CAROTID BODY TUMOURS

WITH CASE PRESENTATION AND ANGIOGRAPHIC DEMONSTRATION

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There exists a considerable difference of opinion about tumours arising from the carotid body. Part of the controversy concerns the degree of malignancy, if any, of the tumours. This has a direct bearing on the treatment since resection of these tumours at the stage when the patients usually present themselves is technically difficult and frequently hazardous. Further controversy centres around the feasibility of removing the tumours without sacrificing the carotid bifurcation, and the dangers inherent in triple ligation of the common, internal and external carotid arteries, if resection of the bifurcation is deemed necessary.

Carotid body tumours are rare, and it is unlikely that any surgeon will encounter more than a few in the course of his career. As a result, one should benefit from the experience of others, as Monro[15] has indicated, and a periodic review of the situation, with reorientation of one's ideas, if necessary, is in order. The reader is referred to excellent reviews by Morfit *et al.*[16] and Byrne.[23] One further case is presented below, and the value of arteriography in diagnosis is indicated. This procedure has received scant attention, only 4 references to it having been found in the literature (Lichtenauer,[13] Idbohrn,[9] Wetzel[22] and Lipschitz[14]).

ANATOMY AND PHYSIOLOGY

The carotid body was first described by Von Haller in 1743 as the 'ganglion minutum'. It is situated in the carotid bifurcation, surrounded by a capsule which sends fibrous septa into the gland, subdividing it into lobes and lobules. It is supplied, usually from the external carotid artery, through the ligament of Mayer, which attaches it to the bifurcation. The degree of vascularization of the tumour varies very much, with a consequent variation in the consistency of the tumour.

Histologically the normal carotid body consists of polyhedral cells with finely granular cytoplasm, and round or oval, dark, eccentric nuclei. The cells are pervaded by a rich network of capillaries which may show sinusoidal dilatation.

The carotid body functions as a chemoreceptor, playing a part in the control of respiration. It is probably not called into play under normal physiological conditions, 'but as a last line of defence against respiratory failure' (Best and Taylor).

Boyd[1] has shown that the carotid body develops in the adventitial layers of the artery and that, in the mature state, it maintains this relationship. Morfit *et al.*[16] and Byrne[23] emphasize that tumours developing from the cartoid body preserve this relationship, even in the most advanced stage. This is obviously of very great importance from the surgical point of view, because it enables the surgeon to dissect the tumour from the artery in the subadventitial plane, leaving the tunica media inviolate. Gordon-Taylor[8] has described the 'white line' of demarcation which can be demonstrated by very careful dissection.

PATHOLOGY

The carotid body tumour lies in the fork between the internal and external arteries, splaying them apart (Fig. 1). The constituent cells are usually arranged in sheets, but may on occasion show an adenomatous or peritheliomatous arrangement. Great difficulty arises in the attempt to assess histologically malignancy of these tumours. It is apparent from numerous reports in the literature that precise histological criteria of malignancy in these tumours are practically impossible to determine so that more reliance must be placed on follow-up studies to predict their clinical behaviour.

It is in this aspect that there are such conflicting reports. Le Compte,[12] reporting under the auspices of the Subcommittee on Oncology of the National Research Council in America, states that the great majority of these tumours are both clinically and histologically benign, so that it is doubtful whether anything more than a diagnostic biopsy should be done in the symptomless and slowly growing tumours. Lahey and Warren[10] state that malignancy, when it occurs (in 15-20% of cases), is of low grade with involvement of local lymph nodes, but no proved haematogenous dissemination. Harrington, Clagett and Dockerty[8] found 50% (10 of 20) of their cases to be malignant. Monro[15]

stated the incidence of malignancy to be 12% in the cases he reviewed.

The ability of these tumours to produce distant haematogenous metastases, which would constitute unequivocable proof of their malignant potentialities, has been hotly disputed by many authors. Morfit *et al.*[16] have collected a formidable body of evidence from the literature in support of the view that these tumours can, and do, spread by the blood stream. Thus Donald and Crile[3] report a case which spread to the vertebrae, ribs, manubrium and ilium. Goodof and Lischer[5] had a case which spread to the pancreas. Prendergrass and Kirsch[18] report spread to the pelvis, femur, ribs and sternum. Spotnitz[21] reports spread to the cervical glands, skin, kidneys, pancreas, heart and pleura. Grönberger[7] studied a case which spread to the kidney and mediastinum. The case of Gilford and Davis[4] metastasized to the liver. Authors have objected to many such cases on the grounds of inadequate date supplied, or, in some cases, erroneous interpretation of histological appearances. Morfit *et al.*,[16] however, report one of their patients who died with metastases to the heart, lungs, ribs and vertebrae after resection of an advanced primary growth. Sections from these metastases were circulated to many leading pathologists in America, and these concurred, without exception, in the diagnosis of metastases from a carotid body tumour.

This evidence of distant dissemination would appear to be irrefutable; and if one further considers Monro's observation[15] that in 30% of cases left untreated, or treated inadequately, e.g. by curettage, death was directly attributable to the tumour, the direct bearing these facts have on the aggressiveness with which the tumours should be approached becomes apparent.

CLINICAL PRESENTATION

1. *The Swelling*

A carotid body tumour usually presents as a symptomless, very slowly growing lump in the upper anterior triangle of the neck. The lump is round or oval, and characteristically can be moved fairly freely in a lateral direction, but hardly at all in a vertical direction. Monro[15] emphasizes a great variation in the consistency of these tumours, depending on the degree of vascularization. They are usually solid, but, if vascular enough to appear cystic, they should manifest true expansile pulsation, making differentiation from branchial cysts relatively easy. An aneurysm may be closely simulated, hence the value of angiography (see below). In the usual solid type, the pulsation is of the transmitted type.

A very important physical sign is the relationship of the carotid arteries to the tumour. In 1938 Shawan and Owen[20] observed that the external carotid artery can be palpated over the outer surface of the tumour. This was re-emphasized by Vaughan-Hudson in a personal communication to Monro:[15] 'It is the only tumour of this region in which a large vessel can be felt pulsating in front of the tumour.' The internal carotid artery is displaced posteriorly and laterally to the tumour, and in the case presented below, the external and internal carotids respectively could be felt distinctly anterior and posterior to the tumour. In fact, with a thumb and forefinger in these positions, an impression of expansile pulsation was obtained, whereas direct palpation of the outer surface of the tumour with one finger revealed no pulsation whatever.

2. *Thrill, Bruit and Compressibility*

The presence of these signs also depends on the degree of vascularity of the tumour. Compression of the common carotid artery may cause a decrease in size of the tumour if it is vascular enough. These features were present in only 3 of the 18 cases reported by Lahey and Warren.[10]

3. *Pressure on Adjacent Structures*

A. *Nerves.* Palsies of the recurrent laryngeal, the cervical sympathetic (Horner's syndrome), hypoglossal and spinal accessory nerves, have all been reported, but occur rarely.

B. *The carotid sinus.* A carotid-sinus syndrome is uncommon in these tumours. Byrne[23] found it in 3 of his 13 cases. One might expect this syndrome more commonly than it occurs in view of the anatomical situation of the tumour, but the sensitivity of the sinus is probably the determining factor.

C. *Pharynx.* The tumour may bulge into the pharynx and cause dysphagia.

DIAGNOSIS

The clinical features described above should enable a correct diagnosis to be made in the majority of cases if the condition is kept in mind. The slow growth and long history exclude neoplastic and inflammatory lymph-node swellings in many cases, but the differentiation is more difficult in the early case. A branchial cyst should seldom cause much difficulty, but the very vascular type may be confused with an aneurysm.

Biopsy. A formal biopsy will certainly confirm the diagnosis, but the difficulty in deciding histologically whether the tumour is benign or malignant has been mentioned above, since mitoses and bizarre forms are rare and the usual criteria for malignancy are absent (Byrne[23]). In the authors' opinion arteriography is a much more valuable diagnostic procedure.

CAROTID ANGIOGRAPHY

Monro[15] maintains that carotid angiography plays little or no part in the diagnosis of carotid body tumours. Wetzel,[22] on the other hand, maintains that angiography may play a useful role, not only in the diagnosis of carotid body tumours, but also in the diagnosis of other swellings in this region, especially aneurysms.

This wide divergence of opinion regarding the place of angiography in the diagnosis of carotid body tumour may be explained in part by the paucity of recorded cases. Thus a review of the literature reveals that the first angiographic demonstration of a carotid body tumour was by Lichtenauer,[13] the angiogram having been performed during the course of the operation for removal of the tumour. Thirteen years were to elapse before the next reported cases were published in 1951 by Idbohrn.[9] This author reported 2 cases with preoperative angiograms, using the percutaneous technique, and another case where angiography was carried out at operation. In 1957 Wetzel[22] reported a further percutaneous angiographic demonstration of the tumour, and finally in 1958, Lipshitz[14] recorded a further case. The case presented here is the seventh recorded angiographic demonstration of a carotid body tumour.

The angiograms demonstrated (Figs. 1-5): (1) Displacement of the left common carotid artery in a lateral direction, (2) marked splaying of the carotid bifurcation, (3) marked displacement of the internal carotid artery, laterally and

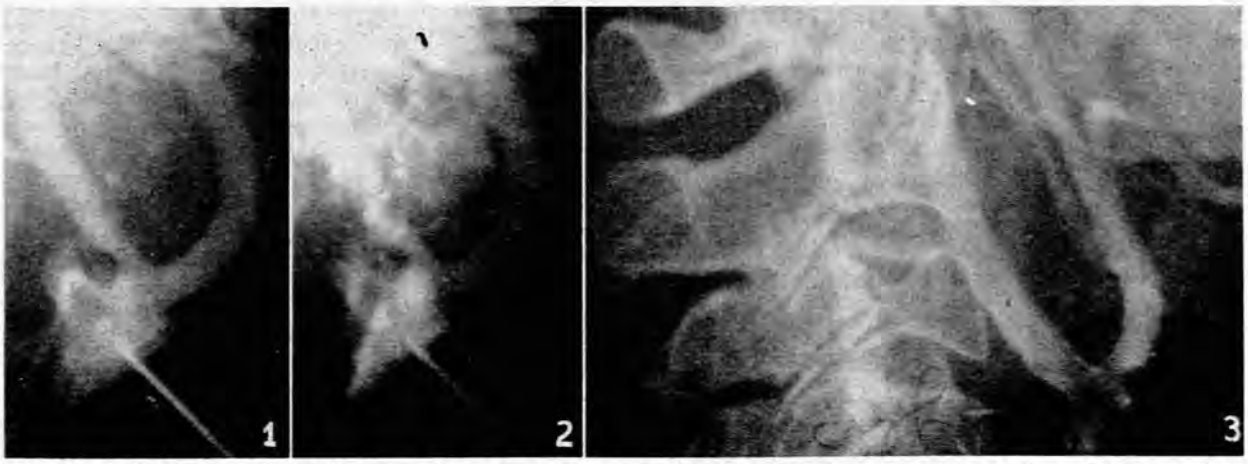

Fig. 1. A.P. Characteristic splaying of the carotid bifurcation with lateral displacement of internal carotid and medial displacement of external carotid. *Fig.* 2. Well-marked vascularity of the tumour. *Fig.* 3. Lateral view. Anterior displacement of the external carotid artery and posterior displacement of the internal carotid artery.

posteriorly, (4) displacement of the external carotid artery, medially and anteriorly—the displacement of this artery is not as marked as that of the internal carotid artery and (5) an extremely rich blood supply to the tumour. The blood supply appears to be wholly derived from the external carotid and consists of well-formed blood vessels with pooling and a 'blush' phenomenon.

The most significant feature of the above appearance is the striking similarity to previously reported cases. The splaying of the artery, the displacements, and the extremely rich blood supply, appear to constitute a characteristic picture. The authors thus feel that, with this characteristic picture, angiography plays a useful role in the early diagnosis and differentiation from other vascular swellings, especially aneurysms.

Furthermore, angiography is extremely useful in demonstrating the size, extent and relations of the tumour to the carotid arteries, which may be of great assistance to the surgeon. In addition, any anomaly of the circle of Willis can be demonstrated at the same examination, and by means of the well-known cross-circulation test, a rough guide to the collateral circulation through the circle of Willis may be obtained, although the latter is not absolutely reliable (see below).

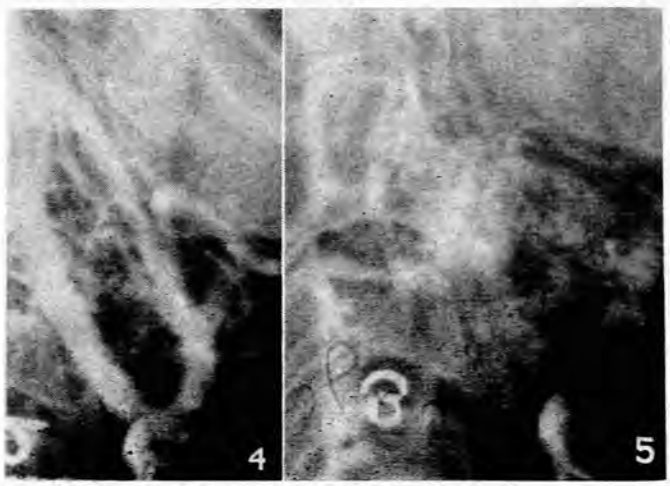

Fig. 4. The rich blood supply to the tumour.
Fig. 5. The blood supply demonstrates a 'blush' phenomenon due to pooling of dye in the highly vascularized tumour.

Finally, by examination of the findings in a sufficiently large number of cases, angiography may provide some help in differentiating the benign from the malignant tumours, bearing in mind that there are well established criteria for doing this in cerebral tumours. This would appear to be a field worthy of further study, in view of the limited value of biopsy.

Radiographic Technique

The examination is carried out on the Lysholm skull table. Conventional A.P. and lateral projections are employed. Note that in the A.P. projection, the tube of the skull table is tilted away from the head to its maximum extent, compared with the 30° tilt to the head (Townes) used in cerebral examinations. This is done in order to obtain a bone-free projection of the carotid bifurcation. 10 ml. of 60% urografin is used, as in cerebral angiography.

The timing of the films calls for special comment. The first film, the arterial phase, is exposed early in the injection, when approximately half the dye in the syringe has been injected. This is in direct contrast to cerebral angiography, where the first film is exposed when three-quarters of the dye has been injected. The reasons for the early exposure are: (*a*) the proximity of the tumour to the site of injection and (*b*) the extreme vascularity of the tumour.

It is also advisable to delay the subsequent films by about 4 seconds each in order to display the 'pooling' and 'blush'. This delay also enables adequate visualization of the cerebral circulation to be achieved.

TREATMENT

It is well established that carotid body tumours, if left untreated, or treated inadequately, can kill the patient. This happens in 30% of cases according to Monro.[15] The development of blood-borne metastases has also been proved. For these reasons it might be thought that an aggressive attack on these tumours was indicated in every case. The difficulty lies in the total reported mortality associated with surgery,

ranging from 15 to 30%. Monro[15] found the mortality in cases dissected off the carotid bifurcation to be 6·4%, compared with a mortality of 29% if the bifurcation was resected. Lahey and Warren[10] lost 2 of their 6 cases in which a 'triple ligation' was done. It is noted that even when they were able to dissect the tumour off the bifurcation, 1 of 5 cases died from a post-operative thrombosis of the internal carotid artery.

Morfit *et al.*,[16] Byrne[23] and Gordon-Taylor[6] have all made a plea for the sanctity of the carotid bifurcation, and have emphasized the feasibility of removing the majority of these tumours without sacrificing the bifurcation. If the tumour is approached in the sub-adventitial plane, aided, if necessary, by the infiltration of a local anaesthetic, a patient and meticulous dissection enables this to be done. Even if the main arterial trunk is damaged, ligation of all the major vessels should never be carried out (Morfit *et al.*[16]). On many occasions repair of a hole in the artery has been performed satisfactorily. If this is not possible, an end-to-end anastomosis between common and internal arteries should be considered. If neither of these methods is feasible, a graft must be used. However Morfit *et al.*[16] have never had to resort to this.

Various methods have been described to assess pre-operatively the adequacy of the collateral circulation across the circle of Willis, in case the carotid bifurcation has to be sacrificed. The Matas manoeuvre of compressing the common carotid artery against Chassaignac's tubercle and observing whether cerebral symptoms appear, has been found to be unreliable by many authors. Lahey and Warren[10] lost 2 of 6 cases who had successfully withstood periods of compression lasting 10 minutes 3 times a day. Lambert Rogers[19] combined this method with electro-encephalography to detect slighter degrees of cerebral ischaemia. Angiography has been mentioned above, but is likewise not foolproof.

Attempts to improve the cerebral collateral circulation either by intermittent digital compression of the carotid arteries or by the preliminary application of a ligature or Crile's clamp round the carotid artery under local anaesthesia (Dandy[2]) are also unreliable. Morfit *et al.*[16] report a case in which a Crile clamp was progressively screwed down over a 10-day period, without any effect, only for the patient to lapse into coma after one further turn of the screw on the 11th day. He concludes that 'there is no method currently available to make carotid ligation safe'.

It is to be noted in passing that carotid artery ligation for neoplasm carries a far greater risk than ligation for intracranial aneurysm. In neoplasm mortalities of 19-58% have been recorded, as compared with 4 - 5·1% inintracranial aneurysm.

In view of the fact that the onset of hemiplegia following resection of the bifurcation may be immediate, or delayed for several days, it has been suggested that thrombosis of the internal carotid artery is responsible for the neurological sequelae. The reason for the thrombosis is stated to be the long (17 cm.) segment without any branches between the bifurcation and circle of Willis. In an attempt to prevent this thrombosis, Pemberton and Livermore[17] used post-operative anti-coagulants in 4 cases after resection of the bifurcation, without mortality. Morfit *et al.*[16] however, state that autopsy studies have seldom demonstrated thrombosis of the distal arterial segment, and that, while anti-coagulants may possibly increase the factor of safety, they certainly do not eliminate the dangers inherent in ligation.

To summarize the position at present, we may conclude that the natural history of these tumours makes their removal desirable as a general principle. Resection of the carotid bifurcation carries a prohibitive mortality, and should never be done. In view of the fact that the carotid body develops in the adventitia of the artery, and that even advanced tumours do not invade the media, meticulous dissection in a sub-adventitial plane will usually leave the bifurcation inviolate. In the few cases where this is not possible, restoration of arterial continuity is mandatory. The following case, however, demonstrates the role of extraneous factors in determining the plan of management.

Case Report

Mr. W., aged 57 years, was admitted to the Johannesburg General Hospital in May 1958. Fifteen years previously he had accidentally discovered a small lump in the left side of the neck. This lump slowly and progressively increased in size throughout the years. Shortly before admission he noticed a certain amount of local discomfort, and also pain in the back of the neck. For 2-3 years he had been having frequent severe frontal headaches. There was no hoarseness or dysphagia.

Examination. A large, rather obese, plethoric looking man, with an oval swelling, 1½ inches by 1 inch, situated in the left anterior triangle of the neck below and behind the angle of the jaw. The lump was firm and smooth and exhibited the type of pulsation described above. The common carotid artery could be traced to the lower pole of the tumour, which was felt to be embraced anteriorly and posteriorly by 2 large arterial trunks. There was free lateral mobility but hardly any vertical mobility. There was no bruit or thrill over the tumour, and no cranial nerve palsies. The left pupil was a trace smaller than the right, but no Horner's syndrome was present. His blood pressure was 170/110 mm. Hg. The patient was also polycythaemic, his haemoglobin being 22·2 g.%. An arteriogram showed the features described above and illustrated in Figs. 1-5.

Management. A firm clinical diagnosis of carotid body tumour was made, and the diagnosis was confirmed by arteriography. The patient made things easy by refusing operative treatment, but if he had consented to operation, we should have been faced by the difficult decision between the general desirability of removing the tumour and the undoubted additional risks in his particular case on account of his age, hypertension and polycythaemia. The case of Lahey and Warren,[10] in which a fatal post-operative thrombosis of the internal carotid artery occurred after dissection of the tumour off the artery, has been quoted, and this patient's polycythaemia would probably increase the chances of post-operative thrombosis. All in all, the hazards of operation in this particular case appeared to outweigh the possible advantages.

SUMMARY

1. The anatomy and development of the normal carotid body, and the pathology, clinical presentation and treatment of carotid body tumours, are reviewed.

2. Of these tumours 12-15% are clinically malignant. Spread to lymph nodes and by the blood stream has been established.

3. Histologically, criteria of malignancy are notoriously difficult to establish, severely limiting the value of biopsy.

4. In principle, carotid body tumours should be removed, but resection of the carotid bifurcation carries a prohibitive mortality. In the majority of cases the tumour can be dissected off the carotid vessels, in view of the fact that it develops in the adventitia of the artery, and does not invade the media even in advanced cases. If the main arterial trunk is damaged, continuity must be restored by arteriorrhaphy, end-to-end anastomosis, or insertion of a graft.

5. There is no certain way of predicting a successful outcome to bifurcation resection. Methods designed to increase the cerebral collateral circulation are unreliable.

6. The role of arteriography in the diagnosis is discussed. The tumours present a characteristic angiographic appearance. A plea is made for the wider use of angiography in the diagnosis of swellings in the upper neck, especially those swellings that appear pulsatile and related to blood vessels.

7. A case is presented showing the typical features of carotid body tumour and illustrating the fact that, whereas removal of these tumours is generally desirable, each case must be considered on its merits. In certain circumstances the hazards of removal outweigh the dangers inherent in leaving a potentially malignant tumour *in situ*.

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