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Multiple glomus tumours

DIMITRIS G. BALATSOURAS, M.D., PANAYOTIS N. ELIOPOULOS, M.D., COSTAS N. ECONOMOU, M.D. (Piraeus, Greece)

Abstract

A patient with multiple, synchronous, non-familial head and neck paragangliomas is reported. There were three primary neoplasms, a glomus tympanicum and glomus vagale on the right side and a glomus tumour of the carotid body on the left. Such a combination has never been reported previously.

The reports of all the series with paragangliomas in the literature, as well as the reports of single cases with multiple tumours during the last three decades, are reviewed. Specific problems in diagnosis and management of multiple glomus tumours are discussed.

Introduction

Glomus tumours are rare, benign neoplasms, arising from paraganglionic tissue, derived from the neural crest. Pearse *et al.* (1973) were the first to incorporate the paraganglia into the diffuse neuroendocrine APUD (Amine, Precursor Uptake and Decarboxylase) system, stating that neoplasms of these tissues are to be regarded as members of the APUDOMA family. Information accumulated during recent years has verified this theory, though the majority of these tumours remain endocrinologically silent (Azzarelli *et al.*, 1988). Other names have been applied to them, such as carotid body like tumours, chemodectomas, and nonchromaffin paragangliomas, stressing some of their properties, but most authors today accept the term 'glomus tumour' and classify them according to the site of the lesion (Lawson, 1980).

Most of them occur in the head and neck region and specifically in the carotid body (Chambers and Mahoney, 1968), the temporal bone (Rosenwasser, 1945), the vagus nerve (Endikott and Maniglia, 1980) and other locations, including the trachea (Zeman, 1956), larynx (Stanley *et al.*, 1986), periaortic area (Lattes, 1950), nose (House *et al.*, 1972), mandible (Werner *et al.*, 1968) and ciliary ganglion of the eye (Ahmed *et al.*, 1969). Temporal bone glomus tumours are further subdivided into *glomus tympanicum* tumours, found on the promontory of the middle ear and in the intrabony canaliculi of the auricular and tympanic nerves, and *glomus jugulare*, tumours arising in the jugular bulb adventitia (Alford and Guilford, 1962; Jackson *et al.*, 1982). Furthermore, two primary *Fallopian glomus tumours* have been reported (Dutcher and Brackmann, 1986; Bartels *et al.*, 1990). Multiplicity (Spector *et al.*, 1975a) and familial incidence (Wilson, 1970) of glomus tumours, sometimes in association with pheochromocytoma (Parkin, 1981), is well established.

We report one patient with a combination of three synchronous glomus tumours, a glomus tympanicum and glomus vagale on the right side and a glomus tumour of the carotid body on the left. No such combination has been reported in the literature so far.

Case report

A 33-year-old woman had noted fullness in the right ear for approximately one year before referral to our department. Other symptoms that may accompany glomus tumours such as vertigo, otalgia, otorrhea, and pulsatile tumours were absent. No family history of glomus tumours was reported. On examination a painless pharyngeal swelling was evident in the right tonsillar area and lateral oropharyngeal wall. No lymph nodes were palpable. No nerve deficits were found and clinical evaluation was otherwise unremarkable. Otoscopy revealed a red, bulging tympanic membrane. Pure-tone audiometry showed a conductive hearing loss. The tympanometric curve was flat and no pulse-synchronous excursions of the tympanic membrane, were recorded.

Blood count, serum chemistry and chest X-ray were normal. Blood pressure was 120–80 mmHg and urinary metanephrine and vanillylmandelic acid levels were within normal limits.

Computerized tomography (CT) examination revealed a single soft-tissue mass on the left side, superior to the bifurcation of the left common carotid artery (Fig. 1) and two masses on the right side, the upper filling the middle ear cavity (Fig. 2), and the lower at the level of the nodose ganglion of the vagus nerve.

Digital vascular imaging (DVI) with arterial injection of the common carotid artery revealed a widening of the carotid bifurcation on the left with a well defined tumour blush, characteristic of the intense vascularity of the tumour (Figs. 3 & 4). This appearance is pathognomonic of glomus of the carotid body.

On the right side, a well defined, highly vascular mass was revealed, posterior to the right internal carotid artery, 3 cm distal to the carotid bifurcation (Fig. 5) compatible to a glomus vagale tumour. The tumour apparently received no blood supply from the internal carotid artery, but displaced it anteriorly. The blood supply was derived mainly from the ascending pharyngeal artery. A smaller, well defined blush appeared superior to this, approximately 7.5 cm distal to the carotid bifurcation, compatible with a glomus tympanicum tumour.

Operation: The glomus tympanicum tumour of the right middle ear was removed surgically. No serious bleeding or other complication occurred and the patient had a normal postoperative course.

Pathology: The histological examination of the excised mass showed the typical features of paraganglioma (glomus tympanicum) (Barnes and Taylor, 1990). In the subepithelial tissue there was tumour with prominent vascular component (Fig. 6). The tumour was composed of distinct lobules and nests of small, uniformly sized, epithelioid cells having finely granular eosinophilic cytoplasm and small round or oval nuclei (Fig. 7). Each cell nest was surrounded by collagen and reticulum fibres and a rich capillary network.

From Department of Otolaryngology—Head and Neck Surgery, Tzanion General Hospital of Piraeus,Greece.
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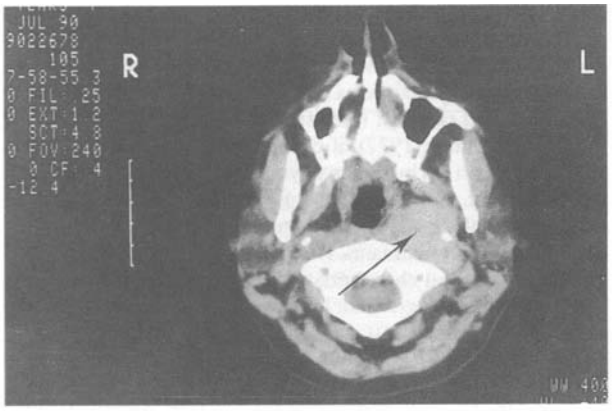

Fig. 1

CT examination—axial cut at the level of C1. An enhancing parapharyngeal mass is shown on the left (arrow), displacing the surrounding tissues.

The patient was advised to accept surgical removal of the other two tumours, as definitive treatment, especially since she was young, but she refused. The possibility of radiotherapy was also discussed. It is now almost one year since she has been diagnosed, and she has not yet decided to continue therapy. The tumour size and the clinical picture of the patient remain unchanged.

Discussion

Glomus tumours of the head and neck region are very rare. Lack *et al.* (1977) reported an incidence of 0.012 per cent in a series of 600,000 patients with various tumours and Wilson (1964) estimated no more than 500 cases in the world literature, over a 73-year period. Despite its rarity, however, no other lesion in the head and neck has been the subject of more controversy than the glomus tumour, which continues to evoke considerable interest because of its location and the technical challenge involved in its surgical removal. Early in this century Mathews (1915) observed: 'This rare tumour presents unusual difficulties to the surgeon and should he encounter it without having suspected the diagnosis, the experience will not soon be forgotten'.

Most authors agree that overall the incidence of multiple glomus tumours is approximately 10 per cent (Spector *et al.*, 1975a). This however increases markedly in familial cases from 30 per cent to 50 per cent (Pratt, 1973; Grufferman *et al.*, 1980; Baars *et al.*, 1981). A few cases of multiple tumours developing metachronously, after excision of the initially diagnosed tumour, have been reported (Oberman *et al.*, 1968), but their exact percentage is unknown.

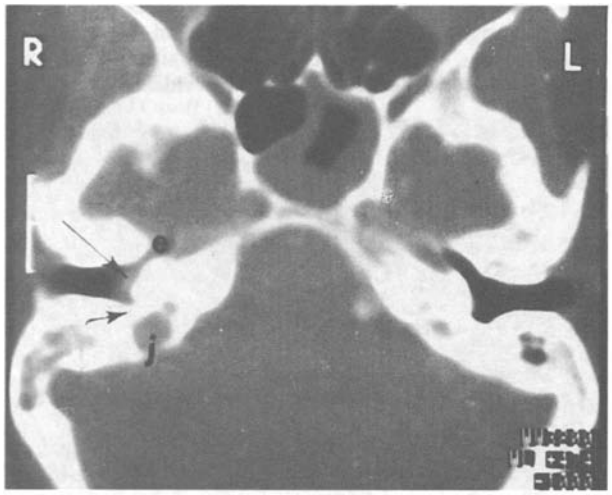

Fig. 2

CT examination—axial cut at the level of the jugular foramen. Soft tissue mass (long arrow) filling the middle ear cavity on the right, extending to the eustachian tube (e). The osseous membrane of the hypotympanum (short arrow) appears intact and the jugular foramen (j) is normal in size and configuration.

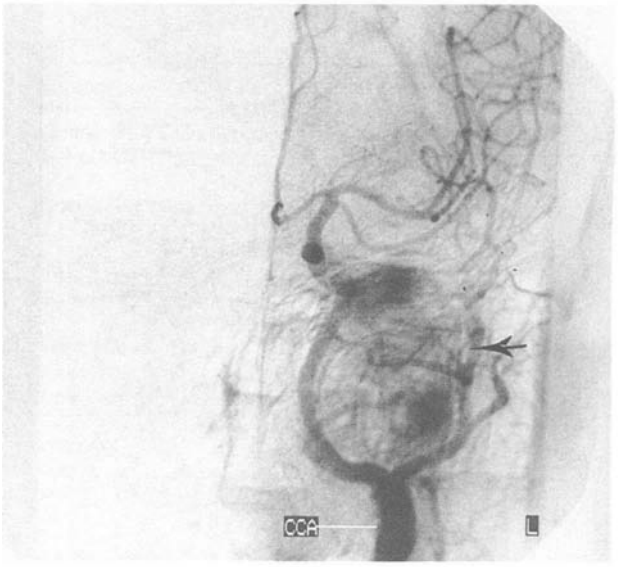

Fig. 3

DVI examination (anteroposterior view). Glomus of carotid body on the left (arrow). Blood supply mainly from branches of the external carotid artery.

A careful scrutiny of the English language literature reveals that in most cases the term 'multiple' refers to synchronous presentation of two glomus tumours, usually bilateral carotid body tumours or a carotid body tumour accompanied by a glomus jugulotympanicum. Only a few cases of three or more synchronous glomus tumours have been reported. In our quest for cases with three or more primary tumours we reviewed the literature of the last three decades, during which carotid arteriography (Irving, 1983) established itself as the main diagnostic tool in recognizing multiple glomus tumours which sometimes remained asymptomatic for long periods.

A detailed review of the previous literature by Alford and Guilford in 1962 revealed only two patients with three primary

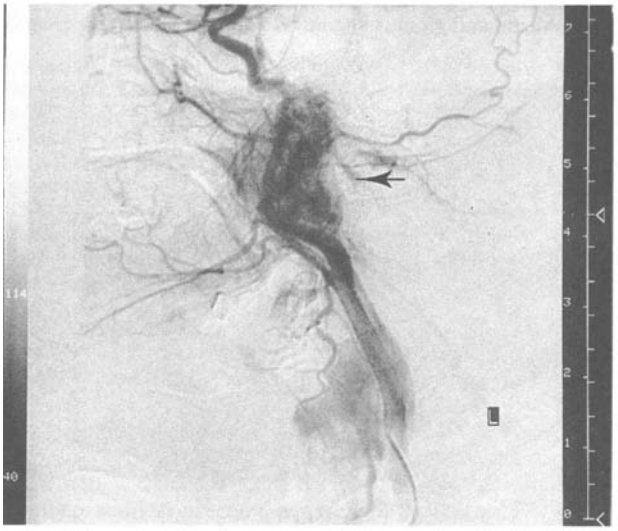

Fig. 4

DVI examination (lateral view). The glomus of the carotid body (arrow) displaces the internal carotid artery anteriorly.

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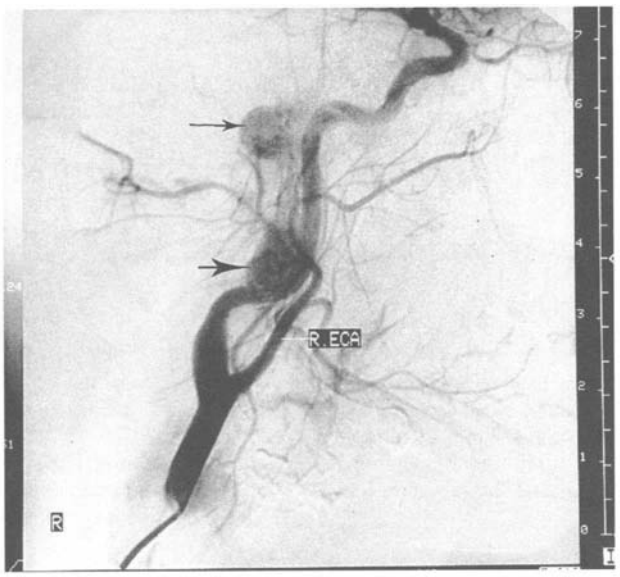

Fig. 5
DVI examination (anteroposterior view). Glomus tympanicum (upper arrow) and glomus vagale (lower arrow) on the right.

synchronous tumours. All the large series of patients since then have been considered and appear in Table I in chronological order, while isolated cases of multiple paragangliomas are listed in Table II. Asynchronous cases and other associated malignancies in other sites sometimes reported to occur (Lack *et al.*, 1977) are excluded from both tables.

It is obvious that very few cases of three or more multiple tumours have been reported. The only exception is the review by Lawson (1980) who reports an unusually high number of cases—up to 2,239 including 249 multiple tumours. The exact locations however are not mentioned and it is not clear if the tumours were primary, recurrences or metastases. Grufferman *et al.* (1980) and Baars *et al.* (1981) report on familial cases, the first one comprising 12 patients with three tumours each (five patients belonging to familial cases) and the second one including six patients, each with three tumours and one with four tumours.

The coexistence of glomus vagale and glomus tympanicum, seems to be a rare phenomenon, having been reported only twice (Spector *et al.*, 1975a; Davidson and Gullane, 1988). Our case, a combination of three synchronous glomus tumours, a glomus tympanicum and glomus vagale on the right side and a glomus tumour of the carotid body on the left, is the first reported case in the literature, as far as we were able to ascertain.

Glomus tumours are usually of benign nature and a long delay before onset of symptoms is common. However, malignancy is observed in 10 per cent (Heinrich *et al.*, 1985) to 30 per cent (Gaylis and Mieny, 1977) of cases, depending on the location, and is characterized by regional lymph node metastases (in about 50 per cent of the malignant tumours) and distant metastases usually to the lungs, liver and bone and in a few cases to other sites (Zbaren and Lehmann, 1985). Lack *et al.* (1979) described certain histological characteristics of malignant tumours. However, most authors agree that the only proof of malignancy is the presence of metastases (Gaylis and Mieny, 1977).

The diagnosis of a glomus tumour of the temporal bone is sometimes obvious on clinical grounds alone, since typically a patient suffering from conductive hearing loss, pulsatile tinnitus and sometimes neurological deficits, presents with a red mass behind the tympanic membrane. However, the clinical picture may vary (Spector *et al.*, 1975b) and differential diagnosis between many other entities has to be made in cases without typical features (Alford and Guilford, 1962). Such pathological states include an abnormally high jugular bulb, cholesterol granuloma, ceruminous adenoma, secretory otitis media, inflammatory polypi, angioma, angiosarcoma, fibroma, granular cell myoblastoma, endothelioma, acoustic neuroma, nasopharyngeal carcinoma, chordoma, carcinoma of the ear or sphenoid, metastatic carcinoma, meningioma, neurinoma of cranial nerves V to XII, Ewing's tumour, eosinophilic granuloma, Hand-Schuller-Christian disease, cholesteatoma and otosclerosis with a prominent Schwartze's sign.

An incisional biopsy confirms the diagnosis, but most surgeons avoid it (Glasscock *et al.*, 1979), since they consider that it may be complicated by profuse haemorrhage.

The critical point in the diagnosis of glomus tumour if surgical treatment is to follow, seems to be the estimation of the precise location and extent of the tumour (Spector *et al.*, 1979). Although a CT scan is a useful non-invasive method of identifying the anatomical location and general vascularity of glomus tumours (Phelps and Stansbie, 1988), the role of carotid arteriography cannot be overemphasized (Spector *et al.*, 1979); it establishes the diagnosis, demonstrates the existence of multiple vascular tumours, allows accurate estimation of the tumour size, and delineates its blood supply. Lately, digital vascular imaging (DVI) has developed into a method that is equally diagnostic, safer and quicker to perform (Irving, 1983).

Retrograde jugular venography (Spector *et al.*, 1979) is sometimes useful in revealing the inferior border of the tumour and displaying intraluminar invasion or extraluminal compression of the jugular vein. Furthermore, invasion of the sigmoid sinus, lateral sinus or inferior petrosal sinus may be evidenced and differ-

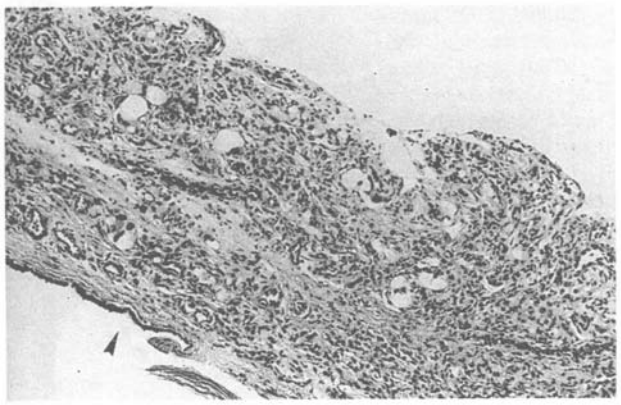

Fig. 6
Narrow zone of connective tissue separates mucosa of middle ear from glomus tympanicum tumour (arrow). Nest of cells and prominent vascular component are observed (haematoxylin—eosin, ×100).

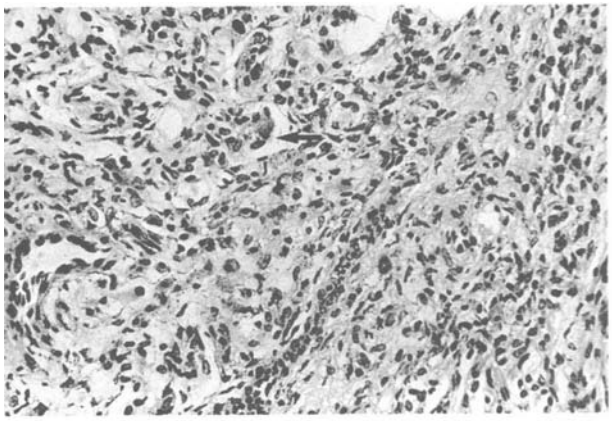

Fig. 7
Small clusters of epithelioid cells arranged in an organoid pattern (nests), some of which protrude into vascular spaces (arrow) (haematoxylin—eosin, ×220).

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TABLE I
MULTIPLE GLOMUS TUMOURS

Authors	Patients	Two sites	Three or more
1. Alford and Guilford (1962)	316	12 (5 GJT-CB ipsi, 5 GJT-CB contra, 1 GT-GJ ipsi, 1 GJ-GJ)	2 (GJT-CB-CB)
2. Morris *et al.* (1963)	12	4 (CB-CB)	—
3. Anderson and Scarcella (1963)	15	1 (CB-GJT ipsi)	—
4. Mcilrath and Remine (1963)	64	4 (3 CB-CB, 1 CB-GJ)	—
5. Capps (1964)	44	3 (CB-GJ)	1 (CB-CB-GJ)
6. Conley (1965)	29	—	1 (CB-CB-GJ-GJ)
7. Wilson (1966)	15	1 (CB-CB)	—
8. Staats *et al.* (1966)	10	1 (CB-CB)	—
9. Cordell *et al.* (1967)	10	—	—
10. Farr (1967)	37	1 (CB-CB)	—
11. Javid *et al.* (1967)	17	2 (CB-CB)	—
12. Oberman *et al.* (1968)	40	1 (CB-CB)	—
13. Rosenwasser (1968)	36	1 (CB-GJT contra)	—
14. Chambers and Mahoney (1968)	36	1 (CB-GJT contra)	—
15. Vigor *et al.* (1969)	16	1 (CB-CB)	—
16. Shamblin *et al.* (1971)	90	1 (CB-CB)	—
17. Hewitt *et al.* (1972)	19	—	—
18. Westbrook *et al.* (1972)	20	4 (3 CB-CB, 1 CB-GJ)	1 (CB-CB-GJ)+ +:recurrence?
19. Moore *et al.* (1973)	33	—	—
20. Pratt (1973)	52*	16 (mainly CB-CB)	—
21. McGuirt and Harker (1975)	9	2 (CB-GJ contra, CB-CB)	—
22. Spector *et al.* (1975a)	76	7 (GJ-GJ, GT-GV ipsi, CB-GT contra, CB-GV contra, 2 CB-GT ipsi, CB-GJ ipsi)	1 (CB-CB-GJ)
23. Javid *et al.* (1976) (new version of series 11)	24	3 (CB-CB)	—
24. Dent *et al.* (1976)	15	1 (CB-CB)	—
25. Gaylis and Mieny (1977)	25	1 (CB-CB)	1 (CB-CB-GV)
26. Lack *et al.* (1977)	69	2 (CB-CB, GJT-GJT)	—
27. Irons *et al.* (1977) (new version of series 16)	116	5 (2 CB-CB, 2 GV-GV, 1 GJT-GJT)	—
28. Glasscock *et al.* (1979)	70	6 (GJ-CB ipsi, 2 CB-CB, 1 (?), 1 GJ-CB contra)	—
29. Lawson (1980)	2039-2239	249 multicentric tumours —insufficient data	
30. Grufferman *et al.* (1980)	835	30 (CB-CB)	7 (CB-CB-?)
	88*	22 (CB-CB)	5 (CB-CB-?)
31. Baars *et al.* (1981)	26*	6 (1 CB-CB, 1 CB-GV, 2 GJ-GJ, 2 CB-GJ)	6 (3 CB-CB-GJ, 1 CB-GJ-GV, 1 CB-CB-GJ, 1 CB-GV-GV) 1 (CB-CB-GV-GV)
32. Van Asperen de Boer *et al.* (1981)	42	9 (5 CB-CB, 3 CB-GJ, 1 CB-GT)	—
33. Lees *et al.* (1981)	39	4 (location not specified)	—
34. Rosen *et al.* (1981)	27	4 (3 CB-CB, 1 CB-GJ)	—
35. Jackson *et al.* (1982) (continuation of series 28)	11	3 (GJ-CB, GJ-CB, GJ-GV)	—
36. Krupski *et al.* (1982)	19	1 (CB-CB)	—
37. Padberg *et al.* (1983)	39	—	—
38. Brown (1985)	231	6 (location not specified)	—
39. Sykes and Ossoff (1986)	36	2 (CB-GV ipsi, CB-CB)	2 (CB-CB-GV, CB-GJT-GJT)
40. Dickinson *et al.* (1986)	32	10 (5 CB-CB, 4 CB-GJT contra, 1 CB-GJT ipsi)	2 (CB-CB-GJT)
41. Hodge *et al.* (1988)	59	4 (location not specified)	—
	5*	2 (CB-GV contra, CB or GJT-GV contra)	—
42. Bundgaard *et al.* (1989)	45	—	—
43. McPherson *et al.* (1989)	25	2 (CB-CB, CB-mediastinal paraganglion)	1 (CB-bilateral PH-paraaortic PH)
44. Barnes and Taylor (1990)	23	1 (CB-CB)	1 (CB-CB-GV)

*: familial cases, PH: pheochromocytoma, ?: not specified in text, GT: glomus tympanicum, GV: glomus vagale, GJ: glomus jugulare, CB: glomus of carotid body, GJT: glomus jugulotympanicum (exact location not defined).

entiation between glomus tympanicum and glomus jugulare may be achieved.

During recent years, magnetic resonance imaging (MRI) with gadolinium enhancement (Phelps and Stansbie, 1988; Phelps and Cheesman, 1990) has proved itself a useful adjunct in preoperative diagnosis, contributing to the detection of small tumours confined to the middle ear and excluding involvement of the jugular bulb.

The definitive management of the glomus tumour has been the source of decades of vehement controversy. Nowadays, treatment of these lesions, whether benign or malignant, consists primarily of surgical resection, especially in young patients (Lewis, 1983). However, these neoplasms, especially the large ones, represent monumental challenges to even the most experienced of neurotological surgeons (Jackson *et al.*, 1982). A team approach of head and neck surgeons, neurosurgeons and vas-

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TABLE II
CASES WITH MULTIPLE GLOMUS TUMOURS

1. Lattes (1950)	(R)GV-(R)CB-glomus of the aortic body
2. Marcuse and Chamberlin (1956)	(R)CB-(L)CB-2(L)GV, superior and inferior to (L)CB
3. Bogdasarian *et al.* (1979)	(L)CB-2(L)GV (at the levels of left thoracic vagal-T1 and left cervical vagal) - pheochromocytoma
4. Wilson (1979)	(L)CB-(R)CB-(R) subclavian paraganglion tumour
5. Demarino *et al.* (1990)	(R)-CB-(R)GV-(L)CB-(L)GJ.
6. Present case (1991)	(L)CB-(R)GV-(R)GT.

CB: glomus of carotid body, GV: glomus vagale, GJ: glomus jugulare, GT: glomus tympanicum, L: left, R: right, T1: 1st thoraric vertebra.

cular surgeons is considered necessary, especially in skull-base surgery. Although pre-operative embolization of the tumours (Murphy and Brackmann, 1989) may significantly increase morbidity, it has proved useful under certain circumstances in reducing blood loss and operative time.

In the past many local recurrences had been observed after initial treatment because of incomplete removal of the tumour and a long follow-up was recommended. Today, great advances in technical and surgical capabilities, especially in the field of skull base surgery, ensure excellent results as compared with radiotherapy (Glasscock *et al.*, 1979). However, recurrences after successful initial treatment may still occur. Radiotherapy definitely holds a place in the management of glomus tumours, either in conjunction with surgery or as single treatment in advanced unoperable cases (Cole, 1979).

It appears that multiple glomus tumours pose specific problems in therapy. Frequent local and distant recurrences are observed in these patients after surgical excision of the initial lesions, resulting in a high rate of metachronously developing tumours in multiple sites.

In order to maintain a widely accepted therapeutic approach to patients with multiple glomus tumours, most authors should specify the adopted method of treatment and reevaluate the results after long-term follow-up. These results may be quite different from those of patients with solitary glomus tumours.

References

Ahmed, A., Dodge, O. G., Kirk, R. S. (1969) Chemodectoma of the orbit. *Journal of Clinical Pathology,* **22:** 584–588.

Alford, B. R., Guilford, F. R. (1962) A comprehensive study of tumours of the glomus jugulare. *Laryngoscope,* **72:** 765–787.

Anderson, R., Scarcella, J. V. (1963) Carotid body tumours. *American Journal of Surgery,* **106:** 856–859.

Azzarelli, B., Felten, S., Miyamoto, R., Muller, J., Purvin, V. (1988) Dopamine in paragangliomas of the glomus jugulare. *Laryngoscope,* **98:** 573–578.

Baars, F. V., Van Den Broek, P., Cremers, C., Veldman, J. (1981) Familial non-chromaffinic paragangliomas (glomus tumours): Clinical aspects. *Laryngoscope,* **91:** 988–996.

Barnes, L., Taylor, S. R. (1990) Carotid body paragangliomas: A clinico-pathologic and DNA analysis of 13 tumours. *Archives of Otolaryngology—Head and Neck Surgery,* **116:** 447–453.

Bartels, L. J., Pennington, J., Kamerer, D. B., Browarsky, I. (1990) Primary fallopian canal glomus tumours. *Otolaryngology—Head and Neck Surgery,* **102:** 101–105.

Bogdasarian, R. S., Lotz, P. R., Arbor, A. (1979) Multiple simultaneous paragangliomas of the head and neck in association with multiple retroperitoneal pheochromocytomas. *Otolaryngology—Head and Neck Surgery,* **87:** 648–652.

Brown, J. S. (1985) Glomus jugulare tumours revisited: A ten-year statistical follow-up of 231 cases. *Laryngoscope,* **95:** 284–288.

Bundgaard, T., Tandrup, O., Elbrond, O., Nordentoft, A. M. (1989) Treatment of glomus tumours: A retrospective survey. *Clinical Otolaryngology,* **14:** 155–160.

Capps, F. C. W. (1964) Carotid body tumours and their surgical management. *Proceedings of the Royal Society of Medicine,* **57:** 954–955.

Chambers, R. G., Mahoney, W. D. (1968) Carotid body tumours. *American Journal of Surgery,* **116:** 554–558.

Cole, J. M. (1979) Panel discussion: Glomus jugulare tumours of the temporal bone: Radiation of glomus tumours of the temporal bone. *Laryngoscope,* **89:** 1623–1627.

Conley, J. J. (1965) The carotid body tumour: A review of 29 cases. *Archives of Otolaryngology,* **81:** 187–193.

Cordell, A. R., Myers, R. T., Hightower, F. (1967) Carotid body tumours. *Annals of Surgery,* **165:** 880–886.

Davidson, J., Gullane, P. (1988) Glomus vagale tumours. *Otolaryngology—Head and Neck Surgery,* **99:** 66–70.

Demarino, D. P., Mueller, D. P., Maves, M. D., Yuh, W. T. C., Dolan, K. D. (1990) Imaging case study of the month: Multiple paragangliomas. *Annals of Otology, Rhinology and Laryngology,* **99:** 85–86.

Dent, T. L., Thompson, N. N., Fry, W. J. (1976) Carotid body tumours. *Surgery ,* **80:** 365–372.

Dickinson, P. H., Griffin, S. M., Guy, A. J., McNeil, I. F. (1986) Carotid body tumour: 30 years experience. *British Journal of Surgery,* **73:** 14–16.

Dutcher, P. O., Brackmann, D. E. (1986) Glomus tumour of the facial canal: A case report. *Archives of Otolaryngology,* **112:** 986–987.

Endikott, J. N., Maniglia, A. J. (1980) Glomus vagale. *Laryngoscope,* **90:** 1604–1611.

Farr, H. W. (1967) Carotid body tumours: A 30 year experience at Memorial Hospital. *American Journal of Surgery,* **114:** 614–619.

Gaylis, W., Mieny, C. G. (1977) The incidence of malignancy in carotid body tumours. *British Journal of Surgery,* **64:** 885–889.

Glasscock, M. E., Jackson, C. G., Dickins, J. R., Wiet, R. J. (1979) Panel discussion: Glomus jugulare tumours of the temporal bone: The surgical management of glomus tumours. *Laryngoscope,* **89:** 1640–1654.

Grufferman, S., Gillman, M. W., Pasternak, L. R., Peterson, C. L., Young, W. G. (1980) Familial carotid body tumours: Case report and epidemiologic review. *Cancer,* **46:** 2116–2122.

Heinrich, M. C., Harris, A. E., Bell, W. R. (1985) Metastatic intravagale paraganglioma: Case report and review of the literature. *American Journal of Medicine,* **78:** 1017–1024.

Hewitt, R.L., Ichinose, H., Weichert, R. F., Drapanas, T. (1972) Chemodectomas. *Surgery ,* **71:** 275–282.

Hodge, K. M., Byers, R. M., Peters, L. J. (1988) Paragangliomas of the head and neck. *Archives of Otolaryngology—Head and Neck Surgery,* **114:** 872–877.

House, J. M., Goodman, M. L., Gacek, R. R., Green, G. L. (1972) Chemodectomas of the nasopharynx. *Archives of Otolaryngology,* **96:** 138–141.

Irons, G. B., Weiland, L. H., Brown, W. L. (1977) Paragangliomas of the neck: Clinical and pathologic analysis of 116 cases. *Surgical Clinics of North America,* **57:** 575–583.

Irving, J. D. (1983) Angiography in the investigation of tumours of the ear: Value of digital vascular imaging. *Journal of Laryngology and Otology,* **97:** 319–331.

Jackson, G. G., Glasscock, M. E., Harris, P. F. (1982) Glomus tumours: Diagnosis, classification and management of large lesions. *Archives of Otolaryngology,* **108:** 401–406.

Javid, H., Chawla, S. K., Dye, W. S., Hunter, J. A., Najafi, H., Goldin, M. D., Serry, C. (1976) Carotid body tumour: Resection or reflection. *Archives of Surgery,* **111:** 344–347.

Javid, H., Dye, W. S., Hunter, J. A., Najafi, H., Julian, O. C. (1967) Surgical management of carotid body tumour. *Archives of Surgery,* **95:** 771–779.

Krupski, W. C., Effeney, D. J., Ehrenfeld, W. K., Stoney, R. J. (1982) Cervical chemodectoma: Technical considerations and management options. *American Journal of Surgery,* **144:** 215–220.

Lack, E. E., Cubilla, A. L., Woodruff, J. M. (1979) Paragangliomas of the head and neck region: A pathologic study of tumours from 71 patients. *Human Pathology,* **10:** 191–218.

Lack, E. E., Cubilla, A. L., Woodruff, J. M., Farr, H. W. (1977) Paragangliomas of the head and neck region: A clinical study of 69 patients. *Cancer,* **39:** 397–409.

Lattes, R. (1950) Nonchromaffin paraganglioma of ganglion nodosum, carotid body, and aortic-arch bodies. *Cancer,* **3:** 667–694.

Lawson, W. (1980) Glomus bodies and tumours. *New York State Journal of Medicine,* **80:** 1567–1575.

Lees, C. D., Levine, H. L., Beven, E. G., Tucker, H. M. (1981)

Tumours of the carotid body: Experience with 41 operative cases. *American Journal of Surgery,* **142:** 362–365.
Lewis, J. S. (1983) Surgical management of tumours of the middle ear. *Journal of Laryngology and Otology,* **97:** 299–311.
Marcuse, P. M., Chamberlin, J. A. (1956) Multicentric paragangliomas: Case report with demonstration of intravagal paraganglionic tissue at a previously undescribed level. *Cancer,* **9:** 288–292.
Mathews, F. S. (1915) Surgery of the neck. In *Operative therapeusis* (Johnson, A. B., ed.), Vol. 3, Appleton-Century-Crafts, Inc., New York. p. 315.
McGuirt, W. F., Harker, L. A. (1975) Carotid body tumours. *Archives of Otolaryngology,* **101:** 58–62.
Mcilrath, D. C., Remine, W. H. (1963) Carotid-body tumours. *Surgical Clinics of North America,* **43:** 1135–1144.
McPherson, G. A. D., Halliday, A. W., Mansfield, A. O. (1989) Carotid body tumours and other cervical paragangliomas: Diagnosis and management in 25 patients. *British Journal of Surgery,* **76:** 33–36.
Moore, G. R., Robbins, J. P., Sedle, D. L., Eitz-Hugh, G. S., Constable, W. C. (1973) Chemodectomas of the middle ear: A comparison of therapeutic modalities. *Archives of Otolaryngology,* **98:** 330–335.
Morris, C. G., Balas, P. E., Cooley, D. A., Crawford, E. S., Debakey, M. E. (1963) Surgical treatment of benign and malignant carotid body tumours: Clinical experience with 16 tumours in12 patients. *American Surgeon,* **29:** 429–437.
Murphy , T. P., Brackmann, D. E. (1989) Effects of preoperative embolization on glomus jugulare tumours. *Laryngoscope,* **99:** 1244–1247.
Oberman, H. A., Holtz, F., Sheffer, L. A., Majielski, J. E. (1968) Chemodectomas (nonchromaffin paragangliomas) of the head and neck: A clinicopathologic study. *Cancer,* **21:** 838–851.
Padberg, F. T., Cady, B., Persson, A. V. (1983) Carotid body tumour: The Lahey Clinic experience. *American Journal of Surgery,* **145:** 526–528.
Parkin, J. L. (1981) Familial multiple glomus tumours and pheochromocytomas. *Annals of Otology, Rhinology and Laryngology,* **90:** 60–63.
Pearse, A. G. E., Polak, J. M., Rost, R. W. D., Fontaine, J., Lelievre, C., LeDouarin, N. (1973) Demonstration of the neural crest origin of type I (APUD) cells in the avian carotid body, using a cytochemical marker system. *Histochemie,* **34:** 191–203.
Phelps, P. D., Cheesman, A. D. (1990) Imaging jugulotympanic glomus tumours. *Archives of Otolaryngology Head and Neck Surgery,* **116:** 940–945.
Phelps, P. D., Stansbie, J. M. (1988) Glomus jugulare or tympanicum?: The role of CT and MR imaging with gadolinium DTPA. *The Journal of Laryngology and Otology,* **102:** 766–776.
Pratt, L. W. (1973) Familial carotid body tumours. *Archives of Otolaryngology,* **97:** 334–336.
Rosen, I. B., Palmer, J. A., Goldberg, M., Mustard, R. A. (1981) Vascular problems associated with carotid body tumours. *American Journal of Surgery,* **142:** 459–463.
Rosenwasser, H. (1945) Carotid body like tumour involving the middle ear and mastoid bone. *Archives of Otolaryngology,* **41:** 64–67.
Rosenwasser, H. (1968) Glomus jugulare tumours: Monograph. *Archives of Otolaryngology,* **88:** 29–66.
Shamblin, W. R., Remine, W. H., Sheps, S. G., Harrison, E. G. (1971) Carotid body tumour (chemodectoma): Clinicopathologic analysis of 90 cases. *American Journal of Surgery,* **122:** 732–739.
Spector, G. J., Ciralsky, R., Maisel, R. H., Ogura, J. H. (1975a) Multiple glomus tumours in the head and neck. *Laryngoscope,* **85:** 1066–1075.
Spector, G. J., Ciralsky, R. H., Ogura, J. H. (1975b) Glomus tumours in the head and neck: III. Analysis of clinical manifestations. *Annals of Otology, Rhinology and Laryngology,* **84:** 73–79.
Spector, G. J., Sobol, S., Thawley, S. E., Maisel, R. H., Ogura, J. H. (1979) Panel discussion: Glomus jugulare tumours of the temporal bone: Patterns of invasion in the temporal bone. *Laryngoscope,* **89:** 1628–1639.
Staats, E. F., Brown, R. L., Smith, R. R. (1966) Carotid body tumours, benign and malignant. *Laryngoscope,* **76:** 907–916.
Stanley, R. J., Weiland, L. H., Neel III, H. B. (1986) Pain-inducing laryngeal paraganglioma: Report of the ninth case and review of the literature. *Otolaryngology—Head and Neck Surgery,* **95:** 107–112.
Sykes, J. M., Ossoff, R. H. (1986) Paragangliomas of the head and neck. *Otolaryngologic Clinics of North America,* **19:** 755–767.
Van Asperen de Boer, F. R. S., Terpstra, J. L., Vink, M. (1981) Diagnosis, treatment and operative complications of carotid body tumours. *British Journal of Surgery,* **68:** 433–438.
Vigor, W. N., Rainer, N. G., Basque, G. (1969) Cervical chemodectomas: Clinical consideration in 16 cases. *American Journal of Surgery,* **118:** 976–980.
Werner, P. E., Crockett, H. A., McSwain, B. (1968) Malignant carotid body tumour with mandibular involvement. *Oral Surgery,* **25:** 774–789.
Westbrook, K. C., Guillamondegui, O. M., Medellin, H., Jesse, R. H. (1972) Chemodectomas of the neck: Selective management. *American Journal of Surgery,* **124:** 760–766.
Wilson, H. (1964) Carotid body tumours: Surgical management. *Annals of Surgery,* **159:** 959–966.
Wilson, H. (1966) Carotid body tumours. *Surgery,* **59:** 483–493.
Wilson, H. (1970) Carotid body tumours: Familial and bilateral. Annals of Surgery, **171:** 843–848.
Wilson, M. A. (1979) Multiple chemodectomas. *Journal of Nuclear Medicine,* **20:** 905–906.
Zbaren, P., Lehmann, W. (1985) Carotid body paraganglioma with metastases. *Laryngoscope,* **95:** 450–453.
Zeman, M. S. Carotid body tumours of the trachea (1956) *Annals of Otology, Rhinology and Laryngology,* **65:** 960–962.

Address for correspondence:
Dimitris G. Balatsouras,
40–42 Charilaou Trikoupi,
18536 Piraeus,
Greece.

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