

**Melanie:** Welcome to the podcast series from the specialists at Penn Medicine.

I'm Melanie Cole, and joining me today is Dr. Giorgos Karakousis. He's a Professor of Surgery, and Endocrine and Oncologic Surgery at the Hospital of the University of Pennsylvania. He's here to highlight retroperitoneal sarcoma.

Dr. Karakousis, it's a pleasure to have you join us again today. Please start by telling us a little bit about retroperitoneal sarcoma, the prevalence of it, what you've been seeing in the trends and the scope of what we're talking about here today.

**Dr. Giorgos Karakousis:** Well, first off, thank you, Melanie, for having me on this podcast again. So as you mentioned, I'm a surgical oncologist here at the University of Pennsylvania, and so that means I treat a variety of cancers that are found throughout the body. But one of my clinical interests and specialty is specifically with retroperitoneal sarcomas.

Retroperitoneal sarcomas are a subtype of sarcomas that are found in the back of the abdomen. And they're fairly uncommon. They represent maybe 10% to 20% of all sarcomas and less than 1% of solid malignancies in adults. So, these are rare tumors that can occur in patients. And as a result, they oftentimes need expertise from specialized centers that have experience in taking care of these.

**Melanie:** Are there different types, doctor?

**Dr. Giorgos Karakousis:** Yeah. So sarcomas in general, there's over 70 different histologic subtypes of sarcoma. And similarly, there are various subtypes of retroperitoneal sarcoma. The most common being a subtype called liposarcoma, which are tumors that arise from the fatty tissue in the back of the abdomen and leiomyosarcomas, which are tumors that arise from the smooth muscle cells in the body. Those are the two most common types.

There's other types as well. And one, for instance, type is these so-called IVC leiomyosarcomas. These are tumors that actually arise from the walls of major blood vessels. And so depending on the histologic subtype, depending on the location, that can affect and influence what kind of decision we make for these particular types of tumors.

**Melanie:** Based on the fact that these are pretty rare, what symptoms do patients typically present with?

**Dr. Giorgos Karakousis:** So, that's an excellent question. You know, many times actually patients won't experience any symptoms, particularly when these are small in size. It's often not until they get large that patients will notice maybe some vague abdominal symptoms or actually feel a mass in their abdomen, and that's what alerts them to something being awry.

These tumors, unlike carcinomas, when we talk about carcinomas like garden variety colon cancer or stomach cancer or pancreas cancer, which are adenocarcinomas which tend to be infiltrative, meaning they invade into nearby organs causing things like bleeding or obstruction or severe pain. These sarcomas are often displacive, meaning they kind of push things to the side and can grow at quite large size before causing any significant symptoms. So, the symptoms are often not there until these are large. And as a result, they can sometimes go undiagnosed for quite a period of time until they get to a large size.

**Melanie:** Well then, speak about diagnostic criteria and the process for diagnosing these.

**Dr. Giorgos Karakousis:** So when a patient begins to experience any such symptoms, oftentimes we get some sort of cross-sectional imaging test, most commonly a CT scan of the abdomen and pelvis that will show us the tumor and its relationship to nearby organs and other structures and help us in our decision-making as to how to best treat the tumor. In some cases or situations, an MRI may be helpful to better delineate the extent of the tumor.

Additionally, since these tumors can commonly spread to other locations, most commonly to the lung, we oftentimes also obtain a CT scan of the chest to complete staging of patients with these tumors to ensure that there's no evidence of metastases.

**Melanie:** And if there is no evidence of metastases, which I imagine would change your treatment criteria and modality, what does treatment look like for patients with retroperitoneal sarcoma?

**Dr. Giorgos Karakousis:** So, the main stay of treatment still remains surgery for these patients. So if surgery's feasible, then that's by far the best treatment for these tumors. The surgery involves removing the tumor in its entirety, and also en bloc resection of any contiguously involved organs. So, commonly, that may be intestine, oftentimes the large intestine, the colon, which these tumors can be involving, or other structures. In some cases, vascular structures. If there's vessel involvement, it may involve resection of blood vessels as well. So, that's the mainstay of treatment.

That being said, there may also be a role for radiation for these tumors. If radiation is given, generally, we recommend radiation upfront or what's called neoadjuvant radiation. Because when these tumors are removed, it's very difficult to provide radiation afterwards, since a lot of the intestines and other organs shift around in the abdomen, making it very hard to spare those organs from receiving the radiation.

Recent studies have not shown a tremendous impact in radiation benefit for these particular types of tumors. Although some subgroup analysis have shown there may be some advantages to radiation for local control. And so, decision for things like radiation are really made on a case to case basis based on the histology, the size of the tumor, location, and those sort of factors.

Chemotherapy is generally not recommended upfront for these tumors if they're resectable. But in some situations, again, if there's a concern of difficulty of achieving a complete resection, we can consider

chemotherapy as well to try to shrink these tumors and make the surgery more feasible.

**Melanie:** This is very comprehensive, doctor. So, what makes the Penn Medicine Abramson Cancer Center so unique when it comes to diagnosing and treating retroperitoneal sarcoma? I'd like you to speak about your team and the importance of that multidisciplinary approach for these patients.

**Dr. Giorgos Karakousis:** The Penn Medicine Abramson Cancer Center really has an outstanding team of clinical specialists to treat these rare tumors. And that ranges from the surgeons like myself and other surgical oncologists that are part of the team; other surgeons that are oftentimes involved in these cases like vascular surgeons, who may sometimes have to reconstruct blood vessels after we remove these tumors; to plastic surgeons – sometimes these tumors can involve abdominal wall and involve reconstructions of the abdominal wall to repair.

And then, other disciplines of course. So, radiation oncology can play an incredibly important role for these tumors. And here at Penn, we're fortunate to have proton therapy, which is a particular type of radiation therapy that can focus the radiation beams to specific areas and therefore spare collateral damage of the radiation to other nearby tissue. And for these particular types of tumors, this can be very important because these lie in an area where there's oftentimes other organs nearby like the kidney or the colon. And so, really having ability to do focused radiation can be very important.

Medical oncologists can also play an important role for these tumors in cases where, for instance, these tumors have recurred or metastasized. And again, we're very fortunate to have really an outstanding team of medical oncologists who can offer their expertise to patients and also offer clinical trials to patients which are not available in other places.

So, this is really I think what makes Penn unique. We have a really multidisciplinary approach for every patient that comes in with a sarcoma to review their imaging with our radiologist, our pathologist. Many times, these patients will get biopsies upfront. And so, review of the pathology with specialists who really have a clinical focus in sarcoma specifically to coming up with clinical decision-making with our clinical team. And so, this, I think is incredibly important for these tumors, which are rare, and for which outcomes can really vary depending on the expertise of the center.

**Melanie:** Doctor, is there any research at Penn? You mentioned clinical trials. I'd like you to speak about any research that you're performing in the retroperitoneal sarcoma space.

**Dr. Giorgos Karakousis:** Yeah. So, as I mentioned, we have a variety of clinical trials open for sarcoma, which are rare tumors, and they're inclusive of patients with retroperitoneal sarcoma specifically. So, this is something that I think affords patients who fail conventional therapy to be able to be treated with. Beyond that, there's a lot of basic scientist collaborators that we have at Penn who are working specifically on sarcoma here on campus and in association with our veterinary school to better understand the biology of these tumors and make further advancements into finding new treatments for these tumors.

**Melanie:** As we wrap up, doctor, what would you like to let other other physicians know, the key takeaways from this podcast on retroperitoneal sarcoma and when you feel it's important that they refer their patients to Penn Medicine?

**Dr. Giorgos Karakousis:** So, I think the take home points are these are rare tumors. And so when they're found on imaging either incidentally or based on symptoms, early referral to a center like Penn that has expertise is really critical for improving the outcomes of the patients.

The first step, even from initial biopsy – to be done by an expert team that is familiar with doing these biopsies – to the review of the

pathology, to the multidisciplinary team to review the plan of care, to the surgical expertise to be able to remove these safely to try to minimize the chance of recurrence and injury to other nearby organs are all things that Penn Medicine can offer.

The takeaway points: when making a diagnosis of a retroperitoneal sarcoma, early referral to a place like Penn is incredibly important to improve the outcomes of these patients.

**Melanie:** Thank you so much for joining us today and sharing your incredible expertise.

To refer your patient to Dr. Karakousis at Penn Medicine, please call our 24/7 provider only line at 877-937-PENN or you can submit your referral via our secure online referral form by visiting our website at [penncancer.org/referyourpatient](https://penncancer.org/referyourpatient). That concludes this episode from the specialists at Penn Medicine. I'm Melanie Cole.