Primary tracheal tumours

When the many unanswered questions about the treatment of bronchogenic carcinoma—this most common of neoplasms—are considered, it is no surprise that we have few data on primary tracheal tumours. In addition to the rarity of these tumours, effective and safe techniques for tracheal resection and primary reconstruction have only been available for about 30 years. In this issue of Thorax Gelder and Hetzel (pages 688–92) have made an admirable attempt to collect such information in their survey of primary tracheal tumours seen in the UK over a 10 year period.

The survey, however, has serious limitations in that, as far as we can tell, it is a survey by recollection. Location and extent of the tumours were not determined by reviewing radiographs or CT scans. In clinical practice it is all too common to find that visual description of the level and extent of a tumour, particularly by flexible bronchoscopy without specific measurement, is in error. The histological diagnosis was not checked by a review of slides. It is also startling that 13-7% of the patients listed in this survey had no diagnosis at all. The reasons given (“poor condition” of the patients and fear of biopsy of vascular tumours) are not acceptable. It is possible to perform bronchoscopy safely in almost any patient. If there is severe airway obstruction it should be carried out under appropriate conditions of anaesthesia with a rigid bronchoscope by a competent operator prepared to establish an airway. In the rare tumour, such as an occasional carcinoid which appears to be hypervascular on precise examination performed with a magnifying telescope through a rigid bronchoscope, biopsy may be deferred and primary resection performed. Such tumours are almost always mechanically amenable to resection and reconstruction. The more common malignant tumours such as squamous cell carcinoma and adenoid cystic carcinoma can almost always be safely examined by biopsy.

This series is unusual for its histological composition. The extraordinarily high incidence of squamous cell carcinoma relative to adenoid cystic carcinoma, and the relatively high incidence of large cell and small cell carcinoma and adenocarcinoma as primary tumours of the trachea, makes one suspect that many of these tumours may have been invasive bronchogenic carcinomas penetrating the tracheal wall. In the absence of data about the configuration and location of the original tumour this suspicion must remain. It seems unlikely that this is a geographical difference between North America and Europe. In three of the larger single institutional studies of primary tracheal tumours, two in North America and one in Russia, adenoid cystic carcinomas predominated significantly. If the three series are looked at together, of 377 cases reported there were only two cases of adenocarcinoma and one of oat cell carcinoma. This present survey shows a 6% incidence of small cell carcinoma. The absence of carcinoid and mucoepidermoid tumours in a collection of over 300 primary tracheal tumours is also puzzling. One “bronchial adenoma” is mentioned. This term, of course, has been variously used to describe both of these types of tumour and adenoid cystic carcinoma as well. Because of its very imprecision in lumping together three tumours of very different behaviour and histological characteristics the term has largely been abandoned. One might also speculate that some of the oat cell carcinomas may well have been carcinoid tumours of lesser differentiation. Once again, precise histological data would be necessary to determine what the material really means.

One of the great problems in deducing anything from data presented on primary tracheal tumours is the very heterogeneity of the tumours and, hence, the small number in any single group. In addition, as recognised by the authors, the fact that adenoid cystic carcinoma has a very long and insidious clinical course makes five year survival figures worth very little. This also makes it difficult to assess the effect of surgical treatment and radiotherapy or the combination of the two. It has been our clinical experience that these tumours are almost always very sensitive to radiotherapy but that they almost uniformly recur in 5–7 years, usually precisely at the point where the maximum bulk of tumour lay.

The preferred methods of treatment for this type of tumour remain under discussion. There would seem to be no question that a benign tumour, or one of very low malignant potential such as a typical carcinoid, should be excised surgically with primary reconstruction. For most tumours which are of relatively limited extent the procedure is tested and safe in experienced hands. Unfortunately, the techniques of tracheal resection and reconstruction are not widely used and there are relatively few centres in the world where expertise is available. It seems likely that similar treatment should be applied to malignant tumours which are of limited extent. The extraordinarily low resection rate of 10% reported by Gelder and Hetzel suggests that few of the patients were referred to centres where such expertise might have been available. Surgical resection rates in the series studied at the Massachusetts General Hospital were 75% for adenoid cystic carcinoma, 63% for squamous cell carcinoma, 90% for other tumours (including many benign lesions), and 74% for the entire series. As advised by Gelder and Hetzel, the relative role of radiotherapy certainly needs to be investigated further, particularly with higher dosage. It is hoped that their prospective study will shed some real light on this topic. However, such a study will have to be very precisely organised and detailed data obtained. Meanwhile the following cautionary information should be noted. A number of our patients had tumours of such longitudinal extent that surgical excision was not thought to be safe or advisable. The actual bulk of the tumours was not necessarily much greater in lateral dimension than many of
those resected. All patients who underwent resection received full dose mediastinal irradiation postoperatively whether the margins were positive or negative or the adjacent lymph nodes positive or negative. In the squamous cell group 11 patients who had undergone resection and received irradiation died of carcinoma and 18 survived for more than one year without carcinoma. Of those treated by irradiation alone 16 died of carcinoma and only one survived. With adenoid cystic carcinoma seven of those undergoing resection and irradiation died and 38 were alive without carcinoma at the time of the study. On the other hand, nine treated by irradiation alone had died and only three were alive.

A further concern was that, of eight patients with squamous cell carcinoma who had positive nodes at the time of resection and had irradiation postoperatively, six died from cancer and two survived. Of those with invasive cancer evident microscopically at the margins of resection, four died of cancer and one survived. In contrast, when nodes and margins were negative three died from cancer and 12 survived. With adenoid cystic carcinoma, given its remarkable radiosensitivity and its prolonged course in any case, the figures were somewhat more encouraging. Of those with positive nodes and margins there were three deaths and 16 survivors; this was only a little less favourable than the group with negative nodes and margins of whom four died from cancer and 22 survived. Unlike squamous cell carcinoma, this might reflect an ability to cure small deposits of adenoid cystic carcinoma. Figure 3 in the paper by Gelder and Hetzel would suggest that some differentiation in the results of treatment is indeed appearing, even in the small numbers of cases studied.

The current approach of using surgery combined with irradiation is based upon the following facts. Historically radiotherapy led to palliation but not to cure. Admittedly this may change with the higher doses and more effective irradiation now used. When surgery was initially applied alone adenoid cystic carcinoma was seen to recur very many years later even where margins and lymph nodes had been histologically negative at the time of surgery. Tracheal resection is always limited by the short length of the trachea and the technical difficulties of approximating as the length of resection grows. Given the narrow resection margins, it has appeared judicious to apply irradiation in addition to the surgery. It would seem that Gelder and Hetzel’s survey primarily reflects an era when diagnosis was made with a certain lack of precision and completeness and surgical therapy was not widely or safely available. The attempt to set up a study which will evaluate the lesions appropriately for measured management would therefore seem to be a possibly clarifying step. In our experience it has become unacceptable to randomise treatment between irradiation and surgery in patients in whom surgical resection is feasible.

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