

T-cell Lymphoblastic Lymphoma

What is T-cell Lymphoma?

Lymphoma is a cancer of the lymph glands or tissue and is broadly divided into Hodgkin's or non-Hodgkin's lymphoma (NHL). T-cell lymphoblastic lymphoma (T-LBL) is a NHL that is usually found in children and adolescents. It grows quickly and presents as a mass in the chest, and patients may have bone marrow involvement and circulating T-lymphoblasts and are treated as T-cell ALL. It is estimated that over 100 cases of childhood NHL are diagnosed per year in Gujarat, and around 15-20 of those are T-LBL.

What is the cause of T-cell lymphoma?

We don't know what causes T-LBL but it is not inherited, nor can it spread to other members in the family. There is nothing you did to cause this.

How are patients diagnosed?

Because there is often a mass in the chest compressing the airway, it is important to make an accurate diagnosis quickly, and treat aggressively with life-saving chemotherapy to achieve the best outcome. Steroids and low dose radiation may be needed to shrink the chest mass. Biopsy of a piece of the tumor is essential to diagnosing T-LBL and can be difficult because of the location of the tumor, needing CT guidance. Sometimes fluid from the chest can provide the cancer cells needed, or bone marrow sample.

A careful history and physical exam, in combination with review of the tumor slides by an experienced pathologist, with special stains, and characteristic changes in blood chemistry LDH, uric acid, potassium and phosphate usually allows a diagnosis to be made quickly. T-LBL lymphoma cells resemble T-lymphoblasts with surface markers for T-cells such as CD3, CD4 and CD8. Cytogenetics do not play much of a role in risk stratification.

How are patients staged?

Staging tells us how far the lymphoma has spread, and how aggressive treatment needs to be. Tests include total-body imaging and tissue sampling such as:

- CT, and/or MRI scans
- PET scans (although this does not change treatment in children so use is uncertain)
- Bone marrow biopsy and aspiration to distinguish T-LBL from acute leukemia
- Lumbar puncture to look for evidence of brain and spinal cord (CNS) involvement, which is rare. and but Bone Marrow (BM) involvement is common.

What urgent treatment is needed in newly diagnosed patients?

- The presence of a large mass in the chest presses on the airway and blood vessels and causes superior vena cava syndrome, shortness of breath and even respiratory arrest. Pleural or pericardial effusions may be present. It is important to keep the patient sitting upright, but he/she may need oxygen or may even need to be put on a ventilator.
- Tumor lysis results from rapid breakdown of cancer cells, causing blood abnormalities such as high potassium, uric acid and phosphate. This can cause kidney failure or heart rhythm problems. High IV fluid volumes to flush out these substances, and medications like allopurinol or rasburicase reduce the risk of kidney failure.

What is standard treatment?

All patients with T-lymphoblastic lymphoma will need chemotherapy and standard protocols are available from both the American (COG-A5971) and European (BFM-NHL-95) groups. Patients with low-stage (stage I or stage II) lymphoblastic lymphoma respond to a protocol treating Acute Lymphoblastic Leukemia approach and induction, consolidation, and maintenance therapy for a total of 24 months, with long-term survival rates of around 90%. Patients with high-stage (stage III or stage IV) lymphoblastic lymphoma have long-term survival rates of around 80%. Mediastinal radiation is not necessary for patients with chest masses, except in the emergency treatment of some patients with blood vessel or airway obstruction where steroid treatment is not providing rapid relief.

Multiple chemotherapy drugs are given, mostly into the vein (intravenous), although some are taken by mouth and others may need to be put directly into the spinal fluid by performing a lumbar puncture. The drugs include prednisone, dexamethasone, vincristine, daunorubicin, doxorubicin, L-asparaginase, cyclophosphamide, cytarabine, methotrexate, 6-mercaptopurine, 6-thioguanine, but radiation is omitted unless patients have lymphoma in the brain or spinal fluid. Your doctor will provide you side-effect sheets for the chemotherapy drugs given.

What are long term outcomes?

As mentioned above despite its initial scary presentation, outcomes with the right treatment are surprisingly good with 80-90% patients having a long-term cure. Discuss with your doctor any short and long-term side effects from treatment you may have concerns about.