CHEMODECTOMA (NONCHROMAFFIN PARAGANGLIOMA) OF THE MEDIASTINUM*

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TUMORS SIMILAR TO those arising at the carotid bifurcation have been found infrequently in other locations. Chemoreceptors, from which these tumors originate, are known to be present in the glomus jugulare,[6] aortic arch,[2] and ganglion nodosum of the vagus nerve of man[8] and in the orbit of the chimpanzee.[5] It is probable that this type of tissue exists in additional areas, and we wish to report a case in which a chemodectoma was found in a site other than those mentioned above.

Mrs. U. J., a 38-year-old white female, was admitted to St. Mary's Hospital, Duluth, Minnesota, on September 10, 1953, in active labor. No prenatal care had been sought. She was delivered of a normal living male without incident. Routine admission chest photofluorogram indicated a widening of the superior mediastinal shadow. The patient stated that she had a goiter for a number of years, although in the report of a physical examination 20 months previously, her physician mentioned that there was no thyroid enlargement. She said that during the 6 months preceding this hospital admission she had experienced an aching pain in the right side of the neck. The patient was told that a mobile unit photofluorogram taken one year previously was negative. (Efforts to obtain this film for review have been unsuccessful.)

Physical examination revealed a mass in the right side of the neck, the upper border of which was firm, non-tender, and at the level of the superior pole of the thyroid gland. The inferior border could not be felt, and extended below the level of the clavicle. The mass did not move on swallowing, and could not be elevated out of the mediastinum. Figure 1 demonstrates displacement of the trachea and esophagus by the tumor. Tracheoscopy and esophagoscopy failed to reveal intraluminal involvement. These findings suggested a tumor of recent origin which was thought to arise from the thyroid gland. However, the posterior location was unusual for a thyroid tumor.

Three weeks after delivery, the patient was explored through a cervical collar incision. The strap muscles were divided on the right side, exposing a firm, encapsulated, lemon-size tumor projecting from the mediastinum along the bodies of the upper thoracic and lower cervical vertebrae. It displaced the common carotid artery and thyroid gland anteriorly. However, the intrathoracic portion was not fixed to the innominate artery or the aorta. Although it was possible to enucleate the mass, troublesome bleeding was encountered. The tumor was fixed posteriorly and medially in the region of the vertebral bodies, but was relatively free elsewhere. A Penrose drain was placed in the large defect in the mediastinum and the wound closed. The patient's postoperative course was uneventful, and she was discharged from the hospital on the fifth postoperative day (Fig. 2).

The tumor weighed 70 Gm. and measured 7.0 by 7.0 by 3.5 cm. It was ovoid and completely enclosed in a thin, fibrous capsule, which also covered a few small nodules on its otherwise smooth surface. The parenchyma was light tan, homogeneous and rubbery, without significant lobulation or trabeculation. There were no necrotic or hemorrhagic areas. Histologically (Fig. 3), the tumor was composed primarily of a rich network of thin-walled blood vessels and sinusoids, adjacent to which were small groups of cells. In some of these the cytoplasm was deeply stained, while in others it was clear. Most nuclei were pyknotic, but a few bizarre forms were found. Mitotic figures were rare. The microscopic appearance was identical with that of tumors arising in the carotid body.

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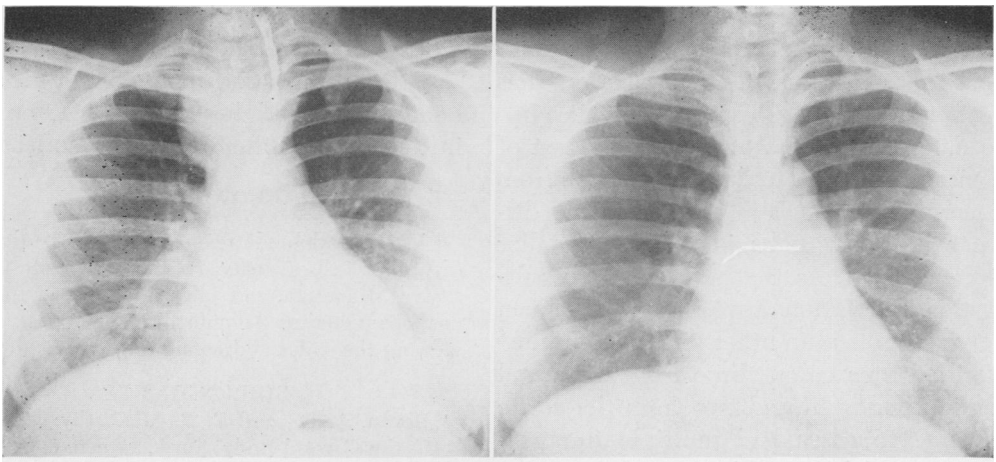

FIG. 1 FIG. 2

FIG. 1. Roentgenogram taken September, 1953, after barium swallow, showing displacement of esophagus to the left at the level of the tumor.
FIG. 2. Roentgenogram, after surgical removal of tumor.

DISCUSSION

In the past, due to the unusual histologic characteristics, many of these tumors were treated as malignant vascular neoplasms. Bevan and McCarthy,[1] in 1929, were among the first to emphasize the innocent nature of these tumors. They collected 134 cases from the literature and reported a mortality of 33.8 per cent due mainly to ligation of the carotid arteries. In addition, 43.3 per cent of the survivors were left with major disabilities because of damage to the brain or a cranial nerve.

Excellent studies, detailing experiences with large series of cases, have been presented by Lahey and Warren[7] and, more recently, by Pettet, *et al.*[10] These and numerous additional reviews indicate that the benign clinical course has not been appreciated fully. Rather, the malignant histologic appearance has been used as an erroneous guide to therapy. Study of frozen sections at the time of surgery is of value in identifying this tumor, and may forwarn the surgeon against an unduly hazardous procedure.

For many years, our knowledge of the normal structures from which these tumors arise was limited to the carotid and aortic bodies.[2] Recently, similar structures have also been found in the glomus jugulare,[6] base of the innominate artery, ganglion nodosum of the vagus nerve,[8] and the orbit.[5] Although originally thought to be part of the paraganglionic system of the adrenal medulla, these structures are now recognized as an independent tissue with a chemorecep-

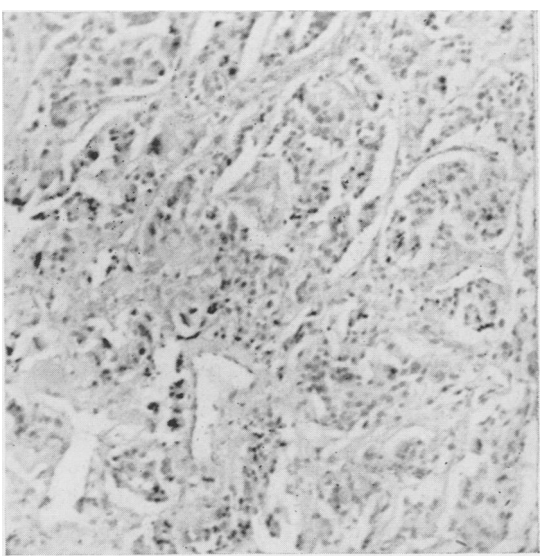

FIG. 3. Histologic section of tumor showing rich network of thin-walled blood vessels and sinusoids. Small groups of cells, some with clear and others with dark staining cytoplasm, are seen adjacent to them (H & E x 120).

tive function altering respiration by responding to chemical changes in the blood.[3] Consequently, the term chemodectoma has been suggested[9] as an appropriately descriptive name for tumors of this type. Most of the tumors reported to date have arisen from the carotid body (314) and, since the discovery of the glomus jugulare, a smaller number (19) from that area.[10] Tumors fitting this description, apparently, have been rarely recognized in other locations. Lattes[8] described four cases, three from the vagal ganglion nodosum and two from the aortic bodies (two cases had multiple tumors). Duncan and McDonald[4] reported two cases of mediastinal origin, and Fisher and Hazard[5] found one in the orbit. Stout[11] described a single tumor in the vagal ganglion nodosum. Thus, only 11 tumors in eight patients have been recorded arising in locations other than the carotid body or glomus jugulare; four of these were found in the chest. In the case presented here, we believe the tumor arose in a location in which no chemodector has been described. Lattes[8] reported two mediastinal tumors which clearly arose from the aortic bodies. Duncan and McDonald[4] described two cases in which they were unable to relate the tumors to a location where chemoreceptors had previously been described. Although our experience is limited to a single case, we wonder whether the extent of chemodector tissue is fully recognized. It is hoped that further reports will clarify this question.

SUMMARY

1. A case is presented in which a chemodectoma was found in the superior mediastinum.

2. Chemodector tissue may exist in previously undescribed areas.

3. Recognition of this fact, as well as the benign nature of these tumors, is important in determining therapy of chemodectomas.

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