Location of extra-adrenal catecholamine secreting tumours by selective venous sampling and nuclear magnetic resonance scanning

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Summary: Iodinated meta-iodobenzylguanidine (MIBG) scanning is a safe non-invasive method for locating extra-adrenal paragangliomas. However, a positive result is dependent on the presence of an uptake mechanism in the tumour. We report two cases of MIBG negative intracranial catecholamine secreting tumours successfully located by selective venous sampling and nuclear magnetic resonance scanning.

Introduction

Selective venous sampling accurately identifies pheochromocytoma in the difficult sub-group of patients without an adrenal mass on computed tomography (CT).¹ Meta-iodobenzylguanidine (MIBG) scanning is also valuable in this situation,² where it is the investigation of first choice since it is non-invasive. False negative MIBG scans do occur however.² We report two such cases: one, a tumour of the glomus jugulare, and one, the first described instance of a catecholamine secreting paraganglioma in the pterygo-palatine fossa. In each, MIBG scanning was negative while selective venous sampling successfully located the tumours, which were further defined by nuclear magnetic resonance scanning (NMR).

Case reports

Case 1

A 40 year old man had a blood pressure of 260/140 mm Hg. Urinary vanillyl mandelic acid (VMA) was raised (56 µmol/24 h; normal range < 10–35). Plasma noradrenaline was elevated (5.4 ng/ml; normal range 0.2–0.8). Plasma adrenaline and abdominal CT scanning were normal. MIBG scanning of head, thorax and abdomen was negative. Selective venous

sampling demonstrated high concentrations of noradrenaline in both internal jugular veins (Figure 1a). A partial saturation (PS 300/22) scan showed a tumour in the region of the jugular bulb (Figure 1b). Angiography showed that this was supplied by the left vertebral and left external carotid arteries. It was embolized (Dr B. Kendall) and partially removed (Professor L. Symon). The patient underwent radiotherapy and is normotensive on labetalol. Histologically the tumour was a chemodectoma.

Case 2

A 35 year old woman gave a 2 year history of hot flushes, headaches and palpitations. Her blood pressure was 215/140 mm Hg. Urinary VMA was 123 µmol/24 h. Plasma noradrenaline was 6.5 ng/ml. Plasma adrenaline was normal, as was CT scanning of abdomen and thorax. MIBG scanning of head, thorax and abdomen was negative. Selective venous sampling (Figure 1c) demonstrated high concentrations of noradrenaline in the left external jugular vein. A spin-echo (SE 544/44) scan showed a tumour in the left pterygo-palatine fossa, extending into the posterior maxillary sinus (Figure 1d). Arteriography showed that the major blood supply was a branch of the maxillary artery. The tumour was embolized and removed completely (Mr A.D. Cheesman). The patient is now normotensive without medication. Histologically the tumour was a paraganglioma.

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Figure 1 Regional venous noradrenaline concentrations (ng/ml) and NMR scans from two patients with intracranial pheochromocytoma. In one, noradrenaline concentrations (a) were markedly raised in both internal jugular veins (I.J.V.) compared to the arterial concentration (9.5 ng/ml). The PS 300/22 scan (b) showed a tumour in the jugular foramen (arrow) with compression of the cerebellum and distortion of the brainstem. In the second patient noradrenaline concentrations (c) were most elevated in the left jugular bulb (J.B.) and external jugular vein (E.J.V.) compared with the arterial concentration (10.3 ng/ml). The corresponding SE 544/44 scan (d) showed a tumour (arrow) in the left pterygo-palatine fossa.

Discussion

Urinary VMA is a useful screening test for pheochromocytoma. Most adrenal tumours are associated with elevated plasma adrenaline concentrations, and can be located by CT scanning.¹ Extra-adrenal tumours often secrete noradrenaline rather than adrenaline, as in our cases, and are more difficult to localize. MIBG scanning is therefore the most valuable non-invasive technique when the plasma noradrenaline is elevated but the adrenaline concentration is normal.² However, MIBG scanning can only be positive if an uptake mechanism is present. The

processes of amine uptake and secretion are sometimes dissociated, as in the glomus jugulare tumour and pterygo-palatine fossa paraganglioma described. The latter tumour is thought to arise from a fetal remnant of paraganglionic tissue surrounding the maxillary artery,³ which supplied the tumour in our case. Thirteen cases have been described previously,⁴ but none that has been secretory. The present cases indicated the importance of selective venous sampling in MIBG negative catecholamine secreting extra-adrenal tumours. The partial saturation sequence used in the first case results in cancellation of the signal from red marrow at the base of the skull, highlighting

the tumour. The spin-echo sequence SE 544/44 displays moderate T_1 dependence. In neither case did the tumour display the low signal areas described by Olsen *et al.* in 12 cases of paragangliomas greater than 2 cm in diameter.⁵ This may have been a consequence of the relatively short echo time values used in these cases (22 and 44 ms) which did not cause sufficient dephasing of the blood signal to produce a signal void.

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