITL and FTCL, abnormal immune cells might produce too many antibodies (also known as 'immunoglobulins'). Antibodies are proteins that B cells usually make to help you fight off infection. If you have AITL or FTCL, you might make abnormal antibodies that react against healthy cells. This is called an 'autoimmune reaction'. It can cause a variety of symptoms, including:

- a low red blood cell count (**autoimmune haemolytic anaemia**), which might make you feel tired or short of breath
- low platelet levels (**immune thrombocytopenia**), which might make you bleed or bruise more easily than normal
- painful, swollen joints
- inflammation of your blood vessels
- thyroid problems.

Treatment of AITL and FTCL

Treatment of AITL or FCTL varies from simple treatment with **steroids** to combination chemotherapy.

The most common chemotherapy **regimen** used for AITL and FTCL is CHOP, which consists of:

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

I had six 3-week cycles of CHOP, which followed a pattern. In week 1, I had low appetite and felt nauseous. In week 2, I felt a bit better but my back and hips ached. In week 3, I started to feel normal-ish. By the end of the second cycle of CHOP I could see a bit of improvement. My lymph node swelling was reducing, and I was tolerating the chemo.
Owen, affected by angioimmunoblastic T-cell lymphoma

Some lymphoma specialists add a chemotherapy drug called **e**toposide to CHOP (CHEOP), or you might have a different chemotherapy regimen altogether.

Several **targeted drugs** are being tested 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 help test new treatments and to find out what the best treatment is for AITL and FTCL.

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**.

Some people with AITL or FTCL have a high number of abnormal B cells in their body. If this is the case, you might have **antibody treatment** such as **rituximab** as well as chemotherapy to diminish the abnormal B cells.

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).

If you are not fit enough to have 'standard' CHOP-like chemotherapy, you might have less intensive treatment, such as gemcitabine, bendamustine or cyclosporine.

Relapsed or refractory AITL and FTCL

It is common for AITL and FTCL to come back (**relapse**) after being in remission. Sometimes, AITL and FTCL don't respond to treatment (refractory lymphoma). In these cases, your doctor might consider:

- a different more intensive **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 chemotherapy and you are well enough
- a **targeted drug**, usually through a clinical trial.

Research and targeted treatments

Many new treatments are being tested to see if they can help people with T-cell lymphoma. Drugs that have been tested or are currently being tested in AITL and FTCL include:

- azacitidine with CHOP chemotherapy
- **antibody–drug conjugates** such as **brentuximab vedotin**
- **immune checkpoint inhibitors** such as avelumab
- **proteasome inhibitors** such as bortezomib or carfilzomib
- **HDAC inhibitors** such as vorinostat, romidepsin or belinostat
- a chemotherapy drug called **pralatrexate**.

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.

Acknowledgements

- With thanks to Professor Tim Illidge, Professor of Targeted Therapy and Oncology and Honorary Consultant Oncologist, University of Manchester, the Christie NHS Foundation Trust, for reviewing this information.
- We would like to thank the members of our Reader Panel who gave their time to review this information.

Content last reviewed: August 2022
Next planned review: August 2025
LYMweb0210AITL2022v3

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