The Journal of Laryngology and Otology
December 1995, Vol. **109**, pp. 1200–1203

Medullary carcinoma of the thyroid metastatic to the temporal bone

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Abstract

We report a case of proven thyroid medullary carcinoma, metastatic to the temporal bone. Review of the otolaryngology literature demonstrates that this is the first such report. The patient presented with symptoms, physical examination, and imaging studies suggestive of a glomus jugulare tumour. The pre-operative diagnosis of metastatic thyroid medullary carcinoma, however, was made, based on a history of medullary carcinoma of the thyroid with previous metastases as well as an elevation in calcitonin levels. Histological examination of the surgical specimen confirmed the diagnosis.

Key words: Thyroid neoplasms; Carcinoma, medullary; Neoplasm metastasis; Temporal bone

Introduction

Medullary carcinoma of the thyroid, metastatic to the temporal bone, has not been reported previously. The otology literature of the last 30 years contains four major reviews of metastases to the temporal bone (Maddox, 1967; Schuknecht *et al.*, 1968; Hill and Kohut, 1976; Nelson and Hinojosa, 1991). Although the thyroid gland is listed among sites of primary neoplasms, in no review is the thyroid tumour type specified. A further search of American and European literature identifies no cases of medullary carcinoma of the thyroid metastasizing to the temporal bone. We, therefore, present the first such case in order to update the otologist's roster of neoplasms which can metastasize to the temporal bone.

Case report

A 58-year-old woman presented to the University of Iowa Department Otolaryngology/Head and Neck Surgery with six months of headache, left ear fullness, dysphagia, and diarrhoea. Medullary carcinoma of the thyroid had been treated with surgery and radiation therapy 22 years before. A right neck recurrence 12 years thereafter was treated with a functional neck dissection. A right upper lobe pulmonary recurrence eight years after that was treated with lobectomy and radiation. Calcitonin levels remained elevated in the 700's and 800's picograms/millilitre (normal less than 200), increasing to 1100–1670 at the times of her two recurrences. Screening of her family members for endocrine neoplasias was negative.

Physical examination found pareses of the left cranial nerves 9–12. Facial function, ear examination, and hearing were all normal. Magnetic resonance imaging of the brain and brainstem showed a lesion which resembled a typical glomus jugulare tumour (Figure 1). This large enhancing mass arose from the inferior posterior aspect of the petrous bone (the jugular bulb region), and expanded into the posterior cranial fossa. In view of the patient's history, however, metastatic medullary carcinoma was suspected.

Pre-operative angiogram with embolization was performed and the patient underwent a Fisch infratemporal fossa type-A approach to tumour resection (Fisch and Mattox, 1988), with rerouting of the facial nerve, and temporary trans-section of the external auditory canal. The tumour involved the occiput adjacent to the foramen magnum, the occipital facet, and the inferior surface of the temporal bone, directly medial to the pars nervosa of the jugular bulb. The intradural space was clear of tumour.

Histopathological examination of the resected specimen confirmed the diagnosis of medullary carcinoma of the thyroid (Figure 2). The carcinoma was characterized by closely adherent nests of tumour cells within dense fibroconnective tissue. Focal trabecular cell growth was noted. The tumour cells had epithelioid as well as a spindled appearance with pale eosinophilic, minimally granular cytoplasm. Their nuclei showed a clumped chromatin pattern and small nucleoli but minimal pleomorphism or mitotic activity. Tumour cell necrosis was evident in some of the larger islands. The tumour was not associated with amyloid production.

Immunohistochemical stains for calcitonin, chromogranin, and low and high molecular weight keratins were positive.

Discussion

Pathological examination of the surgical specimen from our patient showed the characteristic features of a neuroendocrine carcinoma, including a granular cytoplasm, and a trabecular growth pattern. The mixture of spindled and epithelioid morphology in the tumour are

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Accepted for publication: 23 July 1995.

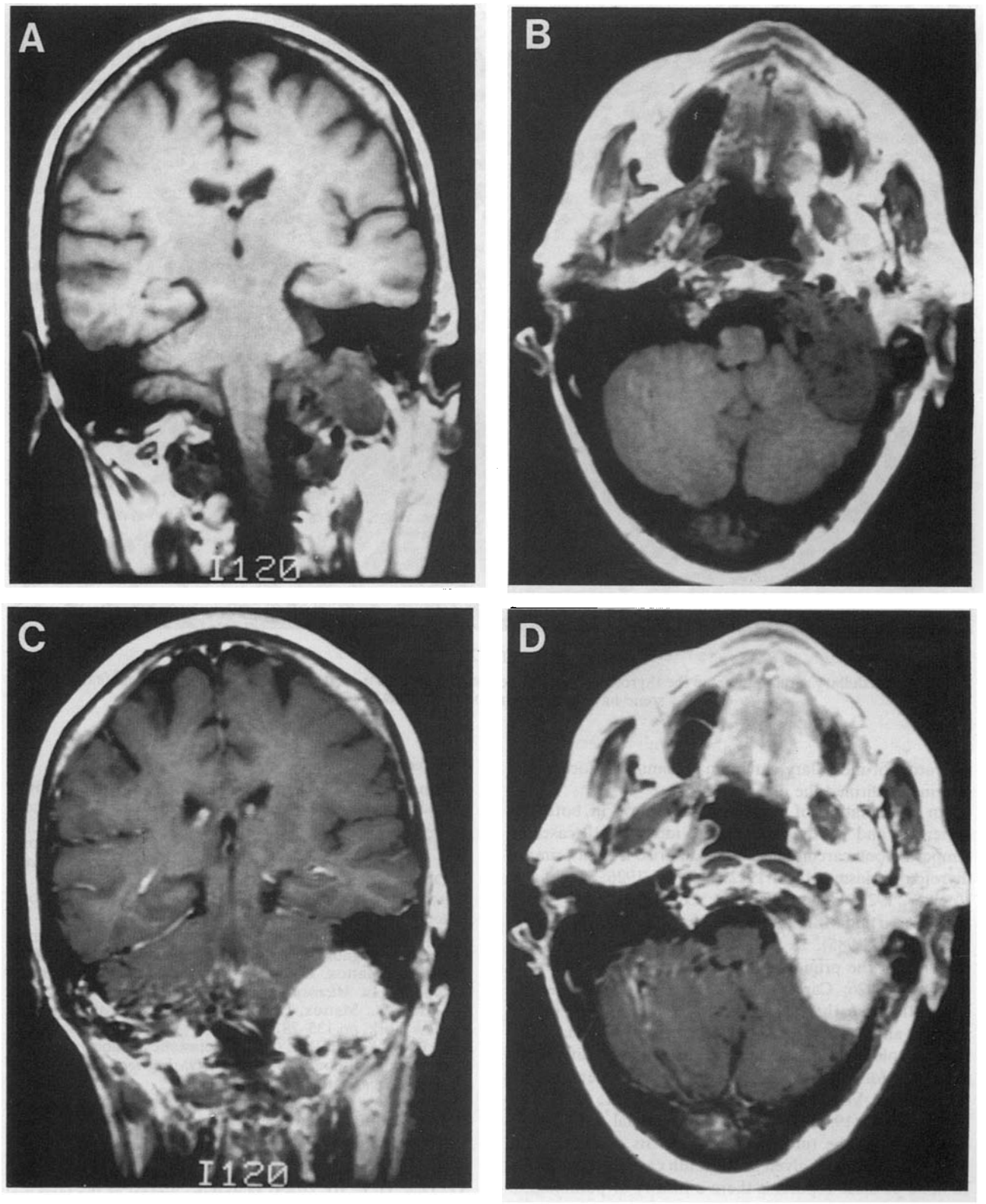

FIG. 1

MRI-imaging of thyroid medullary carcinoma metastasis T1-weighted magnetic resonance imaging (MRI) in the coronal (A and C) and axial (B and D) projections. The mass arises from the posterior inferior aspect of the right temporal bone, in the region of the jugular bulb, and expands into the cerebellopontine angle, compressing the cerebellum and brainstem. Gadolinium enhancement is well seen in both coronal (C) and axial (D) projections.

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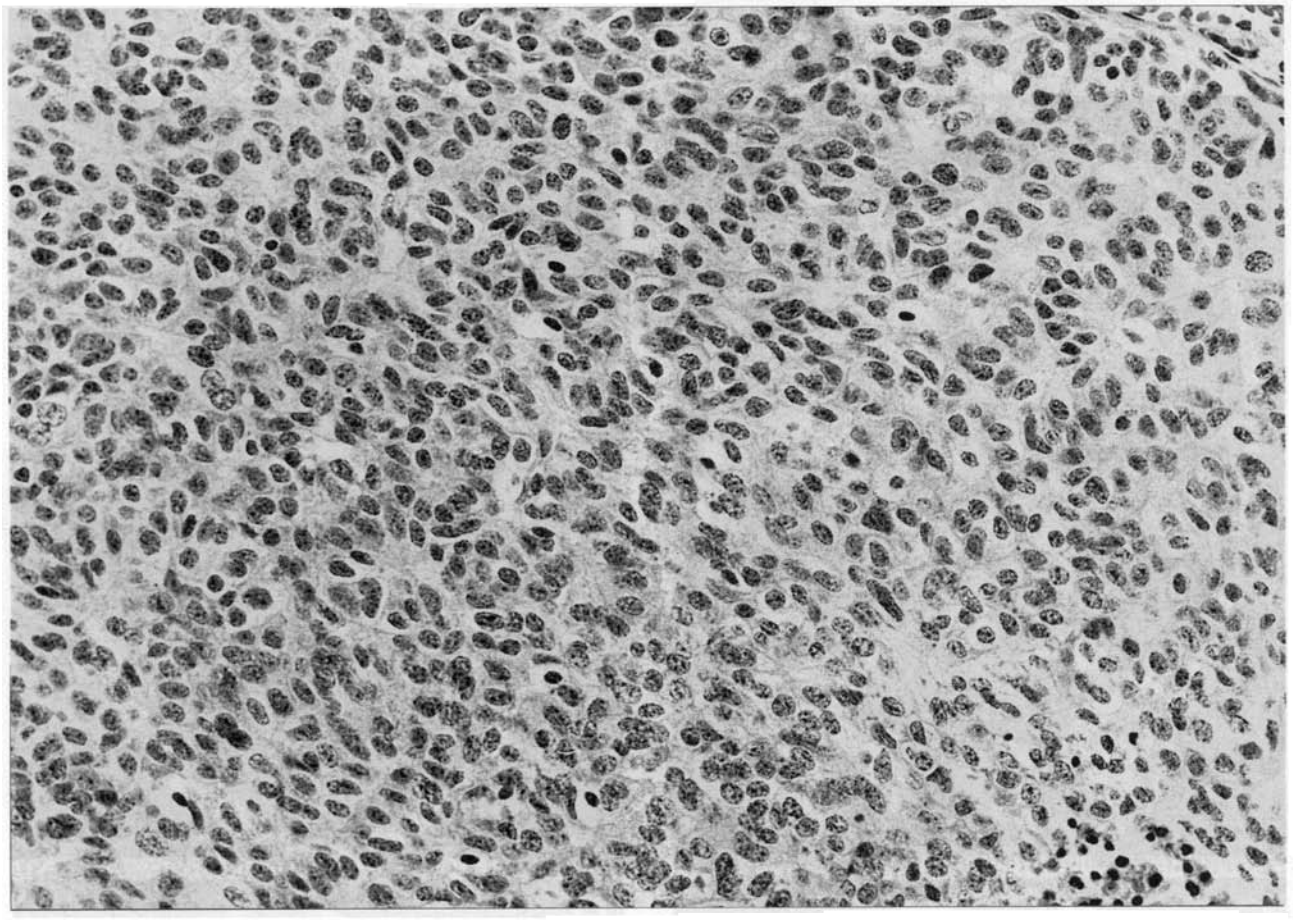

FIG. 2

Metastatic medullary carcinoma from the thyroid. An ill-defined nesting pattern is seen. Some cells have epithelioid features while some have spindled morphology. (H&E ×240).

indicative of medullary carcinoma. Immunohistochemical staining confirmed the diagnosis.

An examination of original case reports in both the American and European literature identifies 10 cases of temporal bone carcinoma thought to arise from a primary thyroid neoplasm (Mayer 1922; Wegelin, 1926; Specht and Voelker, 1929; Ungerecht, 1951; LeMoyne, 1954; Fabre, 1955; Carco and Motta, 1958; Adams *et al.*, 1971; Kelemen, 1977; Belal, 1985). In only three of these cases is the presence of the primary tumour in the thyroid confirmed (Wegelin, 1926; Carco and Motta, 1958; Adams *et al.*, 1971). In these three cases, the histological type of the primary and the metastasis is identified as follicular. The histopathology of the metastasis is described in nine other cases as follicular, folliculopapillary, or adenomatous (Specht and Voelker, 1929; Ungerecht, 1951; LeMoyne, 1954; Fabre, 1955; Kelemen, 1977; Belal, 1985). No description of the tumour, other than its diagnosis as a thyroid neoplasm, is given in the tenth case (Mayer, 1922). In none of these reports is there a history or pathological evaluation diagnostic of medullary carcinoma of the thyroid.

Therefore, the case reported herein remains the only instance of proven medullary carcinoma of the thyroid with a proven medullary carcinoma metastasis to the temporal bone. The scientific merit of this report is simply that the otologist's list of possible metastases to the temporal bone must be continually updated.

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https://doi.org/10.1017/S002221510013244X Published online by Cambridge University Press