Chairperson's Introduction

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The multi-disciplinary teams who look after gynaecological cancers have within their spectrum a large number of rare and uncommon cancers. This symposium will focus on a limited number but illustrates some of the challenges required to manage these cases. Of course, the rarity is a relative factor and for a gynaecological oncologist working in a large regional cancer centre, these cases may be seen more frequently. It would be hoped that the clinician working in a smaller district hospital would at the very least discuss the further management and hopefully refer on these unusual cases for centralised management. It is of course not simply the surgery or the follow up that may require specialist team approach but the imaging and histopathology with access to immunocytochemistry may be very important in helping to distinguish some of these rarer cancers.

Furthermore, it might be argued that it is only by the regional or supra-regional teams seeing a lot of these rare and uncommon cancers that they will develop the expertise to manage these patients better. Allied to this are important issues such as having a registry or database to record the incidence of these tumours. Perhaps if we can accurately correct the data, we may realise that some of these tumours may be more or less common than we expected and we can also look for any changes of trends of incidence. The Gynaecological Cancer Inter-Group (GCIG) had an optimistic aspiration to try and introduce a register/database and a series of guidelines which could be used as an educational resource to help those specialists especially in the smaller cancer centres. Issues of confidentiality and security regarding data transmission have frustrated the attempts to develop this register but nevertheless it is hoped that within each nation initiatives will be introduced to allow something similar.

In this issue we focus particularly on mucinous tumours of the ovary, small cell carcinoma of the cervix and ovary, and thirdly, sex cord stromal tumours of the ovary which include granulosa cell tumours. Whilst most ovarian cancers are of the epithelial types, it will be seen from Dr. van der Vijver's overview that there are many uncommon and rare subtypes. His review tries to help us clarify the classification. This excellent overview focuses initially on the common subtypes and a variety of the rarer types which have been addressed by the clinical papers within this section. The role of pathologists in this area is critical and they are essential members of the multi-disciplinary team.

Mucinous tumours of the ovary remain contentious and it is disputed whether they are indeed true ovarian epithelial tumours or whether they may be better considered as arising in the gastro-intestinal tract. It is important to distinguish a metastatic tumour as the conventional treatment would involve different chemotherapy. Rigorous attention to immunocytochemistry and careful pathological analysis may help to distinguish these tumours. Until now, treatment of primary mucinous ovarian tumours has focused around conventional regimes which are platinum or platinum/taxane based. However, reference is made to the forthcoming MEOC-1 study and GOG studies in which conventional chemotherapy will be compared with a schedule more associated with gastrointestinal tract origin. The next 5 years will hopefully help to clarify whether these are truly ovarian tumours or not.

In the article by Isabelle Ray-Coquard, the team from Lyon describe their approach to the management of the sex cord tumours. Their group have helped set up a French gynaecological cancer rare tumour registry and this article describes some of the pathways in managing these patients. These are tumours with sufficient rarity that it is strongly recommended that their management is centralised. Attention is drawn to the website which they have set up which has made an important impact in the management of these rare cancers in France and French speaking countries.

The third article discusses the management of small cell cancers. Yet again there are controversies as to whether these are separate entities from small cell cancers arising at other sites, most commonly the lung. The importance of good pathology and radiology review is essential. As a generalisation, these tumours do tend to behave very aggressively and

require multi-disciplinary management, and although it may be debated, combination therapy including surgery, chemotherapy and radiation may confer the best results.

In conclusion, whilst the selection of these three rare tumour types may seen arbitrary, they do all illustrate a variety of practical points about the management of these rare and uncommon cancers. They key thread must be a combination of multi-disciplinary team working and in the first instance, discussion with centralised or regional teams and referral to these teams to carry out the key management processes in their care. We must learn more about the incidences and biology of these tumours and again we must develop national databases and registries to record the frequency of these tumours. Along with this there should be opportunities to establish tumour and tissue banks so that further studies can be carried out when new information becomes available. These can be virtual tumour banks rather than requiring physical storage. Finally, we need local champions for each tumour type who will help to oversee the necessary developments to improve patient care and management.

Conflict of interest statement

None declared.