

Diagnosis of soft tissue sarcomas in children and young people

A summary for healthcare professionals



Consider a soft tissue sarcoma in any child or young person presenting with:

- Unexplained lump
- Swelling
- Pain
- Skin changes over the lump/swelling

Associated symptoms:

- Limbs: lump, limp, restricted movement or altered function
- Head and neck: localised eye swelling, cranial nerve deficit, impaired vision, ear swelling or pain
- Abdomen and pelvis: urinary symptoms, abdominal /flank distension, genital lump/swelling, vaginal bleeding, change in bowel habit, obstructive jaundice
- Systemic symptoms: weight loss, fever, malaise
- Bruising/bleeding, especially in CYP aged under one year
- Back pain (neck to sacrum) with or without signs of spinal cord compression

Ask about predisposing factors

- Neurofibromatosis 1
- Li-Fraumeni syndrome
- DICER1
- Family history of breast or ovarian cancer at a young age
- A sibling with childhood cancer

Unexplained lump

- Soft tissue sarcomas can occur anywhere in or on the body and present as a lump.
- They tend to be fixed and hard on palpation.
- They can be associated with pain, but pain is not always present.
- Sometimes they can have overlying changes in the skin.
- Ask about the presence of the other symptoms of a soft tissue sarcoma (swelling, pain, skin changes, fever, weight loss, back pain, bowel/bladder/erectile dysfunction, birth marks e.g café au lait spots) in a CYP presenting with an unexplained lump.
- Depending on where the lump is, ask about the presence of other associated symptoms including head and neck symptoms (localised eye swelling, cranial nerve deficit, impaired vision, ear pain and swelling or hearing loss), respiratory symptoms (new onset of swallowing difficulties / choking, stridor or snoring) and abdomen/pelvis symptoms (urinary symptoms, abdominal distension, genital mass, vaginal bleeding).

Imaging required for:

- Unexplained persistent* and growing lump
- Unexplained lump with another symptom from the symptom checklist.

**persistent = present for two weeks or more*

(In primary care, requesting an ultrasound should not delay referral/discussion with paediatrics)

Diagnostic pitfalls:

- Attributing lump to an injury without a clear history
- Attributing lumps to lipomas or haemangiomas without typical appearance. If there is diagnostic uncertainty and no improvement, always discuss with a paediatrician or a paediatric surgeon for a second opinion or review.
- Misdiagnosis of a benign mass on imaging, leading to an incorrect surgical procedure. Imaging should be reported by paediatric radiologists, and any diagnostic uncertainty should be discussed at the appropriate paediatric oncology MDT prior to surgery.

Swelling

- Swelling in soft tissue sarcoma can occur on any part of the body.
- Ask about the presence of the other symptoms of a soft tissue sarcoma (lump, pain, skin changes, birth marks (café au lait spots) fever, weight loss, back pain and bowel/bladder/erectile dysfunction).
- Ask about the presence of head and neck symptoms (localised eye swelling, cranial nerve deficit, impaired vision, ear pain or swelling or localised hearing loss) if the lump is in the head/neck area.

Imaging required for:

- Any persistent swelling* rapidly increasing in size, including an eye or ear swelling.
- Any persistent swelling* not resolving despite treatment with regular anti-inflammatories OR antibiotics.

**persistent = present for two weeks or more*

(In primary care, requesting an ultrasound should not delay referral/discussion with paediatrics.)

Diagnostic pitfalls:

- Attributing a red warm swelling to infection despite no improvement with antibiotics. Arrange to see the CYP at the end of the course of antibiotics to assess response. If there has been no response, consider discussion with secondary care for advice or referral.

Assess these children with:

History:

- Associated symptoms
- Any predisposing factors
- Family history

Examination of:

- Lump and regional lymph node
- Skin (birth marks e.g. café-au-lait spots)
- Neurological examination
- Age-appropriate weight and growth

Assessment pitfalls

- Symptoms are usually progressive and can cause other seemingly unrelated symptoms due to pressure on other structures, such as respiratory distress, spinal cord compression or bowel obstruction.
- In primary care, requesting an ultrasound should not delay referral/discussion with paediatrics.

Referral from primary care

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- Discuss all suspected cases with paediatrics via telephone to advise best route to be seen.

Imaging:

- Be aware that ultrasound imaging is not always recommended for a soft tissue sarcoma.
- In secondary care, discuss with paediatric radiologist to decide most suitable imaging.

Pain

- In soft tissue sarcomas, pain is often associated with the lump or swelling.
- Depending on where the lump or swelling is, pain can be neuropathic.
- Be aware of referred pain.
- Be aware that children aged younger than four years, or those with communication difficulties, are frequently unable to describe or localise pain; their behaviour, e.g. withdrawal, holding their leg, not weight bearing, may indicate bone pain. Look for these signs on examination.
- Ask and examine for the other signs and symptoms suggestive of a soft tissue sarcoma (lump, swelling, skin changes, birth marks (café au lait), fever, weight loss, back pain and bowel/bladder/erectile dysfunction if back pain is present) in CYP with pain.

Imaging required for:

- A painful, persistent* or progressive lump or swelling.

**persistent = present for two weeks or more*

(In primary care, requesting an ultrasound should not delay referral/discussion with paediatrics)

Diagnostic pitfalls:

- Not performing a full examination for those presenting with pain that may be referred.

Skin changes over the lump/swelling

- Skin changes can include erythema, thinning of the overlying skin, ulceration, hyperpigmentation, fungating lesion.
- In under 1s, there is risk of consumptive coagulopathy which can cause petechiae, bruising and bleeding.
- Be aware that these skin changes can look different across different skin tones.
- Ask about the presence of the other symptoms of a soft tissue sarcoma in a CYP presenting with skin changes over a lump or swelling pain (fever, weight loss, back pain and bowel/bladder/erectile dysfunction if back pain is present).

Imaging required for:

- A CYP who has a persisting * or progressive lump/swelling with overlying skin changes.

**persistent = present for two weeks or more*

(In primary care, requesting an ultrasound should not delay referral/discussion with paediatrics.)

Diagnostic pitfalls:

- Be aware that soft tissue sarcomas in under 1s have a risk of consumptive coagulopathy (Kasabach-Merritt syndrome). A full blood count is recommended in children under one with a suspected soft tissue sarcoma, especially if presenting with bruising and petechiae.
- Be aware of differences in skin presentations across skin tones.