Malignant Intrathoracic Hurthle-Cell Tumor: Report of a case with 12-year survival following three palliative operations

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Report of a case with 12-year survival following three palliative operations

Conrad R. Lam, MD*
Although no exhaustive review of the literature was made in the preparation of this report, I believe it likely that the lesion is a rare entity, that of a Hurthle-cell tumor presenting as an intrathoracic tumor rather than as a goitre. Of Horn’s 22 cases of Hurthle-cell carcinoma, only one (No. 18) was said to have a substernal component, the clinical diagnosis being “nodular goitre.”

As compared with Horn’s series at the University of Pennsylvania, comparatively few thyroid tumors have been classified or at least indexed as Hurthle cell tumors at the Henry Ford Hospital. The computer indexing system produced only eight cases of benign adenomas for the period of 1964-74, and the tumor registry listed two earlier cases. Both sources produced only nine cases of malignant Hurthle cell tumors, including the case reported in this paper.

In four of these, the primary thyroidectomies were done elsewhere. It is of interest that three of the four patients whose primary goitre operations were done at the Henry Ford Hospital are living and well after more than five years!

Case Report

A white male, age 55, consulted Dr. Earl Congdon, of Flint, MI, in November, 1948, with his chief complaint being a “rush of blood to the head” when lying down, and difficulty in swallowing. He had noted enlarged veins over the upper part of the chest, and thought this part of the chest had become more prominent. He reported some shortness of breath and spells of tachycardia after playing 18 holes of golf.

The chief finding on physical examination was distention of the neck veins and the varicosities over the upper anterior chest. The neck was full, but no definite thyroid gland was palpable. The heart rate was rapid (102 per minute at the first examination). Films of the chest showed a large tumor occupying the upper part of the mediastinum, displacing the trachea to the left and esophagus posteriorly.

(Figure 1A and B.) The basal metabolic rate was plus 29, and hence the tentative diagnosis was intrathoracic goitre, with mild hyperthyroidism. He was referred to the Henry Ford Hospital for operation.

Figure 1A

Figure 1B

Preoperative films showing large superior mediastinal tumor. A, PA view. Note deviation of the trachea to the left. B, lateral view. There is a small amount of barium in the displaced esophagus.
The operation was done on November 19, 1948. After exploration through a second interspace incision on the right, the sternum was transected transversely at this level and by median sternotomy through the manubrium with the Lebsche knife. This provided adequate exposure of the large tumor. The superior vena cava lay laterally and anteriorly, where it was compressed so that little blood passed through it. The tumor had a definite capsule and every effort was made to preserve it as the numerous vascular connections were divided. When the tumor was enucleated and the posterior attachments were divided, the capsule was broken in several places and some pieces of tumor were extruded. A frozen section of one of these was examined by the pathologist (Dr. Frank Hartman), who reported that the tissue was composed of rather large round cells resembling those of a clear cell carcinoma of the kidney. He was of the opinion that the tumor was malignant. Therefore, an effort was made to remove any remaining bits of capsule from the bed of the tumor. The T-sternotomy was closed with heavy silk sutures. The patient's postoperative course was uncomplicated, and he was discharged in two weeks.

The specimen was described as follows: "It consists of a partially encapsulated mass measuring 12 x 18 x 8 cm and weighing 560 grams. On the surface, there are several nodules which at one point appear to have broken through the capsule. The mass is brownish in color. On the cut surface, there is tan, fairly firm tissue with areas of cystic degeneration and necrosis. The tissue is soft and almost encephaloid in character.

"Microscopic pathology (Figure 2A): Sections from various portions of the tumor are all similar except for greater or lesser amounts of necrosis. The tumor is made up of uniformly sized, deeply eosinophilic cuboidal or low columnar epithelial cells. These cells have uniformly sized round nuclei which stain fairly well and possess rather coarse stippling to their chromatin. Mitotic figures are found without difficulty. The cytoplasm in many places is finely granular. In none of the lumina of the tubules or alveoli is there any colloid... The epithelial cells of this tumor resemble in some ways the cells of the liver, of the adrenal cortex and of the renal epithelial tubules. They also resemble the oxyphil cells of the parathyroid. To our mind, they most nearly resemble the cells of uncertain status and origin which are known as Hürthle cells. We believe that this is an instance of Hürthle cell tumor, although to our know-
ledge none has ever been recorded in the mediastinum. Because of past experience with this type of tumor, the presence of mitotic figures and the size, it should be considered a tumor of low grade malignancy. Impression: Substernal Hürthle cell carcinoma."

Postoperative irradiation was recom-
mended and this was done at the Hurley Hos-
pital in Flint, 300 r being given through each of two ports in January, 1949. A progress film of the chest on March 10, 1949 showed a pleasing postoperative picture (Figure 3). During the next four years, there was no radiologic evidence of recurrence, but on December 8, 1954, six years after the operation, a nodule was observed in the left lung (Figure 4). Immediate thoracotomy was done and the metastatic nodule in the medial segment of the left lower lobe was removed. Several smaller satellite nodules between this and the hilum were also excised. The histologic picture of the metastatic tumors was identical with that of the primary lesion (Figure 2B). Six months later, on May 5, 1955, a nodule was visible in the right lung. On July 14, 1955, the nodule in the right lung had not enlarged, but it appeared that there were other lesions in the left lung (Figure 5). In May of 1956, the nodules had not changed much, but there was some fluid at the right base (Figure 6). Five months later, on October 25, the fluid had largely disappeared, and another nodule at the base was visible. In March, 1957, there had been no progression of any of the lung lesions, and at this time, it was elected to remove a mass which had been present in the right pectoral muscle for three years. This had not been growing, but was assumed to be a tumor implant from the original operation eight and one-half years before. The histological picture was the same familiar one (Figure 2C). Dr. Horn described it as follows: "The tumor is composed of eosinophilic cells which are quite large. These cells are arranged in columns, cords and nests. In many areas, they form gland spaces. In no area is there noted colloid formation... The eosinophilic-staining cells are typical Hürthle cells. Diagnosis: Metastatic adenocarcinoma, thyroid gland...""

Five months later, on August 15, 1957, the chest appearance was remarkably good (Figure 7). By this time, nine years after the operation, the patient was 64 years old. During these years, he had been asymptomatic except for the disability from the two operations for recurrences. He worked at his profession of photography, played golf in the season in Michigan and during the winters in Florida. He continued on this program for another three years, although a year later, on July 10, 1958, there appeared to be considerable tumor infiltration in the right lung (Figure 8). On February 19, 1959, the radiologist com-
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Figure 5
Film of July, 1955, showing nodules in both lungs.

Figure 7
Film of August, 1957. There is apparent regression of some of the nodules.

Figure 6
Film of May, 1956. The nodule in the right lung is larger; there is fluid at the base.

Figure 8
Film of July, 1958, with infiltration of lower lung field on right.

It was noted that some of the nodules had decreased in size, and he wondered if there had been some specific therapy. (There had been none). A year later, on March 10, 1960, a film showed increase in the size of multiple nodules. In August, although there had been little change in the radiologic picture (Figure 9), the patient was no longer asymptomatic, but was having hemoptysis. From that time, his course was progressively downhill and he expired on January 2, 1961, a little more than 12 years after the original operation.
Autopsy was done at the Lapeer (MI) County Hospital. It showed infiltration of the mediastinum by dense tumor tissue which extended into the neck and pressed upon the lower poles of the thyroid gland, but did not invade it. There were numerous nodules in both lungs measuring up to 3 cm in diameter. There were metastatic tumors in the liver, spleen and left adrenal. A section of tumor tissue from the chest showed Hürthle cell tumor and fibrous tissue (Figure 10).

Comments

This case is reported because of several interesting features. The Hürthle-cell tumor arose in the mediastinum, obviously from a thyroid anlage, and no tumor was ever found in the thyroid gland in the neck. The malignant nature of the histology of the tumor was evident from the first, but the clinical course was remarkably “benign” for twelve years. With no anti-cancer treatment being administered, tumor nodules in the lungs were observed to remain stable for many months. In some instances they appeared to regress, at least temporarily. The implication is that in the management of certain cancer cases, a measure of optimism is justified.

References

2. Hürthle K: Cited by Horn