In 1998 Travis definitively classified neuroendocrine tumours of the bronchopulmonary tract as three types: typical carcinoid, atypical carcinoid and small cell neuroendocrine carcinomas. The typical carcinoid is a malignant tumour whose clinical behaviour is non-invasive and does not definitely involve lymph nodal organs. However in atypical carcinoid such involvement occurs in at least 50 per cent of cases and is almost invariably found in the case of small cell neuroendocrine carcinomas the behaviour of which is, in contrast, highly aggressive.

About the therapeutic approach of carcinoid tumours of the lung, the surgical resection is still the primary goal. Many aspects, however, remain to be clarified. The surgical questions include the type of resection to be adopted, whether it is to be conservative or extensive involving major pulmonary resection. There is furthermore the question of whether it is useful to perform lymphadenectomy routinely or only in those cases where N+ is present; and finally there is the role of “mini-invasive” surgery and that of multidisciplinary treatment.

The aim of the present work is to evaluate, through a review of our caseload, the various problems associated with therapy and the patient survival rates obtained as a result.

Methods

At Rome’s “La Sapienza” University in the Department of Surgical Sciences and Applied Medical Technologies, from 1969 to 1994, we observed 18 patients with carcinoid tumours of the lung: 13 typical carcinoid (TC) and 5 atypical carcinoid (AC).

We performed 3 conservative and 10 extensive resections on typical carcinoid and 5 extensive resections on atypical carcinoid tumours.

Formal lymph node dissection was carried out on all our patients except in the cases of those with typical carcinoid tumours without enlarged hilar and mediastinal lymph nodes.

The efficacy of adjuvant chemotherapy in carcinoid tumours treatment is controversial and will be confirmed by further trials.

In bronchial carcinoid tumours the long-term prognosis is excellent. In our series the ten-year survival rate is 77 per cent in typical carcinoid and 40 per cent in atypical carcinoid cases.

Key Words:
Bronchial carcinoid tumours, Neuroendocrine neoplasms of the bronchopulmonary tract.
Applied Medical Technologies, during the period 1969 to 1994, there were treated eighteen patients affected by carcinoid tumours of the bronchopulmonary tract. Of these, 14 were men and 4 were women. The age of our patients ranged from 19 years to 63, with a median of 32.

The records of our patients were evaluated with particular attention to the location of the tumour, the type of resection, postoperative TNM classification, lymph node involvement, microscopic evaluation of the resection margins and survival rates.

When a tumor is located above a segmental bronchus, it is defined as centrally located. All other tumors are defined as peripheral.

The therapeutic strategy adopted towards carcinoid tumours in our sample was determined by the histological type and by the clinical stage reached.

Of the 18 patients treated, we were unable to follow-up 2. Of the remaining 16, we have eliminated from this study those patients who were not post-operatively followed-up over a period of at least ten years.

Results

Of these 18 patients, 13 were diagnosed as having typical carcinoids, 5 as having atypical carcinoids (Table I).

Among typical carcinoid cases, 9 patients were at stage I (3 at stage Ia and 6 at stage Ib), 3 were at stage II (2 at IIa and 1 at IIb) and one patient was at stage IIIa (T3N0). Of the atypical carcinoid cases, 2 patients were at stage I, 2 at II and one at stage IIIa (Table II).

When defining “conservative” surgical procedures, we have included bronchoplastactic techniques, segmentectomies and “wedge resections”, while “extended” surgical procedures include the standard lobectomies, bilobectomies and pneumonectomies.

In cases of typical carcinoids we carried out exeresis, using surgical therapy conservatively where possible, and aimed at parenchyma saving.

For the various forms of atypical carcinoids, we adopted the principle of an extensive surgical approach, followed in cases where N+ occurred by one or more courses of adjuvant chemotherapy.

The standard lymphadenectomy was performed on all our patients, save on those cases involving typical carcinoids in which there was no clinical evidence of N+.

The surgical treatment administered to those of our patients who were affected by carcinoid tumours is set out in Table III.

There were no postoperative deaths. Two patients experienced prolonged air leaks. Postoperative atelectasis occurred once after bilobectomy.

The ten-year survival rate among those of our patients affected by typical carcinoids was 77%, while in cases involving atypical carcinoids the rate was 40%.

Discussion

The common neuroectodermic origin of neuroendocrine tumours of the respiratory apparatus does not shed light on the extreme variability of the degree of malignancy or the clinical behaviour of such tumours.

Precise histological type classification may in some cases present difficulties. Endoscopic biopsy, although involving some risk of haemorrhage, is the simplest approach except in cases where the lesion is located peripherally, in which case a useful method is that of fine-needle biopsy CT-guided. Nevertheless, it was not possible to obtain a pre-operative histological diagnosis in approximately 21 per cent of centrally located and in 86 per cent of peripherally located tumour forms. The accuracy of an extemporaneous histological examination ranges between 60% to 40% of cases.

Resection surgery remains the principal therapeutic approach for these tumours.

There is no doubt that in cases of typical carcinoids, which affect mainly young patients, the principal aim of therapy is to carry out, where possible and in particular in N-
forms of disease, a conservative surgical ap-
proach of the “parenchyma-saving” resection
type5-6. However, survival rates do not appear
to be affected by the dimensions or location
of the neoplasia or by whether the surgery
performed was conservative or extensive2,10,11.
“Conservative” types of surgical operation
mainly include segmentary resection and
wedge resection. In certain very special cases
it may also be possible to remove the tumour
by means of a simple bronchotomy or by
means of other bronchoplastic procedures,
while making sure, however, that the
bronchial resection be kept at a distance of
no less than 1 cm from the apparent boun-
dary of the tumour5.
A minor therapeutic role is also played by
VATS-Video-Assisted Thoracic Surgery. This
method, which through small access points in
the thorax enables surgery to be carried out
in the same way as by means of traditional
access, can be adopted only in cases of pe-
ripheral lesions in their first stage “a” (T1
N0) in patients whose pulmonary functions
have been damaged, who require minor pul-
monary resection8,12.
Even lymph nodal spread does not seem,
in cases of typical carcinoids, change the sur-
vival rates of patients which are, indeed, ex-
cellent in these cases. In some cases the pres-
ence of N+, which ranges from 2 per cent to
11 per cent of cases, may be responsible for
local recurrence2,3,7. Martini et al, in a sample
of patients affected by typical carcinoids with
N+, indicate a survival rate over ten years of
76 per cent; for this reason, in the opinion of
these authors, lymphadenectomy should not
be routinely carried out except for cases of
clinical N+.6
A s for atypical carcinoids, however, surgi-
cal exeresis should invariably be of the exten-
sive type, by means of radical resection; “con-
servative” resection should only be carried
out on patients who would be placed at risk
by extensive surgery13.
Lymphadenectomy is necessary for such
patients owing to the high rate of lymph
nodal spread and to enable more accurate
post-operative staging. Among such patients,
lymph nodal spread reduces the ten-year sur-
vival rate to 24 per cent6.
It is worth emphasising that, owing to the
remarkable histological similarity, between
atypical carcinoids and small cell neuroen-
docrine carcinomas, disagreeable post-surgi-
cal surprises can occur6,7.
The role of adjuvant chemotherapy as an
adjuvant treatment in atypical carcinoid is
still controversial for I or II stages; however,
it would appear to be justifiable in stage III

<table>
<thead>
<tr>
<th>Tumour type</th>
<th>Stage I</th>
<th>Stage II</th>
<th>Stage III</th>
</tr>
</thead>
<tbody>
<tr>
<td>Typical carcinoids</td>
<td>a</td>
<td>b</td>
<td>a</td>
</tr>
<tr>
<td>A typical carcinoids</td>
<td>3</td>
<td>6</td>
<td>2</td>
</tr>
<tr>
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</tbody>
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Table II. TNM staging of carcinoid tumours of the bronchopulmonary tract (18 patients).

<table>
<thead>
<tr>
<th>Tumour type</th>
<th>Stage I</th>
<th>Stage II</th>
<th>Stage III</th>
</tr>
</thead>
<tbody>
<tr>
<td>Conservative</td>
<td>3 (2)*</td>
<td>1</td>
<td>5 (3)*</td>
</tr>
<tr>
<td>Segmentectomy</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Wedge resections</td>
<td>2</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Extensive</td>
<td>10 (5)*</td>
<td>5 (3)*</td>
<td>1</td>
</tr>
<tr>
<td>Lobectomies</td>
<td>6</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Bilobectomies</td>
<td>2</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Pneumonectomies</td>
<td>2</td>
<td>1</td>
<td>3</td>
</tr>
</tbody>
</table>

*Peripheral location.
whereas its efficacy in terms of survival rates should be the subject of cooperative clinical further trials.

In conclusion our results confirm that in carcinoid tumours of the bronchopulmonary tract surgical resection is still the primary goal. The surgical conservative resection of the “parenchyma-saving” type in TC is always preferable. The extensive resection is instead recommended in AC whereas the conservative resection is suggested only in patients with damaged pulmonary functions.

The VATS can be applied only in first stage “a” of disease and in peripheral location.

The lymph node dissection in TC is advisable if is present the node involvement but is always necessary in AC, for the high rate of nodal spread.

A bout adjuvant chemotherapy in advanced stages the same treatment criteria that apply to lung cancer should be applied to atypical carcinoid tumours, which is comparable to well-differentiated carcinomas of the lung.

References


