

**What You Need to Know About Anaplastic Large Cell Lymphoma**

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Q & A With Andrei Shustov, M.D.

By Jo Cavallo

Anaplastic large cell lymphoma (ALCL) is a rare aggressive T-cell lymphoma comprising only about 3 percent of all lymphomas in adults and between 10 percent and 30 percent of all lymphomas in children and can present in either the systemic or cutaneous form. We talked with Andrei Shustov, M.D., assistant professor of medicine, Division of Hematology at the University of Washington in Seattle, about what you need to know about ALCL and what's on the horizon for new treatment.

**LRF:** What is anaplastic large cell lymphoma?

**Dr. Shustov:** Anaplastic large cell lymphoma (ALCL) is a rare type of aggressive T-cell lymphoma that is characterized by a unique appearance under the microscope. These are large cells that are different in size and shape and what really defines this disease is a uniform expression of a special marker on the lymphoma cells called CD30. Anaplastic large cell lymphoma belongs to the T-cell lymphoma group meaning that it arises from the T-cell type lymphocytes and is considered to be an aggressive lymphoma, so treatment is required upon diagnosis. Without treatment the disease may progress quickly.

Recently, the discovery was made that ALCL is actually a combination of two different diseases and even though they look the same under the microscope, what separates the two groups is the expression of another protein called anaplastic lymphoma kinase (ALK). That discovery has made a big difference in our understanding of the disease because the prognosis and outcome in patients who express this protein, ALK positive, and in those who don't express this protein, ALK negative, is strikingly different. Besides prognosis and outcomes, there are specific clinical features that characterize each type. So biologically, these are two different disorders even though both of them are grouped together into the anaplastic large cell lymphoma classification.

**LRF:** So the patient has to be certain he's getting the right diagnosis?

**Dr. Shustov:** That's correct.

**LRF:** How common is the disease?

**Dr. Shustov:** It's actually quite uncommon. Only about 3 percent of all lymphomas in adults represent anaplastic lymphoma. In children it's a little bit more prevalent. Ten percent to 30 percent of childhood lymphomas are anaplastic lymphomas. And if we talk about the two different subgroups, the ALK positive and the ALK negative disease, about 60 percent of patients with ALCL will have the disease that expresses the ALK protein.

**LRF:** What are the symptoms?

**Dr. Shustov:** Generally, ALCL presents with symptoms typical for aggressive lymphomas, or what we call B symptoms or constitutional symptoms, that include weight loss, significant sweats at nighttime and unexplained fevers. Patients may notice enlargement of the lymph glands in different parts of the body, most commonly in the neck or armpits.

**LRF:** What treatments are available for this disease?

**Dr. Shustov:** The outcome for ALK positive disease and ALK negative disease is quite different. However, despite that difference they both are approached in the standard fashion and still the standard of care is CHOP chemotherapy. The majority of lymphomas are treated with this combination upfront with more or less different results. Unfortunately for T-cell lymphomas, we do not have the supplemental biologic therapies, such as Rituxan, that are now the standard of care for the B-cell lymphomas, so CHOP alone given on a 21-day basis for six to eight cycles, is still the standard of care.

**LRF:** What happens when patients relapse?

**Dr. Shustov:** Treatment after relapse is certainly a challenge. The standard of care in that situation would be to give patients rescue chemotherapy, which is typically more aggressive than frontline treatment. After the lymph nodes shrink again or the disease gets less bulky, we typically offer patients an autologous stem cell transplantation procedure and can salvage probably about 50 percent of relapsed patients.

**LRF:** Is there new research in clinical studies for this subtype of non-Hodgkin lymphoma?

**Dr. Shustov:** Let me go back to the prognostic definition. We have learned that the ALK positive disease is actually quite good as far as outcome is concerned and up to 80 percent of patients will have a very long disease-free survival with frontline CHOP therapy. Unfortunately, the outcome for patients with ALK negative disease is not as good with about 60 percent to 70 percent of patients relapsing within five years. We're quite satisfied with the results of treatment for ALK positive disease but need new strategies for ALK negative patients. There are a couple of new agents that are being developed for T-cell lymphomas in general and anaplastic lymphoma in particular. What is being tested in clinical trials at this point is a new monoclonal antibody called SGN30. This antibody targets the molecule on the surface of the anaplastic cells called CD30. The result of a recent trial has shown a very promising response rate of about 60 percent of patients who have relapsed after initial treatment. However, we have to be careful interpreting these results because the study was small with only 17 patients who could be evaluated. But it certainly showed activity and my prediction is that SGN30 will probably be used in the treatment of anaplastic lymphoma in combination with chemotherapy in the near future in both newly diagnosed

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disease and relapsed disease.

**LRF:** What advice do you have for ALCL patients and survivors?

**Dr. Shustov:** For patients with ALK positive disease who achieve a complete response or complete remission after therapy, the prognosis is quite good and the chances are very high that they will not have to deal with the disease again. For patients with ALK negative disease, I would encourage them to follow-up very closely with their oncologist or hematologist so they can seek therapy as soon as possible if they relapse. I would also like to encourage both groups that new treatments are one their way and antibody therapy and some other agents are being evaluated that will probably enter late-phase clinical trials in the next couple of years.

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