

Yale CANCER
CENTER

answers

WNPR Connecticut Public Radio



Hosts

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Learning About Sarcoma

Guest Expert:
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Yale Cancer Center Answers

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Welcome to Yale Cancer Center Answers with doctors Francine Foss and Lynn Wilson. I am Bruce Barber. Dr. Foss is a Professor of Medical Oncology and Dermatology, specializing in the treatment of lymphomas. Dr. Wilson is a Professor of Therapeutic Radiology and an expert in the use of radiation to treat lung cancers and cutaneous lymphomas. If you would like to join the conversation, you can contact the doctors directly. The address is canceranswers@yale.edu and the phone number is 1-888-234-4YCC. This evening, Francine and Lynn welcome Dr. Gary Friedlander. Dr. Friedlander is the Wayne O. Southwick Professor of Orthopedics at Yale School of Medicine and he joins us this evening for a conversation about sarcoma. Here is Francine Foss.

Foss Could you start us off by explaining to our audience what a sarcoma is?

Friedlander In general, cancer is referred to in two senses, one are the carcinomas. Those are the far more common variant which include origins in the epithelial or lining cells, or glandular cells. These include tumors arising in the breast, prostate, lung, or kidney. The sarcomas arise from the support structures. Some of them are in soft tissue; they may be fatty in origin or fibrous in origin, such as muscle. Others are hard tissue like bone and cartilage.

Wilson How common are sarcomas and are certain types more common than others?

Friedlander They are relatively uncommon. My life revolves around them so for me they seem to be relatively frequent, but in fact, they are only 1% of all of the cancers. Of the 1.5 million cancers that arise in the United States each year, only 1% of those are sarcomas. Some of them are a little bit more common than others, and some of them are more common in children and others more common in older ages.

Wilson What are some of the common types and are those different between children and adults?

Friedlander Yes, children's sarcomas are very frequently Ewing sarcomas or osteosarcomas, sarcomas that arise in bone. In middle age and later in life, we more commonly see cartilage related, or chondrosarcomas, some of the fibrous sarcomas, malignant fibrous histiocytomas and their variants are seen in middle and later years.

Foss Gary, you are an orthopedic oncologist, so you are basically an orthopedic surgeon specializing in oncology. Can you explain what your role is in the management of these sarcomas and how you interface with other members of the multidisciplinary team?

Friedlander It's a great question and very important. I began my career as an orthopedic surgeon. I did my training in orthopedic surgery, but I had an interest in tumors very early in my training and at some point, I did an additional fellowship in musculoskeletal oncology, which is a variant of surgical oncology related to the musculoskeletal system. But I could not possibly do what I do and certainly not as well as I am able, if it wasn't for the team approach, and I work every day with colleagues like yourselves, of course, from medical oncology to radiation oncology, the radiologist, the pathologist, the entire support team is absolutely critical to doing this correctly.

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- Foss Can you just tell us how patients present with sarcomas?
- Friedlander Sarcomas, like most other lumps and bumps are either perceived as pain or as something growing, as a lump. Many people notice the pain when they bump their lump and so they frequently will go to their primary care physician. Most people do not like to think they have a tumor. When they hurt themselves or they feel a lump or a bump, they generally relate this to some form of injury, but when those symptoms do not go away, do not respond the way a usual bruise, strain or sprain might, they often seek help from their primary care physician and those diligent individuals send them on to people like myself.
- Wilson What happens at that point Gary, what sort of evaluation do you do? What sort of testing? How is the diagnosis actually made or confirmed?
- Friedlander These people go through very much the same process as for any disease or disorder. They need to be heard. You need to listen to them and hear what it is they feel or perceive, what they are concerned about and some people are concerned about cancer and do not have any other symptoms and just need someone to listen and examine them and examine them carefully for lumps, bumps, changes that might signal something going on. That leads generally to some imaging studies, which are very helpful. They are remarkably good, and especially sensitive today. It might be a plain x-ray to begin with and depending on levels of suspicion, we move on to other studies such as CAT scans or MRIs to help define that there is in fact something going on. All tumors are not malignant, there are benign tumors and there are malignant tumors, but once the tumor is identified, image wise, and sometimes by feel, a biopsy often helps. The combination of the story they give, the physical finding they present with, and the imaging pictures that display some characteristics, you can often get very close to the diagnosis and understand what is going on, but often it requires, not always but often, a biopsy.
- Wilson For the biopsies of sarcomas, tell us a little bit about some of the techniques and the important considerations, because I understand that biopsying a sarcoma or a suspected sarcoma may require different types of biopsy or procedures than making the diagnosis of breast cancer, for example.
- Friedlander That's correct. The two options in biopsies are with a needle, some novocaine, and a special needle and that can be very very effective, especially in the hands of the well-trained radiologist that we have here and at most major medical centers. It retrieves a very small piece of tissue if it is placed in the correct orientation, so that it does not interfere with surgery that might be required, that small piece of tissue can often give us tremendous clues. We have pathologists who are used to looking at very small samples and coming to very accurate conclusions. We have done some studies on that. The other option is called an open biopsy and that is done generally in an operating room, either in a radiology suite or in an office. Open biopsies are done in the operating room with a small incision. Again, it has to be placed properly, that incision, so it does not interfere with the ability to take the tumor out with the least amount of impairment later on, but

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that gives us a much bigger amount of tissue to work with if the needle biopsy is insufficient or inappropriate. Then, again, it is a matter of looking under the microscope, and a well-trained pathologist familiar with unusual disorders and diseases with the benefit of multiple techniques generally can confirm the diagnosis.

Foss Gary, can you tell us how often, when a patient presents with a lump in a muscle or bone, how often is it cancer and how often is that benign? What are the benign things that could do that? That is a tough question, but I think an important question for people listening.

Friedlander That is extremely important and in fact, it is one of the highlights of my day when somebody comes in concerned about a bump and I can tell them, yes there is a lump, but this is harmless, this is benign, this is not going to do the things that you are concerned with and the majority of lumps are benign. The vast majority of lumps are benign. I could put a figure on it but I must admit I am guessing, at least three quarters of the lumps that I see turn out to be benign. They often need to be followed. They can weaken the bone, they can cause impairments, they can be frustrating and they can limit activities, but they are benign and ultimately their treatment is different that it is for high-grade malignant tumors, and these high-grade malignant tumors can also be handled much more successfully today than in years past.

Wilson Do these tumors spread to other locations of the body sometimes, if they are malignant?

Friedlander To me, one of the characteristics of a malignant tumor is its ability to spread, to go to other parts of the body, and the tumors that I see, the malignant sarcomas that I see, have a tendency to go to the lung and then to other organs within the body and to other bones, or bones in particular. Just to be clear, people talk about bone tumors and may be referring to one of two situations. One is what is termed a primary bone tumor, something that starts in that bone, osteosarcoma being the classic, the prototype. Another form of, if you will, bone cancer and far more common are tumors that came from other parts of the body, lungs, breasts, prostate, and came to bone secondarily or are metastatic lesions.

Foss Another point with respect to that is we have been talking a lot about lumps that we can kind of feel in the arms and legs and some of these soft tissue sarcomas can occur inside the body as well and it can be harder to pick these out.

Friedlander That is absolutely correct and people know their bodies, and often have a sense that something is not correct and if those feelings persist, I think they really should check with their primary care physician. Very often, a diligent evaluation will help them understand that it is nothing to be concerned about, but this is the way many cancers are found.

Foss You also talked a little bit about high-grade sarcomas, is there a grading system for some of these tumors? Can you tell us a little bit about that and what that means to you as a surgeon and to us as a medical oncologist?

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Friedlander Benign is on one end of the spectrum and what is preferred to as high grade, is on the other end of the aggressiveness spectrum with intermediate stops. In addition to benign, there is low grade, then intermediate grade, and high grade. The primary significance for me as a surgeon is how those lumps, how those tumors are best removed. When you want to remove a high-grade tumor, you want to be as certain as possible that you got every last cell out of this tumor and they do not always have very smooth well-defined margins, so you take out the tumor with an envelop of normal tissue. The more aggressive and high-grade, the thicker that envelop must be. As tumors get more towards the benign end of the spectrum, that envelop can be rather thin. So I need to know which end of the spectrum the tumor is on. It turns out that there are really only two approaches to taking these tumors out, one with the very thin envelop, one with a very thick. It turns out that benign and low grade can often be handled with a rather thin envelop and intermediate and high grade need to be treated more aggressively.

Wilson Briefly, what are some of the risk factors for sarcoma?

Friedlander It is not a brief answer. For example, there are some diseases like Paget's disease that older individuals get in the bone that predispose to cancer. There are some people that you and I see together who have required radiation for one very good reason or another, but that does predispose to tumors later on, things of that nature.

Wilson We are going to take a short break for a medical minute. Please stay tuned to learn more information about sarcoma with Dr. Gary Friedlander.

*Medical
Minute*

Breast cancer is the most common cancer in women. In Connecticut alone, approximately 3000 women will be diagnosed with breast cancer this year and nearly 200,000 nationwide, but there is new hope for these women. Earlier detection, noninvasive treatments, and novel therapies provide more options for patients to fight breast cancer. In 2010, more women are learning to live with this disease than ever before. Women should schedule a baseline mammogram beginning at age 40 or earlier if they have risk factors associated with the disease. With screening, early detection and a healthy lifestyle, breast cancer can be defeated. Clinical trials are currently underway at federally designated comprehensive cancer centers such as Yale Cancer Center to make innovative new treatments available to patients. A potential breakthrough in treating chemotherapy resistant breast cancer is now being studied at Yale combining BSI-101 a PARP inhibitor for a chemotherapy drug, irinotecan. This has been a medical minute brought to you as a public service by the Yale Cancer Center. More information is available at valecancercenter.org. You are listening to the WNPR Health Forum on the Connecticut Public Broadcasting Network.

Wilson Welcome back to Yale Cancer Center Answers. This is Dr. Lynn Wilson. I am joined by my co-host Dr. Francine Foss. Today, we are joined by Dr. Gary Friedlander and we are discussing sarcoma. Gary, we have actually covered a fair amount of detail on sarcoma already but let's shift

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over to treatment information and discuss with our listeners some of the different treatment options and how those are integrated into the treatment of a patient with a sarcoma.

Friedlander This is a very exciting area. I began my interest in tumors about 40 years ago, and our opportunities, our success rates are so much better today that it is truly very gratifying. The surgical component of treatment, in partnership with the many I have described and will come back to later, the surgical component first and foremost is to get rid of the tumor to remove the part that has the disease in the soft tissue or bone. Let me talk about bone for a moment. When I began, if a tumor was in a portion of a bone, the treatment recommended almost invariably was an amputation. Amputation is still very important and can be a very good operation for certain circumstances, and again, we can come back to that, but far more often with earlier detection and with the imaging studies that are available, it is possible to remove the diseased part of the skeleton and replace it with something very functional, what we call limb-sparing tumor resection.

Wilson Could you give us an example of the type of procedure that might fit into that category?

Friedlander For the moment let's envision a tumor that involves the bone just above the knee, the thigh bone is called the femur and towards the knee, may, for example, have a tumor in it. And I or my colleagues can remove that end of the thigh bone which joins together with the shin bone, or the tibia, to make the knee, and depending on the size of the tumor, there may be a gap created by that removal of several inches or a considerable number of inches and I have two major ways to repair that gap. One is to take a bone graft that was donated at the time of someone else's demise to a bone bank where it is properly screened and available and I can get another portion of the thigh bone that looks like the one I removed and put that in place and there is a process by which the body accepts it and heals it. It is a lengthy process, but it works quite well. The other option is to make up that difference with metals and plastics, very similar to the knee replacements, hip replacements, shoulder replacements, elbow replacements, and ankle replacements that we do for arthritic disease, but customized in a way that allows really excellent, not completely normal, but very excellent function, cemented it in place and it is able to be used within a relatively short period of time.

Foss Are those cadaveric bone grafts available everywhere or is that only available in a specialized center?

Friedlander Today, most of these cadaveric or bone donated parts are retrieved from centers around the United States, tissue banks that are specifically set up to acquire, process, and distribute these products and can be obtained almost anywhere. The people who are familiar with their implantation tend to be in academic medical centers like our Smilow Cancer Hospital.

Foss What about if the patient has a tumor in muscles, say a functional muscle like the biceps in the arm, do you have to remove that muscle? If you remove it, what can you do to improve function?

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Friedlander You have to go back to job #1, get rid of all the tumors regardless of what that means in terms of function. If you start to take the tumor out and spent too much time thinking about the function, you might cut corners that leave tumor. It does not mean you have to take things unnecessarily and that's where skill and experience comes in and it does not mean you have to take out the entire muscle. Fortunately, there is a great deal of redundancy in the body and often other muscles can help compensate for getting rid of a nasty problem. Under some circumstances, you can rearrange muscles, you can take a muscles that does one thing and reattach it in a way that it does something that is more important.

Wilson How has the prognosis changed, Gary, over the last decade for sarcoma? Obviously it has to do with treatment advances, surgical advances, multidisciplinary care, but discuss with our listeners how things have changed?

Friedlander When I first became interested, and I have already admitted it was about 40 years ago, the likelihood of surviving osteosarcoma, bone cancer, primary bone cancer, was about 10% at two years. Today, it is probably 70% to 80% of people that have osteosarcoma that are cured.

Wilson That is impressive.

Friedlander It is not all my fault. I would like to think I can contribute to that but people like Francine and my medical oncology colleagues, and my radiation therapy colleagues, have really made the difference. Multidrug chemotherapy has been absolutely spectacular and the imaging studies we have for early detection both of the primary tumor, of metastatic disease, enormous differences today.

Foss Are you alluding to the role of PET scan to detect metastatic disease here?

Friedlander PET scan is one of the very very superb ways for looking for metastatic disease, many of the sarcomas that I deal with metastasize first, very often, to the lung. CT scans have been around for a while, but they can be extremely helpful in surveillance after tumors have been removed. CAT scans and occasionally MRIs. We use MRIs very frequently here at the site of the original tumor to be certain that it is not recurring and the recurrence rates now are down in the 1% and 2% range, if that much.

Foss Can you talk a little bit about the role of radiation therapy and whether you would, in some instances, radiate before you do a surgical procedure?

Friedlander This is choosing between two correct options. My colleague Dieter Lindskog, trained at an institution for his tumor experience that preferred preoperative radiation. In my training in Boston, in that regard, tended to use radiation after the resection and there are some good reasons one might do it before or after, routinely, and there are some specific reasons why you should do it

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before or after, but that's why I will call Lynn and his colleagues and say, what do you think?

Wilson There is obviously a lot of variability depending on the location and the size and how it will affect your procedure, and it can be a complex decision which gets back again to the multidisciplinary aspect of things and how critical it is for all of us to work together with specialists taking care of these patients.

Friedlander We meet regularly for this particular purpose to make sure that we have that kind of communication. We get together as medical oncologist, radiation oncologist, surgical oncologist, pathologist, and radiologist, on a routine basis and go over cases even though we know what we are likely to say and what each other are likely to determine, but that keeps that face-to-face communication very much in our minds, and as you know, when you are clustered together in a superb environment like Smilow, it is just a matter of turning around and finding the right person or picking up the phone and clarifying some of these joint decisions that need to be made together.

Foss Do most of your patients end up seeing the medical oncologist prior to their surgery or does it usually happen after the surgical procedure?

Friedlander If at all possible, I very much prefer them to meet the other members of the team prior to surgery. There are times when the surgery must proceed rather quickly, but for the prototype sarcoma that I was referring to before, very often we consider preoperative chemotherapy. This is a little bit more difficult to explain to the patient because intuitively they come to a surgeon and want to cut it out, they want it removed. The first thing is they just want to get rid of it, they want it out, even though they are going to need chemotherapy. Well, it is a one-two punch, and whether you hit them first with your right or your left hand, can make a little bit of a difference because what is extremely important is to make sure the tumor does not spread, and the best way to do that is to start the chemotherapy. Surgery has no effect on spread other than taking away the primary, and when I see patients there may already be cells in the body and I am depending on you to make sure our chemotherapeutic agents hunt those down and kills those cells. We can also use other medicines that make it hard for those cells to implant and grow in other parts of the skeleton.

Foss So most patients with high-grade sarcomas will get chemotherapy either before or after?

Friedlander Most of those patients where we have good evidence, and we often do, the chemotherapy makes a difference, get that form of treatment, and often get at least part of it before surgery.

Foss Could we just touch on the issue of surgery in a patient who already has metastatic disease? If you see somebody, you make the diagnosis, and then you do your scans and you find out that there are already metastases, do you still try to approach the primary tumor by resecting it?

Friedlander This has to be individualized, but that brings up another very exciting opportunity and where metastatic disease used to be considered irretrievable, if you will, that's clearly not the case any

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more. For example, metastatic disease to the lung often can be cured by resection and it may require even more than one resection, but it is not a zero game at this point, probably at least 20% of people with metastasis to the lungs, if they are relatively few, can be turned around, and in conjunction with chemotherapy. The other thing tumors do to bones is weaken them and we are very sensitive to wanting people to enjoy their life opportunities, and we have to be thoughtful, sympathetic, compassionate, and know when to operate and when not to operate.

Wilson What sort of rehabilitation is associated with this? I know it is a difficult answer to give depending on what part of the body gets an operation, but I would suspect most of your patients need to go through a rehabilitation program, which could be fairly lengthy for them.

Friedlander They do. For the metallic implants, the rehabilitation gets started quicker and probably does not need to last quite as long. Many of these can be home exercises. For the bone grafts, the donated bone, they need a lot more protected time as biology heals.

Dr. Gary Friedlander is the Wayne O. Southwick Professor of Orthopedics at Yale School of Medicine. If you have questions or would like to share your comments, visit yalecancercenter.org where you can also subscribe to our podcast and find written transcripts of past programs. I am Bruce Barber and you are listening to the WNPR Health Forum on the Connecticut Public Broadcasting Network.