



Forum

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Fall 2007

From the Executive Director 2

Stress Management 3

Cutaneous Lymphoma Patient Educational Opportunities..... 4

Fundraising for the Cutaneous Lymphoma Foundation..... 5

Cutaneous B-Cell Lymphoma Overview..... 6

Research Discovers CTCL Incidence Rising in US

The Cutaneous Lymphoma Foundation has funded an important epidemiology study that shows that the number of new cases of CTCL has doubled. In this study, published in the July 2007 issue of the *Archives of Dermatology*, Drs. Vincent Criscione and Martin Weinstock of Brown University report that there are approximately 1,600 new cases of CTCL diagnosed each year in the United States. This represents over double the number of new cases in the past fifteen years.

In their study, Criscione and Weinstock describe incidence trends (or the rate at which this disease occurs) for CTCL in the United States from 1973 to 2002. They determined that the overall annual incidence of CTCL was 6.4 per million, a total of 0.14 percent of all cancers, and 3.9 percent of non-Hodgkins lymphomas. This represents a more

than doubling of the incidence since the last study that was conducted in 1992. Cases of CTCL was higher among blacks and men. Racial differences decreased with age and have not changed over time. Gender differences in incidence increase with age and have decreased over time. Geographic variation in incidence was found and correlated with high physician density, high family income, high percentage of the population with a bachelor's degree, and high home values.

The exact cause for the observed

increase in CTCL incidence is unknown and cannot be determined from this epidemiology study. However, the findings do provide an important framework for investigators to design new clinical and experimental studies. It also draws much needed attention to CTCL and may contribute to greater resources for research.

The study did not address other important epidemiologic factors of CTCL such as death rate, prevalence (number of patients with CTCL) and factors influencing survival. Case-control studies of the 1990s have demonstrated that a complete response in patients diagnosed with Stage IA CTCL (less than 10% body surface area involvement)

...there are approximately 1,600 new cases of CTCL diagnosed each year...

correlates with a lack of disease progression and a normal life span. Recent combinations of biologic therapies appear to have prolonged survival

in advanced stages of CTCL. Future studies are needed to determine death rate, prevalence and the impact of treatment on CTCL survival.

Because this study was funded by the Cutaneous Lymphoma Foundation, it represents tangible evidence of the Foundation's capacity to partner with physicians and scientists to advance the agenda of CTCL. The Cutaneous Lymphoma Foundation looks forward to continuing its relationship with investigators and funding meritorious research in the future.

Forum

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Cutaneous Lymphoma Foundation is not responsible for the medical care or treatment of any individual.

From the Executive Director



I am so pleased that the Cutaneous Lymphoma Foundation was able to fund the epidemiology study by Drs. Vincent Criscione and Martin Weinstock of Brown University. It has made a large impact in the cutaneous lymphoma community. We knew that lymphoma in general had increased, but had not been able to confirm that this was true of CTCL

until now. We will continue supporting projects to gather more information about our disease.

When we first started the Mycosis Fungoides Foundation, almost 10 years ago, we didn't realize that there was no organization specific to the other cutaneous lymphomas. We have put together a lot of information about CTCL, considering this is a rare disease. It has been more difficult to gather information about CBCL, which is even less common. The article in this issue by Dr. Steven M. Horwitz is a start, and we will compile additional resources in the future.

In this issue of *Forum*, Leora Lowenthal writes about stress. As I was reading it, I wondered if she was writing about me. I have tried tapes, books and lists and found that it took too much time and was stressing me out! Now I just take one day at a time and don't spend a lot of time worrying about what may happen tomorrow.

When I look ahead at our schedule for this fall, I am amazed that we were able to put together six educational events along with all the other medical meetings we will be attending. We have two scheduled for the same day! We are able to accomplish this because we have a couple of staff people, Amanda and Holly, who are very efficient and competent in their jobs. We also have a working Board of Directors who volunteer their time to make sure we are on track to "make sure each person with cutaneous lymphoma gets the best care possible." We are fortunate to receive grants from companies to help cover the expenses involved in bringing this opportunity to patients to interact with specialists and learn more about their disease. We are planning our 2008 schedule to include more geographical locations to make it easier for people to attend. I hope you all will get to attend one of our educational forums in the near future.

By the time this newsletter goes to press, a friend of mine will have said goodbye to her 19 year old son for the last time. He has a rare pediatric disease that is always fatal. They have no hope...everything that could be done has been done. It is a stark contrast to what is being done in cutaneous lymphomas. In the past few years, we have had better diagnoses and treatment and fewer patients are dying from our disease. There is a tremendous amount of research out there and more than ever before, there is hope for the future.



Stress Management



Leora Lowenthal sits on the Cutaneous Lymphoma Foundation Board of Directors and is a Senior Social Worker at the New York

University Cancer Institute.

This past June I was invited to speak at an educational forum which was being presented by the Lymphoma Research Foundation and the Cutaneous Lymphoma Foundation. The topic I was asked to address was “managing stress” which initially seemed like a very straightforward matter. Most oncology social workers learn about this in their training both for the benefit of their clients and themselves; it’s considered an essential “trick of the trade.” So, I confidently set about working on my slides and started trying to think of personal anecdotes and references I could use to demonstrate the fine art of stress management. That’s when I first noticed some patterns in my history.

I found myself remembering the “relaxation CD” I purchased which featured the sound of breaking waves. Unfortunately I discovered that the sound of waves crashing through my apartment caused me to feel quite jumpy. Then there was the time I took a yoga class but was so chronically overtired that I would fall asleep as soon as we began floor exercises. I even kept a journal for a while but was consumed by paranoid fears that someone would find it and read it. You get the picture; I was a study in failed attempts at stress management

I also contemplated the many pieces of advice that I offer almost daily which I don’t seem able to apply in my own

life. I am a great believer in the value of making lists and schedules in order to avoid the stress of being forgetful.

Yet in fact

I almost always forget to make lists and when I do I tend to misplace them before

any items have been crossed off. I also advise people that it is important to observe and respect the natural limits of our bodies. If you’re tired, I say take a nap or go to bed early! I usually say this as I am gulping down a caffeinated beverage and silently cursing my own foolish tendency to stay up well past anyone’s bedtime. In truth, throughout the 1990s my favorite form of stress management was chain-smoking far into the night (ironically, quitting smoking was extremely stressful.)

Needless to say after a few hours of musing I was convinced that I should never speak on stress management again. I imagined hordes of people leaving the workshop armed with new, disastrous techniques sure to guarantee a life full of stress and exhaustion. There was only one thing which suggested a reason for hope and it was this: in general, despite being a slightly neurotic, insomniac workaholic, I am a pretty happy person with well-managed stress levels.

So, I won’t bother to describe what does actually work for me here but I give a lot of the credit to my beloved parents, my incredible sister with her equally incredible husband and children, my devoted friends and my brilliant, compassionate colleagues. And, in case you’re wondering, I went on to present at the workshop and thankfully no one has called yet to say that I ruined their life with my wretched advice. I did offer

up a lot of the usual suspects in stress management but I added the suggestion that everyone do a little search through

their own memory archives. Look at your past efforts to manage stress and consider

what hasn’t or hasn’t worked for you. Know your own strengths and exploit them. Never be afraid to ask for help or to take suggestions; people in the audience offered up some great ideas that I had never considered. And, most important of all, never let lymphoma become the most important thing in your life. You don’t have to like lymphoma but you can like, maybe even love, your life with it. Wishing you all a very lovely, stress-free autumn.

Lymphoma Awareness



Worldwide network of lymphoma groups

World Lymphoma Awareness Day is held on September 15 every year and is a day dedicated to raising awareness of lymphoma. WLAD is a global initiative

hosted by the Lymphoma Coalition, a non-profit network organization of patient groups from around the world. For further information, visit www.lymphomacoalition.org.

15 SEPTEMBER
WORLD
LYMPHOMA
AWARENESS
DAY

Cutaneous Lymphoma Patient Educational Opportunities



September 15, 2007
San Francisco, CA

Lymphoma Workshop, presented with the Lymphoma Research Foundation

Speakers: Youn Kim, MD, Stanford University, additional speakers TBA

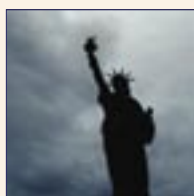


September 22, 2007
Columbus, OH

Cutaneous Lymphoma Patient Educational forum

Speakers: Pierluigi Porcu, M.D, Ohio State University, Christiane Querfeld, Northwestern University, John Zic,

Vanderbilt University



October 12 – 14, 2007
Brooklyn, NY

North American Educational Forum on Lymphoma presented by the Lymphoma Research Foundation

Speakers: David Straus, MD, Memorial Sloan Kettering Cancer Center, David

Ramsay, MD, New York University School of Medicine, Kenneth Hymes, MD New York University School of Medicine, JoAnn Latkowski, MD, New York University School of Medicine, Leora Lowenthal, LCSW, OSW-C, New York University Cancer Institute



October 20, 2007
Pittsburgh, PA

10th Annual Jegasothy Support Group for CTCL

Speakers include: Larisa Geskin, MD, University of Pittsburgh Medical Center, Sue McCann, MSN, RN, University of

Pittsburgh Medical Center, additional speakers TBA



October 20, 2007
Calgary, AB

Living with Lymphoma: The Journey Forward, presented with the Lymphoma Foundation Canada

Speaker: Richard Haber, MD



November 3, 2007
Minneapolis, MN

Lymphoma Workshop, presented with the Lymphoma Research Foundation

Speakers: TBA

Webinar: Focus on Topical Therapies

Date and Time:

Monday, September 17, 2007, 2pm ET • 11am PT

Program Chair:

Stuart Lessin, MD, Fox Chase Cancer Center

Registration Instructions:

This program is free-of-charge; however, pre-registration is required. Visit www.clfoundation.org/THwebinar

What is a webinar?

A webinar is an interactive online seminar. All you need is a telephone and an internet connection. Once you have registered, you will be provided with instructions for how to log on to and participate in the webinar.

What will happen during this webinar?

This webinar will provide an overview of topical therapies that are used to treat cutaneous lymphoma. Both patients and caregivers are welcome to attend this free event. Dr. Stuart Lessin will speak about the treatments. This will be followed up with a question and answer session and include a brief presentation by the Cutaneous Lymphoma Foundation.

For more information about these events, please visit our website, www.clfoundation.org, or email us at education@clfoundation.org, or call (248)644-9014

Patient Educational Forums provide exceptional opportunities for people with cutaneous lymphoma to receive accurate information about treatment options, access experts in the field and connect with other people with similar experiences.

Cutaneous Lymphoma Foundation Patient Education Forums key features:

- Half-day to day-long event
- Held in cities throughout North America
- Professional and lay speakers
- Format includes lectures, Q&A sessions and small-group discussions
- Objectives:
 - Develop a better understanding of diagnostic tests.
 - Learn about treatments available for different stages.
 - Identify resources for treatment and support.

These educational opportunities are made possible by generous unrestricted educational grants from:

Platinum Sponsors: Eisai Inc. and Ovation **Gold Sponsors:** Therakos and Merck Oncology **Silver Sponsor:** Gloucester

Fundraising for the Cutaneous Lymphoma Foundation



Christopher Shipp is Vice President for the Cutaneous Lymphoma Foundation and a Senior Consultant at a leading health care marketing research company

You may recall hearing about Scott, a young man living with CTCL who decided to run a half marathon this summer in order to increase awareness and to raise money for the Cutaneous Lymphoma Foundation. Scott completed the half marathon and raised over \$22,000 for the Foundation.

These funds will serve to educate and support patients, provide continuing education for

physicians and nurses, and fund ongoing clinical research. Because the Cutaneous Lymphoma Foundation is a small non profit organization and because we depend on your donations to continue to serve people with CTCL, donations and fund raising are our life blood and contributions such as Scott's are vital.

As a CTCL patient, I was inspired by Scott's effort, and as a Cutaneous Lymphoma Foundation board member, I was awed by the amount of money he was able to raise for the Foundation. After a few email and telephone conversations with Scott and his family, I came to the conclusion that Scott's accomplishment could be replicated by other CTCL patients and/or their loved ones if they so desired.

To test my theory, I decided to try it out for myself. My first task was to choose an event, so I signed up for a

triathlon (I have not done one of these since being diagnosed in 2004, so wish me luck). The second step was to go to www.firstgiving.com. This is a website that works in conjunction with nonprofit organizations to accept donations on behalf of people participating in some type of fundraising event. It is easy, it costs you nothing, and all you have to do is follow the simple step-by-step instructions and Firstgiving will host your personal fundraising page that allows you to accept donations on behalf of the Cutaneous Lymphoma Foundation without having to handle any of the money personally. Once your page is setup, the next step is to

email the link to anyone you think would be willing to donate. It is that simple. You can check out my page as an example at www.firstgiving.com/cutaneouslymphomafoundation. It is fun to monitor my progress with family and friends and it is inspiring to see how generous people truly are.

I realize not everyone is able or inclined to compete in athletic events to raise money for the Cutaneous Lymphoma Foundation. If this is the case, and you still want to help the Foundation by fundraising on your own, you are only limited by your initiative and creativity. Examples of fundraising activities done on behalf of other non profit organizations include; bake sales, car washes, wine tastings, silent auctions, garage sales, raffles, etc., where the proceeds go directly to the non profit organization.

A single person can make a difference. Scott has proven that. I hope the ideas and information provided in this article, will give others the tools they



Scott single-handedly raised over \$20,000 for the Cutaneous Lymphoma Foundation running a half marathon in New York this summer.

need to follow in his footsteps. It is the Foundation's goal to continually enhance and expand the services we provide to patients and the only way we can do that is through your support.

Cutaneous B-Cell Lymphoma Overview



Steven M Horwitz, M.D., is an oncologist at Memorial Sloan Kettering Cancer Center in New York City

Among all the types of non-Hodgkin's lymphomas, B-cell lymphomas are much more common than T-cell lymphomas. However, in the skin, it is the B-cell lymphomas that are more unusual. B-cell lymphomas which are completely confined to the skin are called primary cutaneous B-cell lymphomas or CBCL. It is important to note that systemic or nodal B-cell lymphomas can secondarily involve the skin and these would not be called primary cutaneous or CBCL. Under the microscope, CBCL look like the more common nodal B-cell lymphomas but often act differently. In the past these differences were not always well recognized. Recently, a consensus classification system between the World Health Organization (WHO) and European Organization for the Treatment and Research of Cancer (EORTC) systems was published. This new system lists three primary types of CBCL: primary cutaneous marginal zone B-cell lymphoma (CMZL), primary cutaneous follicle center lymphoma (CFCL), and primary cutaneous diffuse large cell lymphoma, leg type (LBCB-L).

Indolent Primary Cutaneous B-cell Lymphomas

CMZL and CFCL are generally very slow growing and are thus considered indolent. They respond well to treatment, often mild treatments

when it is needed, but often return or relapse. These relapses of the indolent CBCL are almost always confined to the skin only and are rarely life-threatening. Because these lymphomas usually grow slowly and often return, even after effective therapy, sometimes no treatment is needed.

Primary Cutaneous Follicle Center Lymphoma

Primary Cutaneous Follicle Center Lymphoma (CFCL) is the most common B-cell lymphoma to occur as a primary tumor of the skin. CFCL most commonly presents on the head and neck or trunk with the legs rarely involved. Lesions appear as pink or red nodules or tumors that develop slowly over months to years. They rarely develop open sores or ulcers. Some patients may have nodules in many places, more often a single tumor or small group of papules or nodules is seen.

Marginal Zone Lymphoma

Primary cutaneous marginal zone lymphoma (CMZL) is slow growing B-cell lymphoma that is similar to a type of non-Hodgkin's lymphoma called an extranodal lymphomas of Mucosa Associated Lymphoid Tissue (MALT) type. CMZL are the second most common form of CBCL. People with CMZL present with pink or red papules, nodules, and/or tumors. Ulcers are rare. CMZL can occur at anywhere on the skin but have a predilection for the extremities, particularly the arms, and the trunk. Single or multiple lesions may be present. Lesions may present acutely or may be present for years before diagnosis.

Aggressive Primary Cutaneous B-cell Lymphoma

Diffuse large cell lymphoma, leg type or LBCL-L is a rare and potentially more dangerous type of CBCL that differs from the indolent types by its appearance on the skin and under the microscope.

Primary Cutaneous Diffuse Large B-cell Lymphoma, Leg Type

LBCL-L was initially recognized as a more aggressive disease often showing up in the legs of elderly women, which is where its name comes from. We now can recognize this type of lymphoma on careful pathology review including some unique molecular features that distinguish it from the more common indolent subtypes. While LBCL-L most often shows up on the legs we must remember that this type can occur at any site of the skin and the indolent types can first show up on the legs so site alone is not enough to make the diagnosis. LBCL-L often develop into large tumors with extension deep into the fat. They tend to grow quickly, over weeks to months and frequently develop open sores. Unlike the indolent types, they have a high likelihood of spreading outside the skin.

Primary Cutaneous Diffuse Large B-Cell Lymphoma Other

Primary Cutaneous Diffuse Large B-Cell Lymphoma, Other, describes a group of rare diffuse large B-cell lymphomas that do not fit into any other categories. These lymphomas, while at times showing up in the skin are more often systemic lymphomas that have gone to the skin.

Diagnosis

Appropriate biopsy of skin involvement is critical for the diagnosis and classification of CBCL. Whenever possible, one should perform an incisional biopsy or large punch rather than a small punch or shave biopsy because more tissue is often needed for detailed pathology studies.

Staging evaluation

Once a diagnosis of B-cell lymphoma has been established from a skin biopsy, complete clinical staging is indicated to exclude disease outside the skin and define the process as a primary cutaneous lymphoma. This should include a physical examination with attention to the entire skin and lymphatic system. Laboratory evaluation should include complete blood counts and serum chemistries as well as measurement of serum lactate dehydrogenase (LDH). Imaging studies should include a CT scan of the chest, abdomen and pelvis. The neck should be imaged as well, for disease that presents in the head or neck region. PET scans are sometimes useful. A bone marrow biopsy is often but not always done.

Prognostic and predictive factors

The prognosis for people with CBCL is usually excellent. Recently, the Italian Study Group for Cutaneous Lymphomas reviewed 467 patients with CBCL. For people with CMZL and CFCL about 96% were alive at 5 years after their diagnosis and 90% were alive 10-years later. People with the more aggressive LBCL-L had a more difficult time with 73% alive 5 years after their diagnosis and 47% alive 10 years later. While the chance of the lymphoma becoming life threatening is higher in people with LBCL-L, one should keep

in mind that many people with this disease are diagnosed in their 80s or 90s and survival rates at 10 years includes people who have died from any cause.

Despite the excellent survival rates, relapses are common with almost 50% of people having the lymphoma come back after an initial complete response to treatment. However relapses are almost always confined to the skin, and this in general does not affect prognosis or survival. Less than 10% of people ever develop lymphoma outside the skin (the chance of this is somewhat higher for people with LBCL-L).

Management

Indolent Types: Cutaneous Follicle Center Cell and Cutaneous Marginal Zone Lymphoma

Radiation therapy works very well and is often preferred for people with localized disease. Some people with very small single lesions may be treated with excision only. Topical therapies are also used at times. While radiation therapy is a recommended approach for localized therapy, a preferred approach to systemic therapy is less clear. In general,

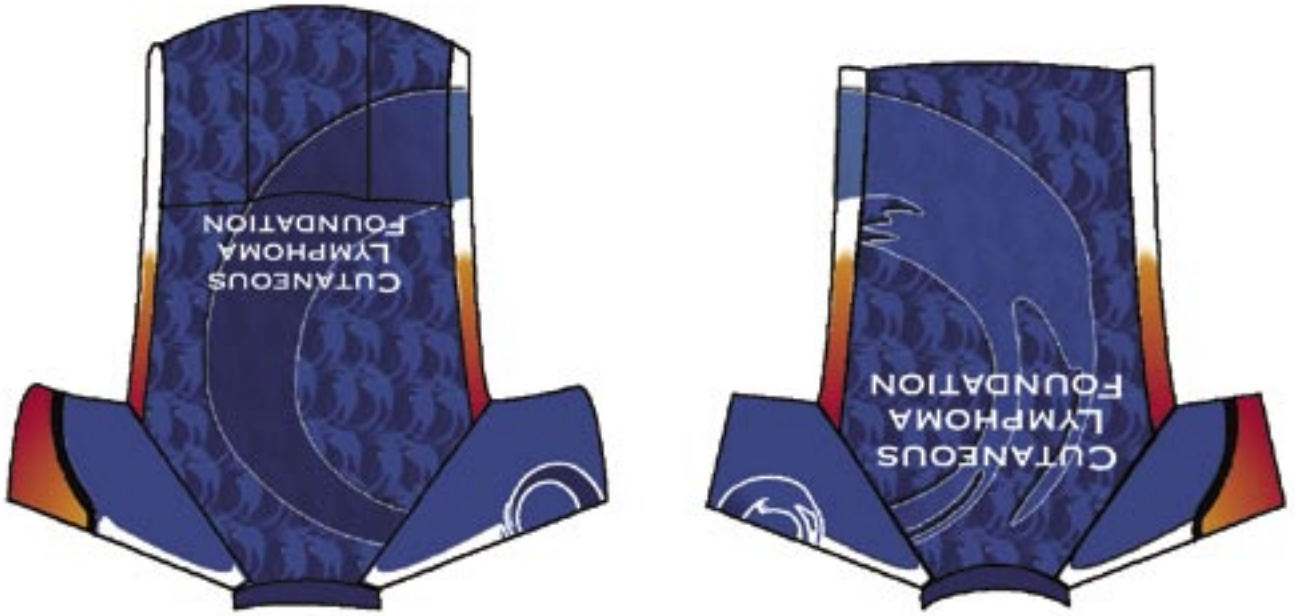
systemic chemotherapies of various types as well as immune therapies such as monoclonal antibodies and interferon have all been used successfully for people with multiple spots. It is important to note that many therapies can treat the lymphoma that is present leading to remission, however no therapy has been shown to be superior in preventing the common recurrences. Sometimes no treatment is needed and people are observed.

Aggressive Type: Diffuse large cell lymphoma-leg type

Given the more poor prognosis for people with CBCB-L, combinations of chemotherapy are often recommended. In people with a single tumor, radiation can be very effective. Often radiation and chemotherapy are given sequentially and can be successful and lead to remission. However, even with successful treatments, relapses are common and there is no known best treatment strategy.

Primary Cutaneous B-cell Lymphomas	
Behavior	Subtype
Indolent	Primary cutaneous marginal zone B-cell lymphoma
	Primary cutaneous follicle center lymphoma
Aggressive	Primary cutaneous diffuse large B-cell lymphoma, leg type
	Primary cutaneous diffuse large B-cell lymphoma, other

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