Glomus jugulare tumours

(A series of 21 cases)

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Glomus jugulare tumours are locally invasive tumours in the region of the middle ear. Guild (1941) described structures in close relation to the bulb of the jugular vein which he named the glomus jugulare, and Rosenwasser (1945) demonstrated a relationship between these structures and highly vascularized tumours in the ear. Glomic tissue may be found not only in the jugular bulb, but also along the course of the tympanic nerve in the middle ear and along the course of the auricular branch of the vagus nerve to the stylomastoid foramen.

The sites and ultrastructure of the normal glomic tissue have been described by Kjaergaard (1973).

Glomus jugulare tumours are generally considered benign, although they exhibit invasive growth and have a marked tendency to recur. Metastasizing cases are on record. Reviewing the literature from a seventeen-year period Friedmann (1974) found ten cases with metastases to regional lymph nodes, liver, bones, and lungs.

The physiological factors have not been elucidated, but it is believed that the glomus cells are chemoreceptors, and therefore the tumours have also been termed chemodectomas.

The symptoms and signs vary. When the tumour affects the region of the middle ear there will most often be hearing impairment and tinnitus as well as facial palsy, whereas a tumour in the bulb of the jugular vein may cause a foramen jugulare syndrome with paresis of the IXth, Xth, XIth, and XIIth cranial nerves.

The therapeutic possibilities, viz. surgical extirpation, radiation, or possibly a combination of the two, are still being discussed.

The object of the following, retrospective analysis was to describe the symptoms, signs, clinical course, and prognosis. The material is not large enough to permit definite conclusions regarding the therapeutic principles, but the analysis may also be interpreted as a contribution to the discussion concerning the treatment.

Material

During the period 1959 to 1973 a total of twenty-one patients with glomus jugulare tumours were treated in the Radium Centre and E.N.T. Department of the Århus Municipal Hospital.

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In seventeen cases the diagnosis was confirmed histologically. In two cases the biopsy material was not sufficient for a definite microscopic diagnosis, but the findings were compatible with that of glomus tumour. In another two cases it was not possible to obtain biopsies for histological diagnosis, but the diagnosis was based on typical clinical symptoms and findings.

Fig. 1 gives the age distribution and sex ratio. The age summit was between 60 and 70, somewhat higher than previously reported in the literature. The age range was 27 to 74 years. As in other series, there was a pronounced female preponderance, there being eighteen females and three males.

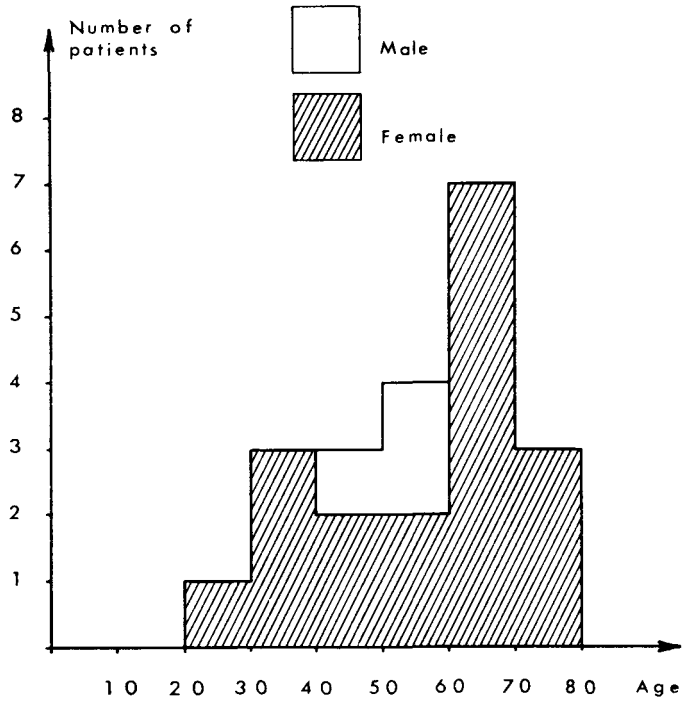


FIG. 1.
Distribution by age and sex.

Table I lists the initial symptoms and signs. The duration of symptoms was from a few months up to thirteen years. Five patients had had symptoms for more than ten years before the diagnosis was made. The most common symptoms were gradually increasing hearing impairment and tinnitus, as a rule of the pulsating type. These symptoms occurred in eighteen and sixteen patients respectively. In most cases the hearing

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TABLE I

Primary symptoms	No. of patients
Impaired hearing	18
Tinnitus	16
Otorrhoea	3
Aural bleeding	2
Vertigo	2
Pain	1
Facial palsy	5
IXth cranial nerve affected	3
Xth cranial nerve affected	3
XIth cranial nerve affected	3
XIIth cranial nerve affected	4
Jugular foramen syndrome	2

impairment was conductive in type, but in two cases there was total deafness.

Five patients had more or less severe facial palsy. Six had neurological defects corresponding to one or more of the IXth, Xth, XIth, or XIIth cranial nerves. Two had a full-blown foramen jugulare syndrome. In one of these patients the initial sign was paresis of the accessory nerve which had been present for thirteen years before the patient was referred for treatment.

The prolonged course contributes to the appreciable diagnostic difficulties often posed by this disease. It may be mentioned that in five cases paracentesis—and in two of them also tubulation—had been performed before the final diagnosis was made. In one case mastoidectomy had been performed because of a suspicion of mastoiditis. Lastly, one patient had undergone a facial nerve decompression operation for the facial palsy, without the tumour being detected on this occasion.

A retrospective classification into three groups was done by a slightly modified Bickerstaff and Howell (1953) method. Group I comprises patients having only aural symptoms and patients who also had more or less marked facial palsy as the only neurological defect. Group II comprises patients in whom the aural symptoms were followed by a complete or incomplete foramen jugulare syndrome. Group III consists of those patients in whom a complete or incomplete foramen jugulare syndrome preceded aural symptoms.

The distribution by this classification is given in Table II.

Otoscopic examination showed abnormalities in all cases, in the majority a localized red swelling of the lower part of the drum or a red, polypoid tumour in the innermost part of the auditory canal.

Radiology showed normal appearances at the site of the foramen jugulare in eight patients, but in three of them, there was destruction in the middle-ear area. In the remaining thirteen patients osseous changes were found in the area of the foramen jugulare. Table III presents the distribution of such bony changes in relation to the classification.

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TABLE II

Clinical classification		No. of patients
Group I	Aural symptoms with or without nerve palsy	15
Group II	Aural symptoms with complete/incomplete jugular foramen syndrome	5
Group III	Complete/incomplete jugular foramen syndrome preceding aural symptoms	1
Total		21

TABLE III

DESTRUCTION OF THE JUGULAR FORAMEN

Group	No. of patients
I	7/15
II	5/5
III	1/1
Total	13/21

Within Group I destruction was found in about half the cases (7/15), whereas all six patients of Groups II and III exhibited destruction. In eight cases retrograde jugular phlebography was carried out (Gejrot *et al.*, 1963), which confirmed the diagnosis in five cases. In the other three the phlebographic study showed normal appearances, in accordance with the tomographic findings.

Treatment

Table IV sets out the treatment used in the present series.

Six patients—all of Group I—aged 33 to 72 years, had primary surgery. In four the tumour affected the middle-ear region and could be radically removed.

In two patients spread of the tumour to the jugular foramen had been demonstrated preoperatively, and an extended operation was performed, resecting the jugular bulb. One case was complicated by transient peripheral facial plasy and paresis of the soft palate. In the other case, to be

TABLE IV

PRINCIPLES OF TREATMENT

Group	I	II	III	Total
Surgery	6			6
Surgery + irradiation	5			5
Irradiation	4	5	1	10
Total	15	5	1	21

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described in more detail below, total facial palsy occurred, as part of the vertical segment of the facial nerve had to be resected to remove the tumour. Preoperatively this patient had had partial facial palsy. Apart from these two cases there were no operative complications. Of course, bleeding was profuse in some of the cases, the blood loss ranging from 400 to 4500 ml.

In five cases surgical treatment was tried, but radical excision had to be abandoned, int. al. because of haemorrhage, and the patients were thereafter treated by irradiation.

In the remaining ten cases primary radiotherapy was performed. This applied to all six patients of Groups II and III and to four of Group I in whom operation was not felt to be indicated because of advanced age and/or a poor general condition. In all cases the treatment was by cobalt^{60}, and the tumour dose was 5000–6000 rad/5–6 weeks. The treatment was well tolerated in all cases, without complications. Regression of the tumour seems to have been observed in most of the cases during or immediately after the treatment.

Results

During the follow-up period, extending from a few months to eleven years (mean six years) only four cases of recurrence or progression occurred. In the other seventeen cases there had not been any progression of the tumour during the follow-up period which was one to eleven years (mean six years) for this group.

Table V lists the four cases with recurrence or progression of the tumour in relation to treatment and classification. In all four cases the tumour had originally been restricted to the middle-ear area (Group I).

TABLE V

NO. OF RECURRENCES

Group	I	II	III	Total
Surgery	2/6			2/6
Surgery + irradiation	0/5			0/5
Irradiation	2/4	0/5	0/1	2/10
Total	4/15	0/5	0/1	4/21

Two of the patients had been considered as having had radical surgery, but fourteen months and five years respectively after the operation they developed an incomplete and a complete foramen jugulare syndrome, and were treated with cobalt irradiation. During the follow-up period thereafter, three months and two years respectively, there has been no progression of the symptoms. The other two patients had primarily received radiotherapy, but developed recurrences in four and eight months respec-

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tively. In the former case the recurrence was found at otoscopy in the course of a follow-up examination, whereas the other patient developed increasing and disabling tinnitus. Both patients were subjected to operation. In one of the cases the radicality was in doubt, but during the subsequent follow-up period of ten years there have been no signs of recurrence or progression. In the other case there have also not been signs of recurrence or progression during a follow-up period of ten years, and the symptoms have decreased.

In the present series metastases have not been observed, and there have been no deaths from the tumour during the follow-up period.

Case report

A woman, aged 33, with no previous aural symptoms, had for one year been suffering from pulsating tinnitus, and for the past six months there had been progressing facial palsy on the right.

At admission to the E.N.T. Department otoscopy showed a tumour protrusion in the innermost part of the auditory canal, partial right-sided peripheral facial palsy, with a 2 mm. closure defect of the right eye. Tomography showed destruction in the hypotympanum and anteriorly in the foramen jugulare. Retrograde phlebography revealed a tumour of the wall immediately below the bulb of the jugular vein. Hearing was normal.

At operation the tumour was found in the posterior auditory canal wall, ensheathing the facial nerve. It occupied the entire hypotympanum, continuing in the wall of the jugular bulb. All grossly visible tumour tissue was removed, together with the bulb of the jugular vein and the entire vertical segment of the facial nerve. About one year later, nerve grafting was done, the continuity of the facial nerve being re-established by a graft from the great auricular nerve. About six months after this operation, there was incipient function in the area innervated by the facial nerve, and since then there has been gradual improvement.

Incidentally, the patient stated, when last seen about six years after the first operation, that she was feeling well. She had no tinnitus. The hearing impairment amounted to about 30 dB.

Fig. 2 shows that quite a good tonus has been achieved in the area innervated by the right facial nerve. There is only mild asymmetry when the face is at rest, but attempts at wrinkling the forehead, screwing up the eye, pursing the lips, etc. reveal that still there is a distinctly reduced function on the right, with distortion and dyskinesia. It must be admitted that cosmetically the result is not quite satisfactory, but it is as good as can be expected after nerve grafting. It may be added that the patient herself is satisfied.

Discussion

As is apparent from the review of the present series of glomus jugulare tumours and from the literature, these tumours often run a very prolonged clinical course. This renders it difficult to assess and compare the results of the therapeutic principles employed—surgical excision, irradiation, and a combination of the two.

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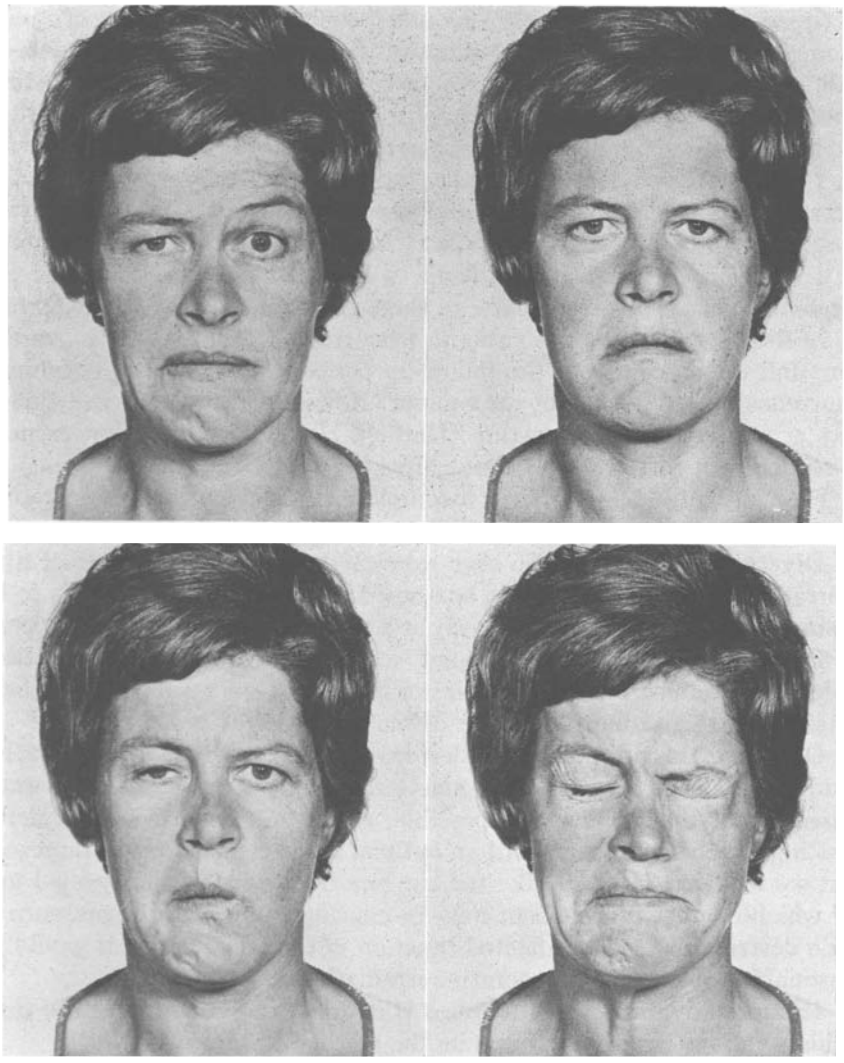


FIG. 2.
Facial function 6 years after nerve grafting.

It seems to be almost generally accepted that surgical excision is preferable, at least in the case of fairly small tumours, restricted to the middle ear. However, some authors (House and Glasscock, 1968) feel that surgery is indicated also for the more extensive tumours involving bone destruction. Let it be mentioned, however, that although surgical excision has apparently been grossly radical, recurrences are not uncommon (Hatfield *et al.*, 1972). In the present series, recurrences appeared in two out of six patients who had been thought to have had radical surgery.

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Opinions have been highly divergent concerning the effect of radiotherapy upon glomus jugulare tumours. By now, it seems to have been realized by most authors that these tumours are fairly radioresistant (Spector *et al.*, 1974). After radiotherapy the stroma shows marked fibrosis and hyalinization, but according to Spector *et al.* (1974), less vascular sclerosis. These are the changes assumed to inhibit tumour growth after radiotherapy. This seems to be confirmed by numerous reports in the literature (Moore, 1973) on the clinical results of treating glomus jugulare tumours by irradiation, although it is difficult to assess the results because of the frequently very protracted course of these tumours—also when untreated.

In the present series ten patients were treated exclusively by irradiation, and in the course of the follow-up period only two cases developed recurrence or progression of the tumour. However, it must be mentioned that according to the literature (Hatfield *et al.*, 1972) recurrences may be seen fifteen to twenty to twenty-five years after treatment.

Five patients had combined incomplete surgical excision and irradiation. There have been no recurrences in this group.

Divergent views have also been expressed concerning the optimal time of irradiation given as a link in combined treatment—whether it is to be postoperative (Hatfield *et al.*, 1972) or preoperative (Spector *et al.*, 1974).

From a surgical-technical point of view, preoperative irradiation, which may reduce bleeding and leave fibrosis of the stroma, might help obtain a safer and more accurate dissection (Spector *et al.*, 1974).

Of course, it is not possible to decide in retrospect whether preoperative irradiation in the case reported above might have rendered an accurate dissection of the facial nerve possible, obviating the need for a nerve grafting which can never afford an optimal functional and cosmetic result. But we feel that in cases, like the one reported, classified in Group I and for which operative treatment may be considered even in the presence of bone destruction and an affected function of the facial nerve, it would be reasonable to consider preoperative irradiation.

In more advanced cases (Groups II and III) radiotherapy in the form of high-voltage irradiation must be the method of choice.

Summary

Twenty-one cases of glomus jugulare tumours were analysed in retrospect. Six had been treated surgically, ten by irradiation, and five by a combination of incomplete surgical excision followed by irradiation. Recurrences appeared in four cases, but no patient has died of the tumour during the follow-up period which is from a few months to eleven years. Metastases were not found in any case. The optimal time of irradiation in combined treatment is discussed.

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