

Yale CANCER  
CENTER

answers

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*Hosts*

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## Rare Skin Malignancies

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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](mailto:canceranswers@yale.edu) and the phone number is 1-888-234-4YCC. This evening, Lynn welcomes Dr. David Leffell to the program. Dr. Leffell is the David Paige Smith Professor of Dermatology and Surgery and Deputy Dean for Clinical Affairs at Yale School of Medicine. Here is Lynn Wilson.*

Wilson Let us start off by having you describe what skin cancer is.

Leffell Skin cancer represents a broad category of malignant or cancerous tumors or growths of the skin. I think that many listeners are most familiar with the terms basal cell cancer and squamous cell cancer, and that is largely because these are the most common cancers that we encounter. In fact, basal cell cancer itself is the most common cancer in humans, but in addition to those two cancers commonly seen in the skin, melanoma, malignant melanoma, is also considered a skin cancer. It is a malignancy or a cancer of the pigment cells of the skin, and it also is unfortunately more common than we would like.

Wilson In what age groups do we typically see basal or squamous cell carcinomas?

Leffell Lynn, that is a great question. It used to be that basal cell cancer and squamous cell cancer was seen in individuals in their 50s, 60s and later, but we are seeing patients with basal cell cancer at a much younger age and, in fact, we have a research study going on now to try to understand why basal cell cancer is developing in individuals under 40.

Wilson What are some of the lesser known types of cancers?

Leffell It is interesting, the human body is an amazing piece of machinery and it is made up, of course, hundreds of thousands of complex parts, organs, and organs are made up of cells, and cells even have little structures within them that help us function biologically. But the opposite side of that coin is that each of these cells potentially could go sour. In some ways that is what cancer is. When we talk about the skin, the skin is a complex organ, believe it or not, and we like to think that it is the largest organ in the body and it includes many types of cells. It includes the epidermal cells, or the top-layer cells, and from the epidermis basal cell cancer and squamous cell cancer develop. I already mentioned that malignant melanoma can develop from the pigment cells that are also found in the top layer of the skin, but beneath the epidermis, which in reality is only about the thickness of a sheet of paper, is the dermis, the second layer of the skin, and there are many different cells types there, and here is the rub, some of those cell types can at times develop into cancer, not very often, and in fact as a group they represent rare skin cancers, and these include cancers or malignancies of the skin that are called, and get ready because these are long names and we in medicine actually tend to abbreviate them which maybe we will do for purposes of this discussion, but the first one is an atypical fibroxanthoma, abbreviated as AFX,

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dermatofibrosarcoma protuberans, abbreviated as DFSP, and Merkel cell carcinoma, which fortunately is brief enough that it does not need an abbreviation.

Wilson Is Merkel cell carcinoma, or any of these other problems, are they related to sun exposure or viral exposures? What sort of risk factors do we know about?

Leffell Let's talk a bit about Merkel cell carcinoma. It is a very rare cancer. There are only about a thousand cases a year in the United States, and it occurs in men generally over 60 on sun-exposed areas, just like basal cell cancer and squamous cell cancer, but what is very interesting and reflects our developing knowledge and the benefits of all the investment that we as a society are making into biomedical research, is that with Merkel cell carcinoma, as we now know, there is a virus called the Merkel cell polyomavirus that probably plays an important role in causing this particular cancer. The virus itself is not the whole story, because we know that the malignancy, the cancer, develops mainly on sun-exposed areas. So clearly, the story of ultraviolet radiation having a negative impact on the skin and setting the stage perhaps even stimulating cancer in the case of Merkel cell carcinoma has to be considered.

Wilson Do you feel that if someone lives long enough, that it is highly likely that they may get a skin cancer, a basal cell carcinoma, for example, since there are so many cases of this disorder?

Leffell Clearly the cause of basal cell cancer and squamous cell cancer is related in many ways to the cumulative exposure to sunlight, ultraviolet radiation. We have not talked really about the risk factors for skin cancer, but now might be a good time. The people that are most at risk for developing skin cancer are those with light complexion, those with fair hair, blue, green, or gray eyes, certainly those people who have a family history of skin cancer. I should point out that many people think that if they have a darker complexion, they are immune to skin cancer, and I have got to tell you that here at Yale where we see patients from all over the state, there are a fair number of people now developing skin cancer who do not fit the typical risk profile, but who have had extensive sun exposure throughout their life, which of course, is the one common factor in patients that develop most of the squamous cell cancers and basal cell cancers.

Wilson Can these cancers spread to other parts of the body, or to lymph nodes?

Leffell Basal cell cancer typically does not. It can be treated readily by your physician, and although there is typically a 40% chance of getting another one within five years after your first, it is very manageable. Squamous cell cancer is thought of by the general public as more serious than basal cell cancer, and in some ways that is true, but in the majority of instances, squamous cell cancer is diagnosed at an early, very treatable stage, just like basal cell cancer. Having said that, in certain rare circumstances, squamous cell cancer of the skin is not trivial and it can potentially spread to the lymph nodes, and from the lymph nodes to the lung and other areas, but in terms of Merkel cell carcinoma, it is a serious cancer, and let me talk a bit about it. First of all, how do you know that you have one? Well, the problem with Merkel cell carcinoma is that it comes up often like just a

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red bump and it is very hard to distinguish it from a pimple early on, or a basal cell cancer or another type of skin lump, bump, or growth, and so it is very important that if you notice a new lump or bump that you get it checked out by your dermatologist as promptly as possible. Having said that, Merkel cell carcinoma in some ways can behave like melanoma, like malignant melanoma, and it can travel to other organs, first, of course, to the lymph nodes. Part of the treatment, once the diagnosis has been made, or in the process of diagnosis and treatment, includes removal of the skin cancer and also an evaluation of the lymph nodes through a technique called sentinel lymph node biopsy. This is a technique that allows your doctor to evaluate whether the lymph nodes are involved by the cancer. The good news about Merkel cell carcinoma is that if you get it, and it is treated and you do not develop a recurrence or spread of the cancer to other parts of the body for three to five years, the odds of developing those problems after that time period are extremely low. Generally we think of three years to five years as a cure period for a Merkel cell carcinoma.

Wilson David, tell us a little bit about Smilow and the Yale University skin cancer treatment history. You are, obviously, regarded as an international expert in these diseases.

Leffell I am most familiar with the program at Yale. I was actually recruited back in 1988 to start the first skin cancer, or cutaneous oncology program, and we have been able to grow substantially since that time and we have a very comprehensive program that includes clinical care where we put a great focus on patient service and quality as well as clinical research where we are actually leveraging all of the clinical information that we develop to advance science and our understanding of skin cancer. Our team helped discover the skin cancer gene in 1996, which has had many implications for our understanding of other cancers as well. The other component of the skin cancer program at Yale Cancer Center and Smilow Cancer Hospital, is our melanoma program, and malignant melanoma, as a disease is increasing and is potentially lethal and is of special concern in Connecticut. I am delighted to mention that we just recently recruited a national leader in melanoma surgery, Dr. Mark Faries who is playing a central role in our melanoma program. He is a surgical oncologist and brings with him, from the John Wayne Cancer Center, many advanced techniques and extensive expertise in the management of malignant melanoma. I would say that we run the gamut. For example, the rare cancers that we began to discuss earlier in the show, Merkel cell carcinoma, AFX, and DFSP are treated at the cutaneous oncology program in the dermatology department, but frankly, we collaborate extensively in a multidisciplinary program with the other experts at Smilow. For example, one of the key treatments for Merkel cell carcinoma after surgical excision is radiation, and that is well established, and we collaborate very closely with our colleagues in therapeutic radiology, not just with Merkel cell carcinoma but, as you know Lynn because we worked together on some of these projects, with complex cases of squamous cell carcinoma for which there is not broad expertise elsewhere, and that is one of the challenges. I actually wish sometimes that there were others that were more expert than we are, because these cases can be very challenging, and in the case of extensive skin cancers, such as DFSP and even basal cell cancer, we have the ability to collaborate very closely with

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reconstructive plastic surgeons at Yale who are also part of Yale Cancer Center and Smilow Cancer Hospital. While most of the cases of skin cancer are easily treated in our unit, we are able to at any time pick up the phone and usually have a patient seen by one of our colleague consultants the same day, which as you can imagine is a great satisfier.

Wilson Absolutely! David, you are a dermatologic surgeon, tell our listeners a bit about that, your training background, and what is Mohs surgery?

Leffell Titles and labels are very complicated at times but they actually represent, in most cases, a wide range of extensive training. Dermatologic surgery is a subspecialty of dermatology, the same way cardiology is a subspecialty of internal medicine, and individuals first have to finish training in dermatology and become board certified in that specialty. After that they pursue a fellowship, a full year of training, specifically in skin cancer surgery and reconstruction, and we at Yale actually sponsor such a fellowship. The fellowship is centered on a technique called Mohs surgery. Mohs surgery is actually named after Frederic Mohs who was a surgeon at the University of Wisconsin who developed a technique to remove skin cancer in a layered fashion, and the advantage of that is that the cancer is removed in as conservative a fashion as possible.

Wilson And how many Mohs surgeons or dermatologic surgeons do we have here at Yale?

Leffell I am pleased to say that our program now has four dermatologic surgeons. Our most recent recruit, Allison Hanlon from Vanderbilt University is an MD, PhD who in addition to performing Mohs surgery and handling a wide variety of skin cancer patients, is developing an active research program in skin cancer.

Wilson Terrific! We are going to take a short break for a medical minute. Please stay tuned to learn more information about skin malignancies with Dr. David Leffell.

*Medical  
Minute*

*This year, over 200,000 Americans will be diagnosed with lung cancer, and in Connecticut alone there will be over 2,000 new cases. More than 85% of lung cancer diagnoses are related to smoking, and quitting, even after decades of use can significantly reduce your risk of developing lung cancer. Each day, patients with lung cancer are surviving, thanks to increased access to advanced therapies and specialized care. New treatment options and surgical techniques are giving lung cancer survivors more help than they have ever had before. Clinical trials are currently underway at federally designated comprehensive cancer centers, like the one at Yale, to test innovative new treatments for lung cancer. An option for lung cancer patients in need of surgery at Yale Cancer Center is a video-assisted thoracoscopic surgery, also known as a VATS procedure, which is a minimally-invasive technique. This has been a medical minute. More information is available at [YaleCancerCenter.org](http://YaleCancerCenter.org). You are listening to the WNPR Health Forum on the Connecticut Public Broadcasting Network.*

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- Wilson Welcome back to Yale Cancer Center Answers. This is Dr. Lynn Wilson. Today, we are joined by Dr. David Leffell, and we are discussing skin malignancies. Let's return our attention to some of the more unusual malignancies of the skin that you have mentioned already, but let us get into a little bit more detail about them, how they present, what sort of physicians should be involved in their treatment and how they should be managed?
- Leffell I think we can start off by talking about the atypical fibroxanthoma, the AFX, and once you get to the level of rare and infrequent cancers, you really butt up against the diagnostic criteria, how do you know what kind of cancer it is and can you be sure and what is the right treatment? Ultimately, the diagnosis of skin cancer is made by a biopsy, and a biopsy is a very simple office procedure where the skin is numbed with a little bit of lidocaine, similar to what a dentist might use, and the specimen is either shaved off or punched out of the skin and sent off to a skin pathology lab, and I want to emphasize this, it is very important to make sure that your skin biopsy is read by a certified dermatopathologist or skin pathologist, especially when we are talking about these rare malignancies, which frankly many pathologists do not have a chance to see that often. You really want to make sure that if the dermatologist is concerned that it may be one of these rare cancers that it is read by individuals, frankly, at a university center.
- Wilson Talk to us about the management of these various, more unusual disorders?
- Leffell Again these are skin cancers that develop in older people, elderly men and most often on the scalp, and that is corroborated by the literature, and they come up in a relatively nondescript fashion. They can look like a red bump or a crusted bump and may not even alert the patient that there is something wrong. Let us assume that you are seeing your dermatologist on a regular basis because you are at risk for skin cancer and the diagnosis is made by biopsy. The treatment for AFX is the Mohs surgery, it is the treatment of choice because you are able to remove the skin cancer in the office setting layer by layer, and once it is all complete, again, in the office setting, if the wound is such that you can perform the reconstruction then and there, it is all taken care of at once. AFX has a relatively low potential for spreading to the lymph nodes and into the blood stream. Although in my experience, I have seen it happen. When it occurs, one really has to wonder whether the diagnosis was an AFX or a cousin of AFX, which occurs less often on the scalp but does have a potential to metastasize, but we are talking now about very arcane issues but it is important for the listening public to know that we, as physicians, do live in a world where we need to be alert to many of the nuances and complexities of things that are not routine. Once AFX is treated by the Mohs technique and the area is repaired, or skin grafted or whatever is required, the risk of recurrence, at least with the Mohs technique, is very low; maybe 1% or 2%, and the patient can be reasonably sure that they are not going to get into trouble from that. On the other hand, individuals with AFX are also at risk for the more common skin cancers that we talked about at the top of the show, squamous cell cancer and basal cell cancer, for example. So AFX is relatively simple, the key thing there is making sure that the diagnosis is correct and that the treatment has been thorough and complete. Another entity, which I mentioned briefly but is also a mouthful, is dermatofibrosarcoma protuberans, which we like to refer to as DFSP for purposes of

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simplicity. This is not a sun-related skin cancer, and it is a bit tricky because it develops very slowly, actually, in many cases over many years so slowly, in fact, that people may not even be aware that they have it, and it is a malignancy of fibrous tissue, which presumably originates in the second layer of the skin, again, that I described in the first half of the show. The treatment for DFSP is excision. How do you know you might have one? Well, they can look like anything and I do not say that to induce a level of broad paranoia, but rather to make people realize that if they have an area that they thought was a scar or something firm on their arm, on their chest, really it can occur anywhere and you do not have a good explanation for it, this should probably be checked out because DFSP is often slightly raised, maybe a little purple in color, a little red in color, sometimes even brownish or tan in color. Sometimes, you can put your fingers around it and pinch it and actually grasp it if it is of sufficient size, and it feels when you do that, like it is embedded within the second layer of the skin, within the depth of the skin itself. The good news is, it almost never spreads to the rest of the body and it is, again, easily treated once the diagnosis is made, with excision, either conventional surgical excision with an appropriate margin of safety or with the Mohs technique where again you are studying the margins immediately and tracing out the cancer in all of its directions until you are certain that it has been completely removed.

Wilson When you do that, David, with the Mohs technique, you are doing that work yourself?

Leffell Yes. One of the unique things about the Mohs technique is that the Mohs surgeon is trained to do the surgical procedure to map the tissue but also to examine it under the microscope, and one of the advantages of this, and I think it is also one of the reasons that accounts for the high cure rate of the Mohs technique, is you have an unbroken chain of events from the time that the surgeon takes the specimen to the time that they evaluate it under the microscope and correlate the findings under the microscope with what is on the map that the surgeon has drawn. Now, I should point out that Mohs surgeons are not acting as pathologists. We are not diagnosing cancer. We would never operate on a cancer that does not already have a diagnosis. We are following an existing malignancy pattern and we have available to us, of course, many experts in dermatopathology. When we confront complicated cases, we again engage the teamwork approach in diagnosing complex situations.

Wilson I would like to add that many patients that I have seen over the years, especially who have had basal or squamous cell carcinomas in their head, neck area, some of those patients I have seen have had relatively large operations and have some cosmetically unattractive scars. When I have talked to these patients, I have found that they have not had the Mohs procedure, and so I think another advantage, see if you agree, is that not only do you have a very confident sense that you have removed all of the cancer, but also cosmetically, if we can remove less tissue, and with your reconstructive skills, make things look good, that sounds very advantageous for a patient, especially if they have a lesion on their face, for example.

Leffell I cannot comment on cases that are done elsewhere, I can only comment on the work that we do and on the principles and the rationale that underlie the Mohs technique. It is, as I mentioned,

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and as you imply, a technique for tissue preservation. Because we are studying the tissue right away, we only have to remove the malignancy and can spare as much normal tissue as possible. What does that mean? Well, let's just say that you have a skin cancer that is 7 mm, the size of a pencil eraser. If you are going to excise it with conventional margins, you have got to take about 4 mm on either side, which is not a lot, and frankly it is very appropriate in many skin cancers. So, let me be clear, the Mohs technique is really indicated for lesions that are high-risk that are in cosmetically important areas that have been recurrent already where the edges of the cancer cannot be identified easily by the dermatologist or the plastic surgeon. In fact, a vast majority of skin cancers below the neck do not use the Mohs technique, but as you suggest, on the face, you definitely want to take advantage of this conservative, tissue-sparing technique that permits immediate reconstruction, all in the office setting.

Wilson        What sort of follow-up do these patients require? You have done the procedure, it has gone well, the patient is going to go home after the office procedure. When do you see them back again, suture removal, and what sort of follow-up program should they have?

Leffell        We work closely with the referring physician, typically if we do some form of plastic surgery, they return within a week to have the sutures removed, and then follow-up with the local referring physician, the local dermatologist. Very often, at the request of the local dermatologist, patients that get a lot of skin cancers or are high-risk, we will co-manage, we will share the management and I think in those cases patients certainly benefit and we're happy to do that, but the most important thing that we do after the surgery is over is try to educate patients about what they need to do themselves going forward, and that includes an aggressive program of sun protection. Sun protection includes the regular use of sunscreen with a sun protection factor of 30 or higher and that includes protection against ultraviolet A waves, so-called broad-spectrum. Sometime this year, we expect to hear from the FDA on a revised monograph that will define new labeling for sunscreen bottles. It is true that all of this is very complicated and the listener, the consumer, often is not clear, it is generally thought that the higher the SPF number, the better, and that is not necessarily true, in a practical sense. Regular application of sunscreen is important, wearing a brimmed hat, avoiding the sun during the peak hours of 10 a.m. to 4 p.m., certainly, if you are responsible for young children, ensure that they are well protected from the sun since so much of lifetime sun exposure is acquired in childhood. The most common thing we hear from people is, well, doc, I am 50, wasn't most of this damage done when I was a kid? And the answer is, yes, but fortunately, people now are living so long that 50 is yesterday's 30, and I think that we need to make sure that people realize that they need to take care of their skin and protect it from the sun because we are all going to be living a very long time and we can definitely forestall additional damage by ensuring that we protect ourselves against the harmful effects of ultraviolet radiation.

Wilson        David, is glass protective, if I am in my car with the windows up, is that enough?

Leffell        That is a very complex question and you think there would be an easy answer. On the one hand, the film that is used in automobile glass to make it shatterproof does provide UV protection and we

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generally consider automobile glass to be somewhat UV protective. On the other hand, we see patients that come in with predominantly left-sided skin cancers and they do not live in England, and clearly they are exposed to sun. I think that from my perspective, automobile glass is protective, but it is very difficult to say explicitly that you do not need to have any type of sun protection.

Wilson      Jumping back to this sentinel lymph node procedure that you had mentioned, obviously, everything we have discussed thus far can be done in the office, relatively easy for patients, they go home with a sentinel lymph node procedure, is that something that is done as an outpatient as well?

Leffell      I am glad you asked that, because increasingly the sentinel lymph node biopsy procedure has become the mainstay of the management of many types of melanoma, Merkel cell carcinoma, in some cases, breast cancer, and other malignancies, and it is a hospital-based procedure, it is done in the operating room by individuals such as Dr. Faries that are experts at performing it, and it is one of those techniques where experience matters.

*Dr. David Leffell is the David Paige Smith Professor of Dermatology and Surgery and Deputy Dean for Clinical Affairs at Yale School of Medicine. If you have questions or would like to share your comments, visit [YaleCancerCenter.org](http://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.*