

THE CAROTID BODY AND ITS TUMOR

II. Chemodectomas. Report of Five Operated Cases.

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SUMMARY :

Clinical and radiological data on five operated cases of carotid body tumors, chemodectomas, are presented. Surgical techniques, end results and experience gained are reported. National medical literature relating chemodectomas is reviewed.

KEY WORDS :

Carotid body chemodectoma.

INTRODUCTION

Carotid body tumors (CBT) are uncommon lesions relating at least three different surgical specialties: neurosurgery, cardiovascular surgery, otolaryngology. Therefore the literature on the subject is voluminous and dispersed. The first attempt at surgical excision of a carotid body tumor was by Reigner in 1880 (7). Currently there are more than 700 case reports of CBT in the literature (3, 4, 7, 8). Reports of CBT in the Turkish medical literature are scarce (1, 5, 10). In this article five operated cases of CBT are presented.

PATIENTS AND METHODS

Patient Characteristics : Seven patients (5 females, 2 males) with CBT were admitted to the neurosurgery, cardiovascular surgery, and otolaryngology clinics of the Social Security Association Ankara Hospital within a span of two years between 1987-1989. Two patients with definite radiological and pathological diagnoses of CBT deferred surgery. Five patients (3 females, 2 males) aged between 23-41 years (average 31.6 years) were operated.

Symptoms and Signs : The single common to all patients was a slowly growing high cervical mass. Symptom duration was between 2-6 years (average 3 years). None of the patients complained of dysphagia, tenderness, hoarseness, tongue weakness or symptoms that could be attributed to central

nervous system dysfunction. Local examination revealed a firm and relatively immobile neck mass of 5-10 cm dimensions, located below the angle of the mandible in all our patients. Neurological examination was normal in this patient group.

Investigative Procedures : Two patients were biopsied before they were admitted to hospital, and presented with their pathological diagnoses. Biopsy is not performed as the first diagnostic procedure in our hospital.

All our patients had carotid angiograms preoperatively (5/5). Four had preoperative cervical computed tomographic (CT) scans (4/5). Electroencephalograms with compression of the carotid artery at the affected side were performed in all our patients (5/5). Postoperative carotid angiograms and cervical CT scans could be performed on only three patients: one expired postoperatively, one was lost to follow-up.

Surgical Technique : All patients were operated under endotracheal general anesthesia. A standard skin incision along the medial border of the sternocleidomastoid muscle, extending from the angle of mandible to 4 cm above the jugulum was made. This incision was necessarily on the tumor mass. The common carotid artery (CCA) was dissected as the first step at the proximal end of the operative wound. Dissection was continued distally and over the tumor capsule. Branches of the external carotid artery (ECA) were coagulated and divided as needed. The internal carotid artery (ICA) and ECA were dissected at

the most distal border of the tumor, taking care of the lower cranial nerves.

An **en bloc** excision including the tumor and the carotid bifurcation (CB), was performed in two patients. Vascular continuity was restored by a CCA-ICA polytetrafluoroethylene (PTFE) graft in these patients. The CCA and ICA were clamped for 25 and 40 minutes during the grafting procedures. Intraoperative shunt was not used.

A subadventitial plane of dissection was created in the remaining three patients. Total excision was performed in two, and a subtotal excision was made in one patient. The ECA was ligated in one of the patients with total excision. The ECA and ICA were intact in the remaining two.

RESULTS

One patient with **en bloc** resection and CCA-ICA PTFE graft expired in the early postoperative period due to hemorrhagic shock in spite of emergency reexploration and CCA and ICA ligation with removal

of the graft. Hemorrhage which was mainly from the distal anastomosis site was possibly due to a transfusion complication.

The other patient with **en bloc** resection and PTFE graft had postoperative dysphagia, hoarseness, and hypoglossal nerve palsy. Postoperative angiography showed complete obliteration of the graft. The patient did not have any hemispheric deficit.

One of the two patients with total excision had postoperative dysphagia, hoarseness and hypoglossal nerve palsy. The other was neurologically intact.

Our sole patient with subtotal excision was also neurologically intact postoperatively.

The patency of the carotid arteries was demonstrated angiographically in two of these latter three patients. One was lost to follow-up.

Morbidity and mortality figures are 50 % and 20 % respectively in this series of CBT.

A summary of patient data is given in Table 1.

Table 1 : A summary of clinical and surgical findings

Patient	Age/Sex	Symptom Duration	Surgical Disk Group-Shamblin(%)	Surgical Procedure	Follow-up Period	Outcome
1	28 F	6 years	3	en bloc excision CCA-ICA graft	—	Exitus
2	22 F	2 years	3	en bloc excision CCA-ICA graft	19 months	Satisfactory (hoarseness-dysphagia hypoglossal palsy)
3	33 K	2 years	2	total excision ECA ligation	22 months	Satisfactory (hoarseness-dysphagia-hypoglossal palsy)
4	41 M	3 years	2	subtotal excision	12 months	No neurological deficit
5	33 F	3 years	1	subtotal excision total excision	12 months lost to follow-up	No neurological deficit No neurological deficit in the early postoperative period

Histopathological examination showed chemodectoma (non-chromaffin paraganglioma) of the carotid body in all our patients (Figure 1). Microscopic criteria of malignancy were not encountered in this patient group.

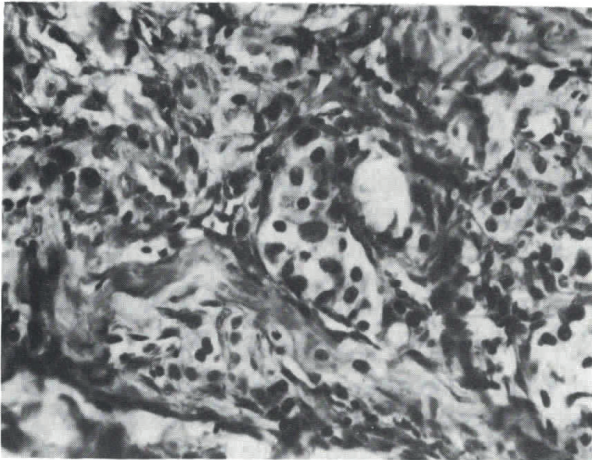


Fig.1 : Chemodectoma (Case 3). Clusters of tumor cells "Zellballen" are visible. Pathological criteria of malignancy are not encountered. (H&E, original magnification X200)

ILLUSTRATED CASE REPORTS

Case 2 : This 22-year-old woman presented with right-sided neck mass of two years' duration. The mass was biopsied in another hospital, and a diagnosis of "paraganglioma" was made. Angiography revealed a CBT (Figure 2a). **En bloc** excision with CCA-ICA PTFE graft was performed. Postoperatively she had dysphagia, hoarseness and ipsilateral hypoglossal nerve palsy. Control angiography showed complete obstruction of the graft (Figure 2b). She did not have any hemispheric symptom or sign.

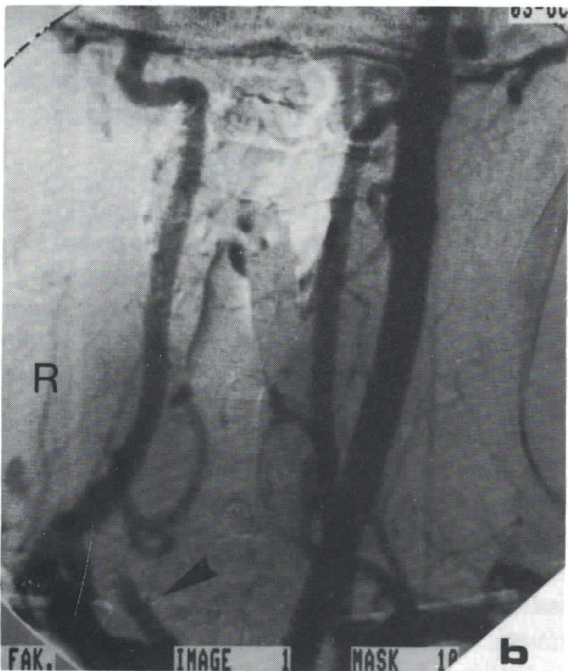


Fig.2 b) : Postoperative aortogram shows total occlusion of the synthetic CCA-ICA graft (arrow).

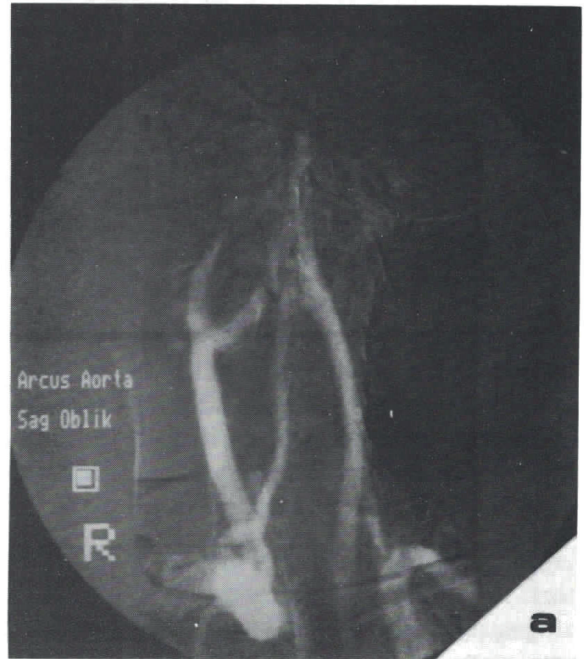


Fig.2 a) : Case 2. Aortogram shows widening of the carotid bifurcation which is diagnostic in CBT. Tumor vascularity is not visible in this early phase arteriogram.

Case 3 : This 33-year-old male patient presented with left-sided neck mass of two years' duration. Previous biopsy in another medical center revealed a "glomus tumor". Carotid angiography showed a highly vascular CBT (Figure 3a). The tumor

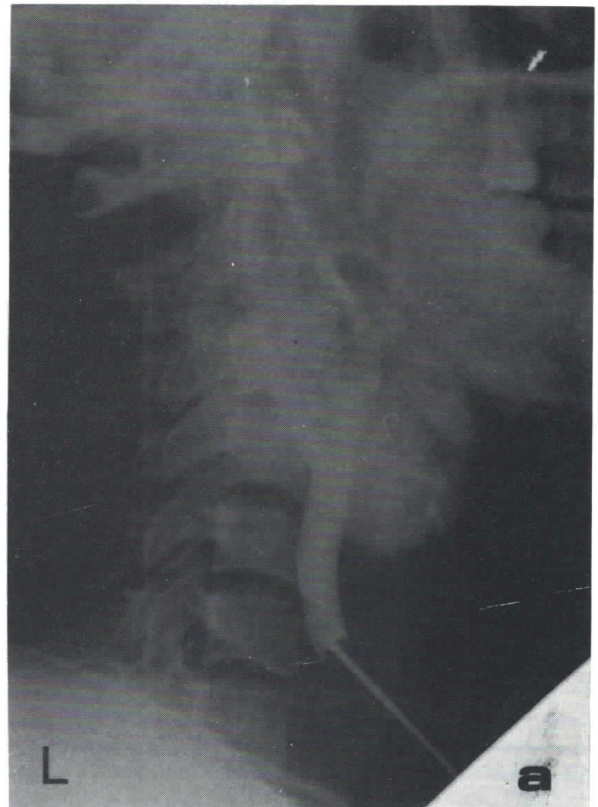


Fig.3 : Case 3. a) Left carotid angiogram shows widening of the carotid bifurcation with a highly vascular globular mass.

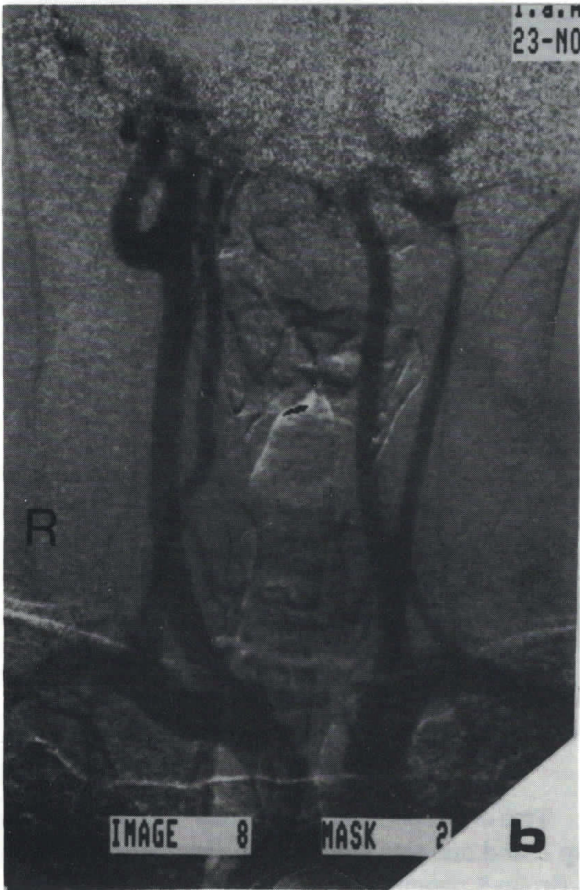


Fig.3 : b) Postoperative aortogram shows CCA-ICA patency. ECA is ligated.

was totally resected protecting the CCA-ICA patency. The ECA was ligated. Postoperatively he had dysphagia, hoarseness, and ipsilateral hypoglossal nerve palsy. Control angiography confirmed the CCA-ICA Patency (Figure 3b).

Case 4 : This 41-year-old male patient was admitted with a right-sided neck mass of three years' duration. Carotid angiography showed a vascular CBT (Figure 4a). A subtotal excision was performed protecting the carotid arteries. Postoperative course was uneventful. Control angiography revealed residual tumor tissue (Figure 4b). Pre-and postoperative cervical CT scans were consistent with the angiographic data (Figure 5a and 5b).

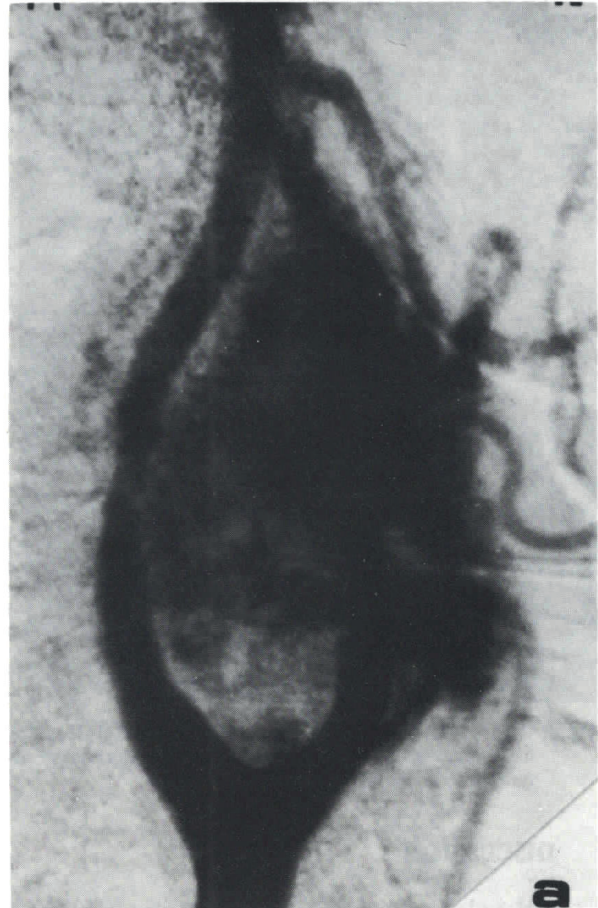


Fig.4 : a) Carotid angiogram displays all the radiological characteristics of a CBT.



Fig.4 : b) Postoperative angiogram shows subtotal excision. Normal configuration of the carotid bifurcation is restored to a great extent, but a dorsally placed residual tumor is diffusely staining.



Fig.5 : Case 4. a) Preoperative contrast enhanced cervical CT of the CBT. Relation of the tumor with adjacent anatomical structures are clearly displayed.

DISCUSSION

A detailed subject review on CBT was published by Meyer et al. (7) in 1986. These authors stated that there were approximately 600 case reports of CBT in the literature. This number has reached more than 700 in the meantime (2, 3, 4).

The morbidity and mortality figures in CBT directly correlate with the extent of tumor growth, and therefore with surgical technique and extent of resection. A classification proposed by Shamblin et al. (9) in 1971 has gained wide acceptance (Figure 6). Group 1 tumors can be resected without significant trauma to the vessel wall or to the tumor capsule. Group 2 tumors surround the vessel wall or to the tumor capsule. Group 2 tumors surround the vessels partially and are more adherent to the vessel adventitia. Nevertheless a total resection is possible with careful subadventitial resection. Group 3 tumors have an intimate and adherent relationship to the entire carotid artery complex, therefore surgical dissection and total removal without injury and/or **en bloc** resection of the vessels is nearly impossible. Surgical repair or reconstruction of the vessel wall becomes mandatory.

Complicated arterial reconstruction (patch, graft or end-to-end anastomosis) increases the postoperative complication rate in Group 3 tumors (4).



Fig.5 : b) Postoperative contrast enhanced cervical CT shows the parapharyngeal residual tumor tissue after subtotal excision.

Two of our patients were Group 3, two were Group 2, and one was Group 1. Our sole mortality was a Group 3 patient with arterial reconstruction. One Group 3 and one Group 2 patient had postoperative permanent cranial nerve deficits. One Group 2 patient was neurologically intact postoperatively due to a limited surgical approach with subtotal excision. Our only Group 1 patient was also neurologically intact after total tumor excision. Stroke was not encountered in this group of CBT. These findings are consistent with literature data (4, 7).

High rates of stroke, 3.8 % and 2.7 % (2,5 respectively) and injury to the lower cranial nerves, 19.2 %, 14.0 %, 40.0 %, 39.0 % (2, 3, 4, 7 respectively) have raised the question of whether an expectant attitude might be more favorable than surgical resection (4). But recent advances in surgical techniques, such as intraoperative monitoring of cerebral blood flow, selective use of intraoperative shunts, and a recently proposed Y shaped incision which allows mobilization of the parotid gland at the distal pole of the tumor seems to further reduce postoperative complications (4, 7). Therefore an expectant attitude is reserved for older patients in poor health, and for expectant attitude is reserved for older patients in poor health, and for patients harboring bilateral tumors with cranial nerve after unilateral resection (7).

Local recurrence after total surgical resection is reported to be 5 to 6 % (4, 8). Regional and/or distant

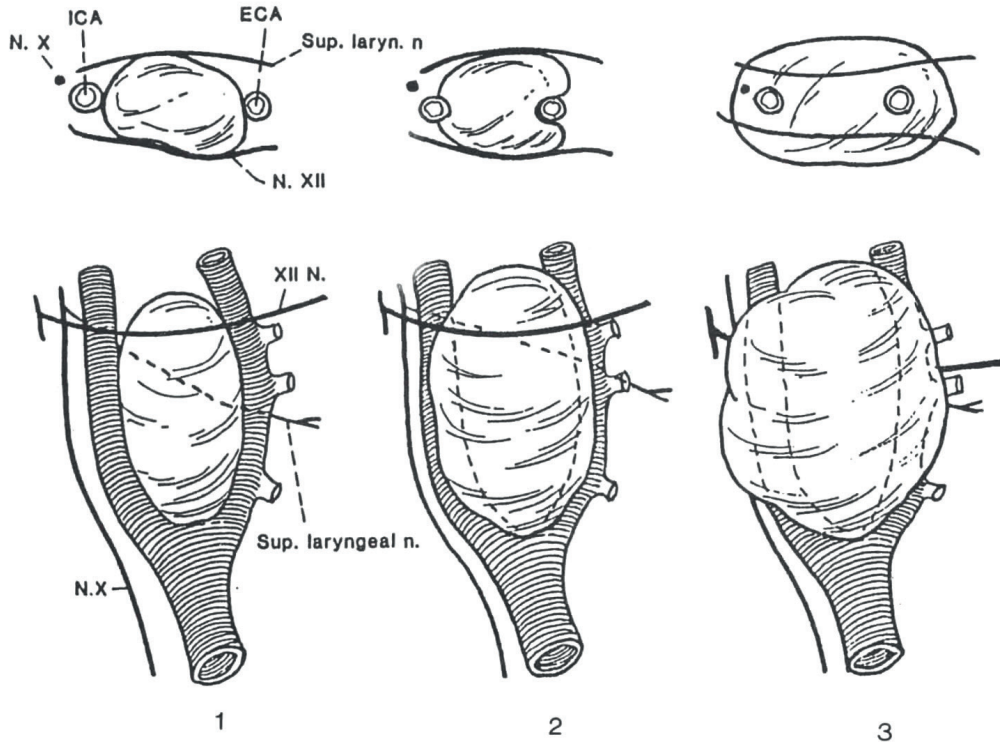


Fig.6 : Shamblin et al's surgical classification of the CBT. (From Meyer et. al. (7))

lymph node, and distant organ metastases are reported to be 28.0, 1.7, 18.0, and 2.0 % (3, 4, 6, 8 respectively). Histopathological criteria of malignancy were not encountered in our group of CBT. Local recurrence, lymph node or distant organ metastases were not detected in the follow-up period either.

We were able to find only three case reports of CBT in the Turkish medical literature (1, 5, 10). Two cases underwent **en bloc** resection without arterial reconstruction and without significant neurological sequelae (5, 10). In one case the tumor was excised totally with preservation of the arteries (1). A spinal extradural metastasis was successfully treated as well in one of these cases (5).

Our series reported in this article represents the preliminary experience of team work on CBT. The high mortality rate (20 %) is due to the limited number of cases. The morbidity rate (50 %) is near the values reported in the literature. Conclusions drawn from the surgical treatment of these patients are as follow: 1) Group 1 tumors can receive surgical resection without vessel injury, 2) Group 2 tumors can also be totally resected with preservation of the vessels, but cranial nerve palsies are a more frequent consequence. Utilization of a Y-shaped incision with mobilization of the parotid gland may prevent cranial nerve palsies, 3) Group 3 tumors must also be carefully dissected taking utmost care of the

cranial nerves. Surgery of Group 3 tumors frequently results in vessel injury which necessitates vascular repair. **En bloc** resection must be attempted as the last resort. Morbidity and mortality rates in this group with or without vascular repair are noticeably higher. Subtotal excision with periodic follow-up examinations may be considered as an alternative to total resection.

REFERENCES

- Aslan R, Kiper H, Kurtaran Y: Paraganglioma. *Anadolu Tıp Dergisi* 2:393-397, 1980
- Dickinson PH, Griffin SM, Guy AJ, et al: Carotid body tumor: 30 years experience. *Br J Surg* 73:14-16, 1986
- Gaylis H, Davidge-Pitts K, Pantanowitz D: Carotid body tumors. A review of 52 cases. *S Afr Med W* 72:493:493-496, 1987
- Hallett JW, Nora JD, Hollier LH, et al: Trends in neurovascular complications of surgical management for carotid body and cervical paragangliomas: a fifty-year experience with 153 tumors. *J Vasc Surg* 7:284-289, 1988
- Keçik C, Uluşu N, Erkus S, et al: Paraganglioma (Carotid body tumor). *Türk Otolarengoloji Arşivi* 26:108-112, 1988
- Martin CE, Rosenfeld L, McSwain B: Carotid body tumors: A 16-year follow-up of seven malignant cases. *S Med J* 65:1236-1243, 1973
- Meyer FB, Sundt TM, Pearson BW: Carotid body tumors: A subject review and suggested surgical approach. *J Neurosurg* 64:377-385, 1986
- Nora JD, Hallett JW, O'Brien PC, et al: Surgical resection of carotid body tumors: Long-term survival, recurrence, and metastasis. *Mayo Clin Proc* 63:348-352, 1988
- Shamblin WR, ReMine WH, Sheps SG, et al: Carotid body tumor. Clinicopathologic analysis of ninety cases. *Am J Surg* 122:732-739, 1971
- Ünlü Ş, Seber N, Timurkaynak E, et al: Karotid body tümörü. *BEGV Dergisi* 1:69-72, 1988