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Editorial

Rare Cancers: How Should We Organize Expertise?

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Specialist medical journals such as *Clinical Oncology* and the *British Journal of Radiology* should include isolated case reports of rare cancers with a literature review. These reports are of educational value to all oncologists and usually provide a reasonably up to date review of the current literature pertaining to an unusual clinical problem. For example, this issue contains a report of the palliative treatment of a malignant melanoma of the cervix that developed probably as a late effect after primary radiotherapy to this region [1]. Other specialist oncology journals such as the *International Journal of Radiation Oncology Biology and Physics* and *Radiotherapy and Oncology*, will no longer publish such single case reports. In the latter case, this is presumably because of the enthusiasm of the editors for evidence-based medicine [2]. Such an attitude may be neglectful of the fact that some cancers are excessively rare and that larger series of patients cannot easily be collated.

There is a tension between the requirements of the common and the rare cancers, not only in literature reporting but also at a more practical level. The initial series of National Health Service reforms in cancer care have, quite correctly, focused on the common cancers. These have allowed for a welcomed recognition of leading cancer clinicians for designated tumour locations in each cancer centre. Such anatomical site specialization carries the responsibility for the development of evidence-based management protocols. These laudable advances will probably produce clear benefits in patient outcomes with time, at least in the case of the common tumours at each site, but what about the rare histological varieties of cancers or when more common histological types of cancer arise in unusual anatomical sites? Even at the larger cancer centres in the UK (and these are very large when compared with those in western Europe and North America), the numbers of patients that have certain rare cancers remain small. A site-specialized consultant may only see a few such patients over a 10-year period. For example, in my own 11-year experience of specializing in gynaecological and central nervous system (CNS) tumours at Clatterbridge Hospital, which served a population of approximately 3 million, I attended to only one patient with melanoma of the cervix, and two with melanoma of the vulva. Two patients had a didelphic uterine system with a carcinoma. In the CNS, two patients with choroid plexus carcinoma were seen and about ten with pineal tumours. This list could be tediously protracted with many further examples. Almost all of these rare tumours will remain unreported in the medical literature and the data are unused by any research organization at present. In addition, most specialists would agree that there can be significant problems in the management of patients with rare cancers. How does one seek further advice in these situations? At present, the following approaches are available:

1. *Perform an Internet literature search*. This will produce series of case reports or the accumulated experience of large centres over a prolonged period of time (sometimes over 20–30 years and using a range of techniques that may not now be considered to be appropriate). Some review articles may be available, but these may have been published several years previously and therefore not include more modern developments.

2. *Books*. In addition to the standard reference books and occasional monographs on rare tumour types, there is the excellent *Textbook of Uncommon Cancers* [4]. There should be a copy in every cancer centre. There is a recent new edition; hopefully it will be updated frequently.

3. *Contact another cancer centre* (in the UK or worldwide) where there is known to be a special interest in the problem. There are clear existing examples of specialists who are major authorities on certain rare cancers and also of some rare cancer databases, as is the case for rare gynaecological cancers occurring in the London hospitals, collected by Dr P. R. Blake at The Royal Marsden Hospital. In some areas on oncology, such as paediatrics and

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ophthalmology, there are already good systems for collecting such data.

Can we improve on this situation? The new structure of cancer specialization in the UK may allow a good opportunity for collaborative ventures between the site-specialized consultants. The creation of a national database of experts in specific rare cancers should now be encouraged. A country with a population of 60 million with designated cancer centres and reliable cancer registries should be able to organize such a list. Continuous collection of the data on all patients with rare tumours and updated management recommendations could be achieved by two or more designated site-specialized consultants (with interdisciplinary participation) over a 10-year period. Whereas it will not be possible to perform large randomized trials in such categories of patients, more information on treatment related outcomes would help to optimize therapy. It should not be forgotten that, where treatment outcome differences are large, smaller patient numbers can produce significant results in randomized controlled trials. For example, Bentzen [3] has calculated that only 10–20 patients are required for a 40% improvement in a two-arm study for a 5% statistical significance level with a power of 80%–90% if the control arm tumour control rate is 35% (see Fig. 2 in Bentzen [3]). Even in the absence of formal trials, variations in treatment (e.g. in time–dose–fractionation) within and between centres can be accounted for in terms of the biological equivalent dose (BED) and an appropriate BED for optimum control determined. There would also be scope for the development of recommended treatment protocols for rare tumours.

A structure of designated experts in specific rare cancers is highly desirable. This could be achieved in several ways, including:

1. Allowing individuals to proceed *ad hoc*. This *laissez-faire* approach would probably lead to duplication and not be sufficiently comprehensive.
2. Formal design of a comprehensive structure with named groups of designated experts working in interdisciplinary teams, with reporting deadlines etc.

The second option could be accomplished by a central co-ordinating system. This could be organized directly, either by the Department of Health or by combined cancer charities through the UKCCR and delegated to professional bodies such as the Royal College of Radiologists/Royal College of Physicians/College of Radiography/British Institute of Radiology as well as the surgical colleges. There are of course multidisciplinary associations, such as the British Gynaecological Cancer Society and the British Neuro-Oncology Association etc. that could also help in the process. The RCR would seem to be uniquely well disposed in this respect and could achieve the desired structure with carefully selected input from invited experts from the other disciplines. Some resource allocation may be necessary, possibly channelled through the existing audit budgets at cancer centres, although overall costs would not be expected to be high in comparison with some forms of cancer research. Eventually, all the necessary information about who to contact for each specific tumour should be available on a rare cancer web site, with regularly updated information on best current management. The latter could also be published.

A highly organized and well-functioning structure for gathering information on rare cancers in the UK could further evolve on a European basis. To offer a strong lead within Europe would provide a considerable boost in morale to the discipline of clinical oncology in the UK, although in the short term some rare tumours may be better treated abroad because of our present technical limitations [5].

It would be prudent if these matters could be discussed within the committee structures of the above organizations. A specific meeting on rare cancers and their future study should be encouraged. An open debate in the columns of this journal about the issues raised would also be welcomed.

References

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