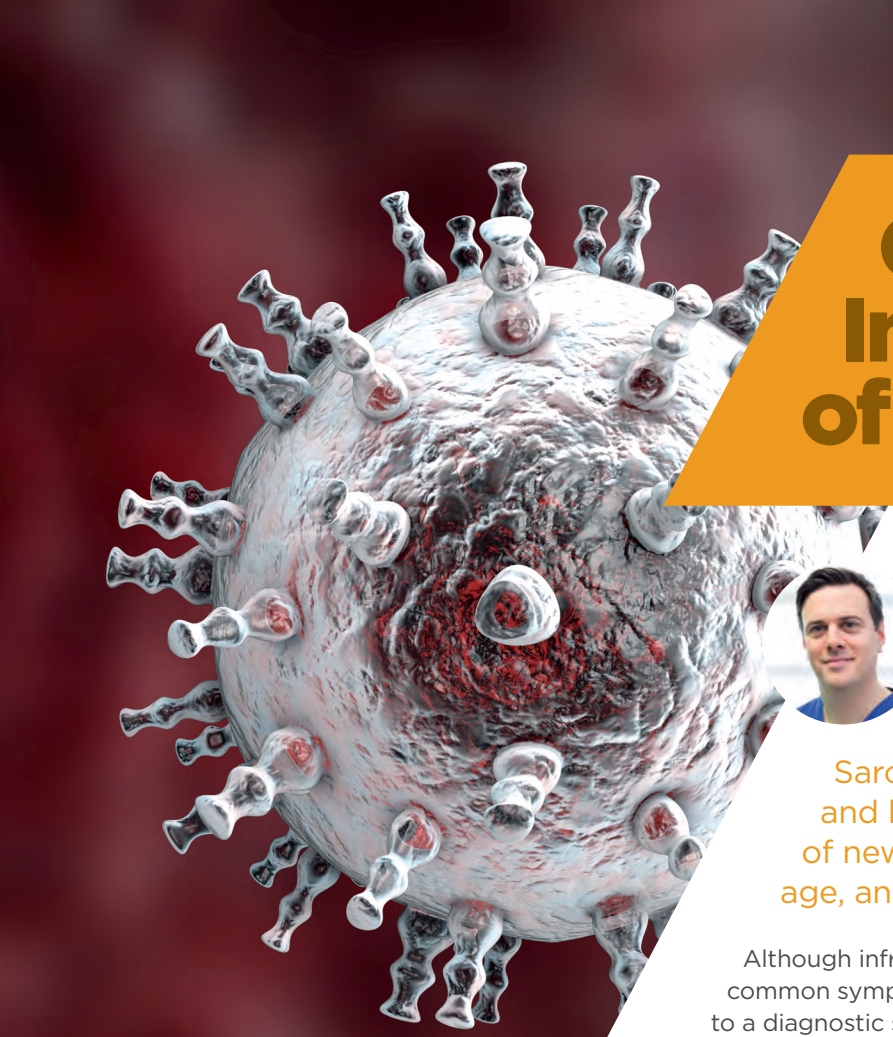

Contemporary Investigation of a Lump

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Sarcomas are rare tumour of soft tissue and bone, which account for about 1% of new cancers, and can occur at any age, and any part of the body.

Although infrequent, soft tissue sarcomas present with common symptoms, and in the UK, referral for ultrasound or to a diagnostic service is recommended if a lump is found with any of the following features:

• Rapid growth • Deep to the skin • Painful • Recurrent lump after previous excision • Larger than 5cms (bigger than a golf ball)

Describing the symptoms and signs of soft tissue sarcoma, led to the development of the Sarcoma UK "On The Ball" campaign which aims to raise awareness of the symptoms of soft tissue sarcoma. This is important as more than half of people in the UK have not heard of sarcoma, and due to its rarity, physicians may not immediately identify a lump as a potential cancer. Early recognition may reduce time to treatment, and catch a sarcoma at an earlier stage.

Investigation is straightforward, with clinical history and examination, ultrasound or MRI, and a core-needle biopsy. Ideally these investigations should occur in the context of a “one-stop” clinic.

Fortunately, only around 10% of patients presenting with these clinical features will have a sarcoma. It is important to note that core biopsy does not increase the risk of recurrence, but it is generally regarded as good practice to perform the biopsy within a predicted future incision, so that the track can be resected at definitive surgery.

A significant proportion of referred cases require discussion at a multidisciplinary tumour board meeting, and other cancers, such as melanoma and haematological malignancies may present as an apparently innocuous lump. However, in the majority of cases, one can be reassured that a lump is not malignant post ultrasound, and proceed with excision if the lesion is symptomatic. Without an ultrasound, it can often be difficult to differentiate a benign from malignant soft tissue lump in lesions >5cm.

There are also a wide range of indeterminate soft tissue masses some of which may have the potential to recur if inadequately excised (e.g. myofibromas) or depending on grade, have a small risk of systemic spread (e.g. granular cell tumours). Others, such as fibromatosis, or nodular fasciitis have the potential to regress. Some benign lesions such as myxomas, intramuscular lipomas and Schwannoma’s (benign neurogenic tumours) benefit from the technical expertise developed by sarcoma surgeons.

The most recent WHO classification has identified over 100 subtypes of sarcoma, with behaviour ranging from indolent (e.g. a well differentiated liposarcoma) to aggressive (high grade undifferentiated sarcoma). Most are pragmatically grouped into low and high grade tumours based on differentiation, mitosis and necrosis, and larger higher grade tumours have the highest propensity for metastatic disease.

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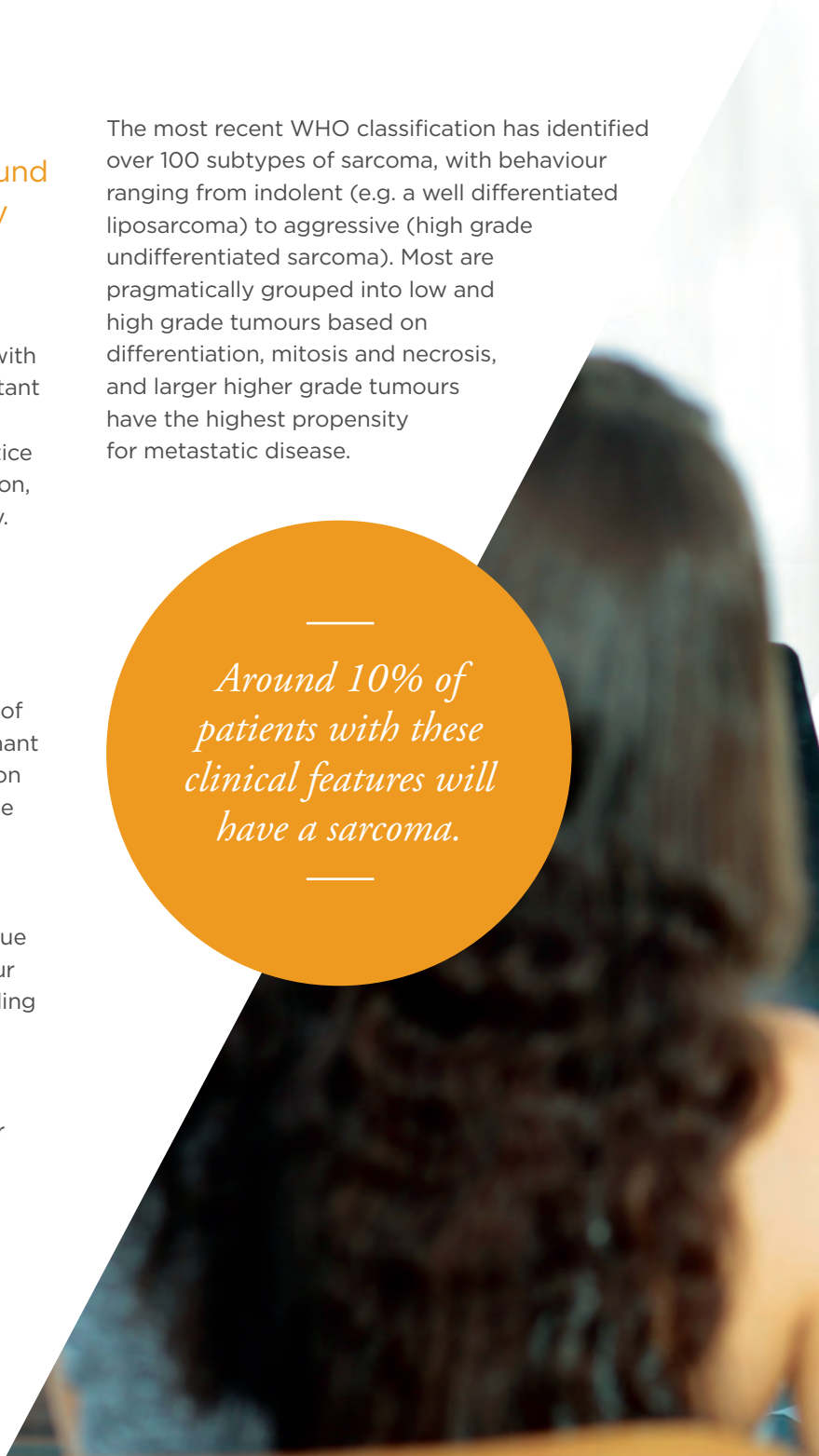

The lungs are the most common site of systemic spread, with almost no lymph node spread, with some specific, rare exceptions (such as clear cell sarcoma) proving the rule. Surgery is the only potentially curative approach, with radiation additive for local control.

Unfortunately adjuvant trials of chemotherapy have not had an impact upon long-term survival, but chemotherapy has an important role in prolonging life in those developing metastatic disease.

Definitive treatment of sarcoma usually requires both surgery and radiotherapy, with physiotherapy extremely important for rehabilitation. The aim of sarcoma resection is to clear the tumour with as much functional preservation as possible. In practice this means a wide excision or muscle compartment resection, with radiation being added if the tumour is large (>5cm), deep and high grade, or if adjacent to a crucial structure such as major vessel or nerve that requires preservation, to reduce the risk of local recurrence.

Plastic surgical reconstruction may be required, in particular for sarcomas arising in the skin or subcutaneous tissue, but thankfully, the need for amputation is minimal with contemporary treatment. Expertise in sarcoma and soft tissue surgery is essential in treatment of these rare tumours, but modern radiotherapy has been the most important advance in adjuvant treatment, enabling the development of limb preserving surgery. The paradigm of contemporary treatment of extremity sarcoma is analogous to the development of breast cancer treatment, with a de-escalation of treatment from amputation to wide-local excision and radiotherapy when indicated. Functional and oncological outcomes are generally good, and survival can be accurately estimated using the Sarculator App (http://www.sarculator.com), based on nomograms developed from leading international sarcoma centres.

It is important to know that the inadvertent excision of a soft tissue cancer may compromise care in two ways.

Firstly, a primary excision without regard to further surgery may require a larger wound defect to definitively clear the affected area.

Secondly, many institutional series have shown inferior cure rates after inadvertent excision. The recurrence rates are up to 3 times higher for sarcomas inadvertently excised when compared to matched primary resections.

The best time to cure a soft tissue sarcoma is with the first intervention – data recently presented by the French NETSARC group at the European Society of Medical Oncology demonstrated that patients who had surgery for sarcoma after discussion at an MDT meeting had a significantly improved rates of adequate resection and local recurrence in a review of over 26,000 cases.

In summary, if a patient presents with a large lump that is bigger that 5cm, deep, painful or recurrent after a previous excision, consider soft tissue sarcoma as a differential diagnosis, and think about referral for ultrasound or direct referral to a soft tissue specialist for assessment.

Outcomes for soft tissue sarcoma are generally good, but initial inadvertent excision compromises local control and survival. Imaging is reassuring in the majority of cases, a significant proportion will benefit from investigation and treatment by a soft tissue specialist, with a small minority requiring treatment for sarcoma, with a generally positive prognosis.

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