Chemodectoma in relation to the aortic arch (aortic body tumour)

A clinical report

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Tumours of the aortic body in man are relatively rare and were first described in 1950. Aortic bodies are found in the region of the base of the heart and great vessels, are thought to be of neuroepithelial origin, and have a role in maintenance of circulatory homoeostasis. A clinical report is presented of a benign aortic body tumour which was removed surgically from a 61-year-old woman.

Tumours of the chemoreceptor organs are comparatively rare, especially those in relation to the aortic arch. The following is a brief résumé of the normal anatomy, physiology, and embryology of the aortic body and a description of a further case of an aortic body tumour.

ANATOMY

Le Compte (1951) points out that the actual anatomical location of the aortic body is not well defined in man. Four localities have been described by Boyd (1937):

1. Between the ductus arteriosus and the descending arch of the aorta ;
2. On the right side and upper surface of the pulmonary artery ;
3. Lateral to the innominate artery root ;
4. To the left side of the antero-lateral aspect of the aortic arch.

PHYSIOLOGY

The exact role of the chemoreceptor organs remains speculative. However, they appear to play a part in maintaining circulatory homoeostasis. It is felt that they monitor changes in the oxygen tension or acidity of the plasma.

Recent electron microscope studies (Grimley and Glenner, 1968) suggest a close structural relationship to sympathetic neuroendocrine glands and autonomic ganglia. The same article details the structural elements of the carotid body to which the aortic body is related both histologically and functionally.

EMBRYOLOGY

According to the much quoted study of Boyd (1937), carotid and aortic bodies originate predominantly from mesodermal cellular condensations found around the vessels of the third and fourth branchial arches. The exact embryological origin of the aortic body is uncertain and Boyd's work does not clarify whether the mesodermal condensations found around the fourth aortic arch precedes the migration of neural element of vagal origin or vice versa. Hammond (1941) studied the development of aortic bodies in cats and his findings indicated that these bodies are essentially neuroepithelial, derived predominantly from vagal elements that migrate early along the depressor nerves. The sympathetic nervous system may add to this secondarily. The mesoderm in his opinion played an insignificant role. Electron microscopic studies help to substantiate the theory that the aortic bodies are of neuroepithelial origin. For further more detailed information the reader is referred to the discussions of Lattes (1950) and Grimley and Glenner (1968).

HISTORICAL REVIEW

Aortic body tumours have been studied in man and animals. Most authors recognize Biedl and Wiesel (1902) as being the first to describe the aortic body. However, their paper dealt with the organs of Zuckerkandl, the paraganglia found in the retroperitoneum of infants near the bifurcation of the aorta. Busacchi (1913) was probably the first to describe aortic bodies in the region of

the aortic arch in humans when he described two 'chromaffin' bodies near the heart of a full-term infant. Rabl (1922) and Penitschka (1931) described an epithelioid structure similar to the carotid body in the tissues at the base of the heart. Palme (1934), Muratori (1934), and Nonidez (1935) also described paragangliomas situated near the heart in man and animals.

Bloom (1943) described two aortic body tumours in dogs. It is probable that the first aortic body tumours in man were detailed by Lattes (1950). Since then more cases have been reported in the world literature. Smithers and Gowing (1965) compiled an excellent summary of all 28 cases reported in the literature to this date. Subsequent to 1965 the authors can find no further cases reported in journals which are available to them. Thus it can be seen that, while aortic body tumours are a rarity in the experience of any individual clinic, this tumour is well described in the literature although as far as the authors can ascertain there have been no reported cases in the Australian literature.

CLINICAL RECORD

This 61-year-old Caucasian woman was found to have a mediastinal tumour when a radiograph was taken by the mobile unit in November 1967. On admission to hospital in January 1968 she complained of a persistent cough, worse at night, for several months. The cough was unproductive of sputum and there was no history of haemoptysis or change in the quality of her voice. However, she would sometimes wake at night with an acute choking feeling, especially if she lay on her left side. On occasions she felt there was a lump in her throat. She had lost 28 lb. (12.7 kg.) in weight but had been on a self-imposed diet. There were no symptoms referable to the thyroid gland nor had she ever lived in a goitre belt.

The patient was a fit elderly woman who was obese. On examination the respiratory system was normal. The pulse rate was 80 per min., blood pressure 130/80 mm. Hg and the heart sounds were physiological. There was no evidence of superior vena caval obstruction or of Horner's syndrome. There were no other physical signs.

The chest radiograph showed a rounded, well-defined opacity projecting from the right side of the superior mediastinum and extending from the second

FIG. 1. Radiograph of chest showing rounded opacity projecting from right side of superior mediastinum.

FIG. 2. Radiograph of chest with barium swallow showing deviation of trachea and oesophagus.

FIG. 3. Section of tumour showing uniform polyhedral cells grouped in 'Zellballen' (H. & E. $\times 120$).

to the fourth thoracic vertebrae. The trachea was displaced to the left and was narrowed at the level of the fourth thoracic vertebra (Fig. 1). In the lateral view the opacity was overlying the trachea and superior vena cava. Tomograms confirmed the presence of an anterior mediastinal opacity which appeared solid and lobulated.

It was thought that the opacity was due to an intrathoracic goitre. A barium swallow showed that both trachea and oesophagus were displaced to the left (Fig. 2); the opacity did not move on swallowing. On bronchoscopy, the air passages were clear. Exploratory thoracotomy was advised.

At operation a mass 5 cm. in diameter was found projecting from the interval between the superior vena cava and the trachea and extending into the right pleural cavity. Vertically it lay between the thoracic inlet and the vena azygos. The right vagus and recurrent laryngeal nerves passed through the posterior part of the tumour. The tumour was adherent to the right subclavian and innominate arteries and the proximal portion of the subclavian artery was displaced downwards. One artery of supply came from the right subclavian while a number of veins drained into the superior vena cava and both

FIG. 4. Section of tumour showing cells with abundant cytoplasm and nuclei with coarse chromatin pattern. There is scanty stroma. (H. & E. $\times 750$).

innominate veins. The right vagus and recurrent laryngeal nerves were sacrificed to remove the tumour.

Postoperatively the patient made a steady recovery. Six months after surgery she had no cough but her voice, though improving, was still a little hoarse

PATHOLOGY OF THE TUMOUR The tumour was discrete and spherical in shape. It was surrounded by a thin pseudocapsule. It was grey in colour and soft in consistency, and the cut surface was haemorrhagic.

Microscopically the tumour consisted of uniform polyhedral cells which were grouped in nests or 'Zellbalen' (Fig. 3). Between these clusters of cells there was scanty stroma and numerous capillaries. The cells had abundant acidophilic cytoplasm and the nuclei had a coarse chromatin pattern. Mitoses were infrequent (Fig. 4). Silver stains to demonstrate argyrophilic and argentophilic properties were negative.

In view of the anatomical site of the tumour, and its pathology, it is regarded as being an aortic body tumour.

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