Multiple malignant paragangliomas

A case report

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Summary

The extra-adrenal paraganglia may be divided into branchiomeric (parasympathetic) and paraxial or para-aortic (sympathetic) networks. A patient is described with two synchronous branchiomeric paragangliomas — a carotid body tumour (glossopharyngeal) and a cardiac chemodectoma (vagus). Both tumours were locally invasive and therefore, by definition, malignant.

The paraganglia are derived from the neural crest region of the embryonic neurectoderm. They form an almost symmetrically organised system closely associated with the segmental autonomic ganglia, and are divided into adrenal and extra-adrenal types, the latter forming an extensive network, being found predominantly connected to the ontogenetic gill arches (branchiomeric) and the paraxial regions of the trunk during embryological development. Although the carotid and the aortopulmonary-coronary paraganglia are classic examples of chemo-receptor tissue, all the paraganglia have similar characteristics on histological and cytochemical examination, as well as catecholamine-generating ability to varying degrees. Hence, the adrenal medulla and Zuckerkrandl’s organ have strong affinity for dichromate ions (chromaffin tissue), whereas the branchiomeric paraganglia react weakly or not at all with these ions (non-chromaffin tissue).

Case report

A 55-year-old man presented with a slow-growing right lateral neck mass in the region of the angle of the mandible. A carotid body tumour was suspected and the diagnosis was confirmed by angiography and magnetic resonance imaging (MRI). While assessing the patient’s fitness for surgery, a continuous murmur was heard over the right precordium. Cardiological evaluation, which included 2-dimensional echocardiography and cardiac catheterisation, demonstrated a coronary-cameral fistula. The echocardiography also demonstrated a large 6 x 6 cm anterior cardiac mass compressing the tricuspid annulus and extending into the right atrium and ventricle. MRI confirmed these findings (Fig. 1). Catecholamine levels were not raised.

The patient was subjected to synchronous excisional surgery of the cardiac and neck tumours. Through a median sternotomy the cardiac tumour was found to be invading extensive areas of the heart including the right coronary artery, thus causing the coronary-cameral fistula. Cardiopulmonary bypass was instituted, with palliative excision of the cardiac tumour and repair of the coronary artery using reversed saphenous vein.

The carotid body tumour was exposed through an incision along the anterior border of the sternocleidomastoid muscle. This tumour was also locally invasive, involving the vagus and hypoglossal nerves and the internal jugular vein and was found to extend through the carotid and jugular foramina into the
Fig. 2. Low-power view of internal carotid artery showing the carotid body tumour invading the media with only a thin rim of intima remaining (left) and the cardiac chemodectoma infiltrating the myocardium (X 150) (right).

skull cavity. Nevertheless, control of the internal carotid could still be obtained between the carotid body tumour and the skull base, so that a palliative resection could be performed here as well. The hypoglossal and vagus nerves and jugular vein were sacrificed because they were obviously invaded by tumour.

In both chemodectomas, obvious tumour infiltration of the surrounding tissues was seen, and nests of tumour cells were definitely left behind in the heart and neck. Postoperatively the patient recovered rapidly and there was no cerebral neurological deficit. The cranial nerves which were sacrificed left the patient with tongue deviation (XIIth cranial nerve) and hoarseness (Xth cranial nerve). Methyldobenzylguanidine scanning demonstrated weak uptake over the heart and neck, but not sufficient to be used as a therapeutic modality to irradiate the residual tumour. The patient remains completely well and very active almost 1 year after surgery.

Histopathological examination of the lesions demonstrated that the tumours were locally malignant by virtue of aggressive local invasion of surrounding structures (Fig. 2). Postoperative MRI showed both tumours to be virtually completely excised.

Discussion

Chemodectomas develop from the chemoreceptor tissue of the carotid body and the aortico-pulmonary-coronary bodies. These are the only branchiomeric paragangliomas that mediate chemosensory reflexes via the IXth and Xth cranial nerves. Fig. 3 shows where the paragangliomas are located anatomically. The patient described had 2 tumours, both of which demonstrated obvious features of local malignant disease, at a gross and a microscopic level.

It is not the cytological examination of the cells that is helpful in the diagnosis of malignant disease, since this is notoriously unreliable with apparently quite benign neoplasms demonstrating the most bizarre cellular morphology. It is the histological examination that may reveal capsular penetration, arterial wall invasion and adherence to surrounding structures, such as the relevant cranial nerves. In addition, the macroscopic behaviour supports the histological evidence, such as the appropriate artery becoming embedded within the tumour, and also the invasion of the skull contents through the foramina at the skull base.\(^4,5\) In our patient the coronary and carotid arteries were embedded within the respective tumours, which were both locally invasive, and the carotid body tumour was found to be infiltrating the basal dura mater through the carotid and jugular foramina.

The patient had no evidence of metastatic disease, and this is the reason for the controversy about whether or not these tumours are malignant. The criteria for malignant disease used by surgical pathologists are local invasion (spread by continuity and contiguity) and metastatic disease (lymphatic and haematogenous spread).\(^4,5\) This patient, as so many others with tumours of the paraganglionic system, had neoplasms which were malignant by virtue of their aggressive local behaviour.\(^4,5\)
The concurrence of malignant chemodectomas of the heart and neck has to our knowledge not previously been reported. Even the occurrence of a solitary cardiac chemodectoma is an extremely rare phenomenon. 8

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REFERENCES

Distal dystrophy — unusual presentations
A report of 2 cases

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Summary

Two men suffering from progressive muscle wasting and weakness are described. The first patient presented with a rapidly progressive myopathy of the right lower leg characterised on histological examination of muscle biopsy specimens by extensive rimmed fibre formation. The second patient exhibited severe muscle weakness and wasting of the lower legs characterised by the accumulation of amorphous sarcoplasmic material and vacuolation seen on histochemical evaluation and confirmed by electron microscopy. There was no clinical or electrophysiological evidence of myotonia in either case.


Distal myopathy was first described by Gowers1 in 1902 and later large families with an autosomal dominant 'Swedish' variety of distal myopathy were described by Welander2 in 1951. This type of distal myopathy is rare outside Sweden. Several other 'non-Swedish' forms with dominant inheritance have since been described. 3-4 Recessively inherited or sporadic cases of distal myopathy have been recognised over the past 21 years 5 and a distinct distal myopathy characterised by rimmed vacuoles and lamella (myeloid) body formation was described by Nonarka et al. 6

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Two patients with unusual dystrophic disorders starting in the lower extremities are described here. One patient had a distal myopathy inherited autosomal dominantly and showed features resembling the autosomally recessively transmitted distal myopathy of Nonarka et al. 8 on histological examination of muscle biopsy specimens. The other patient had a most unusual myopathy affecting his lower extremities characterised by the excessive presence of rimmed fibres.

Case reports
Case 1
The first patient, an unmarried man aged 27 years, had a 4-year history of progressive, painless muscle weakness and wasting, which had started in the right leg and foot and had gradually spread proximally until it involved his right thigh. There was no history of neuromuscular disorder in the family, and the patient was an only child. Both parents were examined and found to be normal.

On examination the patient was a heavily built, muscular man with a mildly lordotic stance. There was loss of muscle in the right lower extremity affecting mainly the intrinsic muscles of the foot, the leg and the thigh and, to a lesser extent, the gluteal muscles. Dorsiflexion of the right ankle joint was weaker than that on the left and, using the British Medical Research Council scale of grading, it was assessed at grade 4. Plantar flexion was grade 3, small muscles of the feet 2, quadriceps 3-4, hamstring 4, and the gluteal muscles on the right 4.

The upper extremities were of normal strength and bulk. Over the chest wall there was evidence of previous scarring from the removal of gynaecomastic tissue bilaterally, performed at the age of 18 years. The chest musculature was normal and although the patient had a mild lordotic posture, the muscles