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Trigeminal Neurinomas
A Series of 111 Surgical Cases from a Single Institution

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Summary

Neurinomas arising from the trigeminal nerve are rare (0.1–0.4% of intracranial tumours; 1–8% of all intracranial neurinomas).

A series of 111 trigeminal neurinomas operated on at the Institute of Neurosurgery "N. N. Burdenko" of Moscow, Russia, during the period 1961–1994 is presented.

Clinical features, diagnostic radiology, surgery and results of treatment are discussed.

We distinguished four groups of trigeminal neurinomas:

a) Posterior fossa tumours; b) Tumours of the Gasserian ganglion; c) "Dumb-bell" supra-subtentorial tumours; d) Neurinomas of the peripheral branches.

The complex clinical symptomatology is related to the actual location of the lesion.

Surgery of trigeminal neurinomas may be very difficult, particularly in cases of dump-bell supra-subtentorial lesions. In this series, 3 cases died postoperatively. Out of the 108 patients surviving surgery, long-term follow-up (min. 13 months, max. 33 years, average 13.5 years) was available in 98 cases. 84 patients (86.7%) showed good-to excellent results, with partial trigeminal deficit as the only surgical sequela.

13 patients (11.7%) had a symptomatic recurrence following incomplete tumour removal. Second surgery, ranging from 1.4 to 9 years (average 3.8 years) following the first operation, was judged radical in 9 cases (69.2), however, the rate of complications of repeated surgery was higher than that occurring after the first operation.

The advent of microsurgery, together with the introduction of the techniques of skull base surgery and of modern diagnostic imaging tools, have improved surgical results in terms of increased radicality and reduced complications.

Keywords: Trigeminal neurinoma; surgical management; skull base surgery.

Introduction

Neurinomas arising from the trigeminal nerve are rare. The actual incidence varies from 0.1 to 0.4% of intracranial tumours [2, 5, 6, 10, 12–19, 21–23] and from 1% to 8% of all intracranial neurinomas in the different series reported by major centres worldwide [2, 6, 12, 13, 16, 18, 19, 21, 23].

A recent careful review of the relevant literature quoted an overall number of 120 cases [13].

The peculiar referral pattern of the Institute of Neurosurgery "N. N. Burdenko" has allowed us to accumulate an unusual experience with these particularly rare lesions. In fact, approximately 150 cases of trigeminal neurinomas have been histologically verified in our Institution since 1935. In this presentation, we analyse the diagnostic data and the management results of the 111 cases of trigeminal neurinomas admitted to the "Burdenko" Institute of Neurosurgery since 1960.

Patients and Methods

In the years 1961–1994, 111 cases of trigeminal neurinoma were operated on in the Moscow Institute of Neurosurgery. There were 79 females and 32 males, age range from 17 to 61 years.

These cases represent 0.3% of the approximately 37.000 intracranial tumours as well as 5.8% of the 1914 intracranial neurinomas operated on during the same period in our Institution.

Classification

We divided trigeminal neurinomas into 4 groups, depending on the location and predominant extension of tumours (Table 1):

1. Posterior fossa trigeminal neurinomas are located in the lateral pontine cistern and the cerebellopontine angle. 2. Trigeminal ganglion neurinomas have a predominant location in the middle fossa. 3. "Dumb-bell" supra-subtentorial neurinomas involve both

*Dr. Aldo Spallone was visiting professor, coordinator of the Skull Base group at the Institute of Neurosurgery "N. N. Burdenko", Moscow, during the period when the present study has been conducted (1993–1994).

Table 1. *Tumour Location*

Posterior fossa	26
Gasserian ganglion	42
"Dumb-bell" supra- and subtentorial	30
Intra-orbital; SOF; infratemporal fossa (Peripheral trigeminal branches)	13
	111

SOF superior orbital fissure.

Table 2. *Size of Tumours*

Location of tumour	Range (cm)	Average (cm)
Posterior fossa	2.5–5.2	3.6
Gasserian ganglion-"Dump-bell"	4.2–9.4	6.2
Peripheral branches	4.9–12	6.8

Table 3. *Plain X-Rays*

	No. of cases
Erosion of petrous apex and/or lateral clivus	58
Erosion of middle fossa floor	16
Widening of SOF	7
Widening of basal foramina	4
Destruction of posterior orbital wall	6

SOF superior orbital fissure.

the posterior and middle fossa. 4. Peripheral trigeminal branch neurinomas expand from the middle fossa through the superior orbital fissure into the orbit; through the foramen rotundum and inferior orbital fissure into the pterygopalatine fossa; through the foramen ovale into the infratemporal fossa.

Clinical Features

Pre-admission clinical history varied from 1 to 6 years, and averaged 3.2 years. Posterior fossa tumours exhibited a shorter clinical history (1–3.2 years, av. 2.2 years) when compared with either Gasserian ganglion - dumb-bell (1.5–6 years, av. 3.4 years) or peripheral branch (1.5–6.2 years, av. 3.8 years) tumours. This matched well with the size of the lesions (Table 2), which appeared remarkable in a large number of our cases.

Subjective or objective signs of trigeminal nerve dysfunction were the general rule in the present cases. Typical trigeminal neuralgia was a complaint in only 11 (10%) of the present patients. Posterior fossa tumours showed rather constantly a sensory deficit in the trigeminal distribution usually accompained by weakness of the masticatory muscles. Auditory and/or facial deficits were present in the majority (20 out 26 cases) of these patients, as was ataxia also. Other cranial nerve palsies occurred less frequently. Eight (35%) of these cases showed a contralateral hemiparesis, whilst an ipsilateral pyramidal weakness was detected in 3 (13%) patients.

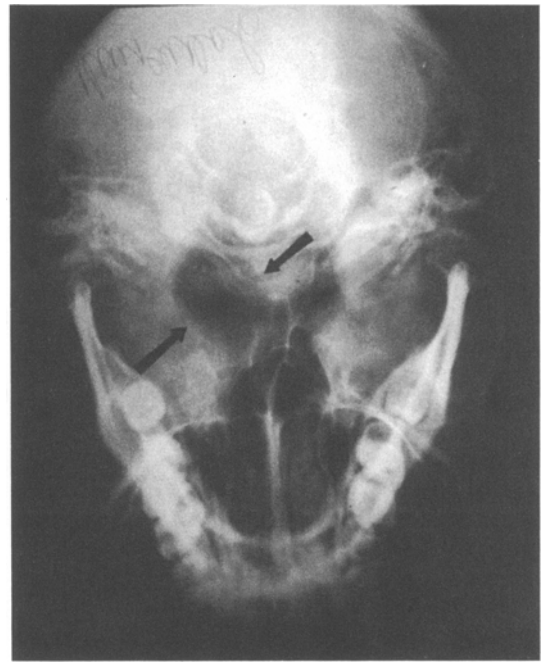


Fig. 1. Pain skull X-Rays demonstrate erosion of the floor of the middle fossa and widening of the foramen ovale

Tumours located either exclusively (42 cases) or predominantly (30 cases) in the Gasserian ganglion had a previous history of epilepsy in most instances. "Typical" temporal epilepsia, with either gustative and/or auditory hallucinations, was a sign and symptom in 23 cases (32%). Hemiparesis was detected in 12 patients (17%), whilst dysphasia was a significant sign in 7 cases (10%) harbouring tumours in the dominant hemisphere. Signs of increased ICP were present in approximately half of these patients (29 cases), as a likely result of the remarkable size of the lesions. Oculomotor nerve palsies occurred not unfrequently in these cases, however, it appeared to be significant – i.e., complete III and/or VI nerves deficit – in only 10 patients (14%).

Tumours of the trigeminal roots often exhibited exophthalmos (4 cases) and deformation of the facial skeleton (5 cases), as the result of the actual location of the individual lesions. Oculomotor nerve palsies of various degrees was a frequent finding also.

Diagnostic Radiology

Traditional radiology was contributory in most cases (Table 3) and showed signs of bony erosion in the middle fossa, the orbit and/or the petrous apex (Fig. 1).

Cerebral angiography was performed in 76 patients, and showed anterior-inferior displacement of ICA in 24 cases, with tumours of the Gasserian ganglion. In 52 patients with either "dumb-bell" or posterior fossa neurinomas a supero-medial displacement of the posterior cerebral and superior cerebellar artery

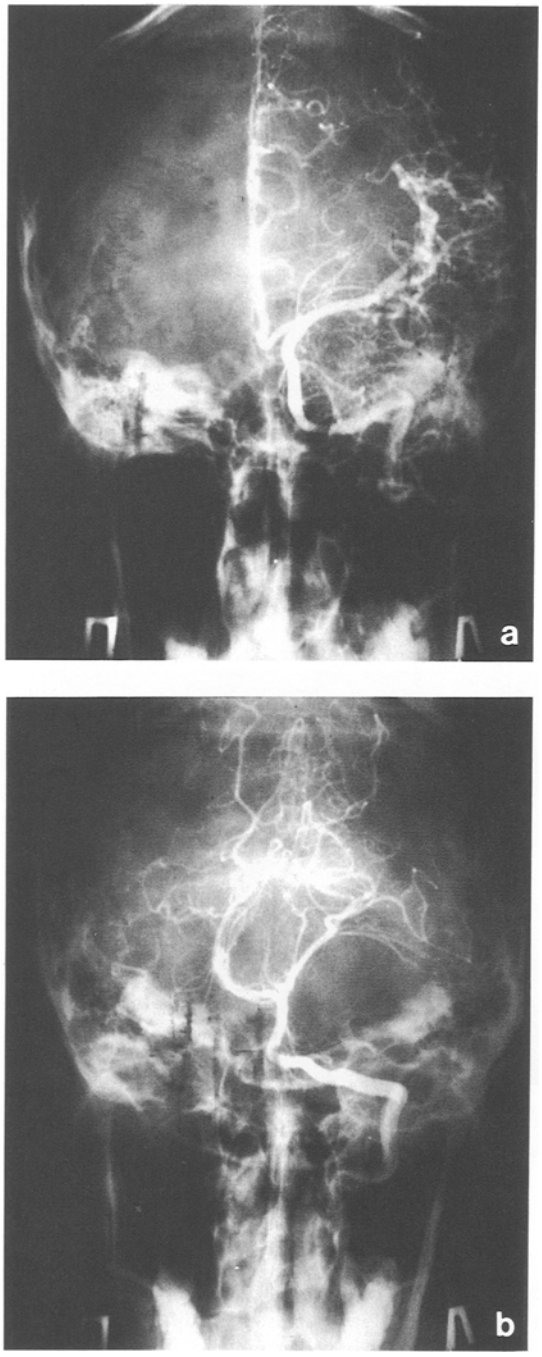


Fig. 2. Cerebral angiography. (a) Left carotid injection shows upward and medial displacement of the MCA due to a large-size "hourglass" neurinoma. (b) Left vertebral injection shows displacement of the superior cerebellar and posterior cerebral arteries. A minimal tumoural "blush" is demonstrated by carotid injections

was demonstrated, together with an inferior displacement of the anterior-inferior cerebellar artery (Fig. 2a, b). A definite tumoural blush was detected only occasionally.

Computed tomography was performed in 84 patients, and totally replaced the previously routinely performed air study in the last

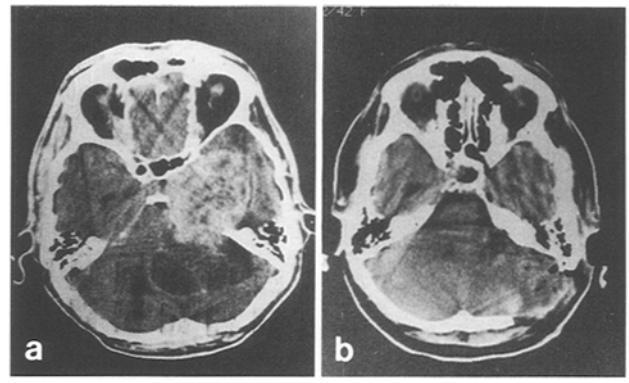


Fig. 3. CT scan following contrast enhancement. (a) A dishomogeneous "hourglass" shaped mass occupying at the same time the left middle and posterior fossa is evident. (b) Postoperative scan demonstrates total tumour removal

Table 4. *Surgical Approaches in 111 Cases*

Approaches	Years		
	1962/1977	1978/1989	1990/1994
Subtemporal[a]	11	20	2
Transtentorial	7	14	6
Suboccipital	8	15	3
Frontotemporal[a]	1	2	3
Subfrontal	1	3	1
Combined sub-temporal-suboccipital	–	5	4
Fronto-zygomatic	–	–	3
Presigmoid	–	–	2
Total cases	28	59	24

[a] Combined with orbitotomy in 5 cases.

80 cases. A typical dishomogeneous hyperdense lesion was shown in 71 cases, whilst 24 cases exhibited areas of decreased density in the context of an hyperdense mass, which suggested a cystic tumour (Fig. 3a, b). Nine cases (10.7%) showed an isodense lesion with faint contrast enhancement.

MRI was performed in the most recent 32 cases, and typically showed an hypo-isodense mass in the T1 sequence (26 cases – 81.2%), which became hyperintense in T2 (28 cases – 87.5%). The lesions highly enhanced with Gadolinium (Fig. 4a, b).

Surgical Treatment

A variety of surgical approaches were used in the present series (Table 4). We analysed 3 different periods: 1962–1977, when the operating microscope was either not available (until early 70s) or still not routinely used in our Institute; 1978–1989, when microsurgery became a rigid routine for basal tumours; 1990–1994, when either combined or skull base approaches were extensively used, if indicated, in the present cases.

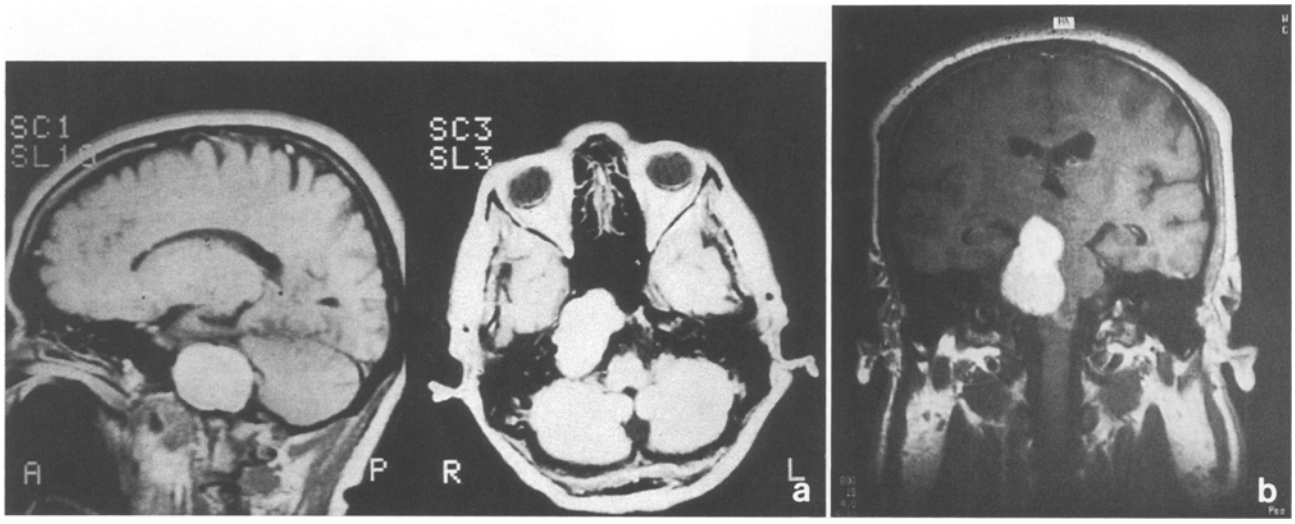


Fig. 4. MRI with Gadolinium, T1 phase. (a) Sagittal and axial scans and (b) coronal scan, give evidence of this huge dumb-bell trigeminal neurinoma arising from Meckel's cave on the right side

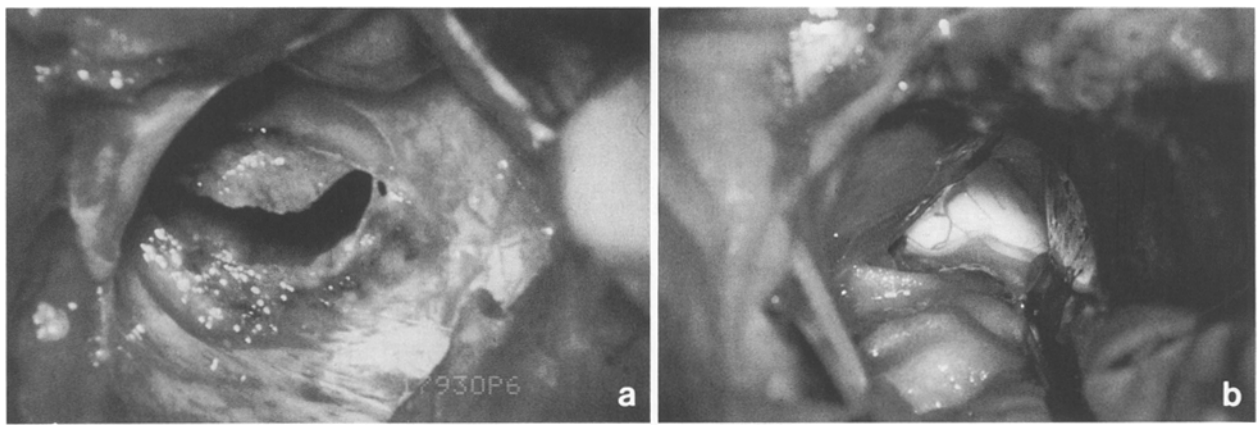


Fig. 5. Same case as Fig. 3. (a) Following left presigmoid approach and tentorial section the tumour is exposed and extensively removed intracapsularly. Retractor holds the left temporal lobe. (b) Following complete tumour removal, the lateral aspect of the brain stem comes into view

Tumours involving predominantly the Gasserian ganglion and/or the peripheral branches were approached using either the subtemporal or the fronto-temporal approach.

Tumours with predominant location in the posterior fossa were removed using the retrosigmoid approach.

Large "dumb-bell" tumours were traditionally removed via a subtemporal route supplemented by a wide tentoriotomy until recent years, when either combined supra-subtentorial, or skull base approaches were adopted with the ultimate idea of achieving a surgical field wide enough for performing a total removal of the lesion. Modern diagnostic imaging tools significantly assisted in planning properly the approach in the more recent cases.

Modern skull base approaches were used in 5 cases. Three were approached via a presigmoid route. As far as the remaining 2 patients, 1 was operated on using a fronto-zygomatic approach as described in the literature [1, 9, 20]. In the remaining one, as well as in a few cases of basal cranio-orbital and cavernous sinus lesions operated on during the last year, we used a modification of the fronto-orbito-zygomatic craniotomy which allows the zygomatic arch to remain attached to the frontal bone in its full length, and makes its replacement and fixation at the end of operation very easy and effective. As a rule, tumour removal was attempted in one stage. Removal of the lesion started with a generous intracapsular debulking, obtained using curettes, sucker and the ultrasonic surgical aspirator in the last 10 years. Then the lesion is gradually mobilized and separated from the adjacent neurovascular structures, and finally removed (Fig. 5a, b). Bleeding from the involved cavernous sinus may be significant, but can be easily controlled with Gelfoam tamponade. Care must be taken when debulking tumour close to the carotid artery, whose location with reference to the lesion must always be borne in mind, in order to avoid inadvertent vessel injury. This occurred in one of our early cases. Several factors, includ-

ing tumour size and location, vascularity, invasion of the cavernous sinus, invasion of bony structures with significant extracranial extension, adversely affected surgical removal. This was thought to be achieved completely in 86 (77.4%) as reported on by the operating surgeon and verified by postoperative CT control, with evident improvement in the more recent cases (Table 5).

Histology

97 tumours were classified as schwannomas, and 14 as neurofibromas. Two cases, one schwannoma and 1 neurofibroma, respectively, showed features of histological malignancy, such as abundant mitoses, cellularity and tendency to infiltrate the adjacent structures including the dura mater (Fig. 6).

Results

Three cases died post-operatively (Table 6). Tumour removal had been radical in but one of these cases. Of the 108 patients surviving surgery, long term follow-up was available in 98 cases. Out of the 10 cases lost to follow-up, 5 underwent a grossly total removal, and 5 an incomplete removal. In the remaining cases follow-up ranged from 13 months to 33 years, and averaged 13,5 years.

Overall, 84 patients (86.7%) showed good to excellent longterm results, with partial trigeminal deficit as the only surgical sequela (Table 7). There was clear improvement with increased experience, particularly after the introduction of microsurgical techniques as is also shown by the analysis of postoperative complications, which are listed in Table 8a, b.

As far as skull base approaches are concerned, we have the impression that tumour exposure and removal was greatly facilitated. However, the cases were too few to make a definite statement.

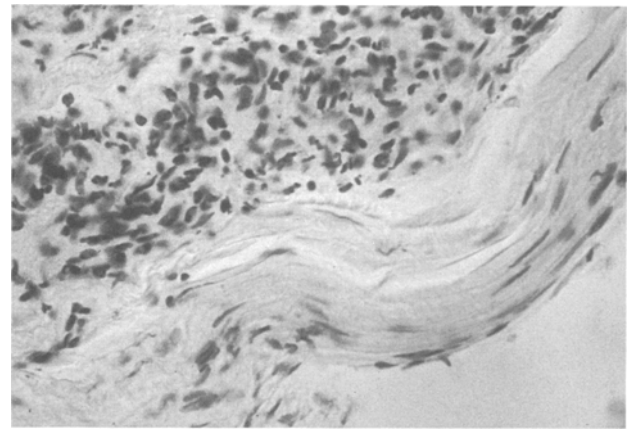


Fig. 6. Photomicrograph (×100), of the histological specimen demonstrating infiltration of the dural sleeves of the Gasserian ganglion

Interestingly, major complications such as postoperative cranial nerve deficits and hemiparesis showed a definite tendency to improve with time.

Recurrences

13 patients (11.7%) had a symptomatic recurrence following incomplete tumour removal, and required further surgical treatment. The approaches used in those cases are summarized in Table 9. Either combined or skull base approaches were used in 4 cases operated on in the last 2 years, 3 for the second time, and one for the third time.

Radicality in cases where a skull base approach was adopted tended to be higher when compared with the remaining cases.

Table 5. *Surgical Radicality*

Year	No. of patients	Radical removal	Mortality
1962–1977	28	19 (67.8%)	2 (7.1%)
1978–1989	59	46 (78%)	1 (1.7%)
1990–1994	24	21 (87.5%)	–

Table 7. *Long-Term Surgical Results in 98 Cases*[a]

Good (no or moderate V c.n. palsy)[b]	84
Unmodified	7
Worsened	7

[a] Follow-up 15 months – 33 years, av. 13.5 years.
[b] Decreased but not abolished sensation in the distribution of the trigeminal nerve, with spared corneal reflex.

Table 6. *Characteristics of the Cases Who Died*

Location	No. of cases	Surgical approach	Tumour removal	Year of operation
Gasserian ganglion – subtentorial	2	subtemporal	1 partial 1 radical	1968 1978
Posterior fossa	1	paramedian	radical	1969

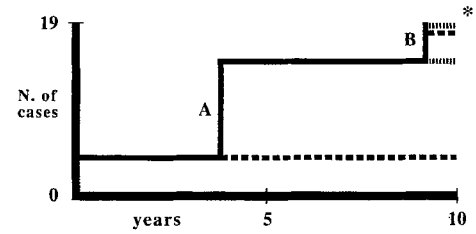
Table 8 a. *Postoperative Complications*

Complications	Years			
	1962–77	1978–89	1990–94	Total
CSF	1 (4%)	2 (3.3%)	1 (4%)	4 (4%)
Hydrocephalus	–	1 (1.7%)	1 (4%)	2 (2%)
Temporal intracerebral haematoma	2 (7%)	–	–	2 (2%)
Subdural haematoma	1 (4%)	–	–	1 (1%)
Temporal lobe oedema	1 (4%)	1 (1.7%)	–	2 (2%)
Hemiparesis worsening	5 (18%)	6 (10%)	–	11 (10%)
Aphasia	1 (4%)	2 (3.3%)	–	3 (3%)
Pneumonia	1 (4%)	1 (1.7%)	–	2 (2%)
Temporal lobe epilepsy	1 (4%)	2 (3.3%)	–	3 (3%)
Brain-stem ischaemia	2 (7%)	2 (3.3%)	–	4 (4%)
4th and 6th c.n. palsy	3 (11%)	7 (12%)	4 (16%)	14 (12%)
3th	1 (4%)	3 (5%)	2 (8%)	6 (5%)
7th	3 (11%)	5 (8.5%)	1 (4%)	9 (8%)
Total	22/28	32/59	9/24	63/11

Table 8 b. *Postoperative Complications.* Comparison of early versus six months–one year follow-up

Complications	Early postoperative period	Follow-up[a]
	No. of cases	No. of cases
5th c.n. palsy	All	88
3th, 4th, 7th, c.n. palsy	29	12
Hemiparesis	12	4
CSF leak	4	–
Hydrocephalus	–	2

[a] Six months to 1 year postoperatively.



▪▪▪ patients with asympomatic residual/recurrent lesions.
ıılıılıı patients in whom tumors were (almost) totally resected at repeated surgery.
* 1 patient died following fourth operation (see text).
A first re-operation.
B second re-operation.

Fig. 7. Kaplan-Meyer's curve depiticting the time course of clinical recurrences in the present series

Table 9. *Surgical Approach at Second Operation*

– Same	6/13	(radical in 3/6)
– Other	4/13	(radical in 3/4)
– Skull base	3/13	(radical in 3/3)

3 cases re-operated on 3 times (radicality 1/3)
1 case re-operated on 4 times[a] (radicality 1/1)

[a] Dead.

The interval between a first incomplete and a second operation ranged from 1.4 to 9 years, average 3.8 years. Repeated surgery was judged grossly total in 9 (69.2%) of the cases. In the remaining 4 patients a third intervention was required, at an average interval of 5.6 years (2.3–16 years) from the 1st re-operation (Fig. 7). Surgery could be radical in only one of these cases. One patient was re-operated on a forth time, and died following an attempt at total removal. This was the only death in this series of patients operated on for recurrences (7.7%). However, the rate of postoperative complications was definitely higher than that of patients operated on for the first time (Table 10). This is not surprising if one considers that repeated surgery in intracranial neurinomas is particularly troublesome due to significant adhesions which require difficult, meticulous and sometimes unavoidably traumatic dissection of the recurrent lesion.

We could not identify any significant factor which would distinguish the 13 patients exhibiting clinical signs of recurrence and requiring re-operation from

Table 10. *Post-Surgical Complication in Second Surgery*

	%
CSF leak	2 (15.4%)
Temporal lobe oedema	1 (7.7%)
Worsening of paresis	5 (38.5%)
Aphasia	1 (7.7%)
Pneumonia	1 (7.7%)
Temporal epilepsy	1 (7.7%)
Brain-stem ischemia	1 (7.7%)
IV and VI c.n. palsy	4 (30.8%)
III c.n. palsy	2 (15.4%)
VII c.n. palsy	3 (23.1%)
Total cases	13

those 6 that harboured an apparently quiescent residual lesion.

Discussion

Neurinomas of the trigeminal nerve are rare, and the total number of cases reported in the major literature does not exceed 200.

The unusual number of cases in the present series reflects our particular pattern of referrals, which enabled us to collect a great number of brain tumours from all over the former Soviet Union.

Generalities

The incidence of our neurinomas as related to brain tumours as well as to intracranial neuromas falls into the range reported in the literature [2, 5, 6, 10, 12–19, 21–23], as does the age of patients too. Although a predominance of female patients has been generally noted [13], this appeared to be rather striking in the pesent series.

The approximately 10% incidence of neurofibromas in the present series matched that of the literature [13], as it does the very low incidence of malignant tumours.

Clinical and Diagnostic Features

Neurological signs were related to the actual location of the lesion. Prepontine predominantly posterior fossa neurinomas occasionally posed problems in the differential diagnosis with intra-axial brainstem tumours, owing to similarity of clinical symptoms and signs, and sometimes of CT appearence also.

The typical CT appearence of the tumours is well known [13]. Dishomogeneous lesions with areas of diminished density in the context of an hyperdense mass were observed in one fourth of the present cases submitted to CT scanning, and represented tumours which underwent partial cystic degeneration. As stated above, this may create problems of differential diagnosis with intra-axial brainstem tumours when neurinoma is located in the posterior fossa only. MRI gives very useful information for surgical planning due to its capacity to outline threedimensional aspects. Although MR angiography is available in our Institution since January 1994, no one of the present cases underwent this examination.

Surgical Management

Surgery of trigeminal neurinoma may be a very challenging task, particularly if the lesion is located both supra- and subtentorially and has attained a considerable size. In general the results of surgical management of these tumours have been rather disappointing in the past [10, 11, 18], due to the limited number of total lesion removals that could be achieved and the consequent high rate of recurrences, as well as considerable operative mortality and morbidity. The advent of microsurgery together with improvement of diagnostic imaging has allowed a definite improvement in the results of surgery [2, 3, 13, 17]. However, large dumb-bell tumours, extending either into more than one intracranial compartment or extracranially, remain very difficult to remove totally, and require in several cases a two-stage procedure to obtain a gross total removal. In general, tumours located in the middle fossa lie extradurally and as a rule have a distinct plain of clivage, whilst posterior fossa trigeminal neurinomas are localized in the subarachnoid space and usually have rather significant adhesions with the surrounding nerves and vessels as well as with the pia mater. The principles of microsurgery include careful dissection of the tumour capsule from the delicate surrounding structures following adequate debulking of the mass. In this respect, the size and the vascularity of the tumour may represent a serious problem for the surgeon, who can be forced to debulk rather quickly a richly vascularized tumour in order to reduce its size in a convenient manner without causing a significant blood loss. This, however, may pose problems in maintaining an adequately clear surgical field for accurate microsurgical dissection. Another problem is represented by the invasion of the cavernous sinus, which was considered a definite limit to total remov-

al until few years ago [10, 14]. However, recent advances in the surgery of the cavernous sinus [7] has made possible total removal of neurinomas even involving the cavernous sinus [22].

The recent introduction of the techniques of skull base surgery have allowed pioneers in this field to obtain extremely good results in terms of radical tumour removal and postoperative morbidity and mortality in limited series of trigeminal neurinomas [4, 8, 16, 22]. The philosophical principle of the skull base approach is to give to the surgeon a wider operative field and a shorter working distance for manipulating a deep-seated lesion. This applies certainly to trigeminal neurinomas also, however, these advantages must be balanced against the shortcomings of the lateral skull base approach and its sequelae, i.e., the length of the procedure and the morbidity related to the approach itself in terms of unadvertent injury of neurovascular structures and CSF leak. Modern skull base approaches are obviously mandatory for the treatment of the rare trigeminal neurinomas with large extracranial extension [4], which represent a definite minority in neurosurgical cases [16] including ours.

In our experience, the size of the lesion was a significant issue in dictating surgical tactics. In fact, if "traditional" approaches are used adequate debulking of large tumours may allow obtaining enough space for microsurgical dissection and removal of the tumour capsule. Combined and/or skull base approaches appear to be mandatory in "dumb-bell" neurinomas in order to obviate a two-stage procedure. Deep-seated lesions of smaller size which would require a significant amount of brain retraction for exposure and to properly deal with them would definitely benefit from targeted skull base approaches. We found the presigmoid approach very appropriate for these neurinomas, since it allows an easy and complete tentorial section which offers an unobstructed view of the target area. The transzygomatic middle fossa approach also can be a sound alternative, although it is our impression that full division of the tentorium with this approach would pose a significant risk of injuring the fourth nerve unless the fifth nerve is fully exposed in Meckel's cave, with consequent risk of incomplete dural closure and CSF leak.

We experienced a postoperative CSF leak in one case where the presigmoid approach was used, which required two additional procedures to manage this complication; however, this occurred early in our experience. Another case deteriorated following subtotal removal of a huge dumb-bell neurinoma exposed via a transzygomatic transtemporal approach; however, this was the third operation performed in an incompletely removed tumour, originally located almost exclusively in the posterior fossa, which later progressed to occupy a considerable part of the middle fossa.

In our experience, microsurgery definitely contributed to improvement of surgical results. Our philosophy of attempting a radical removal of the lesion irrespective of the size, as well as familiarity and a considerable experience with the subtemporal transtentorial approach, has allowed us achieving a gross total removal in a large percentage of sizeable lesions treated using microsurgical technique, with a reasonable incidence of postoperative complications. The tendency of those tumours to expand bony foramina and locate subarachnoidally in the posterior fossa makes it very difficult sometimes to achieve a radical removal. Also, an apparent gross total removal of a huge trigeminal neurinoma does not give full assurance that tumour cells are not inadvertently left in situ. However, this would not necessarily mean that such a situation would end up with a recurrence later [13]. It is worthy of note that out of a total of 24 present cases surviving surgery where tumour removal was macroscopically incomplete, only 13 required repeated surgery for symptomatic recurrence.

Conclusion

Neurinomas of the trigeminal nerve are rare. Their complex clinical symptomatology tends to the related clearely to the actual location of the lesion.

Surgery may be difficult, particularly in cases of "dumb-bell" supra-subtentorial lesions. On the other hand a subtotal tumour removal does not necessarily mean a symptomatic tumour recurrence requiring repeated surgery. Radical surgery remains the therapeutic goal in trigeminal neurinoma. Microsurgery, and more recently the techniques of skull base surgery, together with modern diagnostic imaging tools, have definitely assisted in obtaining increased management radicality with an acceptable rate of surgical complications.

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Comments

The interesting aspect of this paper is the exceptionally large series of trigeminal neurinomas, certainly the largest in the world. This series will remain unparalleled for a long time.

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