TUMOURS OF THE GLOMUS JUGULARE

BY

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Although many tumours of the glomus jugulare have now been reported, attention has been mainly directed to the symptoms and signs of aural disease which they produce; comparatively little interest has been shown in their neurological manifestations. In this communication we report the clinical and pathological findings in six patients suffering from tumour of the glomus jugulare who have been treated at the London Hospital. Five of these patients had certain evidence of intracranial extension of the tumour and this was confirmed by necropsy in two.

Anatomical Considerations

The glomus jugulare is a part of the chemoreceptor system, a group of small bodies or glomera which are found in the head, neck, and thorax in relation to the glosopharyngeal and vagus nerves. It has been established that the largest member of the system, the carotid body, is a chemoreceptor sensitive to certain chemical changes in the blood (Heymans and Bouckaert, 1939); it has been inferred on grounds of structural resemblance that the other members of the group have a similar function. Their general distribution is shown in Fig. 1. The structures concerned are the carotid and aortic bodies, glomera in association with the ganglion nodosum and auricular branch of the vagus and the glomus jugulare. They are all composed of rounded "epithelioid" cells in small alveolar clusters separated by narrow capillary blood channels, the whole being surrounded by a little delicate connective tissue (see Fig. 16). Apart from the carotid body they are very small, rarely measuring more than 0.5 mm. in diameter, and varying in number and situation.

According to Guild (1941, 1951) the glomus jugulare consists of one or more bodies lying in the adventitia of the dome of the jugular bulb. Guild also describes similar bodies, which may represent the whole of the glomus jugulare, situated along the course of the tympanic branch of the glosopharyngeal nerve. Lundgren (1949) has suggested the name "tympanic body", or glomus tympanicum, for this latter group, and we have adopted his terminology. The glomus jugulare receives its innervation from the tympanic branch of the glosopharyngeal nerve and its blood supply from the ascending pharyngeal artery. Beyond these facts very little is known of the normal anatomy of the glomus jugulare. A full discussion of the chemoreceptor system and the tumours deriving from it is given by LeCompte (1951).

Fig. 1.—Diagram to indicate the positions of the various bodies in the chemoreceptor system. Modified after LeCompte (1951).
Clinical Histories and Pathology

Case 1.—A woman, age 17 (L.H. No. 43148/17), was first admitted to the London Hospital in October, 1914, with a nine months' history of pain and left aural discharge following an injury. Mastoidectomy was performed. She continued to complain of pain in the left ear, and several operations for the removal of recurrent aural polypi were performed. In January, 1916, left facial weakness was first noted. At this time an attempt was made to remove a mass of growth in the external auditory meatus, but it was abandoned because of haemorrhage. By September, 1916, paralysis of the left vocal cord and left half of the tongue had appeared, and a mass of bluish-red growth, extending from the external meatus to the internal ear, was removed. She was admitted for the last time in December, 1917, complaining of severe pain in and behind the ear. Examination showed left facial paralysis, complete deafness on the left, paralysis of the left vocal cord, wasting and weakness of the left sternomastoid, and left lingual hemiatrophy with fasciculation. She was anaemic (Hb. 40%, R.B.C. 3,200,000 per c.mm.) and the urine contained albumin. She required large doses of morphia for the relief of her pain and died on January 17, 1918.

The necropsy (P.M. 32.1918) may be summarized as follows: Purulent bronchitis; bronchiectasis of the posterior part of the left lower lobe; amyloid disease of the liver, spleen, and kidneys; disseminated secondary deposits in the liver, lungs, and spleen from a primary tumour of the left glomus jugulare; ascites, great general anasarca, and marasmus.

Examination of the Base of the Skull.—A mass of pink elastic granular growth extended from the left pterygoid fossa to the left mandibular articulation and reached below to the left atlanto-occipital joint. Laterally it continued into the middle ear cavity and the mastoid antrum; it formed nodular masses in the dura over the posterior aspect of the petrous bone. Its medial boundary was just lateral to the left fifth and seventh cranial nerves. The left jugular foramen was completely closed by growth and the lumen of the jugular vein at its highest part was occupied by what appeared to be organized thrombus. The left seventh, eighth, ninth, tenth, and eleventh cranial nerves were all surrounded by growth. The right middle ear was unaffected. No abnormality was noted in the brain. There was fibrous atrophy of the left sternomastoid muscle, but no masses were discovered in the neck or thorax.

Secondary deposits (up to 1-8 cm. diameter) were very numerous throughout the liver and lungs, and there was one in the spleen. All showed a finely granular cut surface resembling the primary growth.

Microscopical Examination.—Three sections of the primary tumour only were available. It is regretted that no sections of the metastatic deposits were preserved, an oversight attributable to shortage of staff during the war years.

The tumour is densely fibrous throughout (Fig. 2). Between the collagen bundles are numerous narrow trabeculae and alveoli of rounded and oval cells separated by many narrow vascular channels. The cells and their nuclei are regular in size and shape. Mitotic figures are not seen. The cell cytoplasm is eosinophilic and in Masson’s tri-chrome preparations fine fuchsinophil granules are demonstrable. In one section active infiltration of nerve bundles by tumour cells is visible (Fig. 3).

Case 2.—A man, age 56 (L.H. No. 13534/34), developed right-sided deafness and tinnitus in 1918 following influenza. These symptoms persisted but did not progress. On August 6, 1931, he was admitted to the London Hospital on account of profuse bleeding from the right ear. Questioning revealed that he had noticed dryness of the right side of his tongue, and occipital headaches for several months. Examination showed freely bleeding polypi sprouting from the right middle ear, the drum having been destroyed.

There was partial right facial weakness, right deafness, and loss of taste over the posterior third of the right side of the tongue. He had a right palatal paresis and paralysis of the right vocal cord. There was wasting and weakness of the right sternomastoid and upper trapezius. The
right half of the tongue was markedly wasted. No other abnormalities were found in the nervous system.

Blood pressure was 160/125 mm. Hg. Radiographs of the skull and mastoids were normal. The cerebrospinal fluid contained 90 mg. of protein per 100 ml. but was otherwise normal.

The patient was treated with deep x-rays (D.X.T.), 500 r. t.d., in two applications. He attended regularly as an out-patient, and in 1934 was noted as complaining of headache, vertigo, dysphagia, and recurrent haemorrhage from the right ear. A bruit was present on the right mastoid with visible pulsation of the overlying soft tissues; this could be abolished by digital compression of the right carotid artery. The only change in his neurologic state was that lateral and vertical nystagmus was now present. He was admitted to hospital and the right external carotid artery was ligated in the neck. The bruit disappeared, pulsation in the soft tissue was less marked, and the aural bleeding ceased for a time. Over the period 1932-36 he was in addition treated with further courses of deep x-ray therapy, receiving an estimated total dosage of 6,000 r. t.d.

His condition remained unchanged until 1938 when enlargement of the polypi and recurrent bleeding led to further deep x-ray therapy, an estimated total dosage of 4,000 r. t.d. being administered in courses between 1938 and 1941. The only progression noted in his physical state was that anaesthesia had developed over the right side of the pharynx. The bruit reappeared in 1941, when there was an episode of transient sixth nerve weakness with diplopia. In 1943, the palsies of the seventh, eighth, ninth, tenth, eleventh, and twelfth cranial nerves persisted. It was noted that the facial paralysis and deafness were now complete, but the palatal paralysis had improved. Radiographs of the skull showed for the first time destruction of the petrous temporal bone; there had been numerous previous negative films. In 1945 further aural bleeding was treated by deep x-ray therapy, 3,000 r. t.d. in 31 days, and the insertion of radon seeds into the growth in the internal auditory meatus. The effect of this was to cause the polypi to shrink to a scarred mass and the aural bleeding ceased for the remainder of life. Apart from one episode of right facial pain there was no alteration in the pattern of symptoms or signs during the remaining years. He died at home in 1948, aged 73 years, 30 years after the onset of symptoms, following a respiratory infection. No necropsy was made.

Microscopical examination of the fragments from the right external auditory canal shows fibrous granulation tissue and small pieces of tumour. The latter are composed of rounded cells in small clusters with many narrow vascular channels and a few strands of collagen. The nuclei are rounded and regular. No mitotic figures are seen. The reticulin preparation clearly shows the alveolar grouping of the tumour cells.

Case 3.—A woman, age 37 (L.H. No. P.F. 74/1942), in 1937 developed a high-pitched tinnitus in the right ear; aural examination at that time was negative. In 1940 she noticed progressive deafness on the right and experienced brief attacks of vertigo. Examination revealed a bulging tympanic membrane overlying a solid swelling. Following a particularly severe vertiginous attack a partial mastoidotomy was performed. No operative details are available. After this she was completely deaf in the right ear, but the tinnitus and vertigo disappeared. She remained well until 1942 when there was an onset of acute pain in the right ear. This grew worse and she suffered bouts of vomiting. The right side of her tongue felt stiff and she had a constant desire to swallow. There was no aural discharge or bleeding at any time.

She was admitted to the London Hospital in May, 1943. On examination an aural polyp was seen on the right and a bruit was heard over the right mastoid. There was lateral nystagmus, total deafness on the right, and weakness and wasting of the right half of the tongue. There were no other abnormalities in the central nervous system or elsewhere.

Radiographs of the skull showed a small area of erosion in the right petrous temporal bone. The right labyrinth showed no response to caloric tests.

On May 10, 1943, Mr. W. M. Mollison and Mr. D. W. C. Northfield operated by a mastoid approach. A mass of smooth, red, vascular tumour was found filling the middle ear, hollowing out the petrous temporal bone and extending along the dura of the posterior fossa towards the region of the jugular bulb. The growth separated readily from the bone and portions were removed. Lumbar puncture after the operation revealed a clear, colourless fluid containing 200 mg. protein per 100 ml. but otherwise normal. Her post-operative course was uneventful.

She was treated with deep x-ray therapy, 5,000 r. t.d. in 30 days, that is, from July 12 to August 11, 1943. Her deafness remained unaltered after treatment but all her other symptoms disappeared rapidly. No formal neurological examination has been made for several years, but we know the patient to be symptom-free and leading a normal life.

Microscopical examination of fragments received from the external auditory meatus, middle ear, and posterior fossa all show tumour of the same composition. This consists of small clusters of closely packed cells with ill-defined borders and faintly granular eosinophilic cytoplasm (Fig. 4). Vacuolation occurs in many but not sudanophil lipid is visible in frozen sections. Nuclei are in general regular and rounded though hyperchromatic larger forms are not infrequent. Mitotic figures are very scarce. Separating the alveolar clusters are bundles of collagen fibres and vascular spaces of varying breadth. Large venous spaces occur frequently. Reticulin preparations clearly show the alveolar structure of the tumour (Fig. 5). The aural polypus is entirely composed of tumour except for a covering of squamous epithelium over one surface.

Bone from the mastoid region contains tumour infiltrating the marrow spaces (Fig. 6) of the same composition as elsewhere.
Case 4.—A woman, age 55 (L.H. No. 34760/50), was admitted to the London Hospital on June 29, 1947. Twelve years earlier she had developed constant tinnitus in the right ear. Ten years before she noticed progressive deafness in the same ear and hoarseness of the voice. For six years she had suffered from mild dysphagia with occasional nasal regurgitation of fluids. Seven weeks before admission she experienced headache, vertigo, and unsteadiness of gait which lasted one week.

On examination a bruit was heard over the right carotid and mastoid process. In the right ear a red, irregular swelling was seen protruding from the posterior wall of the external meatus (Fig. 7). There was coarse nystagmus to the right and fine to the left. There was considerable deafness of the right ear. There was loss of sensation over the right posterior pharyngeal wall and posterior part of the soft palate. The palatal and pharyngeal reflexes were lost on the right. There was marked weakness of the right palate and paralysis of the right vocal cord. The right sternomastoid and trapezius showed advanced wasting and weakness. There was gross right lingual hemiatrophy with fasciculation. A mild degree of cerebellar ataxia was observed in the upper limbs, right more than left.

Blood pressure was 170/100 mm. Hg. Radiographs of the skull and internal carotid and vertebral angiograms were negative. The cerebrospinal fluid was under a pressure of 250 mm. of water and contained 160 mg. of protein per 100 ml. Caloric tests showed reduced excitability of the right labyrinth.

On July 22, 1947, acerebellar exploration was performed by Mr. D. W. C. Northfield, exposing a nodular, highly vascular tumour broadly adherent to the dura in the right cerebellopontine angle. It was clearly inoperable, chiefly on account of its extent, as it appeared to pass anterior to the medulla and pons. A biopsy was taken. The post-operative course was uneventful and without alteration in her neurological state, apart from a slight
lower left facial weakness, which has persisted. She was given deep x-ray therapy, 3,500 r. t.d., in 28 days from September 19 to October 17, 1947.

Since that time the patient has remained very well, complaining only of deafness and minimal tinnitus and showing some signs of objective improvement. When seen in August, 1952, there was slight lateral nystagmus and her deafness was unchanged. The right side of the palate remained weak and the sternomastoid was wasted and inactive. The trapezius had normal power. The right side of the tongue showed ridging only and no fasciculation. Her voice remained hoarse, but the dysphagia had disappeared. There was no bruit over the mastoid. Minimal ataxia of the right upper limb persisted. A slight bulging of the posterior wall was the only abnormality seen in the right auditory meatus.

Fragments from the posterior fossa of the skull were examined microscopically.

The tumour is composed of sheets of pale-staining, often vacuolated, cells (Fig. 8), broken up by a few bands of dense collagen. Reticulin preparations show a pronounced alveolar pattern (Fig. 9) not distinctly evident in the haematoxylin and eosin preparations. The tumour cells are poorly outlined. Their nuclei, though round and fairly regular, not uncommonly exhibit larger forms, some of which are hyperchromatic. Mitotic figures are not seen.

Case 5.—A man, age 29 (L.H. No. 29615/49), was admitted to the London Hospital on November 19, 1951. Eight years previously he had been struck on the right ear, after which deafness and aural discharge developed. He attended hospital for treatment; the discharge ceased after five months but the deafness persisted.

In June, 1949, he attended the neurological department of this hospital complaining of double vision which had followed a recurrence of the right aural discharge. Examination revealed a right sixth nerve palsy and no other abnormality. He attended irregularly until he was admitted as an emergency with acute suppuration of the right ear.

Examination showed a polypoid mass in the right ear with much purulent discharge. The right sixth nerve palsy was still present with slight right facial weakness and considerable right deafness. A mastoidectomy was started but abandoned because of profuse bleeding from soft tissue and bone. A biopsy was made of the polyp. Recovery was uneventful. Radiographs of the skull and mastoid region showed no erosion of bone. The cerebrospinal fluid was normal with 20 mg. protein per 100 ml.

Following discharge from hospital the patient again defaulted and has not received the deep x-ray therapy planned.

Microscopical examination of the aural polypus shows fibrous granularity tissue in the centre of which are rounded clusters of cells surrounded by narrow capillary channels (Fig. 10). Higher magnification demonstrates gross vacuolation of the cytoplasm of the tumour cells (Fig. 11), the outlines of which are poorly defined. The nuclei are regular and rounded. No mitoses are seen.
There was to was of both nerve. and progress in all four limbs, right more than left. Her gait was ataxic and she staggered to the right. All the tendon reflexes were exaggerated, the abdominal reflexes were absent, and the plantar responses were extensor. There was no sensory loss in the limbs. Blood pressure was 180/120 mm. Hg and the heart was slightly enlarged.

Lumbar puncture gave a spinal fluid under 220 mm. pressure and containing 140 mg. protein per 100 ml. Radiographs of the skull, including tomograms of the petrous temporal bones, were normal, and vertebral and internal carotid angiograms showed no abnormality.

The neurological state was considered to indicate advancing bulbar compression, and on April 2, 1952, Mr. D. W. C. Northfield performed a cerebellar exploration, revealing a reddish, nodular, highly vascular tumour with a broad sessile attachment to the dura at the site of the right jugular foramen (Fig. 12). There was marked compression and displacement of the right cerebellar lobe. It was felt that only by extirpation of the mass could adequate decompression be achieved, and therefore radical removal of all the tumour presenting within the posterior fossa was performed. It was noted that the right vertebral and basilar arteries were in contact with the tumour mass, but no larger branches entering the tumour were seen. Numerous large veins traversed the growth however, and these had to be sacrificed in the removal. The patient died on the sixteenth day after operation.

Case 6.—A woman, age 63 (L.H. No. 9601/52), was admitted to the London Hospital on March 14, 1952. Four years before this she had developed right-sided deafness, tinnitus, and otorrhoea, which she related to a mild occipital head injury. Her aural symptoms did not improve and one year later a polyp was removed from the right ear. She began to have occipital headaches which progressed and were associated latterly with vomiting and neck stiffness. For 10 months she had been having severe attacks of vertigo, and her legs had felt weak and unsteady. For six months dysphagia, hoarseness, and occasional diplopia had been present. She also noticed numbness of the right face, dryness of the right side of the tongue, and difficulty in coughing.

Examination on admission showed a drowsy, obese woman. There was a purulent discharge from the right ear, which was filled with pinkish-grey polypi. There was a bruit over the right mastoid. There was weakness of both external recti, right more than left. The right corneal reflex was absent and there was impaired sensation in all divisions of the right trigeminal nerve and motor weakness of the same nerve. There was right facial weakness of peripheral type. She had considerable deafness on the right. Palatal and pharyngeal sensation was lost on the right. There was bilateral palatal weakness and paralysis of the right vocal cord. The right sternomastoid and trapezius were wasted and weak and there was right lingual hemiatrophy with fasciculation. Cerebellar ataxia was present in all four limbs, right more than left.

FIG. 11.—High power view on one alveolus (Case 5) edged by flattened capillary endothelial cells. Note the marked cytoplasmic vacuolation. Haematoxylin and eosin × 600.

FIG. 12.—Sketches of Case 6 done during the operation illustrate the size of the tumour and its great vascularity. On the right the tumour has been excavated and nerve roots are seen running into it.
The necropsy (P.M. 159.1952) may be summarized as follows:—Inhalation of vomit; partial infarction of the brain stem; recent operation for partial removal of a tumour of the right glomus jugulare.

Examination of the Skull.—Replacing the dura over an area (2.5 cm. × 2.5 cm. diameter) in the anterior wall of the right side of the posterior fossa was a soft haemorrhagic tumour spreading beneath the dura to reach the porus acusticus internus above, and the intradural course of the right sixth nerve medially; laterally it completely occupied the lower three-quarters of the sigmoid sinus, while below it reached almost to the edge of the foramen magnum, having destroyed the bone between the foramen lacerum and the anterior condylar canal. The twelfth nerve was involved in this medial extension. Serial parasagittal slices of the right petrous bone, made after decalcification in formic acid, revealed that the tumour bulged downwards, expanding the jugular foramen, and invaded the lumen of the internal jugular vein to form a firm pink and cream tongue of tissue (2:3 cm. long × 0:5 cm. broad), resembling antemortem thrombus, lying free within the lumen of the vein, but firmly attached to the main tumour above (Fig. 13a). The ninth, tenth, and eleventh nerves and the jugular vein disappeared into the tumour above (Fig. 13b).

Anteriorly the tumour had destroyed the ridge between the jugular and carotid openings, and, coming into contact with the adventitia of the internal carotid artery, bulged forwards lateral to it towards the pterygoid fossa and the temporomandibular articulation. It had destroyed most of the inferior surface of the petrous bone. The growth continued into the middle ear cavity, which it completely filled. It did not penetrate the thin tegmen tympani but extended outwards to form a polypus in the external auditory meatus. The internal ear was intact. The mastoid bone was densely sclerotic but did not contain tumour.

The brain was hardened for several weeks in formalin. There was a broad depression filled with blood clot and fibrin foam in the region of the right cerebellopontine angle. Running into the clot to become lost in its substance were the right anterior inferior cerebellar artery, a delicate anomalous branch arising 0·4 cm. caudally from the basilar artery, and the anastomotic branch from the posterior inferior cerebellar artery. At the edge of this emerged the flattened nerve roots of the fifth to eleventh cranial nerves inclusive; in its floor lay part of the pons, the inferior and middle cerebellar peduncles, and the adjacent portion of the cerebellum. The whole brain stem had been shifted rostrally and to the left. The right twelfth nerve roots were separated from the depression by the prominence of the olive on that side. The ninth, tenth, and eleventh roots had been severed at operation.

Transverse sections of the cerebellum and brain stem revealed softening around the edge of the cavity in the outer two-thirds of the inferior and middle cerebellar peduncles and the adjacent pons and cerebellum.

No abnormality was found in the rest of the brain.

No metastatic deposits were discovered in any organ. The carotid bodies were of normal size. No aortic or pulmonary mass was found nor was there any abnormality in the left jugular foramen or middle ear.

Microscopical Changes.—Sections of the tumour from the operation specimen were distinctly more congested than those from the post-mortem tissues. The tumour is composed of closely packed irregular cells (Fig. 14) in rounded alveoli or in trabeculae separated by broad vascular channels lined by flattened endothelial cells. The nuclei of the tumour cells are mostly rounded with a finely granular chromatin pattern and are of fairly regular size. Large bizarre and hyperchromatic forms are, however, not infrequent. Cytoplasmic vacuolation is conspicuous but no lipid or glycogen is demonstrable. Numerous fine fuchsinophil granules in the cytoplasm of many tumour cells are seen with Masson's stain, and silver impregnation of the reticulin fibrils demonstrates distinctly the pattern of the tumour cells (Fig. 15). Large venous channels containing a few elastic fibrils in their walls are common, but arterial channels are not
Besides the limits of the tumour attached to the capsule of the ganglion nodosum of the vagus is a normal glomus vagale (Fig. 16).

The area of ischaemic necrosis of the brain stem is confirmed by the sections and microglia are active at its periphery. No tumour was found in direct association with the brain, either in the operation area or elsewhere.

Sections from the pituitary gland, the adrenal gland, the pancreas, both carotid bodies, and the left jugular dome do not show any significant abnormality.

Discussion

The site of the glomus jugulare enables a tumour arising from it to extend through the thin floor of the tympanum or into the posterior fossa. The clinical facts suggest that the middle ear is generally invaded first. In two of our cases the full development of the tumour by local extension was found at necropsy; similar observations have been made by Kipkie (1947), Lattes and Waltner (1949), and Poppen and Riemenschneider (1951). There can be little doubt that these tumours arose from the glomus jugulare. There are also reports of tumours which have been found at operation to fill the tympanic cavity and extend through the tympanic floor to the jugular bulb (e.g. Rosenwasser, 1945; Lundgren, 1949). These neoplasms presumably represent an early stage in the development of glomus jugulare tumours, although they might be regarded as an extension of a third group, namely those in which the tumour is found solely within the tympanum and the tympanic floor is intact. Such cases have been described by several workers. (e.g., Capps, 1944; Winship and Louzan, 1951). Winship and Louzan suggest that these growths arise from the tympanic body, and the term tympanic body tumours, originally proposed by Lundgren, should be reserved for them. Black (1952) states that about one half of all cranial glomus tumours arise from the dome of the jugular bulb and the remainder from the glomera within the temporal bone.

conspicuous in the tumour. In view of this great vascularity it is not surprising that many fine haemosiderin granules occupy macrophages.

At its periphery the growth is circumscribed by a zone of dense collagen, often infiltrated by alveolar clusters of tumour cells. At the jugular foramen the tumour is growing into the lumen of the internal jugular vein where it composes the whole of the tongue of tissue already described. The alveolar arrangement, the vascular spaces, and the bands of collagen are here identical with the tumour elsewhere. This could well have been a potential source for tumour emboli.

FIG. 15.—Preparation from Case 6 showing reticulin fibrils confined to the vascular channels and outlining the clusters of tumour cells. Foot's reticulin impregnation method × 74.

Clinical Features.—Craniocerebral glomus tumours are five times more common in women than in men (Capps, 1952). The age at onset of symptoms has varied from 17 to 80, but the incidence is greatest in the fifth and sixth decades. Although it is unlikely that trauma bears any aetiological relationship to the tumours it is noteworthy that in two of our patients, Cases 1 and 5, symptoms first appeared after trauma to the affected area, and in a third they followed a mild occipital head injury (Case 6). Tumours of the cranial glomera may be associated with growths of the carotid or aortic bodies (Kipkie 1947; Bartels, 1949; LeCompte, 1951). Bartels has reported the occurrence of tumours of the glomus jugulare, the carotid body, or of both in members of three generations of one family.

The clinical picture presented by these tumours naturally varies with the origin of the growth. When it arises from the tympanic body the presenting symptoms are otological. Again, when the tumour originates from the glomus jugulare the initial symptoms are auricular. Exceptionally the presenting complaints are due to intracranial extension, for example hoarseness due to vagal involvement as in Capp’s (1952) Case 5. Thus in tumours of the glomus jugulare and glomus tympanicum haemorrhage from the ear, purulent otorrhea, auricular pain, cochlear and, less constantly, vestibular disturbances are the usual complaints. On auroscopy vascular polypi are seen either deep to or bulging through the drum. With tumours of the glomus jugulare, however, symptoms and signs of intracranial extension frequently arise, though their appearance may be delayed for many years. An analysis of 77 reported cases of tympanic and jugular glomus tumours, including our six cases, shows that 31 (40%) had certain evidence of intracranial extension; 11 of the 31 cases were reported by Bartels (1949) and five are ours. We have excluded our fifth patient as his sixth nerve palsy may well have been due to a venous thrombosis secondary to his aural infection; moreover the site of origin of his tumour is not certain.

The neurological picture presented by our patients is in accord with previous reports, very few of which have been made from the neurological standpoint. Several neurosurgeons have recorded their experiences of single cases (Poppen and Riemen-schneider, 1951; Dockerty, Love, and Patton, 1951; Alexander, Beamer, and Williams, 1951), but these are the only reports from neurological sources. Bartels (1949) gives an exhaustive account of his 11 patients who showed the various patterns of cranial nerve palsies described below. The salient facts regarding our cases are summarized in Table I. The initial symptoms were invariably auricular, neurological symptoms following in from two to 13 years. Nevertheless three patients were referred directly to the neurological clinics (Cases 4, 5, and 6). The earliest neurological complaints have been dryness or stiffness of the tongue, a sensation of irritation in the pharynx, dysphagia, and aphonia. Headaches, vomiting, and failure of vision were the only manifestations of intracranial extension in Dockerty and his colleagues’ (1951) patient. Two patients complained of trigeminal pain or sensory disturbance, symptoms which have been reported by other observers and which may arise in the absence of signs of involvement of the nerve. Two patients also complained of diplopia and ataxia.

Examination has usually revealed paralysis of the seventh to twelfth cranial nerves on the affected side, but the fifth and sixth nerves may also be attacked. Table II shows the frequency of individual cranial nerve palsies in the 31 recorded cases. The

<table>
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<th>Patient</th>
<th>Age at Onset</th>
<th>Sex</th>
<th>Neurological Signs</th>
<th>Aural Polyp Present</th>
<th>Length of Illness (yrs.)</th>
<th>C.S.F. Changes</th>
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<tr>
<td>1</td>
<td>17</td>
<td>Female</td>
<td>7, 8, 9, 10, 11, 12 cranial nerve palsies</td>
<td>Yes</td>
<td>4 Died</td>
<td>—</td>
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<td>2</td>
<td>43</td>
<td>Male</td>
<td>6, 7, 8, 9, 10, 11, 12 cranial nerve palsies; nystagmus; bruit</td>
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<td>90 mg. % protein</td>
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<td>31</td>
<td>Female</td>
<td>*8, 12 cranial nerve palsies; nystagmus; bruit</td>
<td>Yes</td>
<td>15 Alive</td>
<td>200 mg. % protein</td>
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<tr>
<td>4</td>
<td>43</td>
<td>Female</td>
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*In these cases it is uncertain whether nerve deafness was present.
hypoglossal nerve is often attacked before the ninth, tenth, or eleventh nerves, as in our third patient and in others reported by Bartels (1949). This unexpected finding can only be attributed to the fortuitous pattern of nerve compression and invasion by the tumour. Sometimes the ninth, tenth, or eleventh nerves are the first to be involved. Deafness is nearly always present before frank neurological signs develop, but there are occasional exceptions. It is not possible to ascertain from the clinical records how frequently the auditory nerve itself is affected. We did not find papilloedema in any of our patients, nor have we traced any reports of its presence. Nystagmus was a frequent finding.

On the motor side two of our patients showed signs of cerebellar disorder, as did those of Winship, Klopp, and Jenkins (1948) and Alexander and others (1951). Pyramidal signs were found in our last patient and also in two of Bartels’ cases. Poppen and Riemenschneider’s (1951) patient had weakness and hypoesthesia of the contralateral side, presumably the result of bulbar compression, besides nystagmus and cranial nerve palsies.

In all our patients vascular aural polypi were seen in the affected ear, chronic suppuration being present in two instances. A bruit was heard over the mastoid process in four cases, but it was only a source of annoyance in the second patient. Occasionally a bruit is the presenting or dominant symptom (Semmes, 1951).

The variable length of the clinical course pursued by these tumours is well shown by our patients (Table I). Survival for 20 years is not uncommon (Black, 1952). It appears that the length of the illness depends upon the variable local malignancy of the tumours, their rare liability to metastasize, and the nature of treatment. Radical surgery has been associated with a high mortality on the few occasions it has been attempted. Recurrence of glomus tumours involving the middle ear after apparent removal has been frequently observed (e.g. Lundgren, 1949). Presumably the intracranial growths may behave in similar fashion.

The common clinical picture is therefore that of a relatively benign tumour which, after an initial period of otological symptoms, proceeds to a slowly progressive succession of palsies of the lower cranial nerves. Ipsilateral cerebellar signs are sometimes found but pyramidal disturbances are uncommon. Sensory disorders and symptoms of increased intracranial pressure are rare.

Investigations.—The important investigation is biopsy of the aural polyp, care being taken to obtain an adequate specimen. The cerebrospinal fluid may be under increased pressure and the protein content is usually raised (Table I). Radiography of the skull has proved of limited value in our experience. Bartels (1949) and others have described the erosion of the petrous bone and enlargement of the jugular foramen which may be found. The radiographs of our second and third patients showed erosion of the petrous bone. Nevertheless it was not until 25 years after the onset of symptoms that radiological changes became apparent in the second case. Alexander and others (1951) were able to demonstrate the tumour in their patient by external carotid angiography. We have performed internal carotid and vertebral angiograms in two patients without displaying any abnormality of the vascular tree. This finding supports the contention that the blood supply of the tumour is mainly from the external carotid artery.

Differential Diagnosis.—There is usually no difficulty in reaching the correct diagnosis of these tumours as the characteristic aural polypi are generally visible by the time neurological symptoms and signs have appeared. In the earlier stages the polypi may be less easily discernible, there being a mere redness of the ear drum, without bulging of the membrane or protrusion of the tumour. Confusion might conceivably arise when a simple aural polyp occurred in the presence of some other intracranial tumour of similar location, but biopsy would provide the correct answer. From our own experience we would stress the liability of patients to forget their aural symptoms. These were sometimes elicited only after auroscopy had revealed the presence of polypi and further questioning had been undertaken.

Histological Diagnosis.—The relative uniformity of the histological picture of tumours of the chemoreceptor system is one of their outstanding features, particularly when reticulin preparations are examined. From the descriptive standpoint there is little to add to that already written by Lattes and Waltner (1949), Berg (1950) and LeCompte (1951). However, variations do occur—especially in the proportion of vascular to tumour cells. Very vascular tumours have not infrequently in the past
been misdiagnosed as haemangioendothelioma (Berg, 1950; Capps, 1944) and when the tissue is taken from the posterior fossa haemangioblastoma of the cerebellum must naturally be considered as an alternative diagnosis. Case 4 was such an instance. With routine stains it was difficult to discern any clear-cut pattern in this tumour; vascular spaces, endothelial cells and tumour cells appeared intimately mixed. The pattern brought out by reticulin staining identified the true nature of the tumour, for an arrangement such as described is not a feature of haemangioendothelioma. Nevertheless it must be recognized that distinction is at times difficult on purely histological grounds and the situation of the tumour must be taken into account in instances where doubt arises.

A further point of differential diagnosis must be made from angiomatous meningioma since this tumour, when lying in the posterior fossa, most often arises from the region of the sigmoid sinus. Both tumours are exceedingly haemorrhagic, but the difference in the appearances of the cells and the characteristic arrangement of each type of tumour should serve to separate them. Moreover jugular body tumours invariably invade the middle ear; meningiomas must do so very rarely, if at all.

Attempts have been made in this series to correlate degree of invasiveness with atypical appearances of the cells. The material from Cases 2 and 5 was too scanty to be of value, but in the remainder large and hyperchromatic nuclei occurred with about equal frequency, except in Case 1. This latter tumour developed with the same rapidity as had that of Case 6 and had produced widespread metastatic deposits, yet the tumour cells looked the least atypical. Further, atypical nuclei were moderately conspicuous in Cases 3 and 4 both of which, though they showed local invasiveness, were clinically slowly growing neoplasms. It is apparent therefore that in the assessment of prognosis histology may be of little service.

Metastasis and Multiple Tumours.—Metastatic deposits from these tumours are rare. Lattes and Waltner (1949) found deposits in the liver in one case. Winship and others (1948) discovered secondary tumour in cervical lymph nodes in one case that did not come to necropsy. Our Case 1 thus makes the third example. In none of these three cases was the histology in any way remarkable, except a certain excess of collagenous stroma in the case presented here. It is possible that a potent source of secondary, blood-borne deposits is invasion by the tumour of the lumen of the internal jugular vein; this was present in both our necropsied cases.

On the other hand mention should be made of the not infrequent occurrence of multiple tumours, a fact that may cause some confusion unless recognized. Of interest in this connexion is one of the earliest reported examples of a jugular body tumour that was diagnosed as a metastasis from a carotid body tumour on the same side (Lubbers, 1937). Both Kipkie (1947) and Bartels (1949) reported other examples. Aortic and vagal body tumours have occurred together and so have carotid and vagal body tumours (Lattes, 1950). Tumours of both carotid bodies and the left jugular body were described in one patient by Lattes and Waltner (1949). The situations of these tumours should be the chief clue as to their true nature.

Treatment.—When these tumours are diagnosed at an early stage treatment will be in the hands of otologists to whom the patients will usually be referred. When intracranial extension of the tumour brings the patient under the care of the neurologist or neurosurgeon the decision on treatment rests between surgery and radiotherapy (deep x-ray therapy). Our own experience and that of others (e.g. Dockerty and others, 1951; Poppen and Riemenschneider, 1951) leads us to suppose that radical surgery has no present place in the treatment of these cases. The situation and vascularity of the tumours render any surgical approach extremely hazardous. Semmes (1951) stated that he had one patient in whom complete removal of the intracranial part of a glomus jugulare tumour was achieved "as far as could be determined". This is the only recorded example of survival after radical extirpation that we have traced. Suboccipital decompression may well be of value when cerebellar or bulbar compression is present. Ligature of the external carotid artery may be helpful in suppressing a troublesome bruit.

Alexander and others (1951) and Capps (1952) have reported early results of deep x-ray therapy which they regard as encouraging. Our own experience of it has been satisfactory to date. We can draw no conclusions from our second patient to whom deep x-ray therapy was largely given before the development of modern radiotherapeutic techniques, though it is clear that the last course of treatment was effective in reducing the size of the aural growth and in preventing haemorrhage. The favourable results in our third and fourth patients are apparent from the case reports. Objective evidence of improvement in the neurological status is recorded in Case 4. Surveillance for many years is
clearly necessary in view of the character of the tumours.

Summary

The clinical and pathological findings in six cases of tumours of the glomus jugulare are described. The neurological aspects of their natural history are discussed.

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