Chemodectoma of the Aortic Body

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A patient with chemodectoma of the aortic body is presented. The difficulty in management of these tumors by virtue of their location and extreme vascularity is emphasized. A review of the pathologic anatomy, embryology and physiology of the aortic bodies is given.

The first report of aortic body tumor in a human was described by Lattes in 1950. This type of tumor has been variously referred to as aortic body tumor, or chemodectoma. The term chemodectoma derives from the Greek words chemia (infusion), decheshai (to), and oma (tumor), and non-chromaffin paraganglioma. In reviewing the English literature, there have not been more than 37 cases reported. It seems worthwhile, therefore, to report this patient because of the problems encountered in surgical management.

CASE REPORT

A 59-year-old white housewife was admitted to Huron Road Hospital on May 18, 1969, with the complaint of hoarseness and a choking sensation for the past few months. This was accompanied by neck vein distention and productive cough. The patient had lost 40 pounds of body weight by dieting.

Past History

She had had an appendectomy, and fractures of the left 2nd, 3rd, and 4th metatarsal bones in 1964, and an excision of a 3-4 cm supraclavicular lipoma in July, 1967. She stopped smoking 13 years ago. Otherwise, her past history was non-contributory.

Physical Examination

The patient was moderately built, well-developed, and in no acute distress. Blood pressure was 140/90 mm Hg, pulse rate was 84 per minute, and respiratory rate was 20 per minute. There was no edema, cyanosis, or clubbing of fingers. No abnormalities were observed in the ears, eyes, or nose. Indirect laryngoscopy revealed the right cord to be paralyzed. The neck showed marked distention of the cervical veins and no adenopathy. The lungs were clear to auscultation and percussion. There was no shift of the heart position. A2 was greater than P2; no heart murmurs were heard and there was a normal sinus rhythm. The abdomen, genitourinary, and central nervous systems were all within normal limits.

Laboratory Data

The patient's hemoglobin was 10.6 gm; hematocrit 32 vol percent; WBC count 7,800; segs 51; bands 9; L 37; monocytes 3. Urine, FBS, BUN, creatinine, alkaline phosphate, serology, electrolytes, BSP, liver profiles, and prothrombin time were all within normal limits. ECG was normal. Chest film showed bilateral superior mediastinal widening, and a mass 10.6 cm in width. Small hilar calcifications were noted bilaterally. The sterno-aortic space was 0.5 cm in width (Fig 1). Barium enema gave negative findings. Bronchoscopic examination was negative. The bronchial wash specimen was negative for tumor cells and showed normal respiratory flora.

Figure 1. A-P view shows a bilateral superior mediastinal widening and a mass 10.6 cm in width.
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Figure 2. Lateral view.

Esophagoscopy revealed displacement of the esophagus anteriorly. There was no evidence of tumor arising from the esophagus.

The patient underwent surgery on May 27, 1969. From a median sternotomy approach, a large mass posterior to the aorta was found. The mass displaced the superior vena cava to the right and posteriorly and the right branch of the main pulmonary artery to the left. Dissection was started in this area, separating the mass from the pulmonary artery. Massive bleeding was encountered with a very small amount of dissection. It was not possible to control the bleeding adequately because the tumor was posterior to the vital structures. The tumor actually bled from its posterior aspect. This area was packed with large amounts of Oxycel and finally the bleeding was abated. A segment of the tumor was removed for biopsy. The chest was closed in the routine fashion.

Figure 3. Photomicrograph of a section showing rounded to polyhedral cells which are arranged in clusters or large masses forming an organoid arrangement (original magnification × 400).

Figure 4. Photomicrograph of a section showed the numerous vascular channels of the tumor (original magnification × 400).

Microscopically, sections of the specimen showed, in some parts, a cellular pattern whereby the cells were arranged in clusters or large masses forming an organoid arrangement. In most places they seemed to be associated with thin-walled blood vessels. The cells were rounded to polyhedral with moderate amounts of cytoplasm. In some areas there was evidence of old hemorrhage as seen by the presence of pigment-containing histiocytes. Towards the periphery of the tumor, there were distended, vascular channels with thick walls lined by a single layer of endothelial cells. Varying amounts of fibrous trabeculae penetrating the tumor tissue were also noted. In one area there was a slight variation in nuclear patterns, some cells showing slightly larger dark staining nuclei in comparison to the rest of the lesion (Fig 3,4). In view of the pathology and location of the tumor, it was concluded that this was a nonchromaffin tumor of the aortic body.

The patient had right thoracentesis on the fifth postoperative day, which yielded 1,000 ml of serosanguinous fluid. Another 1,000 ml was obtained from the left side on the 11th postoperative day. On the morning of the 15th postoperative day, the patient became very short of breath, cyanotic, and had marked tachycardia and shock. ECG showed right ventricular strain and chest films showed no evidence of bleeding or fluid accumulation. CVP was 15 cm of water, and there was swelling of the right leg with positive Homan's sign. The patient was thought to have had a pulmonary embolus and was given symptomatic and supportive treatment with norepinephrine (Levophed) in parenteral fluid for maintaining blood pressure. The patient recovered slowly and had a series of 11 deep x-ray treatments which began on the 34th postoperative day. In all, she had a total dosage of 4,000 rads; however, the tumor size remained unchanged. The patient suffered another episode of pulmonary embolus and expired on the 51st postoperative day. Unfortunately, necropsy was not permitted.

Discussion

The aortic bodies were controversially termed as paraganglion caroticum inferior in 1922, paraganglion aorticum supracardiale in 1931, paraganglion supracardiale superius and inferior, aortic glomus, aortic arch body, aortico-pulmonary glomus, aortico-pulmonary epithelioid body, cardioaortic body, paraganglion inferior, etc.

The anatomic location of the aortic bodies have
been described by Boyd,11 Comroe,12 Hollinshead,13 Barnard,14 and Nonidez.15 One is found near the innominate artery either lateral to the right subclavian artery or close to the bifurcation of the subclavian and common carotid artery. A second one is situated on the anterolateral aspect of the left aortic arch near the origin of the left subclavian artery. The third one is near the pulmonary end of the ligamentum arteriosum in the angle between the vessel and the descending portion of the arch of the aorta. The fourth one is found on the right side and upper surface of the trunk of the pulmonary artery near the origin of the left coronary artery.

The aortic bodies are derived from the fourth branchial arch arteries and are innervated by the sensory branches of the vagus nerves.11-16 Hammond17 disagrees with the concept of mesodermal origin and has expressed his opinion that they are migrated neuroepithelial sensory cells derived from vagal cells.

The aortic bodies are supplied by the arterial branches of the aorta18 or from the left coronary arteries, and occasionally the right coronary artery.12-19 However, in the newborn kitten, the pulmonary artery invariably furnishes a branch to some of the aortic bodies with the systemic artery.20 In the majority of postnatal development cases, the vessel becomes occluded by the end of the first month. Seldom does the pulmonary artery branch remain patent into adult life.12,16 It is an uncommon variation due to the abnormal persistence of the fetal condition.19,20 The phenomenal switching of the aortic body blood supply from the pulmonary artery to the aorta during normal developmental processes is not observed in birds.21 The venous drainage of the aortic bodies directly goes to the superior vena cava or into the left costocervical vein.18

The carotid and aortic bodies22 are concerned with changes in the chemical constitution of arterial blood (chemoreceptors), whereas, the carotid and aortic sinuses are concerned with the changes in arterial pressure (pressoreceptors). These chemoreceptors are stimulated by: (a) decreased arterial oxygen tension (not oxygen content); (b) increased acidity; (c) increased carbon dioxide tension; (d) increased blood temperature, and (e) certain drugs such as cyanides, sulfides, nicotine, lobeline, papaverine, potassium salts, a variety of choline derivatives and nikethamide.

The response of the chemoreceptors result in: (a) increased respiratory rate, depth and minute volume (the response of the carotid body is seven times stronger than the aortic body);23 (b) increased sympathetic nervous system activity manifested by liberation of adrenalin, (c) increased cerebral cortical activity (the restlessness of anoxia may be due in part to this factor).

The physiologic responses of the glomectomized dogs (excision of carotid bodies and denervation of the aortic bodies) to high altitude acclimatization are characterized by a marked increase in erythrocyte volume, a decrease in plasma volume and a minimal increase in total blood volume which probably is the result of greater hypoxia in the glomectomized dogs secondary to their lack of ventilatory response as compared to controls. The respiratory response to anoxia is markedly decreased and the sensitivity to inspired CO2 at high altitude is unchanged from that of sea level.24 Germant25 found that a selective elimination of the chemoreceptors caused a reduction in the ventilation of about 36 percent.

Chemodectoma of the aortic body occurs in both sexes and is usually a benign, slow growing mass. The patient may be asymptomatic and is found to have the tumor during routine chest x-ray examination or accidental finding in autopsy. The tumor may cause pressure symptoms, as in our case (compression on the recurrent laryngeal nerve). Therefore, chemodectoma of the aortic body should be considered one of the differential diagnoses of tumor of the mediastinum.

The majority of these tumors have been treated by surgical excision. The high mortality rate is due to the location near the vital structures, high vascularity of the tumor with massive bleeding during surgery. On occasion, deep x-ray therapy has been administered for incomplete removal, recurrence and apparent metastasis. The probability of recurrence and metastasis were infrequently cited.26,27 An aortogram may be helpful in diagnosis and evaluating the resectability of the tumor.26,28,29 Multicentric chemodectoma presenting simultaneously in different locations has also been described.1,26,27

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Invention of Tools for Intellectual Proficiency

Leibniz (1646-1716) made important contributions to perfecting symbolism of mathematics. He invented the notation of determinants in solving simultaneous equations, and the dy/dx symbol of calculus. Although Newton (1642-1727) is regarded as the independent discoverer of the calculus, mathematicians prefer Leibniz's notation to Newton's. The idea that economy of symbols is helpful to scientific thought may be traced back to the fourteenth century logician William of Occam, who enunciated the principle known as "Occam's Razor": "Do not multiply hypotheses more than is necessary"—necessary, that is, to explain phenomena or to carry out calculations required in scientific work. Copernicus's chief improvement on Aristotle's and Ptolomy's geocentric systems was to show the greater simplicity of the heliocentric scheme once offered by the Pythagoreans in a cruder form.


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