

# Soft tissue sarcoma

## **Introduction**

This booklet is to give you, your family and your friends some information about this group of illnesses, and how we treat them at The Christie.

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## **The Christie sarcoma team**

The sarcoma team at The Christie is part of the Greater Manchester and Oswestry Sarcoma Service (GMOSS). This is a regional specialist team taking referrals from the Greater Manchester and the surrounding regions, Wales and Staffordshire. Each year this team sees over 100 new patients with soft tissue sarcoma and approximately 50 patients with primary bone sarcoma.

At The Christie the team members are:

- Dr James Wylie, clinical oncologist
- Dr Catherine Coyle, clinical oncologist
- Dr Daniel Saunders, clinical oncologist
- Dr Michael Leahy, medical oncologist
- Dr Laura Horsley, medical oncologist
- Mr David Mowatt, plastic and reconstructive surgeon
- Mr Damir Kosutic, plastic and reconstructive surgeon
- Professor Noel Clarke, professor of urological oncology
- Dr Patrick Shenjere, consultant histopathologist
- Ann Buchan, sarcoma clinical nurse specialist
- Lisa Cooper, sarcoma clinical nurse specialist
- Mandy Bell, retroperitoneal sarcoma clinical nurse specialist
- Maxine Cumbo, sarcoma physiotherapist

The core team is supported by consultants from the pathology and radiology departments who have specialist expertise in the diagnosis of soft tissue sarcomas.

## **What are soft tissue sarcomas?**

Soft tissue sarcomas are an uncommon type of malignant disease, accounting for less than 1 in 100 of all adult cancers. They can appear in supporting tissues of the body, such as muscle, fat, blood vessels or nervous tissues - in fact, they can occur anywhere in the body. They are particularly common around the limbs, but can also occur within the abdominal cavity, the chest cavity, or around the head or neck region.

Soft tissue sarcomas are different from other sarcomas which occur in bone and cartilage.

Soft tissue sarcomas can occur in childhood, but the information here describes the type of disease that occurs in adults.

## **What causes soft tissue sarcomas?**

For the vast majority of people with soft tissue sarcomas, we do not know the cause. Like many other cancers, sarcomas often arise 'out of the blue', even in people who lead a healthy lifestyle.

**However, we are beginning to build up a picture of what might cause soft tissue sarcoma.**

- Research suggests that some sarcomas are caused by a 'spelling mistake' in the instructions contained within the genes that control the activity of supporting tissues.
- Sometimes it is possible to identify the damaged gene by performing molecular analysis of the sarcoma cells in a gene research laboratory.
- Very occasionally the genetic damage that can lead to sarcoma is inherited. This is the case with a condition called Neurofibromatosis which is characterised by multiple skin lumps called neurofibromas. People who carry this condition have a 1 in 10 chance of developing a sarcoma. We are developing a Central Register of people with Neurofibromatosis in Manchester, so that we can help track people who may be related to someone with this condition.
- A rare but important cause of soft tissue sarcoma is damage caused to tissues by previous radiotherapy for another condition.

## What types of soft tissue sarcoma are there?

These are the most common types of soft tissue sarcoma:

- epithelioid sarcoma
- fibrosarcoma
- leiomyosarcoma
- liposarcoma
- malignant peripheral nerve sheath tumour (MPNST)
- rhabdomyosarcoma
- sarcoma NOS (not otherwise specified)
- synovial sarcoma.

There are other rarer varieties of sarcoma.

For information about the treatment of some rarer sarcomas, see the end of this information sheet.

## Making the diagnosis

There are many different types of soft tissue sarcoma, some of which are easier to treat and others which can be more difficult. So before any treatment can begin, we need to establish the exact type of sarcoma.

In cases where the lump is very small or superficial, it may be possible to proceed straight to surgery. But in most cases, it is useful to have a specialist scan (CT or MR scan) before any surgery, followed by a needle biopsy of the lump. This involves placing a needle into the lump (under local anaesthetic) to remove a core of tissue for analysis.

When this tissue is examined under the microscope, it is often possible to classify the *grade and type* of the tumour. Low grade tumours (grade 1) tend to be easier to treat and have a better prognosis than high grade tumours (grades 2 and 3). Some tumours may in fact turn out to be benign (non-cancerous).

Once the diagnosis of sarcoma is confirmed, we usually arrange a CT scan of the lungs, to make sure that the tumour has not spread to this area (the most likely part of the body for these types of tumours to spread to).

## Questions for your doctor

Whenever treatment is discussed with you, remember that it is always up to you whether you have it or not. We would like to help you make a choice by providing as much information as you need before you make that decision. You will always be asked to sign a consent form before starting any treatment. Before you decide, here are some things you might like to think about and perhaps discuss with your doctor.

What are the benefits of this treatment?

What are the side effects or risks (short term and long term)?

Are there any alternatives to this treatment?

What would happen if I did not have treatment?

## How do we treat soft tissue sarcomas?

To treat soft tissue sarcomas effectively, it may be necessary to involve several different specialists, such as surgeons and doctors specialising in radiotherapy (treatment with x-rays) and chemotherapy (treatment with drugs). So patients coming to The Christie are asked to attend a multi-disciplinary clinic where all these specialists are present. We can then agree on a treatment plan, tailored to the individual patient's specific problem.

The treatment we can offer will vary depending on whether the sarcoma is a *primary tumour* (one which has not spread elsewhere) or whether it is more advanced.

### **Treatment for primary tumours: surgery, radiotherapy, chemotherapy**

The recommended treatment is usually a combination of surgery and radiotherapy.

The aim of the surgery is to completely remove all the visible malignant tissue, as well as a safety margin of normal tissue around the tumour. This may sometimes mean removing an entire muscle group, or an organ such as the kidney, or a section of bowel, depending on where the tumour is.

Radiotherapy may be delivered to the area where you had your operation, either before or after surgery. However, for some people surgery on its own is sufficient.

## Surgery

The different types of surgical operations are as follows:

**Wide local excision:** this operation involves removing the sarcoma with a margin of normal tissue around it. Radiotherapy is often given as well, as there is a risk of sarcoma cells remaining after this particular type of operation. This is the operation offered to the majority of patients.

**Compartmentectomy:** the muscles of the arm and leg are divided into separate anatomical groups or compartments. Surrounding each compartment, there is a dense fibrous sheath, which is relatively resistant to the spread of the sarcoma.

A compartmentectomy is an operation to remove the entire sarcoma – containing muscle compartment, together with its surrounding fibrous sheath. This may be offered for larger tumours.

In some cases, the surgeon may need to use a technique which brings new skin (skin graft) and muscle (known as a pedicle or free flap) into the area to repair the wound.

Radiotherapy is sometimes given as well, depending on the likelihood of sarcoma cells remaining after the operation.

**Amputation of limbs:** nowadays, this is only done in a very small minority of cases, where the tumour comes back after the original treatment.

## Radiotherapy

This may be given before surgery (pre-operative radiotherapy) or after surgery (post-operative radiotherapy). Very occasionally, when it is not possible to use surgery, radiotherapy will be the only treatment given. The radiation doctor (a radiotherapist) may discuss both these options with you if both are felt appropriate.

**Post-operative radiotherapy:** This is usually given once the wounds from the operation have healed. The radiotherapist will use information from the surgeon and also from scans carried out before the operation (pre-operative imaging) to determine accurately the area at risk. Normally, the whole site of the operation is treated with a safety margin of approximately 5cm (2 inches).

**Pre-operative radiotherapy:** In certain circumstances, it is better to deliver the radiotherapy before the operation. We will always discuss the pros and cons of pre-operative radiotherapy with you before a final decision is made.

Radiotherapy is usually given daily (excluding weekends) over 5 to 6½ weeks.

## **Chemotherapy**

Some research suggests that some patients may benefit from having chemotherapy given after treatment of the primary tumour. However, the level of benefit obtained is much less than with chemotherapy for other types of cancer. For patients with a low grade tumour, there appears to be no benefit at all. Your medical team will discuss this with you.

## **Physiotherapy**

Many soft tissue sarcomas arise within or very near to muscle tissue. An operation to remove the tumour may involve removing some muscle tissue. This may leave your arm or leg stiff and weak. You will be shown specific exercises to re-establish full range of movement and regain your strength.

If you have difficulty walking after the operation the physiotherapist will assess you to see if you need a walking aid, such as a walking stick or elbow crutches, to improve your mobility.

Radiotherapy may also cause some soft-tissue tightness. A physiotherapist will show you stretching exercises to minimise these effects.

We will also show you some specific massage techniques to reduce the soft tissue tightness and improve the scar tissue. We advise you to continue with this for up to 12 months following the radiotherapy.

## **Treatment for advanced disease**

Advanced disease is when the tumour has spread to other parts of the body. The terms used for this spread are secondaries or metastases. The most common site for the tumour to spread to is the lung, but metastatic disease can also occur in other sites.

If there are only a few lung metastases, we may advise that these are removed surgically. Otherwise, the recommended treatment for metastatic disease is chemotherapy.

There are international studies going on at the moment to try to improve the effectiveness of chemotherapy by using different types and dosages of drugs. At The Christie, we are participating in some of these studies.

We advise patients to report any lumps around the site of the primary tumour, as these may need further investigation. Symptoms such as coughs, chest infections, and pain in the back or abdomen should be reported to the GP in the first instance. In most cases, these symptoms will be due to everyday illnesses which will get better with treatment from the GP. However, if they do not respond to treatment, or if they persist for more than 2 or 3 weeks, you should see a specialist doctor, so that a chest x-ray or other relevant tests can be organised.

## **Follow-up after treatment**

Following the successful treatment of a primary soft tissue sarcoma, we currently recommend follow-up visits initially every few months for most sarcomas. In some lower grade tumours the follow-up is less frequent. After 2 years follow-up will become less frequent but will often continue for 10 years, including a chest x-ray at each appointment.

If chemotherapy has been part of the treatment, follow-up visits may be more frequent.

## **Information about rare soft tissue sarcomas**

### **Desmoid tumours**

**(also known as fibromatosis or musculoaponeurotic fibromatosis)**

Desmoid tumours are not true sarcomas and strictly speaking, they are not malignant either, because they do not spread. However, they do often come back after treatment. They can sometimes occur in young adults, associated with a condition known as familial adenomatous polyposis or Gardner's syndrome.

In some cases, these tumours appear as distinct round lumps, for example, within an abdominal muscle. If so, treatment would be very similar to the treatment of a muscle sarcoma - complete surgical removal, with a wide safety margin.

In other cases, fibromatosis can produce a diffuse mass that spreads through several adjacent tissues. This makes surgical clearance more difficult. We would still aim to treat it surgically, as this does appear to be the most successful form of treatment. Radiotherapy or chemotherapy may also be considered, either with surgery or instead of it.

In some cases, fibromatosis will grow fairly rapidly for a few months, and then stop growing, even without treatment. When this happens, it may be preferable to have a period of observation instead of any treatment.

### **Dermofibrosarcoma protuberans (DFSP)**

DFSP is a tumour arising within the skin. It very rarely metastasises (spreads to other parts of the body), but it can spread through the skin and produce a large and painful tumour or ulcer.

The usual treatment is surgery, and it is essential to make sure that there is a wide clearance of unaffected (healthy) tissue around the DFSP. This may necessitate a skin graft or other type of reconstructive surgery, which can leave scars. Sometimes, radiotherapy is also given after surgery.

### **Retroperitoneal sarcomas**

Retroperitoneal sarcomas are those that arise within the abdominal or pelvic cavities. The commonest type of sarcoma found in this area is a liposarcoma.

As with other sarcomas, we will arrange for careful assessments to be made before any treatment is started. This will include appropriate imaging, and a needle biopsy is often useful.

The usual treatment is surgery to remove the tumour, and possibly other adjacent organs. Radiotherapy and chemotherapy may also be used.

We have a specialist team dealing with these particular tumours.

## **For further information**

Members of the soft tissue sarcoma team are very willing to answer your questions.

|  |                      |
|--|----------------------|
| <b>Ann Buchan, sarcoma clinical nurse specialist</b>                 | <b>0161 446 3094</b> |
| <b>Lisa Cooper, sarcoma clinical nurse specialist</b>                | <b>0161 918 2196</b> |
| <b>Mandy Bell, retroperitoneal sarcoma clinical nurse specialist</b> | <b>0161 918 7000</b> |

In addition, you may also like to see the booklet on soft tissue sarcomas produced by Macmillan. This is available on their website [www.macmillan.org.uk](http://www.macmillan.org.uk) or by phoning their freeline **0808 808 0000**, or visit the cancer information centre at The Christie on the glass link corridor: opening hours Monday – Friday 9:30am to 4:30pm.

Many of The Christie booklets are available on The Christie website – [www.christie.nhs.uk](http://www.christie.nhs.uk)

## **CancerHelp UK**

Cancer Help UK is Cancer Research UK's website for patients. They have excellent explanations of the diagnosis and treatment of sarcoma including "Questions for Your Doctors". The content of this website is carefully approved by doctors who are specialists in the field. The website is at [www.cancerhelp.org.uk](http://www.cancerhelp.org.uk)

## **Sarcoma UK**

Sarcoma UK is a patient support organisation founded by a patient with sarcoma and provides a newsletter, information leaflets, organises meetings and lobbies for patients with sarcoma. Get involved! Their website is at: [www.sarcoma-uk.org](http://www.sarcoma-uk.org)  
An email support group is at [www.groupspaces.com/sarcoma](http://www.groupspaces.com/sarcoma)

## **GIST Support UK**

This is a patient support group specifically for patients with a type of sarcoma of the stomach and intestine called GIST (gastro-intestinal stromal tumour). They provide leaflets and organise patient support meetings. Their website is at: [www.gistsupport.co.uk](http://www.gistsupport.co.uk)

## **Bone Cancer Research Trust**

This organisation is run by parents and patients with bone sarcoma (e.g. osteosarcoma). They have a fund raising programme to support research into bone sarcoma. Their website is at: [www.bonesarcomaresearch.org.uk](http://www.bonesarcomaresearch.org.uk)

## **Research into sarcoma at The Christie**

Partly because sarcoma is such a rare tumour we have a lot to learn about what causes this type of cancer and how best to treat it. The sarcoma team at The Christie is committed to research to best help the patients of today and to try to improve treatment for patients in the future. We have an active clinical trial programme. This means that new treatments may be available in the clinic and you may be invited to join a clinical trial. All clinical trials are completely voluntary and full discussion and information about any trial is always given before entering. If you would like to know more about the clinical trials that we are doing at the moment please ask.

## **Clinic times**

### **Dr Wylie, Dr Coyle, Dr Saunders, Mr Mowatt and Mr Kosutic's joint sarcoma clinic:**

Every 2nd and 4th Tuesday mornings.

Other appointments may be given for follow-up or urgent appointments.

### **Dr Leahy's and Dr Horsley's sarcoma clinic:**

Every Tuesday morning and Thursday afternoon.

### **Prof Clarke's urology clinic:**

Every Wednesday afternoon.

## **Case conferences**

All new cases are presented and discussed at meetings with the whole team.

The GMOSS sarcoma team meets every Wednesday at the Manchester Royal Infirmary. This ensures that all appropriate treatments are carefully considered.

If you have a retroperitoneal sarcoma, your case will be discussed at the Retroperitoneal MDT. This also occurs every Wednesday at The Christie and involves Professor Clarke who is a specialist in the surgical management of this type of sarcoma.

## **Personal data protection**

We carefully collect information about each patient, their tumour, and their treatment. This data is held in a secure computer database to help us monitor and improve our service. Access to the data is restricted to appropriate people.

If you need information in a different format, such as easy read, large print, BSL, braille, email, SMS text or other communication support, please tell your ward or clinic nurse.

We try to ensure that all our information given to patients is accurate, balanced and based on the most up-to-date scientific evidence. If you would like to have details about the sources used please contact **patient.information@christie.nhs.uk**

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Contact The Christie Hotline for urgent support and specialist advice

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