Role of expert centres in the management of sarcomas

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1. Introduction

Sarcomas are rare tumours of the connective tissue which may resemble a variety of tissues – such as muscle, nerve and bone – although many sarcomas have no normal tissue counterpart. The annual incidence of soft-tissue sarcomas (STSs) in England and Wales between 1990 and 2007 was 2300, which equates to about 40 per million per annum. Bone sarcomas are significantly less common, representing only 0.2% of all malignancies. Treatment within specialised multidisciplinary teams (MDTs) is crucial since a body of expertise in all areas of diagnosis and treatment is required to manage them appropriately. Studies have shown that conformity to approved treatment guidelines is improved when patients are treated by an MDT in a reference centre [1].

2. Diagnosis – histopathology, radiology

The risk of a tumour being metastatic at diagnosis, and of subsequent death, is directly related to tumour size [2]. Earlier diagnosis could have a huge impact, and guidelines are now in place in the UK to encourage early referral of suspicious lumps (or X-rays in the case of bone tumours).

Once a tumour is suspected, the two key diagnostic tools are radiology and histopathology. The initial assessment of suspicious lumps will be by physical examination and probably ultrasound, followed by core needle biopsy. Core needle biopsy has an accuracy of >90% as well as the ability to distinguish high-grade from low-grade lesions and in most cases the specific sarcoma subtype [3].

Cross-sectional imaging is required prior to surgery, in order to plan treatment and for staging. This is usually in the form of magnetic resonance imaging (MRI) for the primary disease site and computed tomography (CT) for staging purposes. It is common for the diagnosis of patients referred with a diagnosis of sarcoma to be revised to another subtype, another disease, or even a benign condition [4]. Reported discrepancy rates between referring and expert pathologists are generally in the order of 25%, with a benign to malignant discrepancy of 5%.

3. Sarcoma surgery

The primary management of most sarcomas is surgical excision. Unplanned operations, performed on the assumption that the "lump" is benign, can make the eradication of disease much more difficult. A study demonstrated that patients who had unplanned surgery had a much higher local recurrence rate and poorer long-term disease control, in spite of definitive surgery and radiotherapy [5]. All sarcoma operations should be performed in specialised centres in order to ensure optimum outcomes. For retroperitoneal surgery, where multivisceral resections are common, guidance is available [6]. The NICE (National Institute for Health and Care Excellence) Improving Outcomes Guidance (IOG) for people with sarcoma recommended that specialised centres should treat a minimum of 100 STS a year and 50 in the case of bone sarcomas. The IOG, which also addresses wider issues concerning the sarcoma MDT, can be obtained using the following URL: http://guidance.nice.org.uk/CSG

4. Radiation oncology

Adjuvant radiotherapy improves the local control of high-grade extremity soft tissue sarcomas [7]. Research continues into the appropriate timing, dose and field size of adjuvant irradiation. The complexity of pre- and post-operative radiotherapy for sarcomas is such that specialised centres are best placed to offer the appropriate expertise, in the context of the MDT.

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5. Medical oncology

Chemotherapy for most sarcomas is palliative, but nevertheless valuable. Recent years have seen a significant increase in treatment options and tailoring of treatment to the individual disease subtype. The standard agents, doxorubicin and ifosfamide, remain useful, but other drugs are now in routine use, including gemcitabine plus docetaxel for leiomyosarcoma and pleomorphic sarcoma [8,9], trabectedin for leiomyosarcoma and liposarcoma [10] and paclitaxel for angiosarcoma [11]. The management of gastrointestinal stromal tumour (GIST) was transformed by the introduction of imatinib [12,13], and subsequently sunitinib [14]. More recently another tyrosine kinase inhibitor, pazopanib, has been licensed for treatment of STS [15]. Certain rarer diseases require special approaches: e.g. the use of rapamycin analogues for PEComa, imatinib for chordoma, tamoxifen for fibromatosis and aromatase inhibitors for endometrial stromal sarcoma.

6. Clinical trials and data collection

Clearly, for such a rare group of diseases it is essential that care be concentrated in specialised centres which can treat patients in appropriate clinical trials. These will not be available in smaller centres, putting patients at a disadvantage. The cumulative experience of the MDT together with the amalgamation of clinical and laboratory data also represent a major resource for research and the opportunity to use these data directly for the benefit of patients.

7. The wider multidisciplinary team

In addition to surgeons, radiation and medical oncologists, radiologists and histopathologists, the MDT will have clinical nurse specialists, physiotherapists, dieticians, palliative care physicians and site-specific specialists.

As described, the management of sarcomas is truly multidisciplinary, increasingly complex and, as more molecular targets are identified, more likely to be treated with highly specific targeted therapy. The need for specialised centres has been recognised in the UK, and a process, informed by the NICE IOG, is leading to the concentration of care in a limited number of centres. We hope that earlier diagnosis, fewer unplanned operations and better integrated care will lead to a significant improvement in outcomes, which have not changed over the last 20 years (http://www.ncin.org.uk/publications/data_briefings/soft_tissue_sarcoma). We can only hope to do better.

Conflict of interest statement

None declared.

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