A retroperitoneal tumour of the chemodectoma type

J. N. HARcourt-Webster

From the Department of Pathology, University of Edinburgh

SYNOPSIS A tumour of the chemodectoma type arising in the retroperitoneal space and projecting between the layers of the transverse mesocolon is reported. Attention is drawn to the lack of correlation between the varied histological features and clinical findings in this group of tumours and the inadvisability of forecasting behaviour.

The chemoreceptor cells of the carotid and aortic bodies and the glomus jugulare are sensitive to changes in the pH and the O₂ and CO₂ tensions in the blood and produce reflex changes in respiratory and sympathetic activity (Schmidt and Comroe, 1940; Dripps and Comroe, 1944). Lent. C. Johnson (quoted by Smetana and Scott, 1951) described collections of histologically similar cells in the connective tissue sheath of the femoral vessels. Similar foci have been described in the intra-abdominal and retroperitoneal tissues of rats and mice (Goormaghtigh, 1936; Hollinshead, 1942) but they have not been conclusively demonstrated at these sites in man.

The infrequent tumours arising in the carotid and aortic bodies and the glomus jugulare are well recognized (Lattes, 1950; LeCompte, 1951; Stewart, Ogilvie, and Sammon, 1956). Similar tumours are reported in the organs of Zuckerkandl (Cragg, 1934), the pancreas (Goodof and Lischer, 1943), the orbit (Lattes, 1950), the thigh (Randall and Walter, 1954), the extremities (Sirsat, 1954), the abdominal cavity (Arean and Ramirez De Arellano, 1956), and the retroperitoneal space (Zacks, 1958; Sessions and Scott, 1960). The name of chemodectoma is often applied to such histologically similar tumours believed to arise from chemoreceptor tissues since it emphasizes the physiological properties of the tissue of origin without suggesting a sympathetic or other specific site of development (Mulligan, 1950). For those tumours, however, which arise at sites where no chemoreceptor tissue has been satisfactorily demonstrated the term non-chromaffin paranganglioma may be preferred. The tumour now recorded is regarded as a further example of such a tumour arising in a very unusual site.

CASE REPORT

A married woman of 29 years, who had previously been in good health, presented with an eight-month history of loss of appetite, flatulence, increasing listlessness and lethargy, and a painful swelling in the epigastrium. A firm, slightly mobile, tender swelling was centred in the epigastrium slightly to the left of the midline. The pulsatile character of the swelling was considered to be transmitted rather than direct. There was no lymphadenopathy or abnormality of the cardiovascular or respiratory systems. Blood pressure readings before and after operation were within normal limits. A provisional diagnosis of a pancreatic cyst was supported radiologically.

At operation, a fairly mobile, solid tumour, 8 cm. in diameter, was found projecting forwards from the posterior abdominal wall between the layers of the transverse mesocolon. Numerous dilated veins transversed the surface deep to the peritoneum. A plexus of similar veins invested the pedicle of the tumour which was closely adherent to both the aorta and the inferior vena cava. Extensive haemorrhage from both series of veins proved difficult to control and prevented removal of the tumour.

The urinary catechol amine excretion measured after operation was within normal limits.

The patient was treated by x-ray therapy after which there was a considerable improvement in her general health. One year after the operation she was well and there was no clinical or radiological evidence of metastases.

MACROSCOPIC

The biopsy taken at operation consisted of firm, pale brown tissue measuring 1.2 × 1.2 × 0.7 cm. The cut surface was moderately firm and faintly striated. No capsular tissue was present.

HISTOLOGY

The tumour is composed of polygonal cells of epithelioid type arranged in moderate sized clusters within a stroma formed by abundant elongated vascular channels (Fig. 1). A reticulin preparation shows that in some areas reticular strands link the
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FIG. 1. General pattern of tumour. Clusters or 'Zellballen' of epitheloid type cells with highly vascular stroma. Haematoxylin and eosin × 125.

FIG. 2. Slender reticular strands link the vascular channels and complete the stromal pattern. Gordon and Sweets × 150.

FIG. 3. Pleomorphism of cells and their nuclei; bizarre cells including multinucleate forms. Centrally a collapsed sinusoid with tumour cells adjacent to endothelium. Haematoxylin and eosin × 400.

FIG. 4. Large cell with a giant lobulated nucleus and conspicuous nucleoli. Haematoxylin and eosin × 575.
vessels (Fig. 2) though elsewhere these strands are deficient.

Though there is some variation, the cells generally are large. Many are columnar or fusiform, and their long axes radiate from the walls of the vascular channels. The cell membranes are well-defined and there is abundant, finely granular, eosinophilic cytoplasm. Occasional cells show fuchsinophilia in that the cytoplasm contains numerous, fine, orange granules after staining with Masson’s haematoxylin-Ponceau-fuchsin-light green; the chromaffin reaction is negative. Many nuclei are large, round, or oval and well-defined with a faint chromatin network and one or two nucleoli (Fig. 3). A moderate number are hyperchromatic and there are scanty mitotic figures. There are also a few small nuclei and occasional very large forms, some of which are lobulated (Fig. 4); additionally, there are occasional, widely scattered giant cells with abundant cytoplasm and variably sized and shaped nuclei up to three or four in number.

The vascular channels, many of which are elongated and branched, are lined by a single layer of endothelial cells, some of which have a plumper swollen appearance. The larger vessels (? sinusoids) possess a thin cuff of connective tissue between the endothelium and the tumour cells. In this connective tissue there are small numbers of fusiform cells identical with endothelial cells. No pigmentation was noted and the Perls’ prussian blue reaction was negative.

**DISCUSSION**

The non-chromaffin paragangliomas which have been reported as arising in the retroperitoneal space are summarized in the table. A.P. Stout (1962, personal communication) includes an additional group only one of which metastasized. Of these, three arose in the region of the organs of Zuckerkandl, two in the retroperitoneal tissue between the kidneys, two just below the kidneys, two at the rim of the pelvis or the sacral promontory, and one from the retroperitoneal tissue adherent to the duodenum. This latter example is closest in site to the tumour reported here. Though examples occur in all age groups; the majority arise in the second to fourth decades. There is no significant sex difference.

The histological characteristics of the tumours arising in the carotid body and glomus jugulare are well known (Lattes, 1950; LeCompte, 1951). The features of this case and of those summarized are consistent with those characters: nests or ‘Zellballen’, of pleomorphic large epithelioid type cells within a highly vascular reticular stroma, many cells being closely adjacent to thin-walled blood vessels. Lattes described the occasional presence of haemosiderin. The origin of the abdominal tumours is uncertain owing to the lack of evidence for the constant presence of chemoreceptor tissue in the abdomen of man; further studies to determine the normal distribution of this tissue within the abdomen are necessary.

Similar neoplasms have been diagnosed as granular cell myoblastomas, metastases from a primary renal, hepatic or other carcinoma, lipo-sarcoma and angio-endothelioma (Kolodny, 1927; Smetana and Scott, 1951). Another group having similar characteristics are the alveolar soft part sarcomas (Christopherson, Foote, and Stewart, 1952; MacFarlane and MacGregor, 1958); this group metastasizes particularly to the lungs. If the assumption

<table>
<thead>
<tr>
<th>Author</th>
<th>Sex and Age</th>
<th>Site of Tumour(s)</th>
<th>Behaviour</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cragg (1934)</td>
<td>F 30</td>
<td>On either side of abdominal aorta and lateral to origin of inferior mesenteric artery</td>
<td>No metastasis</td>
</tr>
<tr>
<td>Goodof and Lischer (1943)</td>
<td>M 47</td>
<td>Anterior part body of pancreas. This case also included a carotid body tumour.</td>
<td>No metastasis</td>
</tr>
<tr>
<td>Smetana and Scott (1951)</td>
<td>M 23</td>
<td>Lumbo-sacral region with attachment to sacrum and right psoas muscle</td>
<td>Metastasis to lung and adrenals</td>
</tr>
<tr>
<td></td>
<td>F 27</td>
<td>Invading muscle of lateral and posterior abdominal wall</td>
<td>Metastasis to lungs and liver</td>
</tr>
<tr>
<td></td>
<td>F 9</td>
<td>Over left psoas muscle and to left of great abdominal vessels</td>
<td>No follow-up</td>
</tr>
<tr>
<td></td>
<td>F 39</td>
<td>Slightly separated from upper pole of r. kidney</td>
<td>No follow-up</td>
</tr>
<tr>
<td>Block et al. (1955)</td>
<td>M 19</td>
<td>Right side promontory of sacrum; adherent to, but not invading r. coronary and internal iliac arteries</td>
<td>No metastasis</td>
</tr>
<tr>
<td>Zacks (1958)</td>
<td>M 20</td>
<td>Along abdominal aorta and pelvic brim</td>
<td>No metastasis</td>
</tr>
<tr>
<td>Sessions et al. (1959)</td>
<td>F 48</td>
<td>Extending from diaphragm to aortic bifurcation and</td>
<td>No metastasis</td>
</tr>
<tr>
<td></td>
<td>M 17</td>
<td>vena cava above renal veins</td>
<td>No metastasis</td>
</tr>
<tr>
<td>Sessions and Scott (1960)</td>
<td>M 17</td>
<td>Surrounding aorta and inferior vena cava from bifurcation to renal vessels; thence posterior to aorta up to diaphragm</td>
<td>No metastasis</td>
</tr>
</tbody>
</table>
is correct that the group of tumours, regarded as chemodectomas, are derived from chemoreceptor tissue then it is possible that some of these other, histologically similar, tumours are derived from similar tissue and should also be regarded as chemodectomas.

Regardless of site, the features of many of these tumours in the chemodectoma group, particularly the large bizarre cells, suggest malignancy but in many of these cases neither the clinical findings nor the subsequent behaviour confirms this. There is probably no way, using cytological criteria, of recognizing malignancy in these tumours; the presence of tumour cells within blood vessels is not a valid criterion of malignancy (LeCompte, 1951). Of those cases which have metastasized, the bones, lungs, lymph nodes, and liver are the usual sites with occasional spread to the brain and adrenals.

It is of interest that the tumours of the chemodectoma group arising in sites below the diaphragm metastasize more frequently than those sited above the diaphragm (Pettet, Woolner, and Judd, 1953); the subdiaphragmatic tumours, however, often only metastasize after a prolonged period of time though there is an earlier tendency to local recurrence (Smetana and Scott, 1951).

I am indebted to Professor G. L. Montgomery for criticisms and advice, to Mr. T. C. Dodds for the photomicrographs, and to Mr. E. L. Farquharson, under whose care the patient was admitted, for permission to publish this case.

REFERENCES

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J Clin Pathol 1964 17: 444-447
doi: 10.1136/jcp.17.4.444