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CARCINOMA OF THE LUNG OCCURRING IN PATIENT WITH CAROTID BODY TUMOR: CASE REPORT

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Primary tumors of the carotid body are exceedingly rare neoplasms wihch present difficult and controversial problems in management. Because of their intimate association with the carotid vessels, surgical removal is extremely difficult and operative morbidity, as well as mortality is uncomfortably high.[1,2] In general, these tumors are not radiosensitive[1] but in some instances radiation therapy has been helpful.

This report concerns a case of recurrent carotid body tumor associated with an undifferentiated carcinoma of the lung. A review of the literature failed to reveal a previous report of this combination of tumors although carotid body tumors have been reported as occurring simultaneously with other malignacies.[2,3]

Case Report

Mr. A. S., a 66-year-old Lithuanian steel worker was admitted on July 19, 1964, for evaluation of a mass on the right side of his neck. About six months prior to admission, he began to note an intermittent sensation of fullness and pressure in the right mandibular area, occasional periods of dizziness, and headaches on the right side. Syncopal episodes were denied. These symptoms were associated with the appearance and gradual enlargement of a mass on the right side of his neck. There were no associated constitutional symptoms such as weakness, anorexia, weight loss or fever. He had never smoked.

Review of his past medical history revealed that in 1927, he had a mass similar in appearance excised from the right side of the neck in Lithuania. No diagnosis was available on this tumor. He was well from then until 1961 when he noted a recurrence of a mass in the same location. At operation, the mass was noted to be at the carotid bifurcation. It was excised and a histopathological diagnosis of carotid body tumor was established, (Figure 1). A lymph node removed with the tumor was microscopically free of tumor cells. Chest x-rays obtained at that time revealed no active parenchymal disease. He was asymptomatic from this time until the onset of his present illness.

On physical examination, the only abnormal finding was a 3-4 cm. round, firm, slightly tender mass below the angle of the right mandible, which could be moved laterally but not up or down. There was no associated erythema or drainage. The liver was palpable 2 cms. below the right costal margin in the mid-clavicular line on deep inspiration. Chest x-ray revealed a poorly defined 4 cm. patchy density in the right upper lobe. Gastric washings and 24 hour sputum stains, as well as cultures were negative for acid fast staining bacilli. Skin testing with intermediate strength PPD yielded 5 mm. induration at 48 hours. Intradermal

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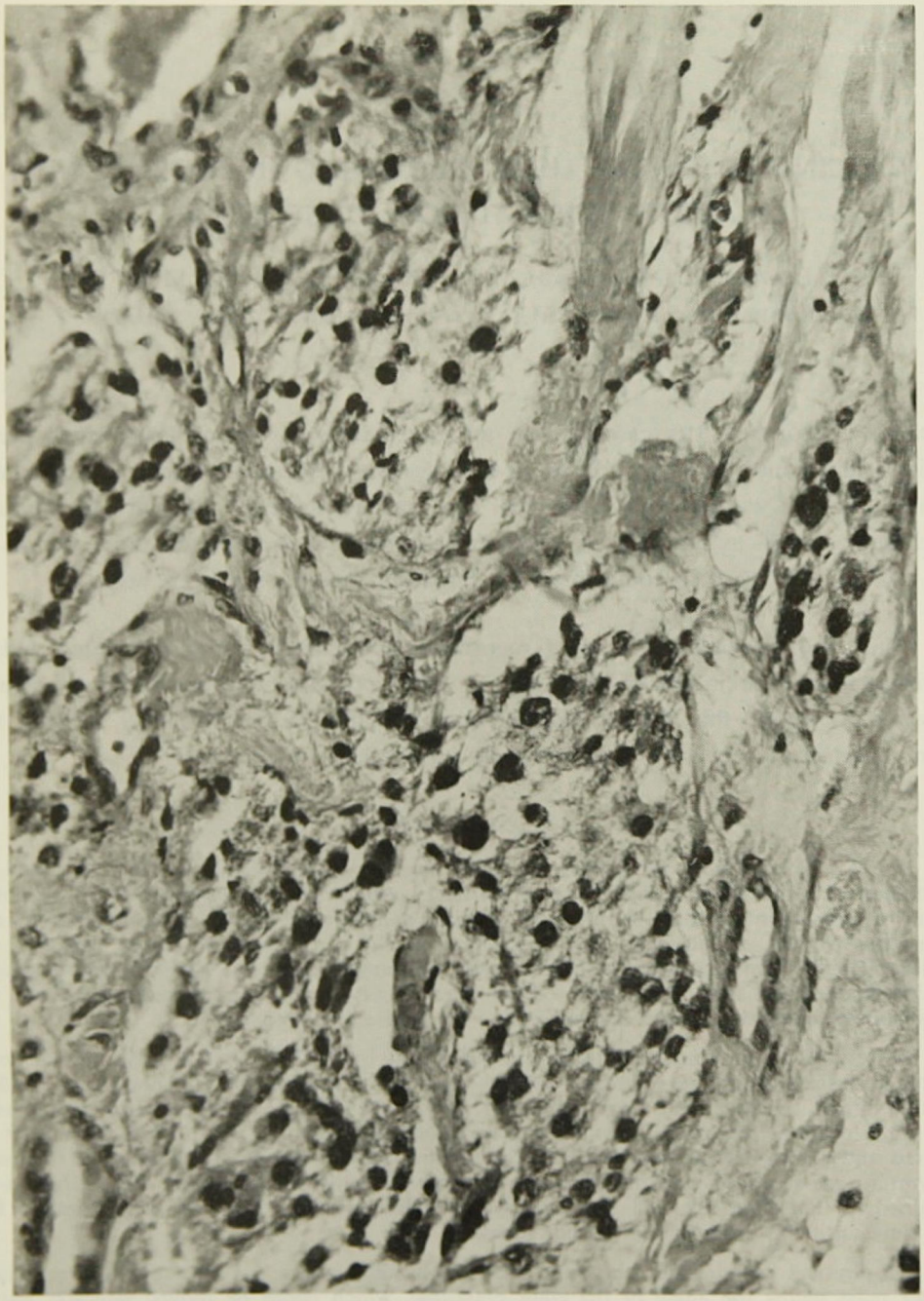

Figure 1

Section of carotid body tumor (x 390) removed September, 1961 displaying nests of cells with round to oval hyperchromatic nuclei with no mitotic figures. This is representative of paraganglioma.

skin testing with antigens of monilia, coccidioidin, mumps, histoplasmin, and blastomycin was negative. Antero-posterior laminograms confirmed the presence of a 3 x 4 cm. mass in the posterior subsegment of the right upper lobe with a nodular infiltrative change distal to the mass. No lesions were noted in the left lung field. Bronchoscopy revealed no significant gross abnormality. Additional laboratory and x-ray studies including hemogram, routine blood chemistries, electrocardiogram, upper gastrointestinal series and barium enema were within normal limits.

On August 8, 1964 surgical exploration revealed the neck tumor to be extensively infiltrative, extending from the carotid bifurcation towards the base of the skull. Excision was considered infeasible. A biopsy revealed the tumor to be a paraganglioma of the carotid body (Figure 2.). The post-operative course was uneventful.

On August 18, 1964 a thoracotomy with right upper lobectomy was performed. Examination of the resected lobe revealed complete occlusion of the lower division of the upper lobe bronchus just distal to the line of resection. The tumor mass measured 3 x 3 x 2.5 cm. and presented as a necrotic, grayish-yellow mass, well demarcated from the surrounding pulmonary parenchyma. Twelve surrounding anthracotic lymph nodes were removed with the resected lobe, none of which were found to contain tumor. Microscopic examination of the tumor revealed it to be a poorly differentiated carcinoma of the upper lobe of the bronchus, (Figure 3). The patient tolerated the procedure well and made an uneventful recovery.

Comment

Carotid body tumors are rare neoplasms about which there is much controversy regarding choice of therapy and degree of malignancy. MacComb[4] reported only 10 cases in a nine-year period on the Head and Neck Service of the Memorial Hospital in New York. Because of their rarity (probably less than 500 cases have been reported), little is known about the natural behavior and therapeutic response of carotid body tumors. Occasionally there are multiple tumors[5] arising more or less simultaneously in sites where the various carotid body-like structures can be found normally, (glomus jugulare, ganglion nodosum and the aortic body). Rarely are they bilateral. There have been several reports of familial occurrence of carotid body tumors.[6,7] It would appear that considering the rarity of this tumor, the frequency of familial occurrence may be significant. Sprong and Kirby[8] reported nine cases in 11 siblings. While there have been reports of carotid body tumors co-existing with other malignancies[2,3] the statistical sampling is too small to permit meaningful interpretation. To our knowledge there have been no previous reports of carotid body tumor, and co-existent cancer of the lung. However, the presence of both tumors in one patient is probably coincidental.

The clinical features of this tumor are few. Most often the patient complains of a painless swelling in the neck of rather long duration. The average duration of symptoms before seeking treatment in many series averages about seven years, the range being between six weeks and 37 years.[9] Occasionally, there may be more dramatic symptoms. Cases have been reported with cranial nerve involvement, severe pain, peripheral and sympathetic nerve involvement, Horner's syndrome, unilateral vocal cord paralysis and encroachment of the pharynx causing dysphagia. Although carotid syncope has been described in association with this tumor, it is surprisingly

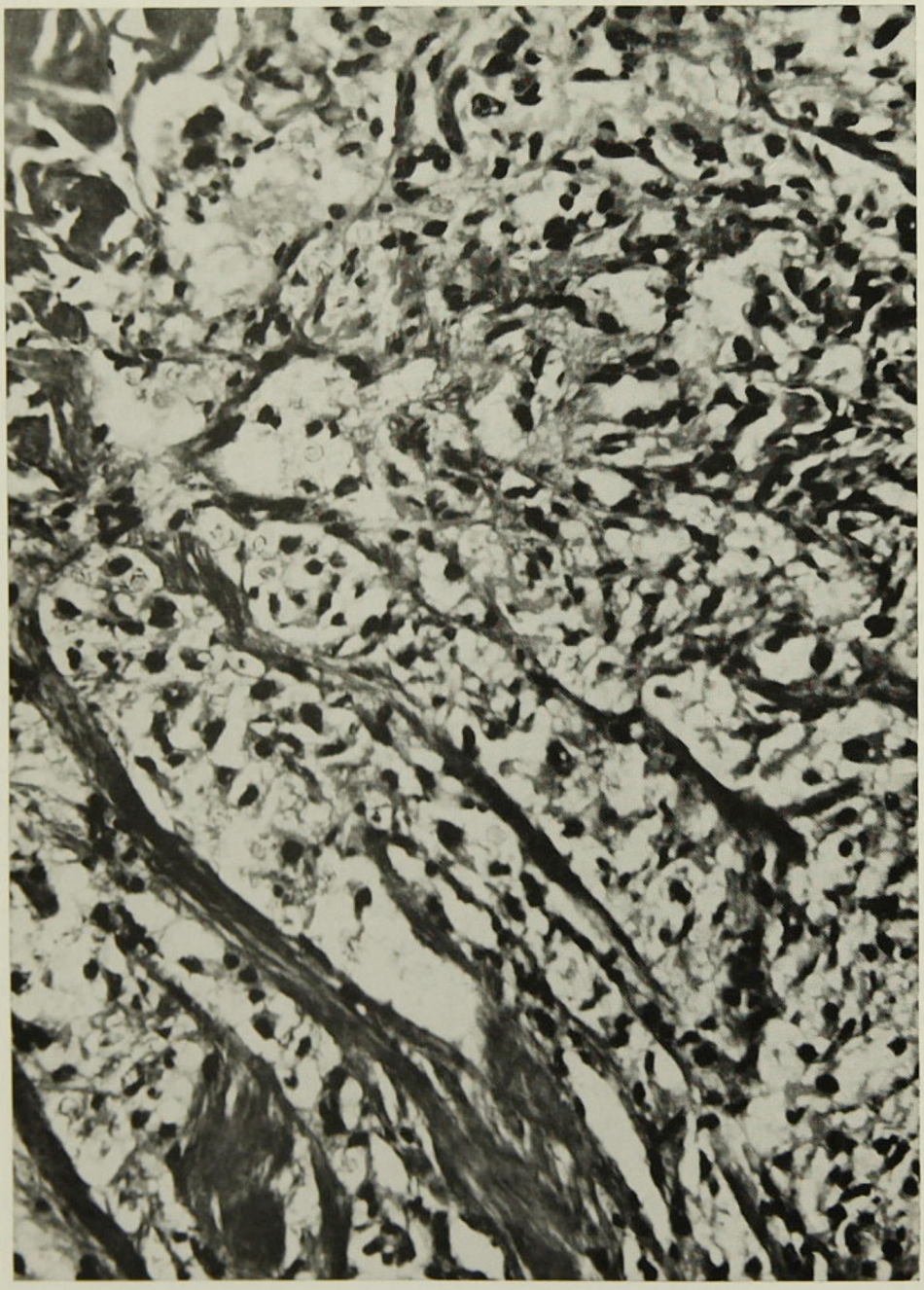

Figure 2

Section of recurrent carotid body paraganglioma biopsied in July 1964 showing very similar histological picture to Figure 1.

rare. In two large series,[2,9] the average age was 34 and 42 years old, with an age range of five months to 76 years of age. Sex incidence in these tumors is approximately equal. Goldberg[10] has proposed five diagnostic criteria for carotid body tumors. These include:

(1) History of several years' duration of a slowly growing, painless mass in the neck.

(2) Firm oval mass in the region of the bifurcation of the common carotid artery.

(3) Full lateral, but none or limited vertical mobility.

(4) Decrease in size on compression of the common carotid artery.

(5) Transmitted, but not expansile pulsation.

The problem of malignant potential is important since surgical removal of these lesions frequently requires ligation of the common or internal carotid artery. Such a procedure is associated with a mortality of approximately 30 per cent, and a high morbidity due to defects of the central nervous system.[2] It is generally agreed that all tests to determine which patients have adequate collateral cerebral circulation, and thus might be expected to tolerate carotid artery ligation, are unreliable. These tests may, however, give assurance that any particular patient will not be able to withstand carotid ligation without serious or fatal results. Angiography has been reported to be helpful in both the diagnosis and management of carotid body tumors.[14]

There is wide variation in the report of the incidence of malignancy in these tumors, the malignant potential being reported as 12 to 50 per cent.[1,2,9,13] This wide variance of opinion is due, in part, to the difficulty of determining valid criteria for malignancy in these tumors.

Le Compte and Willis believe that these tumors are only rarely malignant.[1,11] This view is supported by clinical reviews which claim that approximately 12 to 15 per cent of these tumors will recur or metastasize.[2,9]

Others report that if followed long enough, carotid body tumors are not as benign as previously claimed. They note these tumors may frequently cause serious difficulty due to local invasion and occasionally metastasize, and that at least 30 per cent of patients will die of the untreated disease. In view of this finding, some surgeons[12] recommend removal of carotid body tumors without ligating the artery by dissecting the tumor in the plane of the adventitia of the artery. If the artery is accidentally entered, repair should be accomplished by suture, end to end anastomosis or graft, if necessary.

Histologically, mitotic figures are rare in these tumors, although there is frequently variation in nuclear size and shape. The presence of tumor cells in blood vessels is apparently not a valid criteria of malignancy,[1] and these tumors should not give a chromophin reaction.

Figure 3

Section of tumor of right upper lobe (x 390) demonstrating large, poorly differentiated cells having marked nuclear irregularity and increased mitosis. The histology is not at all similar to Figures 1 and 2.

Le Compte has divided these tumors into three histological types:

(1) Usual type — faithfully reproducing architecture of the carotid body.

(2) Adenoma type — consisting of a tree of cells, plump and rounded, with abundant cytoplasm resembling epithelium and supported by a scant stroma.

(3) Angioma type — chief cells having a spindle or crescent shape simulating endothelial cells. Fundamental pattern as demonstrated by reticulum stains is the same.

Summary

A case of co-existent carotid body tumor, and carcinoma of the lung is presented.

A brief survey of the controversies surrounding the determination of malignancy and proper treatment of carotid body tumors is presented.

This is the first reported case, to our knowledge, of co-existent carotid body tumor and carcinoma of the lung, and this probably is coincidental.

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This patient was referred to Dr. Robert W. Talley by Otonas Vaitas, M.D. of Livonia, Michigan.

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