Neurilemmoma of the intrathoracic vagus nerve

C.J. Davis, E.G. Butchart, A.R. Gibbs

ABSTRACT: A neurilemmoma (Schwannoma) of the left intrathoracic vagus nerve was discovered on a chest radiograph of a 28 yr old woman and was removed via a left thoracotomy. Although neurogenic tumours are the most common tumours of the mediastinum, they rarely involve the intrathoracic vagus nerve.

Neurogenic tumours are the most common mediastinal masses in adults and children and comprise 15–30% of tumours in this location [1]. They may arise from the nerve sheath or nerve cells and are usually located within the posterior compartment. However, in 4–6% of patients they occur in the visceral - or middle - compartment and in such cases, usually arise from the vagus or phrenic nerve and are of nerve sheath origin [2-4]. To date, fewer than 50 neurofibromas and 15 neurilemmomas of the intrathoracic vagus nerve have been reported in the world literature [3-7], the most recent report in the English language being in 1974 [4]. We present a patient with a neurilemmoma of the intrathoracic left vagus nerve.

Case report

A 28 yr old nurse presented with a six week history of weight loss (7 kg), fatigue, and a non-productive cough. Her past history was unremarkable. A chest radiograph showed a large calcified mass in the left middle, superior mediastinum (fig. 1). A chest radiograph taken 10 yrs previously had been reported as normal. A computerized tomographic (CT) scan, with and without intravenous contrast, demonstrated that the mass extended from the brachiocephalic vein to the left pulmonary artery. There was a clearly defined tissue plane between the mass and the aortic arch, but not between the mass.

Fig. 1. – Chest radiograph demonstrating the calcified, left anterior mediastinal mass. (A) Posterior anterior view. (B) Lateral view.
and the pulmonary artery. Although densely calcified, the mass appeared to enhance following administration of intravenous contrast.

At thoracotomy, the mass proved to be completely encapsulated and heavily calcified. It overlaid the aortic arch and subaortic fossa and partly compressed the left pulmonary artery. It had no connection to the mediastinum other than the left vagus nerve, which entered superiorly and exited inferiorly. The left recurrent laryngeal nerve exited the mass separately, inferiorly. Resection of the tumour necessitated sacrifice of the left vagus and recurrent laryngeal nerves.

Macroscopically, the tumour was smooth-surfaced and lobulated. On cut surface, it was grey and rubbery in texture except for focal areas of gritty calcification (fig. 2). Microscopically, the tumour consisted of areas in which spindle cells were arranged in short interlacing fascicles with parallel nuclei (Antoni A). Verocay bodies were present, but loose myxoid areas (Antoni B) were inconspicuous (fig. 3). A prominent feature was the presence of hyalinized eosinophilic areas, several of which were calcified. A nerve was identified traversing the tumour. No histological evidence of malignancy was seen.

The patient made an uneventful recovery. No voice change had been noted preoperatively, or immediately postoperatively, but within one week her voice became weak and she aspirated slightly during swallowing. Indirect laryngoscopy revealed a paralysed left vocal cord. Her voice was restored to normal by teflon injection of the cord. At six months follow-up she was asymptomatic and her chest radiograph was clear.

Discussion

Neurogenic tumours of the thorax can occur at any age [1–9] but are most common in young adults; this is also true for neurilemmomas of the vagus nerve, with most patients presenting from ages 20–50 yrs [4–7]. The ratio of males to females is approximately equal. Most vagal neurilemmomas are asymptomatic and are discovered by a chest radiograph. However, they may cause cough,
chest pain, or dysphagia if they arise near to and compress the trachea, major bronchi or oesophagus. Hoarseness can result from tumour involvement of the vagus nerve proximal to the recurrent laryngeal nerve or of the recurrent laryngeal nerve itself [4, 5].

Radiologically, the masses are typically smooth, rounded and homogeneous with sharply defined borders. Occasionally they are calcified and lobulated [1, 3]. A CT scan of the chest, with and without intravenous contrast, can help delineate the extent of the tumour and its relationship to the surrounding structures.

These tumours can occur anywhere along the vagus nerve but are most commonly found in proximity to the aortic arch and are twice as common on the left as on the right. This may be because of the anatomy of the vagus nerve within the chest [4]. The thickest part of the nerve, which contains more neurons and connective tissue, is located proximal to the points where the recurrent laryngeal nerve and the pulmonary and oesophageal plexuses leave its main trunk. Since the recurrent laryngeal nerve on the left arises at the level of the aortic arch and on the right at the root of the neck, the proximal intrathoracic vagus nerve is thicker on the left than on the right. Theoretically, tumours are more likely to arise from the bulkier section of the nerve.

Neurilemmomas are encapsulated tumours consisting of two components: a highly ordered cellular component (Antoni A) and a loose myxoid component (Antoni B). The tumour in our patient showed the typical features of a neurilemmoma but also contained very prominent foci of calcification, which is characteristic of degenerate neurilemmomas. Degenerate neurilemmomas can be mistaken as malignant because of their cellular pleomorphism. However, malignancy is exceptional and is usually associated with the neurofibromas of von Recklinghausen’s disease.

The treatment of choice of any intrathoracic vagal tumours is surgical removal [1, 3, 6, 9]. Only by thoracic exploration and excision can the diagnosis be made accurately and malignancy be excluded. Removal of the tumour necessitates sacrifice of the vagus nerve; however, resulting dysfunction to the heart, bronchial muscles, and gastrointestinal tract has not been observed [5]. Sacrifice of the vagus nerve results in vocal cord paralysis only when the tumour is located proximal to the take-off of the recurrent laryngeal nerve. Recurrence of vagal neurilemmomas is rare [1, 3].

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References