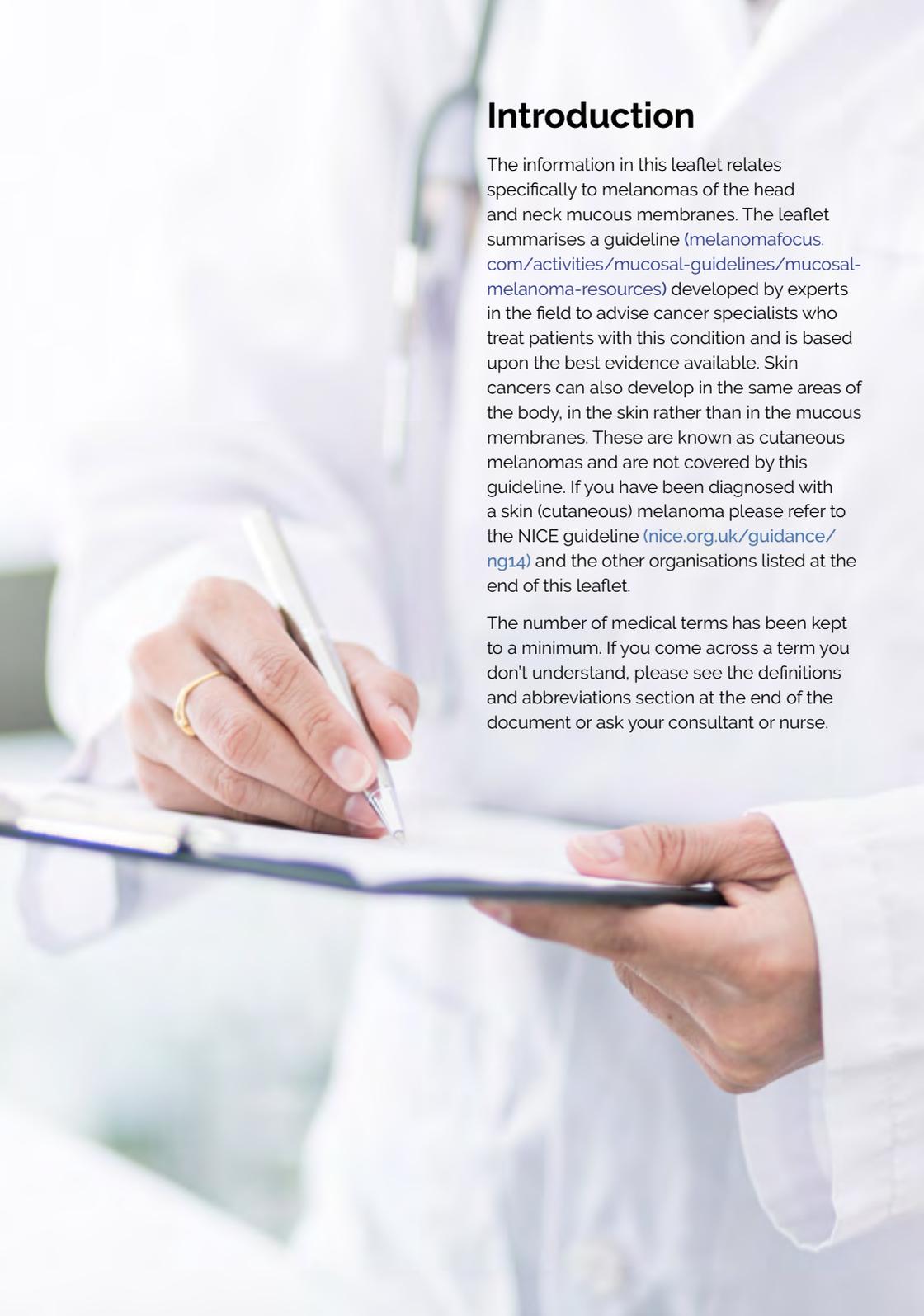


Head and Neck Mucosal Melanoma

Information for patients
and carers



A close-up photograph of a doctor in a white lab coat, wearing glasses and a gold ring, writing on a clipboard with a silver pen. The background is blurred, showing a clinical setting.

Introduction

The information in this leaflet relates specifically to melanomas of the head and neck mucous membranes. The leaflet summarises a guideline (melanomafocus.com/activities/mucosal-guidelines/mucosal-melanoma-resources) developed by experts in the field to advise cancer specialists who treat patients with this condition and is based upon the best evidence available. Skin cancers can also develop in the same areas of the body, in the skin rather than in the mucous membranes. These are known as cutaneous melanomas and are not covered by this guideline. If you have been diagnosed with a skin (cutaneous) melanoma please refer to the NICE guideline (nice.org.uk/guidance/ng14) and the other organisations listed at the end of this leaflet.

The number of medical terms has been kept to a minimum. If you come across a term you don't understand, please see the definitions and abbreviations section at the end of the document or ask your consultant or nurse.

Head and Neck Mucosal Melanoma

What is it?

Melanoma develops if there is uncontrolled growth of melanocytes, the cells responsible for pigmenting (darkening) the skin. Mucosal melanoma is a kind of melanoma that occurs in mucous membranes. These are the moist surfaces that line cavities within the body. Mucosal melanomas can occur in the mouth (oral mucosal melanoma), nasal passages (sinonasal mucosal melanoma) or very occasionally the throat (pharyngeal/laryngeal mucosal melanoma) as well as in other parts of the body including in the eye or around the genital and rectal areas. This leaflet discusses mucosal melanomas of the head and neck. There are other guideline documents by Melanoma Focus which cover eye melanomas (melanomafocus.com/activities/um-guidelines-resources) and ano-uro-genital mucosal melanomas (melanomafocus.com/activities/mucosal-guidelines/mucosal-melanoma-resources).

Mucosal melanomas are very rare: in the Surveillance Epidemiology and End Results (SEER) database which registered melanomas over 22 years, only about 1 melanoma in 100 was a mucosal melanoma. The majority of melanomas grow within the skin. Mucosal melanomas can be more complicated to treat than skin melanomas as they are often diagnosed later. This is because they are in less visible places and because they are often not pigmented (darker), which makes them even more difficult to spot.

These melanomas are different in several ways from skin melanomas. For example, while the risk of getting skin melanoma is increased by too much exposure to the sun, there appears to be no link between sunlight and mucosal melanomas. No specific causes or links with lifestyle have been found for mucosal melanoma and as far as we know there is nothing you can do to prevent it. It also appears that mucosal melanoma tumours are different from skin melanoma tumours. This means that some treatments for skin melanomas may not be as effective for mucosal melanoma and the outlook (prognosis) for mucosal melanomas is not as good as for skin melanomas. The prognosis will be different depending on whether the tumour can be fully removed surgically and whether it has spread. If you would like more information about your individual situation, you should discuss this with your clinical team.

What are the symptoms?

The most common places to have a mucosal melanoma of the head and neck region are in the nose, including the sinuses, or the mouth. More uncommonly they occur in the larynx. As with skin melanomas, there may be an area which is pigmented (darker) but this is NOT always the case. The symptoms are those often associated with less serious conditions and include:

- Nosebleeds out of one nostril only that have occurred several times over at least a 3-week period
- Blockage of one nostril, which nose drops haven't helped and which have occurred several times or for a long time over at least a 3-week period
- A lump on the tongue or in the mouth that may or may not bleed or be darkly pigmented that is growing bigger and has been there for at least 3 weeks
- A mouth ulcer which hasn't got better in 3 weeks
- Unexplained hoarseness which hasn't got better in 3 weeks
- Swollen glands in the neck which haven't got better in 3 weeks

If you have any of these symptoms and your GP cannot find another cause, you should be referred quickly (normally within about two weeks) to a specialist head and neck clinic with experience of this condition, and usually to a head and neck surgeon, for tests and further evaluation. It is most important that the team you see regularly treats patients with this condition. As it is so rare, you may need to travel to a specialist centre rather than attend your local hospital.

Going to the hospital

Tests and examination

At the hospital the consultant will examine you, including the lump, and your neck. You should also have a CT or MRI scan and a biopsy where they take a small sample of the lump to examine under a microscope.

If the consultant makes a diagnosis of head and neck mucosal melanoma, you would normally have a PET-CT scan of your body to check whether the cancer has spread. There are several specific genes which have been identified that may be involved in some, but not all, mucosal melanoma tumours. The pathologist should test the tissue sample from the biopsy to see if one of these identified genes is involved in your cancer as this will help decide the best treatment for you, now and in the future.



Diagnosis, staging and prognosis

The clinical team will make a diagnosis based on the results of your tests and medical examination. If you are diagnosed with a mucosal melanoma, your cancer will be evaluated according to a process known as *staging*. The stage indicates whether the cancer has spread in your body and, if so, where. This staging process will help you and the cancer team make decisions about your future treatment.

For the staging, the UICC (Union for International Cancer Control: www.uicc.org) **TNM** staging system is used. In addition to other information the team should record in particular:

- Location of the primary (original) tumour
- Primary Tumour size and extent of tumours
- Lymph Node involvement (whether or not the cancer has spread to the nearby lymph nodes (also called glands))
- Presence or absence of distant **Metastasis** (whether or not the cancer has spread to distant areas of the body).

The cancer team will make a prediction of the outcome of your cancer – your prognosis – based on the results of your tests and examination and on what happened to others who have had similar cancers.

If you and/or a relative or carer want to have a full discussion about your prognosis, make this clear to someone from the cancer team. It will mean that your appointments will be more informative and will enable you to be more involved in planning your treatment and other aspects of your life. On the other hand you may wish to take it 'one step at a time'. Whatever your decision, you will be fully supported by the cancer team.

Questions you may wish to ask about your diagnosis and prognosis

What are you looking for in the biopsies?

What is the difference between a CT scan, MRI scan and a PET scan – what do each show?

What stage is my cancer at and what is my prognosis?

Are any gene mutations involved in my cancer?

What is the best treatment available to me?

What sort of health problems might I have? And what are the plans I need to put in place to be prepared?

What supportive services are available to me and may I have their contact details for future reference?

Can you recommend leaflets or websites with information on my condition?

Are there any clinical trials available that I might be suitable for?

Questions you may wish to ask about your visits

How often will I have appointments at the cancer centre?

What sort of support will I need at these appointments?

How long will I be at the centre each time?

What further signs and symptoms should I look out for?

What symptoms should I report urgently?

How and who should I contact if I need to do so urgently?

- How to make an appointment with the consultant quickly should you run into problems
- About referral to support services (for example, supportive care services and support groups) should you need them at any point in your treatment
- Written information

Treatment

The first line of treatment for a mucosal melanoma is surgery, which should be done by a surgeon who is very experienced in complex head and neck surgery and is based in a centre where there is support from an experienced skin cancer team.

If the cancer hasn't spread beyond the primary site, it is likely that the tumour will be removed by surgery. If the tumour is in your nose or throat it is more difficult to access than a mouth cancer, and the operation should, if possible, be done endoscopically ('key-hole surgery') to minimise any problems. If possible, the surgeon will biopsy the lymph nodes near the tumour. In any case, the aim of surgery is also to remove a little of the surrounding healthy tissue to ensure that all of the cancer is removed. This is called having 'clear margins'. In planning the operation, the consultant should discuss your current health and how the surgery may affect your quality of life. If it is likely to have an impact, the consultant should discuss other treatment options with you to agree what is best for you.

If the cancer hasn't spread and the margins are clear, this may be the end of your treatment (see Rehabilitation, page 5) or, depending on the stage and tumour, you may be offered adjuvant therapy (see below).

Some questions you may wish to ask about your treatment

How should I expect to feel after my surgery?

What will the recovery time be?

Will I be able to carry on with life as normal?

What will be the effects of my surgery?

What symptoms should I look out for, indicating whether the surgery has worked or not?

What support will I need after surgery?

Am I likely to need rehabilitation?

Will I need adjuvant therapy? If so, what is the reason for this and what will it involve?

What are the side-effects of adjuvant treatment that I may experience?

Adjuvant (Extra) therapy

You are likely to be offered adjuvant (extra) therapy to reduce the likelihood that the cancer comes back after it has been removed entirely by surgery. (A 'belt and braces' approach!). Recently there have been advances in the treatment of skin melanomas with immunotherapy. Immunotherapy stimulates your body to fight the cancer. Immunotherapy doesn't work for everyone and there can be potentially severe, but manageable, side effects.

However, because mucosal melanomas are so rare, scientists are still uncertain whether these treatments work in the same way as with skin melanomas. You may be asked if you would like to enter a clinical trial to help doctors answer this question. As doctors learn more, you may be offered immunotherapy or other targeted therapies. (See definitions below for more information.)

Similarly, you may be offered adjuvant radiotherapy if there is a very high risk of local recurrence in your case but there is no evidence to show that it benefits everyone.

Rehabilitation

If the surgery has affected your teeth or eyes or your appearance, you should be referred to the appropriate services. You should be offered a referral to special psychological services before and after surgery to talk through your condition and your emotions, should you wish.

What to expect from your care

A specific specialist cancer team will look after you during your treatment. As this is a rare cancer which has similarities to both skin and other head & neck cancers, your care needs the expertise of both the skin and the head & neck oncologists. The guideline advises teams on specific ways in which they should work together.

When discussing your diagnosis and treatment you should be told:

- The name and contact details of your consultant and of your cancer clinical nurse specialist
- Who your key worker is (this person is your first point of contact should you have questions or problems and is usually the cancer clinical nurse specialist. You also need to make sure you are given the contact details of the person to contact if your keyworker isn't available)

After treatment

After your initial treatment, you should be offered regular follow-up appointments at the hospital to check that no cancer has returned, either to the original site or elsewhere in your body.

You should be given information on how to make an appointment quickly at any time after your treatment if you are concerned.

The schedule recommended in the guideline if you stay well is:

YEAR 1

Examination by the doctor: every 6-8 weeks
Scan to make sure the cancer hasn't spread: every 3 months
Brain scan: every 6 months*

YEARS 2-3

Examination by the doctor: every 3 months
Scan to make sure the cancer hasn't spread: every 6 months
Brain scan: every 6 months*

YEARS 4-5

Examination by the doctor: every 6 months
Scan to make sure the cancer hasn't spread: every 12 months
Brain scan: every 12 months*

After 5 years the consultant may see you once a year or discharge you, since the risk of the cancer returning reduces with time. But you should have rapid access to advice if you are concerned.

*You may be offered an MRI scan of your sinuses at the same time as the brain scan.



If you would like to read more about the advantages and disadvantages of having scans, refer to the NICE option grid set out in skin cancer guideline: [nice.org.uk/guidance/ng14/resources/followup-with-regular-ct-scans-yes-or-no-pdf-250598416](https://www.nice.org.uk/guidance/ng14/resources/followup-with-regular-ct-scans-yes-or-no-pdf-250598416).

You should be told how to make an appointment quickly at the clinic if you are concerned at any time in the future.

Questions you may wish to ask about your treatment and prognosis

Is there a chemotherapy, targeted therapy or radiotherapy available in my situation?

If so, what is the likelihood that it will reduce my symptoms and/or extend my life?

Is it available on the NHS?

Are there side-effects with this treatment and, if so, what are they?

What clinical trials are available?

What is the aim of the clinical trial in question?

What support will I need while on this treatment?

How will I be able to tell whether it's working or not?

If it is not working, how soon can I move on to something else?

Will this treatment prolong my life?

Will I have a reasonable quality of life while on this treatment?

Are surgery or radiation therapy options for me?

If your cancer recurs or spreads (metastasis)

If the cancer recurs (returns near the same spot as the original cancer) or it has spread elsewhere you may be offered systemic therapy similar to what you may have had before (see Adjuvant (extra) therapy, above). As described above, new treatments are constantly being developed and your consultant should tell you about their success rates, as well as the risks and benefits in your particular case. The cancer team should discuss the options with you and the availability of any clinical trials you could enter. There is little evidence that the routine use of chemotherapy or bio-chemotherapy is effective in the treatment of metastatic mucosal melanoma and it is not currently recommended by the guideline.

Depending on numerous factors relating to your tumour, you may be offered surgery or radiation therapy. Your consultant will discuss these types of treatment with you.

Following treatment for a recurrence or metastasis, you should be offered regular appointments as detailed on page 6.

If the disease progresses, there are other options to make you more comfortable. Refer the NICE guideline Cancer of the upper aerodigestive tract: assessment and management in people aged 16 and over (NG36) – [nice.org.uk/guidance/ng36](https://www.nice.org.uk/guidance/ng36) for further information on managing symptoms and to Palliative and supportive care in head and neck cancer – ([cambridge.org/core/journals/journal-of-laryngology-and-otology/article/palliative-and-supportive-care-in-head-and-neck-cancer-united-kingdom-national-multidisciplinary-guidelines/543AF37F96D5A88E8123EA86320F792F/core-reader](https://www.cambridge.org/core/journals/journal-of-laryngology-and-otology/article/palliative-and-supportive-care-in-head-and-neck-cancer-united-kingdom-national-multidisciplinary-guidelines/543AF37F96D5A88E8123EA86320F792F/core-reader)).

Sources of information and support

Unfortunately, as mucosal melanoma is so rare, there are no websites dedicated to information about this condition.

Melanoma Focus (melanomafocus.com)

is a charity that commissions and funds melanoma research, while providing support & information for patients, carers and healthcare professionals. It funded this leaflet and the associated guideline for health professionals, on which this leaflet is based. It is here *****

Most available information is not specific to mucosal melanomas, but much of the general information on cancers of the head and neck is informative and useful.

British Association of Head & Neck

Oncologists has a patient information area – bahno.org.uk/patients_area/patient_links.aspx.

Cancer Research UK has a great deal of information, including treatments and living with cancer, which is relevant to head and neck cancers.

Mouth cancers:

cancerresearchuk.org/about-cancer/mouth-cancer

Sino-nasal cancers:

cancerresearchuk.org/about-cancer/nasal-sinus-cancer

Nasopharyngeal cancers:

cancerresearchuk.org/about-cancer/nasopharyngeal-cancer

Macmillan (macmillan.org.uk) provides support for people who have cancer.

NHS Choices (nhs.uk/pages/home.aspx) has information on cancer treatments and other aspects of care.

The American Joint Committee on Cancer (AJCC), which publishes the staging manual in use has a patient leaflet explaining staging here: cancerstaging.org/CSE/Registrar/Documents/needtoknow.pdf.

Definitions and abbreviations

Adjuvant therapy

Adjuvant therapy is treatment in addition to that used to treat the initial cancer. The aim is to reduce the possibility that the cancer will come back after it has been removed entirely by surgery.

Biopsy techniques

A biopsy is when a piece of the abnormal tissue is removed so the cells can be examined under a microscope to determine whether it is cancer and if so what type.

There are many ways of taking a biopsy. The ones mentioned in this leaflet are listed here.

Sino-nasal MM about-cancer.cancerresearchuk.org/about-cancer/nasal-sinus-cancer/getting-diagnosed/tests-diagnose

Oral – MM about-cancer.cancerresearchuk.org/about-cancer/mouth-cancer/getting-diagnosed/tests-diagnose

See NHS Choices, from which this information was taken, for more detail nhs.uk/conditions/biopsy/.

CORE BIOPSY

For a core biopsy, the area is numbed with a local anaesthetic, a wide hollow needle is inserted and a sample of the cells taken.

EXCISION BIOPSY

An excision biopsy aims to remove the lesion or lump entirely. It will be examined to determine whether more treatment is needed.

FINE NEEDLE ASPIRATION

A fine-needle aspiration (FNA) biopsy is similar to a core biopsy but isn't as deep and may not need a local anaesthetic. When the needle is in position, it will cut out a small sample of tissue.

SENTINEL NODE BIOPSY

Sentinel node biopsy is a test to determine whether there are cancer cells in the lymph nodes around your groin. The consultant will carry out a small operation to remove some tissue to be tested. There is more information in the links above.

Immunotherapy

Also called biologic therapy or bio-chemotherapy, stimulates the patient's own immune system to fight cancer. There is very helpful information here: about-cancer.cancerresearchuk.org/about-cancer/cancer-in-general/treatment/immunotherapy/what-is-immunotherapy and cancerresearchuk.org/immunotherapy/what-is-immunotherapy.

Lesion

An area of tissue that isn't normal. This term can be used when it is uncertain whether the area is cancer (malignant) or not (benign).

Lymph Nodes

Lymph nodes are often the first place the cancer spreads to. Fuller explanation of lymph nodes and cancer is here: cancer.org/cancer/cancer-basics/lymph-nodes-and-cancer.

Metastasis/Metastatic Cancer

Metastasis is when cancer cells break away from the original (primary) site, travel through the blood or lymph system, and form a new tumour in other organs or tissues of the body. The new, metastatic tumour is the same type of cancer as the primary tumour. For example, if mucosal melanoma spreads to the liver, the cancer cells in the liver are mucosal melanoma cancer cells, not liver cancer cells.

Palliative care support services

More information on what forms of home care are available can be found at: nhs.uk/conditions/social-care-and-support/home-care and cancerresearchuk.org/about-cancer/melanoma/advanced-melanoma/support-home-for-you-your-family.

Pigmentation

Pigmentation is colouring of the skin which can be different shades of brown. Cancerous cells may start producing more pigment, resulting in dark patches. However, some mucosal melanomas are not pigmented.

Scans

For more information on the tests mentioned in this leaflet see the Cancer Research UK website link:

Sino-nasal MM about-cancer.cancerresearchuk.org/about-cancer/nasal-sinus-cancer/getting-diagnosed/tests-diagnose

Oral – MM about-cancer.cancerresearchuk.org/about-cancer/mouth-cancer/getting-diagnosed/tests-diagnose

CT SCAN

Computerised Tomography (CT) takes x-rays from different angles and the computer then puts them together as a three-dimensional picture. See the Cancer Research UK website link above and nhs.uk/conditions/CT-scan for more information.

MRI/MR

Magnetic Resonance Imaging (MRI or MR) takes pictures of the body using magnets and radio waves. These show up soft tissues such as the bowel, liver, lungs etc. better than CT scans. The scan can take up to 1½ hours. See the Cancer Research UK website link above and nhs.uk/conditions/mri-scan for more information.

PET-CT

This combines a CT Scan (see above) with a Positron Emission Tomography (PET) scan. A mildly radioactive substance is injected, which shows up parts of the body where cells are more active. That is, where a cancer might be growing. See the Cancer Research UK website link above and nhs.uk/conditions/pet-scan for more information.

Staging

Staging is an estimate of how much cancer there is and where is located. Staging is used to plan treatment and future options. Fuller explanation here: about-cancer.cancerresearchuk.org/about-cancer/nasal-sinus-cancer/stages-grades/about and about-cancer.cancerresearchuk.org/about-cancer/mouth-cancer/stages-types-grades.

Targeted therapy

Targeted therapy, like chemotherapy uses drugs to treat cancer. There is some very good information here: about-cancer.cancerresearchuk.org/about-cancer/cancer-in-general/treatment/targeted-cancer-drugs/what-are-targeted-cancer-drugs.

ABBREVIATIONS

- CNS** Cancer Clinical Nurse Specialist
- CT** Computed tomography
- MRI** Magnetic resonance imaging
- PET** Positron emission tomography
- SLNB** Sentinel lymph node biopsy



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