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The role of MIBG scintigraphy in the management of a case of metastatic glomus jugulare tumour

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Abstract

Glomus jugulare tumours with metastases are rare as the diagnosis of metastases in this condition can be difficult.

In the past this diagnosis has been made on histology of the metastatic lesions which were identified on clinical grounds. This is one of the reasons why there have been only 20 reported cases in the literature up to 1990 (Johnstone *et al.*, 1990). This case report examines the role of meta-iodobenzylguanidine (MIBG) scintigraphy in the diagnosis of metastases. MIBG scintigraphy is an injected radionucleotide isotope scanning technique which is cheap, non-invasive, sensitive as well as being a specific investigation for identifying sites of ectopic neuroendocrine tissue. It may also have a role in the treatment of these tumours.

Key words: Glomus tumours; MIBG scintigraphy

Introduction

Glomus tumours arise from cells of neural crest origin (Smith *et al.*, 1984; Johnstone *et al.*, 1990; Cornford *et al.*, 1992). The glomus jugulare is a normal structure first described by Guild in 1941 (Taylor *et al.*, 1965). It lies in the dome of the jugular bulb below the floor of the middle ear and is thought to be part of the chemoreceptor organ with chemosensory reflexes but this supposition is unproven (Johnstone *et al.*, 1990; Johnston and Symon, 1992). The first report of this tumour was by Rossenwasser in 1945 and it is now the most common middle ear neoplasm.

Glomus jugulare tumours make up approximately 50 per cent of all the chemodectomas of the head and neck region (Rockall *et al.*, 1990). Malignant lesions are very rare (Taylor *et al.*, 1965; Johnstone *et al.*, 1990; Johnston and Symon, 1992) and constitute five per cent of all chemodectomas. However, metastatic glomus jugulare tumours are extremely rare with only 20 reported cases in the literature up to 1990 (Johnstone *et al.*, 1990). The dilemma in diagnosis is often caused by the inability to ascertain conclusively whether the tumour is a solitary primary or a metastatic form of the lesion. When metastases occur they are usually multiple with spread occurring to the regional lymph nodes as well as the lungs and bone via the lymph channels or blood stream (Urquhart *et al.*, 1994).

This case of a malignant glomus jugulare tumour with distant metastases illustrates the problems involved in making a diagnosis of malignancy and metastatic disease. The role of MIBG scintigraphy in the diagnosis and management of chemodectomas is discussed.

Case report

A 36-year-old patient presented with a two-year history of pulsatile tinnitus and progressive deafness of the left ear. In addition voice change and dysphagia were present for six months. Of note in his past medical history was recent onset of hypertension for which he had been treated with methyldopa and hydrochlorothiazide for a period of 18 months.

Examination revealed a well patient with a vascular mass arising from the left middle ear, eroding through the tympanic membrane and filling the external auditory canal. Tuning fork tests and audiogram revealed a dead ear on the left side. In addition he had paralysis of the IXth, Xth and XIIth cranial nerves on the involved side.

On computerized tomography (CT) scanning a ragged erosion of the left jugular foramen was present with a large mass involving the base of the skull and temporal bone. Angiography showed anterior displacement of the external portion of the internal carotid artery by a vascular mass with involvement of the base of the skull (Figure 1). Urinary vanillylmandelic acid levels (VMA) were elevated to twice the upper limit of normal. Up to this point metastatic lesions were not suspected. MIBG scintigraphy was performed to evaluate the functional aspects of the tumour. This showed multiple areas of abnormal uptake. Including the tumour itself, lesions were seen in the left parasternal region of the chest as well as two areas in the upper abdomen (Figure 2). One of these was present on the medial aspect of the upper pole of the left kidney and the second opposite the lower pole of the left kidney. A CT scan showed the adrenals to be uninvolved. No mass was seen on CT scan of the chest but CT of the upper abdomen revealed a mass of enlarged retroperitoneal lymph nodes (Figure 3).

A diagnosis was made of a malignant glomus jugulare tumour with metastases based on the aggressive behaviour of the primary lesion and the presence of lesions at sites other than where this specialized tissue normally occurs.

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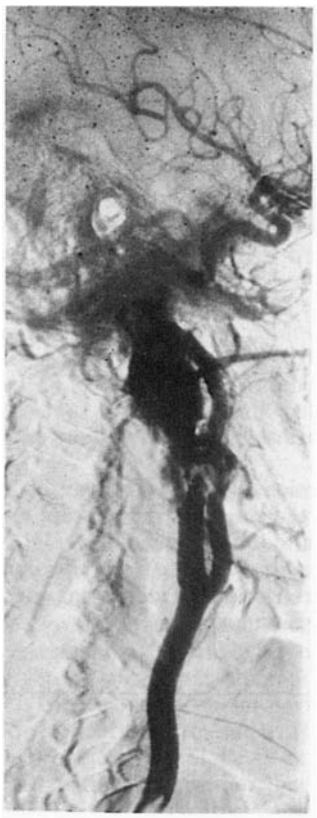

FIG. 1

Angiography shows a vascular mass involving the base of the skull causing anterior displacement of the internal carotid artery.

Discussion

The diagnosis of a suspected glomus jugulare tumour rests on the usual sequence of history, clinical findings and special investigations which usually include PTA, impedance audiometry, calorics and functional assessment i.e. -VMA levels. Radiological evaluations include angiography, CT scanning and magnetic resonance (MR) imaging with or without gadolinium. These investigations should confirm the diagnosis without having to resort to biopsy as well as providing information regarding the stage and anatomical extent of the tumour (Rockall *et al.*, 1990).

The diagnosis of malignancy of the tumour can be difficult from a histological point of view because the appearance of the tumour is variable. Cellular and nuclear pleomorphism do occur but this has not been shown to be a reliable marker of malignancy (Johnstone *et al.*, 1990). Other authors looking at the glomus vagale type of tumour have found histologically that the cells may show atypia with occasional mitoses but no relationship between mitotic activity, perineural or vascular invasion and the clinical course of the disease has been shown (Urquhart *et al.*, 1994).

Malignancy is suggested by firstly, local recurrence bearing in mind that this may just be part of the natural history of an incompletely excised lesion, secondly, occurrence of lesions at sites other than where this specialized type of tissue occurs and thirdly, aggressive behaviour of the tumour (Jackson, 1993).

The diagnosis of metastatic disease needs to be differentiated from multiple primaries and multiple endocrine neoplasias (MEN) syndromes. It is here that scintigraphy is of benefit in allowing whole body scanning to identify other lesions as well as allowing evaluation as to

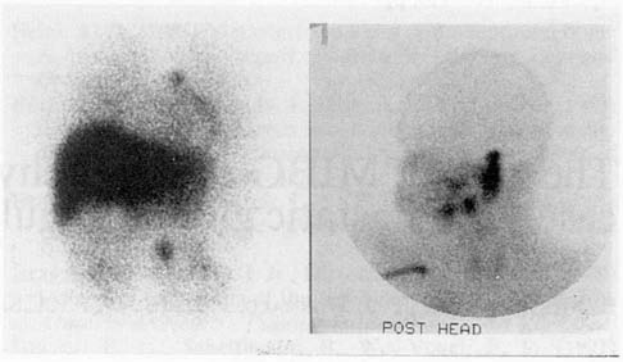

FIG. 2

^{123} I meta-iodobenzylguanidine scintigraphic scan showing multiple areas of abnormal uptake in the left ear, left parasternal region and two foci in the upper abdomen.

whether the other sites are synchronous lesions or metastatic disease based on their site of occurrence.

While some authors feel that all patients with glomus tumours should be screened for evidence of elevated levels of circulating neuroendocrine substances routinely (Von Gils *et al.*, 1990; Kwekkeboom *et al.*, 1993), others feel that only those patients who are symptomatic should be screened with urine testing. Once elevated levels have been found investigations are required to determine whether the origin of these is the primary, multiple primaries or from metastatic deposits. It is here that scintigraphy can be of great value as it has the advantage over conventional radiology of allowing whole-body imaging in an attempt to identify the origin of the excess circulating catecholamines.

The majority of authors feel that when elevated urinary VMA levels are found then scintigraphy with MIBG is indicated (Von Gils *et al.*, 1990; Kwekkeboom *et al.*, 1993; Maurea *et al.*, 1993; Urquhart *et al.*, 1994). It has been suggested that since all glomus tumours contain neuroendocrine substances, MIBG should be used as part of the diagnostic workup (Smith *et al.*, 1984). The reported sensitivity and specificity of I-131 MIBG is high, namely 82 per cent and 100 per cent respectively, for functioning paragangliomas (Maurea *et al.*, 1993).

An additional use for this agent may be in the treatment

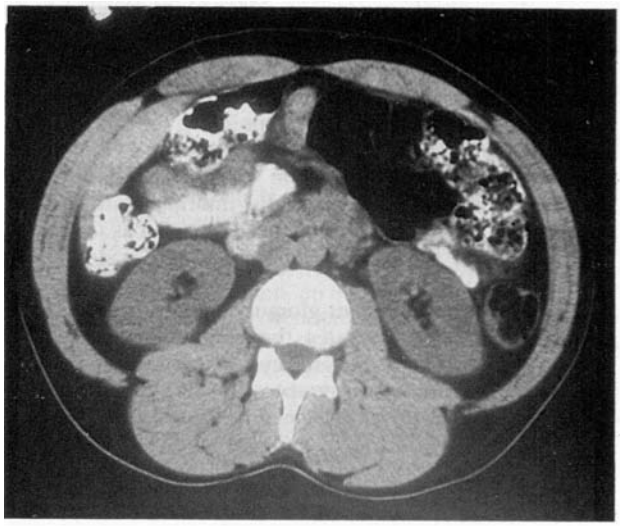

FIG. 3

Axial CT scan of the upper abdomen showing an abnormal mass of retroperitoneal lymph nodes in the same area as the foci on scintigraphy.

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of metastatic disease where, if uptake is shown, it may become a therapeutic option especially in widespread disease. It has been shown to initially control the disease and reduce bone pain from metastases in malignant phaeochromocytomas (Cornford *et al.*, 1992).

Conclusion

Functioning malignant glomus jugulare tumours are rare. The best means to investigate these tumours and to identify concomitant primaries or metastatic deposits remains unclear, however MIBG scintigraphy is a valuable diagnostic and possible therapeutic tool.

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