Intrathoracic Pheochromocytoma*
Report of a Case
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INTRODUCTION
In the posterior mediastinum, tumors of nervous origin are most common. Other diagnostic possibilities include lipoma, enterogenous and bronchogenic cysts, and pheochromocytoma. A thoracic tumor accompanied by chronic and/or paroxysmal hypertension should suggest the diagnosis of intrathoracic pheochromocytoma preoperatively. However, a few proved cases of pheochromocytoma with neither symptoms nor signs of hormonal activity have been described. The 17th case of intrathoracic pheochromocytoma, which demonstrated no prior symptoms or signs, is herein reported.

CASE REPORT
S.B., a 47-year-old white man, was admitted to the University of Minnesota Hospitals on September 9, 1964 because of a “mass in the chest,” which had been initially detected in 1962 on a routine x-ray examination. A slow but definite increase in the size of this mass was noted over the ensuing two years.

Past history was completely unremarkable, with the exception of occasional transitory radicular pain arising in the shoulder and running to the fingers bilaterally. During convalescence, he was re-interrogated concerning his presenting symptoms. Specific questions regarding headaches, vertigo, palpitation, and/or chest pain were asked. All of these symptoms were denied.

Physical examination on admission was also completely unremarkable. Blood pressure was 126/82; pulse 84 per minute and regular. Routine laboratory studies included: Blood urea nitrogen 77 mg. per cent, fasting blood sugar 90 mg. per cent, hemoglobin 14.1 gm. per cent, and a normal urinalysis. An electrocardiogram demonstrated non-specific S-T changes, but was otherwise within normal limits. Chest x-ray films demonstrated a round, 5 by 8 cm., sharply circumscribed mass in the right posterior, superior mediastinum (Fig. 1). Because of the occasional radicular arm pain, a myelogram was per-

Figure 1: Admission chest x-ray film, showing a posterior mediastinal tumor.

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formed, and interpreted as normal. Lateral planigrams of the right hemothorax demonstrated the foramina of the spine to be free from involvement and/or destruction by tumor. An aortogram was also normal, and failed to show any major vascular connection with the mass. The preoperative diagnosis was mediastinal neurofibroma.

At surgery, through a standard right lateral thoracotomy, a round, encapsulated, reddish, soft mass was found high in the posterior mediastinum, firmly adherent to the fourth and fifth ribs. Shortly after dissection of the tumor was begun, the patient's systolic blood pressure rose from 124 mm Hg to 182 mm., accompanied by an increase in pulse from 86 to 132 beats per minute (Fig. 2). At this time, the diagnosis of pheochromocytoma was first entertained. Resection of the tumor was made somewhat tedious by its extremely rich vascular supply, from the second, third, and fourth intercostal arteries, and was attended by a rapid and irregular pulse ranging from 120 to 150 beats per minute. The administration, in several divided doses, of a total of 250 mg. of intravenous hydroxyzine hydrochloride* satisfactorily controlled the hypertension. A frozen section was initially interpreted as showing "tumor of epithelial origin." Permanent sections were definitely interpreted as pheochromocytoma. The postoperative course was entirely uneventful.

*Vistaril, E. R. Squibb & Sons

PATHOLOGIC EXAMINATION

Macroscopic — The ovoid tumor mass measured 13 by 7 cm., weighed 106 grams, had a soft consistency, and was surrounded by a thickened capsule which measured 1.2 cm.

Microscopic — The tumor is made up of highly pleomorphic cells and contains extensive areas of hemorrhage and necrosis. Tumor cells vary from round to spindle-shaped and occasionally show an alveolar pattern. The cytoplasm is finely fibrillar and is quite vascular. Tumor cells are seen to penetrate the capsule (Fig. 3 and 4).

Final diagnosis — pheochromocytoma.

DISCUSSION

In 1958, Maier and Humphreys* collected nine cases of intrathoracic pheochromocytoma and added three of their own. Since then, four, additional cases** have been collected from the literature making this the 17th reported case of intrathoracic pheochromocytoma (Table 1). Clinical information is available in 15 cases, of which eight (53.5 per cent) had chronic hypertension. Six of these eight also had paroxysmal crises only.

Among the remaining five, the two reported by Peiper and Golestan,** and Luna and associates*** had complained of episodes of headache, chest paint, palpitation and

![Intraoperative blood pressure record. Note the changes observed upon palpation of the tumor (P); hypertension and tachycardia which were partially controlled by the administration (in divided doses) of 250 mg. of hydroxyzine hydrochloride intravenously (Vistaril).](image-url)
vertigo for several years. Each time these patients were seen in the hospital, they were normotensive, and were diagnosed as "anxiety reaction." Ultimately, at surgery, pheochromocytoma was found in both cases. In our case, even in retrospect, we were unable to elicit any symptoms, either suggestive of hormonal activity or similar to those of Luna's patient.

It has been estimated that 5 to 10 per cent of pheochromocytomas are in an extra-adrenal position and, of these, a thoracic location appears to be quite unusual. However, a mass in the posterior mediastinum should suggest the possibility of an intrathoracic pheochromocytoma, when associated with chronic hypertension. In such instances, determination of urinary catecholamines may aid in establishing the diagnosis. In addition, a phentolamine (Regitine) test should be performed. If both tests are negative or inconclusive, venous catheterization may be considered to obtain blood samples from both the superior and inferior vena cava, as suggested by Euler and Von Strow, who demonstrated an elevation in serum catecholamines only in samples taken at the ostium of the azygos vein. In patients with documented cri-

Table 1

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Sex</th>
<th>Age</th>
<th>Hypertension—Chronic</th>
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<td>Miller</td>
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<tr>
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<td>+</td>
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*Modified from Peiper and Green.

+Information not available.
ses of paroxysmal hypertension, a histamine test and urine catecholamine estimation should be done.

In obtaining the past history, careful attention must be directed to both signs and symptoms of paroxysmal discharge of hormones, even if the blood pressure is normal at the time of examination. An important percentage of Maier and Humphreys' patients complained of intermittent headaches, profuse perspiration, chest pain, and/or palpitations. Thus, in a patient with a posterior mediastinal mass and such symptoms, even in the absence of documented chronic and/or paroxysmal hypertension, the possibility of intrathoracic pheochromocytoma must be considered.

There are three cases in the group of 15 in which clinical information is available, who had no signs or symptoms of hormonal activity, yet proved to harbor pheochromocytomas at operation. In these cases, the diagnosis was made when the tumor was palpated in the operating room, or at necropsy.

Some clues to the intraoperative recognition of these tumors may be offered by the following:

1. The gross consistency of the tumor: In ten instances the authors agree that this tumor is very soft in comparison with the more common, firm, neurogenic tumors.

2. The very rich vascular supply of these tumors: This blood supply is unusual compared with the other types of tumors, such as neurofibromas and cysts, commonly encountered in the posterior mediastinum.

3. The weight of the tumor: The weight of the tumor was reported in ten instances, the mean being 50.8 grams with a range of 6-106 grams.

In our case, in which hormonal activity was demonstrated only upon palpation of the tumor, hydroxyzine hydrochloride proved to be of great value in partially controlling the effects of epinephrine and norepinephrine upon cardiac rate, rhythm, and blood pressure. It is of utmost importance, of course, to have norepinephrine available for rapid infusion in the event of postoperative hypotension.

The presence of an intrathoracic pheochromocytoma does not preclude the possibility of a similar tumor elsewhere, as noted by Cone and colleagues and Maier and Humphreys. Therefore, if evidence of hormonal activity persists, a search for an additional tumor in other locations ought to be made. It is of histologic interest that there are some tumors (so-called paragangliomas) which can occur in this location and are morphologically indistinguishable from pheochromocytomas. In these cases, a micro-assay of hormonal activity of the tumor tissue should make the final diagnosis.

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References
10. Silvestrini, P.: Mentioned by Green (14).

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