

**TITLE:** Sarcoma – what's in a name?

**AUTHOR:** Prof C. Fisher, Professor of Tumour Pathology

**CENTRE:** Royal Marsden Hospital, London UK

**ABSTRACT:**

Sarcoma is not one disease; the term encompasses many types of mesenchymal tumour. They can be classified by behaviour, by cell size and shape or stromal changes, or by differentiation, i.e. the type of mesenchymal tissue they form. Most sarcomas form only one type of tissue, and they can readily be diagnosed by light microscopy or with the help of adjunctive techniques. Some sarcomas show non-mesenchymal differentiation and many of these have been given erroneous names based on incorrect assumptions of differentiation, or on supposed resemblance to normal tissue or to other tumour types. Examples include malignant fibrous histiocytoma, first described as a histiocytic tumor, and later proved to be fibroblastic and myofibroblastic, and also synovial sarcoma which neither arises from nor forms synovium but which expresses epithelial differentiation as a result of a consistent balanced reciprocal translocation.

Many additional types of sarcoma have been found to have specific translocations. The 2002 WHO classification, a consensus document, incorporates pathology and genetic data and is based on lineage differentiation. Several tumour types, however, remain of uncertain differentiation including many of those with inapt names. A new entity, PEComa, a tumour of perivascular epithelioid cells (for which no normal counterpart cell is known) has been accepted. This subsumes several other tumour types and is a rare example of 'lumping' rather than 'splitting'.

The WHO classification also refines the terms benign and malignant for soft tissue tumours and introduces an intermediate behavioural category, including locally aggressive and rarely metastasizing tumours. An example of the former is atypical lipomatous tumour/well differentiated liposarcoma which has identical morphology and genetics wherever it occurs and which does not metastasize but can dedifferentiate, i.e develop a high grade undifferentiated component that behaves aggressively. Dedifferentiation is rare in the superficial soft tissues, but more frequent in retroperitoneum or mediastinum, allowing for the use of two different terms according to location of the tumour

Names need not be correct but should be used consistently so that the same thing is understood by clinicians and pathologists. A name need not be changed until the true nature of its tumour is clear; once entrenched, however, it might not be changeable.