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Cervical paragangliomas: diagnosis, management and complications

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Abstract

Sixteen patients were diagnosed as suffering from cervical paragangliomas. Eleven patients (68.75 per cent) had twelve carotid paragangliomas (CPs), and five patients (31.25 per cent) had six vagal paragangliomas (VP). One CP (8.33 per cent) originated from paraganglia around the common carotid artery (CCA). Three cases of multiple paragangliomas are presented (18.75 per cent). In 80 per cent (4/5) of VP patients there was widening of the carotid bifurcation similar to that seen with CP. This widening occurred whenever the VP was large enough to grown in between the external carotid artery and internal carotid artery (ECA and ICA). Large VPs may displace the vessels either anterolaterally or anteromedially. Knowledge of the direction of the carotid displacement is essential to avoid intra-operative vascular injuries. Colour flow doppler ultrasound (CFD-US) was found to be a good non-invasive method for diagnosis of vascular neck swellings. It enabled the diagnosis of CP with 100 per cent accuracy, but it was not sufficient for diagnosis of high VP. A transcervical approach, cutting the digastric muscle and the styloid process with the attached ligaments and muscles, was sufficient for excision of most VP. However, midline mandibulotomy might be necessary with high VP. Vascular injuries occurred in 12.5 per cent (2/16) of patients. Superior laryngeal nerve and hypoglossal nerve paralysis occurred, respectively, in (2/11) and (1/11) of patients with CP. Vagal paralysis occurred in all patients with VP. Cerebrovascular accident and post-operative death occurred in one patient (6.26 per cent).

Key words: Carotid Artery, Common; Carotid Body Tumour; Vagus Nerve; Paragangliomas

Introduction

Paragangliomas arise from the extra-adrenal paraganglionic tissue derived from the neural crest which has been recognized as a part of the APUD (Amine Precursor Uptake and Decarboxylation) system.[1] These tumours are named by their site of origin. In the head and neck they commonly arise from the carotid body (carotid paraganglioma), the tympanic plexus (glomus tympanicum), the jugular bulb (glomus jugulare), the vagal ganglia (vagal paraganglioma), and the sympathetic ganglia (sympathetic paraganglioma). Other unusual sites have been reported, including the nose and paranasal sinuses, larynx and orbit.[1,2] Diagnosis of cervical paragangliomas is usually suspected clinically and is confirmed by angiographic demonstration of a hypervascular mass within the carotid bifurcation or in the parapharyngeal space displacing the carotid vessels.[3,4]

Patients and methods

A retrospective study was conducted on 16 patients with cervical paragangliomas who were admitted and treated at the Otolaryngology – Head and Neck Surgery Department in Alexandria University, Egypt and at Fakhry & Al Mohawis Hospital, Al Khobar – Saudi Arabia in the period from 1990–1999. Patients suspected of having cervical paragangliomas were subjected to ENT, head and neck, and cranial nerve examination.

All patients were examined by colour flow doppler ultrasound (CFD-US), contrast computed tomography (CT) of the skull base and neck, and bilateral carotid angiography. Patients were further assessed with: (1) serum catecholamine and 24-hour urinary VMA (Vanilly mandelic acid) to exclude any functional activity; (2) complete medical check up; (3) routine laboratory investigations; and (4) routine chest X-ray.

Patients with glomus jugulare or tympanicum were excluded from the study. Surgical excision of cervical paragangliomas was done in all patients without post-operative embolisation as it was not available. All specimens were examined histopathologically for confirmation of diagnosis. Carotid paragangliomas (CP) were excised via either a longitudinal neck incision anterior to the sternomastoid muscle (two

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patients) or a tranverse upper neck incision along a skin crease (nine patients). Vagal paragangliomas (VP) were excised via either a cervical lip splitting incision with midline mandibulotomy (one patient) or a cervical incision extending from the chin to the mastoid process with excision of the submandibular gland, cutting the digastric muscle and the styloid process with the attached ligaments and muscles (four patients). This was followed by dislocation of the mandible anteriorly to obtain good exposure of the skull base region.

The first step was wide exposure of the tumour, carotid sheath and surrounding cranial nerves. The carotid sheath was then opened with identification of the external carotid artery (ECA), the internal carotid artery (ICA), the common carotid artery (CCA), and the internal jugular vein (IJV) to allow distal and proximal control of the vessels above and below the tumour. Tumour dissection was performed in the subadventitial plane by sharp dissection and bipolar diathermy. In CP the dissection from the ICA was usually started posterolaterally because this was found to be the area least affected by the tumour. The tumour was then dissected from the CCA and ECA. In VP, complete exposure of the tumour and its dissection from the ICA, ECA, CCA, IJV and the surrounding cranial nerves was performed first. The vagus nerve was then cut above the tumour to allow its excision.

Results

Sixteen patients were diagnosed as suffering from cervical paragangliomas. Eleven patients (68.75 per cent) had 12 carotid paragangliomas (CP), and five patients (31.25 per cent) had six vagal paragangliomas (VP) (Table I). Tumours were right-sided in eight patients (six CP and two VP), (50 per cent), and left-sided in six patients (four CP and two VP), (37.5 per cent). There were two patients (12.5 per cent) with bilateral paragangliomas, one with bilateral CP and the other with bilateral VP (Figures 1 and 2). Male to female ratio was 11:5 in all patients, 8:3 in CP and 3:2 in VP. Age of patients ranged from 18–55 years. Three patients (two with CP and one VP) presented with an ipsilateral vagal paralysis.

Location of the tumour

Of CP's 91.66 per cent (11/12) originated from the intercarotid paraganglia and only one tumour (8.33 per cent) originated from paraganglia around CCA (Figure 1a–e). Of VP's 83.33 per cent (5/6) originated from the inferior nodose ganglion and only one tumour (16.66 per cent) originated from the superior nodose ganglion near the skull base.

Multicentricity

Multiple paragangliomas were found in 18.75 per cent (3/16) of patients.

(1) The first patient was a male aged 49 years and presented with CPs around the right CCA and within the left carotid bifurcation (Figure 1a–e), glomus tymanicum, and glomus jugulare tumours in both a synchronous and a metachronous pattern. This patient had had the right glomus tymanicum removed 20 years ago and he had mild lower motor neurone right facial paralysis since that time. In 1992 he presented with bilateral neck swellings of three years' duration and right vagal paralysis of six months' duration. The right swelling (15 × 6 cm) was low in the neck deep to the sternomastoid muscle, and extended from the level of the thyroid cartilage to the clavicle. It did not move on swallowing. The left swelling (4 × 4 cm) was below the angle of the mandible. CT and angiography showed bilateral vascular masses, one surrounding the right CCA posterolateral to the thyroid gland and the other within the left carotid bifurcation (Figure 1a,b,c,d). The right CP was excised first, followed by the left

TABLE I
CLINICAL PRESENTATION AND POST-OPERATIVE COMPLICATIONS OF CAROTID AND VAGAL PARAGANGLIOMAS

N	Sex	Age	Side	Type	Other lesion	FH	Vascular injury	Pre-op NP	Post -op NP	CVA
1	F	18 y	Rt	CP	–	–	–	–	Sup LN	–
2	M	48 y	Bil	Rt CCP Lt CP	Glom j Glom t	–	–	X	–	–
3	M	45 y	Lt	Rec CP	–	–	+	X	Sup LN	–
4	M	35 y	Lt	CP	Glom j	+	–	–	XII	–
5	M	38 y	Rt	CP	–	–	–	–	–	–
6	M	40 y	Rt	CP	–	–	–	–	–	–
7	F	46 y	Lt	CP	–	–	–	–	–	–
8	M	45 y	Rt	CP	–	–	–	–	–	–
9	F	38 y	Rt	CP	–	–	–	–	–	–
10	M	43 y	Rt	CP	–	–	–	–	–	–
11	M	37 y	Lt	CP	–	–	–	–	–	–
12	M	40 y	Lt	VP	–	–	–	X	X	–
13	M	38 y	Rt	VP	–	–	–	–	X	–
14	F	55 y	Lt	VP	–	–	–	–	X	–
15	F	50 y	Rt	VP	–	–	–	–	X	–
16	M	49 y	Bil	VP	–	–	+	–	X	+

FH = Family history; CVA = Cerebrovascular accidents; CP = Carotid paraganglioma; Sup LN = Superior laryngeal nerve; Rt CCP = Rt common carotid paraganglioma; Rec CP = Recurrent carotid paraganglioma; F = Female; M = Male; Y = Years; Rt = Right; Lt = Left; X = Present in 10th cranial nerve; Pre-op NP = Pre-operative nerve paralysis; Post-op NP = Post-operative nerve paralysis; VP = Vagal paraganglioma; Glom j = Glomus jugulare; Glom t = Glomus tympanicus; XII = Present in 12th cranial nerve; + = Present.

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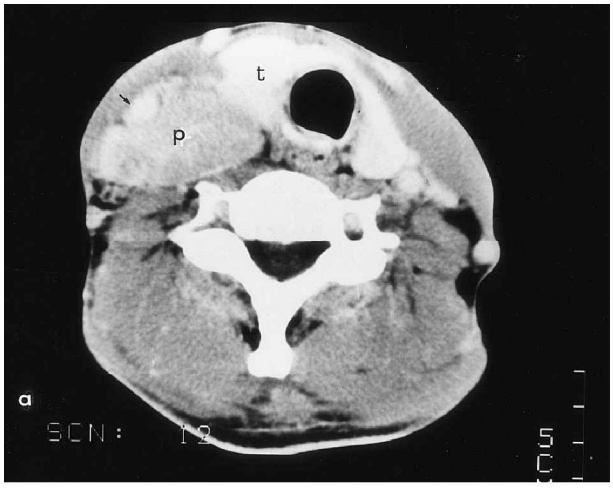

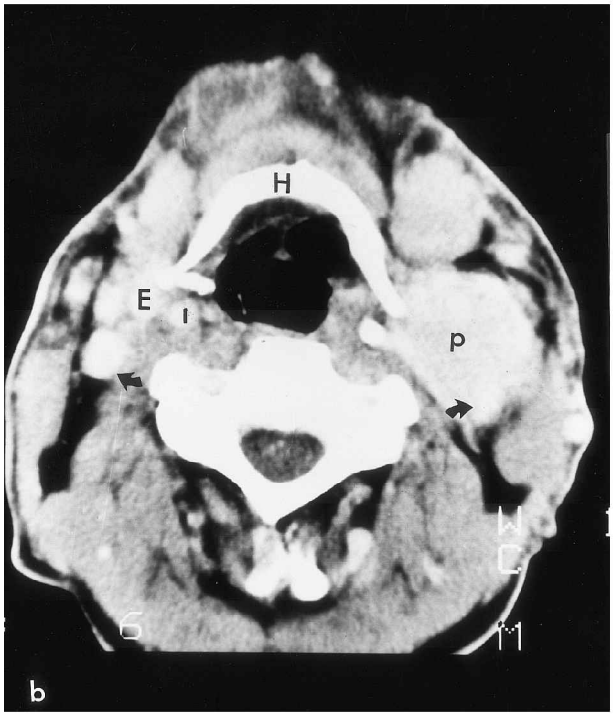

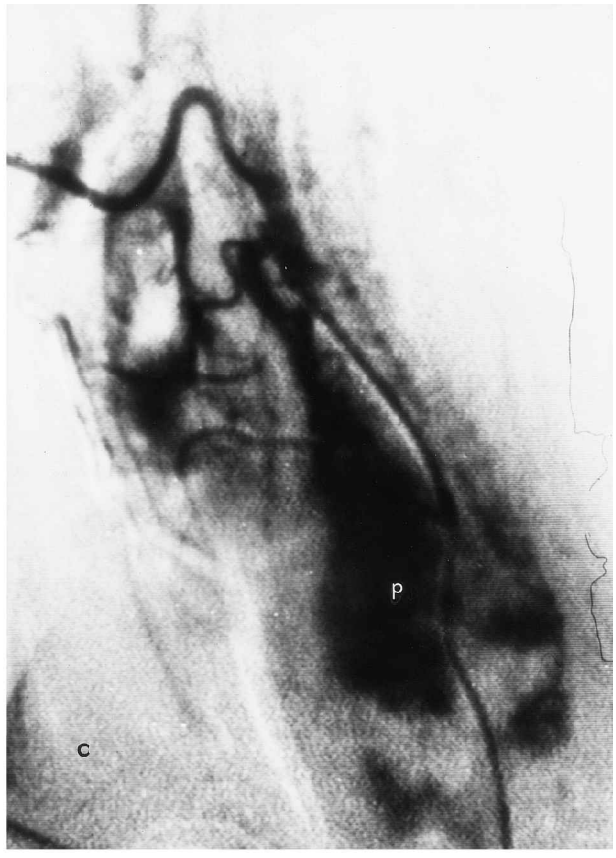

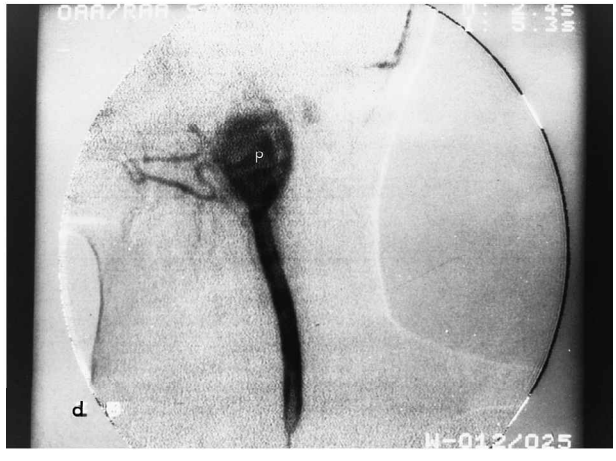

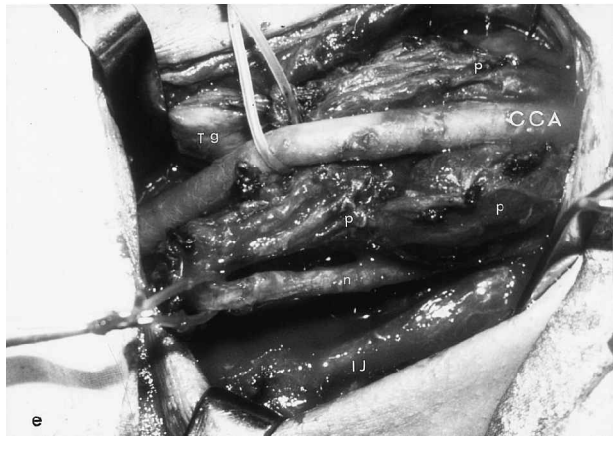

FIG. 1a

CT of patient 1 with bilateral CP showing a mass P around the right CCA (arrow) pushing the thyroid gland t anteriorly.

FIG. 1b

CT of patient 1 at the level of the hyoid bone (H) showing an enhanced mass P at the region of carotid bifurcation. E = external carotid artery, I = internal carotid artery, curved arrow – internal jugular vein.

FIG. 1c

Right carotid angiography of patient 1 showing a vascular paraganglioma P around the right CCA.

FIG. 1d

Left carotid angiography of patient 1 showing a vascular paraganglioma P between the carotid bifurcation.

FIG. 1e

Operative view of patient 1 showing a dissected paraganglioma P around the CCA, vagus nerve (n), internal jugular vein (IJ), thyroid gland (Tg).

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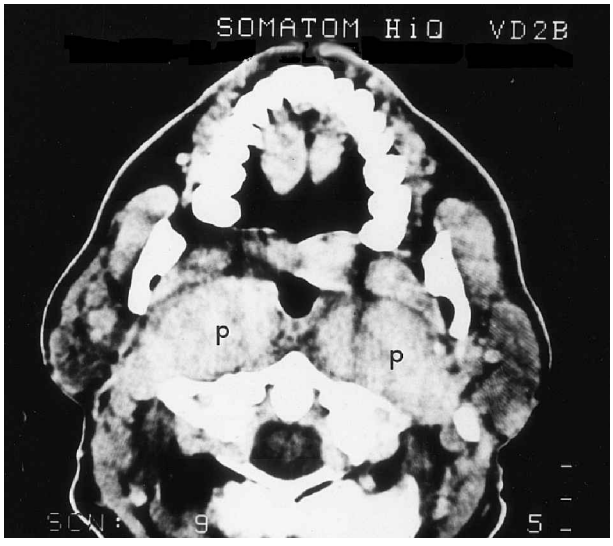

FIG. 2

CT of patient 2 with bilateral enhanced para- and retropharyngeal vagal paragangliomas P encroaching on the airway.

CP six months later. Histopathological examination confirmed the diagnosis of a paraganglioma. In 1995, the patient presented with total right facial paralysis. CT and angiography confirmed the diagnosis of glomus jugulare. The patient refused any surgery, and was sent for radiotherapy.

(2) The second patient was a male aged 50 years and presented with snoring, obstructive sleep apnoea, and huge tonsils. Tonsillectomy was performed but his complaint persisted. Post-operative examination revealed bilateral medial bulging of the pharyngeal walls. CT showed bilateral enhancing retrostyloid para- and retropharyngeal masses (Figure 2). The right tumour was larger and higher than the left. Angiography showed bilateral vascular swellings pushing the carotids anteriorly.

(3) The third patient was a male aged 30 years with a family history of glomus jugulare tumour. He presented with a left glomus jugulare and a left CP. Excision of both tumours was done at the same operation.

Family tendency

One patient with multicentric tumours (12.5 per cent) had a positive family history of paraganglioma (Table I).

Secretory functional activity

No secretory activity was found. Both serum catecholamine and 24-hour VMA were within normal range in all patients.

Size of the tumours

Tumour size after excision was (4–7 cm) × (5-15 cm) in CP and (4–8 cm) × (5–13 cm) in VP. The actual tumour size before excision was usually large due to its engorgement with blood.

CFD-US findings in cervical paragangliomas

CFD-US identified the presence of CP in all patients. It showed a vascular solid mass between the ECA and ICA with widening of the bifurcation. All masses had a hypoechoic and heterogeneous echo pattern with scattered small vascular structures showing pronounced turbulent multidirectional flow (hypervascular tumour flow). In four patients with VP, CFD-US identified the presence of a vascular tumour posterior to the carotid vessels. Its upper limit could not be reached as it extended upward medial to the angle of the mandible.

In the fifth patient, CFD-US failed to identify the bilateral high parapharyngeal VP near the skull base.

CT findings in cervical paragangliomas

CPs were located mainly between the ECA and ICA. In one patient, the tumour surrounded the CCA (Figure 1a–e). All tumours had a highly enhancing homogeneous appearance with well-defined margins except in a patient with a recurrent tumour. Tumour extension to the parapharyngeal space occurred in 18.18 per cent (2/11) of patients. The tumour completely surrounded the ECA in 81.82 per cent (9/11) of patients, the ICA in 27.27 per cent (3/11) of patients, and the CCA in 9.1 per cent (1/11) of patients. VPs were seen as enhancing homogeneous well-defined retrostyloid parapharyngeal masses. In two patients, the tumour had a retropharyngeal extension (Figures 2 and 3a). The carotid sheath was displaced anteromedially in two patients, anterolaterally in two patients, and anteriorly in one patient. In 80 per cent (4/5) of cases, there was a variable extension of the tumour between the ECA and the ICA (Figure 3a). There was no CT evidence of vascular invasion and there was no lymph node enlargement in any case.

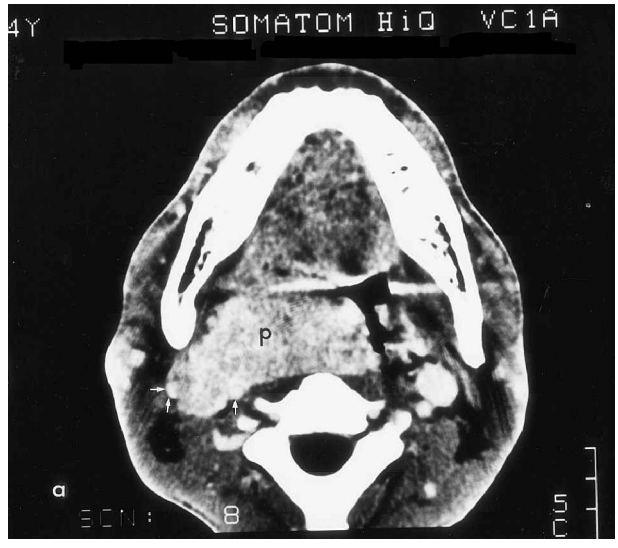

FIG. 3a

CT of patient 3 showing an enhanced retro- and parapharyngeal paraganglioma P extending between the ECA (double arrow) and the ICA (arrow).

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Angiographic findings in cervical paragangliomas

All patients with CP showed highly vascular masses within the carotid bifurcation with its typical widening. In one patient, there was a vascular mass below the bifurcation due to presence of a paraganglioma around the CCA (Figure 1a–e). Blood suply of the tumours was mainly from the ECA. Patients with VP showed a highly vascular mass medial to the mandible, displacing the carotid vessels anteriorly. In 80 per cent (4/5) of patients, there was widening of the carotid bifurcation similar to that seen with CP (Figure 3b).

TABLE II
CLASSIFICATION OF CAROTID PARAGANGLIOMAS[4]

Classification	Description of CP
Group I	Small tumours easily separated from the carotid vessels
Group II	Medium sized tumours that are intimately adherent to the vessels and may be separated from them by subadventitial dissection
Group III	Large tumours that encase the ICA at the bifurcation and often require partial or complete sacrifice of the vessel with replacement

CP = carotid paraganglioma.
ICA = internal carotid artery.

Operative findings

In all CP cases there was ballooning of the carotid sheath at the area of bifurcation. Multiple dilated tortuous vessels and dilated vasa vasourum and nervosa were found usually in the surrounding area. The tumour completely surrounded the superior laryngeal nerve in 63.63 per cent (7/11) of patients, the hypoglossal nerve in 45.45 per cent (5/11) of patients, the ECA in 81.81 per cent (9/11) of patients, the ICA in 27.27 (3/11) of patients, and the CCA in one patient (9.09 per cent). In 72.72 per cent (8/11) of cases, the posterolateral surface of the ICA was not covered by the tumour, so dissection was usually started at this area. According to the classification of Shamblin *et al.*,[4] (Table II), one tumour belonged to group I, seven tumours belonged to group II, and three tumours belonged to group III. We did not include the CCA paraganglioma in this classification as it was below the bifurcation.

Four VPs extended between the carotid bifurcation with ballooning of the carotid sheath simulating CP. The ICA and ECA were displacing anterolaterally in two patients (40 per cent), anteromedially in two patients (40 per cent), and anteriorly in one patient (20 per cent). After dissection of the tumour from the bifurcation it usually appeared as the tip of an iceberg as most of the tumour was found deep in the parapharyngeal space (Figure 3c, d).

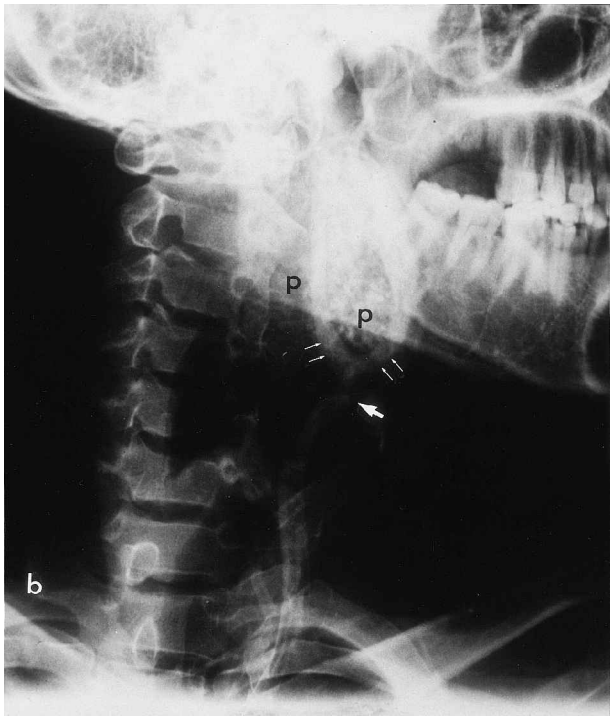

FIG. 3b

Angiogram of the patient 3 showing anterior displacement of the CCA (short arrow) with widening of the bifurcation (long arrows) due to extension of the paraganglioma P between the carotid vessels.

FIG. 3c

Operative view of patient 3 showing a paraganglioma VP arising from the vagus nerve (short arrow) and lying posteromedial to the CCA (curved arrow) and the hypoglossal nerve H, and extending between the carotid bifurcation.

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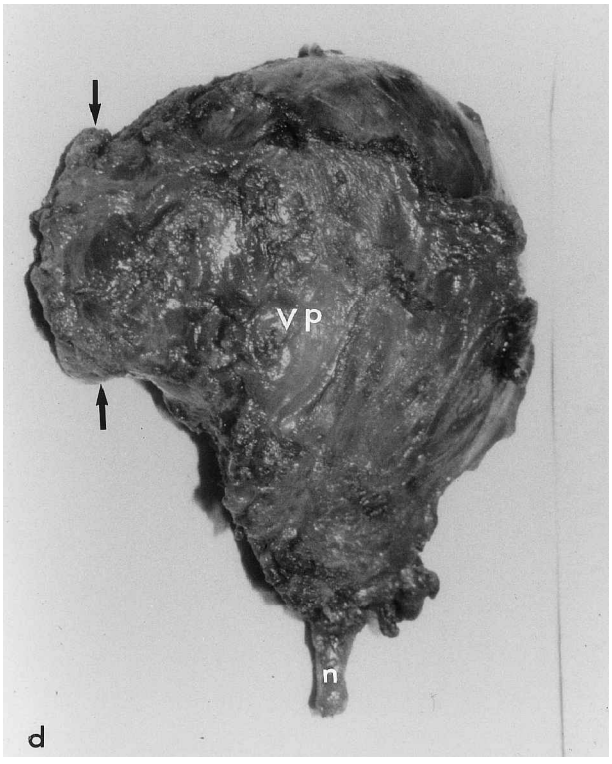

FIG. 3d

The excised specimen of patient 3 arising from the vagus nerve (n) and showing part of the tumour (VP) extending between the bifurcation (arrows).

Complication of surgery of cervical paragangliomas

(1) Vascular injuries and cerebro-vascular accidents: ICA injuries occurred in two patients (12.5 per cent). In the first, the injury was repaired during the operation with the use of intravascular shunt and ICA-CCA anastomosis, and the patient did not develop any neurological deficits (Table I). In the second, the injury was high, near to the skull base, and the ICA was ligated. The patient developed massive post-operative cerebral infarction and died one week later (Table I).

(2) Nerve deficits comprised superior laryngeal nerve paralysis in 18.18 per cent (2/11) of patients with CP, hypoglossal nerve paralysis in 9.09 per cent (1/11) of patients with CP, and vagal paralysis in all patients with VP.

Discussion

Carotid paragangliomas (CPs) are tumours that arise from the carotid paraganglia.[5] They are the commonest head and neck paragangliomas, representing about 60–78 per cent.[2,5,6,7] Equal sex incidence has been reported.[2] Vagal paragangliomas (VP) arise from the paraganglia within or adjacent to the vagus nerve at the level of the inferior nodose ganglion (intravagale), middle ganglion (juxtavagale), superior ganglion, or rarely at any part of the vagal trunk.[5,8,9] They represent 2.5–4.5 per cent of head and neck paragangliomas. Approximately 200 VPs have been reported, with increased female incidence.[8,9]

A heredofamilial tendency in cervical paragangliomas occurs in less than 10 per cent of cases.[6,10,11] Multiple, often bilateral, CPs represents about 10 per cent of sporadic cases,[4] and 25–33 per cent of familial cases.[10–12] Multicentricity occurs in 17 per cent of VPs,[12] whereas bilaterality occurs in four per cent of cases. Eight patients with bilateral VP have been reported.[6,8,13–15] Metastasizing CP occurs in less than 10 per cent and possibly less than 5 per cent of cases.[4,5] Malignancy is more frequent in VP than in CP.[2,16]

Incidence of CP in our study was 68.75 per cent, and that of VP was 31.25 per cent, which is higher than previously reported.[8,12–14] In contrast to previous reports, both CP and VP in our study were more common in males than in females.[2,4,8,9] Incidence of familial tendency in our cases (6.25 per cent) is similar to that reported previously.[10,11] One CP (8.33 per cent) originated below the bifurcation around the CCA in a patient with multiple synchronous and metachronous head and neck paragangliomas. We did not find any similar case in the literature. The presence of a paraganglioma at this location may be due to its origin from ectopic paraganglia on the CCA or due to downward growth of CP. Another explanation may be a rare origin of VP from the vagal trunk low in the neck, but this is a remote possibility because the tumour was found completely separate from the vagus nerve and completely surrounding the CCA.

Bilaterality of CP in our cases (9.09 per cent) is similar to that in previous reports,[4] whereas that of VP (20 per cent) is higher than reported previously.[6,8,13–15] Eight cases of bilateral vagal paragangliomas had been reported up to 1989.[6,8,13–15] Less than two per cent of head and neck paragangliomas have catecholamine secretory function,[16] and we did not find any functioning tumour in our study. Pre-operative vagal paralysis in our study, 18.18 per cent (2/11) of CP and 20 per cent (1/5) of VP, is similar to that reported previously.[2,4,8]

CFD-US has been suggested for an initial evaluation of pulsating neck masses to avoid the unnecessary neurological sequelae of angiography.[17] It identified CP in our patients with 100 per cent accuracy. These findings are similar to those of Barry *et al.* (1993).[18] Whenever a solitary solid, vascular mass is detected within the carotid bifurcation, the diagnosis of carotid paraganglioma should be considered.[17,18]

Although CFD-US can diagnose CP accurately, carotid angiography is still required pre-operatively for detection of multiple head and neck paragangliomas, delineation of the major feeding vessels, differential diagnosis from other vascular and non-vascular tumours, and assessment of circle of Willis and cross-cerebral circulation with the balloon occlusion test.[3,8] In VP, CFD-US could only identify a vascular neck tumour displacing the carotid vessels anteriorly. Diagnosis was suspected when there were clinical, CFD-US, CT, and angiographic findings of a parapharyngeal vascular solid tumour. However, diagnosis was confirmed intra-operatively when the

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tumour was found originating from the vagus nerve. CT features of CP and VP in our cases were similar to those published previously.[8,19,20] However, extension of CP into the parapharyngeal space in our cases (18.18 per cent) was higher than that reported (5–10 per cent).[5,21] This may be due to the large size of tumours in our study. Details of the direction of carotid displacement in VP are of great importance pre-operatively to avoid vascular injury during surgery. We believe that a small VP usually displaces the carotid vessels anteriorly, whereas large VP may displace the vessels either anterolaterally or anteromedially depending on the direction of maximum tumour growth. Extension of VPs between the ECA and ICA in our cases may be also due to the large size of these tumours in our series.

Angiographic findings in our cases were similar to those previously reported;[2,8,13–15] however, 80 per cent of VPs showed widening of the carotid bifurcation. This contradicts the belief of some authors that VP does not cause widening of the carotid bifurcation.[20,21] We think that VP can cause widening of the carotid bifurcation whenever it is large enough to grow anteriorly between the ECA and ICA. Misdiagnosis of VP with widening of the carotid bifurcation as a CP may be the cause of the decreased incidence of VP in literature.[8,9,12–15,20] Shamblin *et al.* (1971) classified CP into three groups; group I, small tumours easily separated from the carotid vessels; group II, medium-sized tumours that are intimately adherent to the vessels and may be separated from them by careful subadventitial dissection; and group III, large tumours that encase the ICA at the bifurcation and often require partial or complete sacrifice of the vessel with replacement (Table II).[4] We used the same classification in our study. Only one tumour of the three classified as group III needed an internal by-pass with ICA-CCA anastomosis due to injury at the bifurcation during dissection. The other two tumours could be dissected from the ICA without any injury. In most CP, the posterolateral surface of the ICA was the only area exposed, and we agree with Conley (1963) that this is the best area to start dissection and release of the ICA.[22]

Histopathological examination of all tumours was consistent with that of paraganglioma with no evidence of malignancy.[5] Lack *et al.* (1979) considered a paraganglioma malignant if it showed two of the following features: central necrosis of the Zellballen, invasion of vascular and lymphatic spaces, and presence of mitotic features.[2] However, most authors agree that the reliable criteria of malignancy are the presence of lymph node involvement or distant metastasis.[4,23,24]

Pre-operative cranial nerve involvement occurs in about 30 per cent of VPs and 20 per cent of CPs.[4,8,25] In our study, pre-operative vagal paralysis occurred in one case of VPs (20 per cent) and two cases of CP (18.18 per cent). One case with CP presented with a recurrent tumour after three operations, whereas in the second case the tumour was around the CCA and adherent to and compressing the vagus nerve. We had only two cases (18.18 per cent) of transient aspiration and superior laryngeal nerve paralysis after surgery for CP. In the first case there was nerve injury in the patient with the recurrent extensive tumour, whereas in the second case the nerve was intact. Paralysis may have been due to neuropraxia caused by manipulation of the nerve, since both cases improved after one month of nasogastric feeding and rehabilitation. Post-operative hypoglossal nerve paralysis occurred in one patient with CP (9.09 per cent), who also had an ipsilateral glomus jugulare. Both tumours were operated upon at the same time, so the nerve paralysis may have been caused by the surgery at the skull base, as the nerve was seen to be intact in the neck. Anand *et al.* (1995) found cranial nerve deficits in 50 per cent of his patients with CP, and in 21.8 per cent in his review of 1181 CP patients.[25] Leonetti *et al.* (1997) found 11 cranial nerve injuries in 50 (8/16) of his CP cases.[26] ICA injury during CP surgery was reported to occur in 22–23.2 per cent.[25] We had only one case of carotid injury at the bifurcation in a recurrent CP which was adherent to the vessel wall. Conley (1963) found that the tumour is most adherent in the region of bifurcation and this is the area where injury is most likely to occur.[22] We did not have any mortality in our patients with CP although the overall mortality rate for surgical intervention in CP was reported to be 3.2 per cent. Central nervous system complications were found in 6.3 per cent of cases.[22]

Post-operative vagal paralysis occurred in all patients with VP because the sacrifice of the vagus nerve was essential for excision of these tumours. Post-operative aspiration was present in all patients, however, it was mild and transient in patients with pre-operative vagal paralysis. All patients had a temporary tracheotomy and nasogastric feeding for two weeks. It has been suggested that intra-operative cricopharyngeal myotomy and Teflon injection of the paralysed vocal fold one week post-operatively would facilitate rehabilitation, improve the voice, and diminish aspiration.[8,12] Death occurred one week post-operatively in one patient with a high VP at the skull base due to injury and ligation of the ICA.

Conclusions

(1) VP can cause widening of the carotid bifurcation when it is large enough to grow anteriorly between the ECA and ICA.
(2) Small VPs usually displace the carotid vessels anteriorly, whereas large VPs may displace the vessels either anterolaterally or anteromedially. Knowledge of the direction of the carotid displacement is essential to avoid intra-operative vascular injuries.
(3) CFD-US is a good non-invasive screening method for differentiation between vascular and non-vascular neck swellings. It enables diagnosis of carotid paragangliomas with 100 per cent accuracy, but its use is not sufficient for diagnosis of high parapharyngeal vagal paragangliomas.

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(4) Transcervical approach, with cutting the digastric muscle and the styloid process with the attached ligaments and muscles and dislocation of the mandible, may be sufficient for excision of large VP. However, midline mandibulotomy may be necessary in high VPs near the skull base.
(5) Carotid paraganglioma may arise below the bifurcation around the common carotid artery.

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