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Yale Cancer Center Answers

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Welcome to Yale Cancer Center Answers with Dr. Francine Foss and Dr. Lynn Wilson. 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 week, Francine and Lynn welcome Dr. Gary Friedlaender. Dr. Friedlaender is Wayne O. Southwick Professor of Orthopedics at Yale School of Medicine and he is here to talk about sarcoma. Here is Francine Foss.

Foss Gary, can you start off by telling us what is sarcoma?

Friedlaender Sarcoma is a form of cancer. There are two huge areas of cancer and one is called sarcoma, which is primarily a disease of the support tissues of the body, the bones, muscles, ligaments, tendons, and other supportive structures as opposed to the huge area of carcinomas which probably occur a hundred times more frequently than sarcomas and cancers of surface areas, cancers of the skin, cancers of the lining of our lungs, breast, prostate, kidney, thyroid, GI tract, things of that sort.

Foss It sounds then like sarcomas can arise from many different areas in the body?

Friedlaender They can, there are bones almost everywhere and there are muscles throughout the body and the support structures that go along with bones and muscles.

Wilson In these sarcomas, Gary, they are soft tissue sarcomas and non-soft tissue sarcomas. Can you give our listeners a little bit more detailed information about those varieties?

Friedlaender Sure, I'd be delighted. For what generally might be called bone sarcomas, these are cancers that start directly in bone and we usually lump those, to start, directly in with cartilage as part of the that group as opposed to those that start in the muscle, tendon, ligaments, blood vessels, fat, or fibrous tissue, the support tissue that is virtually everywhere in the body.

Wilson Can these problems occur in children as well as adults?

Friedlaender All ages.

Foss Can you tell us how common sarcomas are?

Friedlaender There are about 11,000 to 12,000 new sarcomas diagnosed in the United States every year compared to about a million and a half carcinomas.

Foss And are those equally distributed between the bone and the soft tissue?

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Friedlaender Probably more in the soft tissue than bone.

Foss And do most of those occur in children or adults, or is it pretty much split evenly?

Friedlaender There are certain of these sarcomas that are characteristically found in a younger population, rhabdomyosarcoma, a cancer of the muscle in kids, Ewing's sarcoma, osteosarcoma, cancers of bone are typically found in young children, while there are some others that are more typical of middle age and older age.

Wilson Are there are any risk factors, Gary, and are there genetic predispositions to any of these descendant families?

Friedlaender There are, you do see some of these in families. Risk factors come in a variety of categories. One risk factor that does not exist and is commonly misunderstood is trauma. I frequently will see people who recall an injury that draws their attention to their leg or their arm and helps alert them to something going on, but trauma per se, as far as we know, is not a risk factor for sarcomas. Things like radiation, things that we must do in order to successfully treat some forms of cancer have a very small, but clear risk of initiating a new sarcoma at a future date. There are some environmental factors that are associated with cancers, in general. It would be remiss to not point to smoking. It does not necessarily cause sarcomas as opposed to carcinomas, but there are some chemicals that we know have the ability to induce cancers and then there are a variety of gene abnormalities, inherited gene abnormalities, that are also associated with cancers including sarcomas for neuroblastoma and for rhabdomyosarcoma. There are also some individuals who have unfortunate gene abnormalities that make them predisposed to several different forms of cancer, gastrointestinal polyps, Gardner's syndrome, are associated with an increased risk of cancer. So there clearly are situations like that, immunosuppression, relieving the body of its normal defense mechanisms, also seems to have an increased risk of cancer and the story goes on and on, and we are learning more about that at places like Yale and our Smilow Cancer Hospital all the time.

Wilson Have you noticed a predisposition to certain parts of the body? More so in the arms, the legs, or in the trunk or are things generally pretty evenly distributed when these sarcomas occur?

Friedlaender The simple answer is that they occur all throughout the body and that is one of things that I am particularly excited by and my opportunity to help people. I can help kids and more senior people. I can deal with problems in the arms, legs, and everywhere in between, and in this age of super-specialization I have colleagues who concentrate on smaller areas of the anatomy and smaller parts of the population. So for me that makes it even more interesting and more important.

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Foss Gary, you mentioned radiation as a potential risk factor, can you reassure our audiences about say simple chest x-rays and dental radiation? People worry about that and the question is, are they justified in worrying and how often would you see a tumor like a sarcoma after say a routine health care related x-ray?

Friedlaender Thank you for letting me clarify that, firstly. Again, the simple answer is that you never see cancer initiated after a routine kind of x-ray that has become important to our health such as dental x-rays, and I get dental x-rays probably every couple of years and do not worry about that at all. There is no reason to worry about that. You get more radiation in the chest flying from here to Los Angeles by airplane, then in a chest x-ray, if I recall that little factoid, and Dr. Wilson knows these better than I do, but simply put, the doses that are responsible for inducing new cancers are enormous compared to what we accumulate during our lifetime of normal x-rays. Part of it is the x-ray film and techniques, just like photography, we have faster and faster films for photography that require less and less light. We have new techniques in taking these imaging studies that require less than less radiation.

Foss Gary, can we talk a little bit about how a patient presents with a sarcoma? What is the most common thing that you see when somebody walks in with a sarcoma?

Friedlaender Two possibilities, and sometimes both, a lump and pain.

Foss Do they usually present to a primary care doctor or to another physician?

Friedlaender Yes, both of those.

Foss How do you know if you have to worry about a lump? A lot of people have various lumps and people come to me, as an oncologist, as a hematologist, for lymph nodes that have enlarged. When do you actually have to worry about a lump say that occurs in a muscle or on your joint, or another area of your body? When do you worry?

Friedlaender I think when you find a new lump it does not mean automatically that it is a tumor and there are benign tumors and malignant tumors. If I start using the term tumor, some people will jump to the conclusion that it is nasty, but to me it is just a lump. Many lumps are injuries, you bang your leg on the desk or like me, on the bed frame from time to time, and I might get a small lump, but that lump should go away rather quickly. So, a new lump and especially a new lump that enlarges should be something you want to share with a health care professional and sort it out.

Foss Are they painful usually?

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Friedlaender Not always, you cannot depend on pain being a sign of cancer.

Wilson In your role as an orthopedic surgeon, tell us a little bit about what kinds of operations you do, what sort of things are involved from the surgeon's perspective, obviously operating and removing the tumors, but what I am getting at, is talk to the audience about a lot of the planning that goes into these operations and why it is important for someone who is such a specialist such as yourself to actually performed the procedures as opposed to someone who may not have experience in sarcoma surgery.

Friedlaender First of all, why is an orthopedic surgeon, a cancer surgeon? There are not many of us but those of us who have taken a special interest in this important disease, get some additional training. It is built upon a complete residency training program in orthopedics, so I understand the musculoskeletal system. I understand the diseases or disorders that happen in bones, joints, muscles, parts of the musculoskeletal system. I understand how to operate on them and more importantly I understand when not to operate on them. It is much more important in terms of my daily life to decide when an operation is necessary, and how to take care of the person after the surgery.

Wilson What sort of planning goes into that? My understanding is it's not as easy as just doing a scan, seeing the tumor and cutting the tumor. I am trying to get at limb sparing procedures and what sort of geographical planning goes into your operation to try to maintain as much function as possible for the patient?

Friedlaender When one wants to remove a tumor, there should be an envelope of normal tissue surrounding whenever possible and whenever it is required. When the lump or tumor lesion is benign, that envelope can be very very thin and in fact sometimes the envelope is not even necessary, but as tumors get more active and aggressive that envelope needs to be larger, thicker, and as that envelope expands, the potential for other important structures to be within that envelope, structures that we need to be traded in order to get rid of the tumor become an important consideration. So part of my planning is to determine whether an envelope is necessary, how thick it is, what else is in that envelope, and what the consequences will be for the individual by removing those structures. In exchange, we are getting rid of their aggressive tumor. When those trades are reasonable in terms of function, which they are most of the time, tumors can be removed without amputation, but there are some times when the envelope includes structures that make a functional arm or leg impossible and those are times that we have to strongly consider amputation because what we are really trying to do is save someone's life, and there is a lot more to individual's lives than an arm or leg. Not that anyone wants to lose them, but sometimes you have to make a trade.

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Foss Gary, are all of the sarcomas that you take out malignant, or are some of them not as malignant as others?

Friedlaender The word sarcoma implies cancer, or malignant and what makes the tumor malignant as you well know is its ability to spread to another part of the body, but having said that, there are benign growths which are not malignant. There are low grade sarcomas and there are high grade sarcomas and there are some in between. A low grade sarcoma may behave in many ways more like a benign tumor than it does like a high grade tumor, and the things that are required to treat it may be very very different.

Foss We were going to take a short break for a medical minute. Please stay tuned to learn more information about sarcoma with Dr Gary Friedlaender.

*Medical
Minute*

There are over 12 million cancer survivors in the United States right now and the numbers keep growing. Completing treatment for cancer is a very exciting milestone, but cancer and its treatment can be a life changing experience. The return to normal activities and relationships may be difficult and cancer survivors face other longterm side effects of cancer including heart problems, osteoporosis, fertility issues and an increased risk of second cancers. Resources for cancer survivors are available at federally designated comprehensive cancer centers such as the one at Yale Cancer Center, to keep cancer survivors well and focused on healthy living. This has been a medical minute, brought to you as a public service by the Yale Cancer Center. More information is available at yalecancercenter.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 and I am joined by my co-host Dr. Francine Foss. Today, we are joined by Dr. Gary Friedlaender and we are discussing sarcomas. Gary, tell us a little bit about the treatment team, and some of the different treatment options that are involved in the management of patients with sarcomas. You had mentioned a little bit about grade of the tumor, does that have any influence on what sort of treatment program a patient may need?

Friedlaender The grade of the tumor certainly does. It is critical that the patients with aggressive tumors in particular have a team that's thinking about their individual problems and that team goes far beyond any of us as individuals, and I think it is clear that we are colleagues and we work together all the time in teams, which is extremely important. The team to me involves the surgeon, maybe an orthopedist like myself, or it may be a general surgeon or a surgical oncologist or any of the other surgical specialties, as well as medical oncology and radiation oncology which round up the trio of treatment modalities that we usually wind up discussing, but the team also includes the radiologist that helps us understand the imaging to find these tumors, where they are, what they

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look like, and the pathologist who helps us make these diagnoses. Many of these tumors are clarified by biopsies before we treat them, and the way that biopsy is done can have a huge impact on how well we can treat them.

Wilson You mentioned the radiologist, are there particular scans, types of scans that you find most helpful in the diagnosis of sarcoma, CT scan or MRI or are you often using a variety of different types of imaging to really get a sense of the extent of the problem?

Friedlaender The imaging really has to be customized to the individual, and I think that is one of the great advances of cancer treatment in general and at centers like Smilow. Is that individualization. Because a new technique could apply to you as an individual and it may not. There are things I used to do very frequently, an example would be a traditional arteriograms, or angiography, injecting dye into the blood vessels, and we do that today, but we do it far less often because we have approaches like CT scans, MRIs, and PET scans, and a number of evolving technologies like that, but just because somebody has cancer, it does not define specifically what those important imaging modalities are. It takes a team to decide what is in that patient's best interest, and makes sure they are given what they need.

Foss Gary you talked about sometimes unfortunately the need for amputation or removing large parts of a limb, can you talk about some of the new advances in limb sparing surgery and some of the new advances in prosthesis if the patient has to have a limb removed?

Friedlaender Absolutely, one of the things I have really enjoyed over the years is my association with Ted Kennedy, and I do not think he would mind me bringing his name up, but there is an example of an individual who frequently will come and talk, especially to children and teenagers that face this life changing event, to tell them that it is not a life ending event, there is the ability with modern prosthetics to get back to doing a wide range of the very important things people want to do both professionally, educationally and during leisure time. It is very-very gratifying. It is enormously difficult upfront and helping the patient realize how important this operation may be to their safety, their well-being, on one side, and how they are going to be able to do all of this when it's done and behind them and there are people who have gone through this who are very very helpful in explaining this to people at that time of crisis.

Foss And what about the various procedures where you have to say put in pieces of bone, if you have to make a piece of bone out, artificial substances that are now being used to help rebuild these limbs?

Friedlaender Most bone tumors are benign and many of them just require modest operations that do not affect the integrity of the bones, scraping out a tumor and the body has the ability to heal and to regenerate its bone, but as tumors get more aggressive and more malignant, there are times where we will have to take out segments of the skeleton and even whole joints, large portions of bone and

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we have a variety of ways of putting it back together, if you will. Sometimes, it is putting in another piece of bone that has been donated by a generous individual at the time they pass away, just like, kidneys, livers, hearts, and lungs are also provided. They are screened very-very carefully to make sure that they do not possess any diseases and can be extremely helpful in reconstructing someone's skeleton. We also have a growing number of metallic and plastic implants, the designs are improving all the time, and they can also be used to very effectively replace major parts of the skeleton and joints.

Foss Do you ever have a circumstance where a person will donate say a piece of their own bone from one place of their body to go to another area?

Friedlaender Yes, and when that makes sense it is done. It used to be far more common, and what we have learned and with the ability of new molecules that help the body re-grow its bone, we depend on that less and less and keep in mind that in order to take a bone or a part from one area and put it in another, may have some advantages biologically, but it has the disadvantage of a second operation, and removing something that has a function somewhere else. So more and more we are depending both on donated skeletal bone, in particular, and these new biomolecules that help the body repair itself.

Wilson Getting back to treatment for a second, have there been situations where you were considering potentially doing an amputation for a patient, but you considered incorporating pre-surgical radiation, for example, that might help obviate the need for an amputation, has this ever happened? Can you convert someone from a clinical situation where you think you have to do an amputation to possibly avoiding that?

Friedlaender Yes, I almost feel like you are a straight man in that regard, because you helped me out in that regard numerous times and that is why the team approach is so critical. You as a radiation oncologist learn better and better ways to more effectively irradiate some tumors, and as you know, some tumors are more responsive to radiation than others and you are very knowledgeable and help me through those decisions. We often can convert a situation into an easier surgical procedure and often that means limbs sparing as opposed to amputation.

Foss Can you talk about whether there is a role for chemotherapy in the treatment of sarcomas?

Friedlaender I am just trying to think of a word that expresses my awe and admiration to medical oncologists. The things that I get to do and some of them I have gotten extremely creative when it comes to sparing limbs is really because medical oncology has improved so much over the course of my career, my forty years doing this, and we have gotten to a situation where when I started, children with osteosarcoma, the most common bone cancer, having a survival rate of about 10% at 2 years, which was atrocious, to now where the vast majority of those people are cured. It is not a matter

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of a 2-year survival, it is cure because of multi-drug chemotherapy and that gives me the chance to do my limbs sparing tumor approaches so that those individuals who have been saved of their disease will also have a very functional life. So, thank you.

Foss You are welcome. Is it common for say the average sarcoma patient to expect to get chemotherapy or is that only in an exceptional case that you would give chemotherapy as part of the original treatment of the tumor?

Friedlaender It varies, again this is a team decision. There are clinical trials going on all the time to help us answer those questions even better and there was a clinical trial that taught us that giving chemotherapy before surgery for osteosarcoma, what might seem counterintuitive to patients, turns out, to save more people and increase their function. There is a good reason for preoperative chemotherapy, what is dangerous about a malignant tumor is its ability to spread and while I am pleased with what I can do as a surgeon just removing the tumor where it starts, it does not prevent the tumor from spreading, it is the chemotherapy that does that, and so preoperative chemotherapy followed by a well planned surgical procedure and/or radiation is often a superb decision and these individualized options are the results of clinical trials.

Foss In what cases would patients need chemotherapy later on, say if the patient has a metastasis or the tumor comes back, would it commonly be treated with chemotherapy or would surgery be an option at that point?

Friedlaender Again, it could be both or it could be radiation and I can think of examples from the last few weeks that went each of those directions. Certainly, if cancer spreads, chemotherapy becomes even more important and if they have had chemotherapy before this spread occurs, we now have more and more options of other drugs. Just like germs, some germs respond better to one antibiotic and some to another. The same is true of cancer, cancer is not one disease, it is a huge number of diseases that share this feature of spreading.

Wilson Gary, can you comment about the follow-up program for a patient after they get an operation for sarcoma, for example, because obviously the surgery might be very successful, the tumor is removed, but that is probably not the end of the process for a patient, and in fact, for some it might really only be the beginning at that point.

Friedlaender I have a wonderful opportunity to see the patients back for years after their procedure for at least two important reasons. One is, I want to make sure that the tumor does not come back where it was originally located, and that is one of my primary responsibilities. With our team approach the other aspect of their disease we want to keep watch over is whether the tumor shows up anywhere else. Sometimes, I do that part of the job as well, but often my medical oncology or radiation oncology colleagues will play a role in that. I wind up seeing patients, fortunately, for years and years after the original treatment.

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