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Neurosurgery

ROLE OF GAMMA KNIFE RADIO SURGERY IN MANAGEMENT OF VESTIBULAR SCHWANNOMAS-THE INDIAN ARMED FORCES EXPERIENCE

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ABSTRACT Vestibular Schwannomas are the most common tumours of the cerebellopontine angle, with an estimated prevalence of 8-10% of all intracranial tumours and 70-80% of all CP angle tumours. [1]. Conventional Surgery has been the dominant treatment for Vestibular Schwannomas for decades. Gamma Knife surgery (GKS) has emerged as a treatment option that incorporates the benefits of radiation therapy while minimizing the adverse side effects of conventional surgery & conventional radiotherapy. We present our series of 247 cases of Vestibular Schwannoma, treated with GKS over the last 10 years and will present the multimodality treatment algorithms followed in our centre for these complex tumors & would review various treatment modalities currently available.

KEYWORDS : vestibular schwannoma, Gamma Knife Radiosurgery

INTRODUCTION:

Vestibular Schwannomas are the most common tumours of the cerebellopontine angle, with an estimated prevalence of 8-10% of all intracranial tumours and 70-80% of all CP angle tumours. [1].Conventional Surgery has been the dominant treatment for Vestibular Schwannomas for decades.

Radiosurgery has lately emerged as a treatment option that incorporates the benefits of radiation therapy while minimizing the adverse side effects of conventional surgery & conventional radiotherapy. Radiosurgery is equally effective as surgery for tumor control and possesses lower morbidity and mortality rates.

VESTIBULAR SCHWANNOMAS – AN OVERVIEW:

The nomenclature of Glomus tumor evolved with time. It was initially called acoustic neuromas because of its predominant symptom of hearing loss. It was felt however that the above nomenclature should encompass the origin of the tumour from the Schwann cells in the Obersteiner Redlich Zone [2] of the vestibular nerve & was more appropriately christened as Vestibular Schwannomas.

As is obvious, vestibular schwannoma is a histopathological diagnosis. However in view of most cases in this series having been knifed primarily, no histopathological studies are available in these cases. The authors have chosen to label these cases as vestibular schwanomas on the basis of the characteristic and almost pathognomonic radiological features of these lesions in the form of contrast enhancement, acute angle with petrous bone, eroded porous acousticus, extension into internal auditory canal, with or without dural tail & with absence of calcification or hyperostosis.

Epidemiology& Presentation:

70-90% of CP angle tumors have been found to be vestibular schwannoma in various series. In the vast majority of patients vestibular schwannoma will develop as a sporadic neoplasm. These tumors most commonly occur between the ages of 30 and 70 years with a peak incidence between 40 to 60 years. The syndromic patients with neurofibromatosis type 2 often develop bilateral vestibular schwannoma which is sufficient to make the diagnosis of the disease. Approximately 5 to 20% of patient with solitary intracranial schwannomas may have neuro fibromatosis type [2].

The prevalence of Neurofibromatosis type 2 though initially estimated at 1:2000000 has been recently estimated to be as high as 1:60000. [3]
The first report of an acoustic tumor was that in 1777 by Sandifort who recognised it at an autopsy. The first successful surgery was carried out by Sir Charles Ballance in 1894, however it was not until 1917 when Harvey Cushing described the clinical history and the symptoms in details. Paradoxically Cushing himself did not realise that the first symptom could be hearing loss and tinnitus until after he had reviewed his own case material.

There can be a 4 year delay between the onset of first symptom and diagnosis of vestibular schwannoma. The various signs and symptoms of vestibular schwannoma could be unilateral hearing loss, unilateral non pulsatile tinnitus, vestibular dysfunction, Trigeminal hypoesthesia, cerebellar dysfunction, raised intracranial pressure, facial nerve dysfunction, headache, lower cranial nerve palsies and the long tract signs. [4]

Treatment Options:

Observation

It is quite common in this era of ever growing use of MRI for a variety of indications, in combination with the dropping cost, that more and more intracranial lesions are being found incidentally. Approximately two third of Vestibular Schwannoma do not grow during a 5 year observation period which is supposedly a cut off to distinguish growing from non-growing tumor using a volume doubling time model. Approximately 50% of patients maintain good hearing at 5 years observation. Tumor size greater than 1.5 to 2cm is unfit for observation as larger tumor size may predict future tumor growth. The natural history of incidentally found lesions must be weighed against the potential morbidity associated with treatment.[5]

Surgery:

Complete surgical resection is off course the ideal management of Vestibular schwannoma, however, which may come at a formidable price. Surgical excision has been attempted since 1894, with a high incidence of associated complications like hearing loss, facial nerve paresis, lower cranial nerve palsies, meningitis, wound infection, - the list is endless; not to mention a delayed recurrence in cases of incomplete excision. With improving surgical technique, there has been a significant reduction in morbidity rates over the ages.

Radiosurgery, Radiotherapy, Adaptive Hybrid surgery:

Gamma knife radiosurgery has been proposed as an alternative to conventional surgery, whether delivered as a primary or adjunctive therapy. First used by Leksel in 1971, stereotactic radiosurgery is appealing because it does not involve making an incision and is a day care procedure with minimal immediate morbidity. However there are concerns about its long term morbidities like facial paresis, hearing loss, vestibular dysfunction, facial spasm, Trigeminal numbness, brainstem edema and hydrocephalus. Fractionated radiotherapy has shown excellent result as far as tumor control is concerned when compared to stereotactic radiosurgery, however there does not seem to be a significant difference in facial and cochlear nerve preservation whether the radio surgical dose is delivered in a single fraction or over multiple fractions. Fractionated radiotherapy is significantly better in tumors larger than 3 cm when compared to SRS.

The development of an adaptive hybrid surgical technique may offer the best of both the world to the clientele. A real-time intraoperative neuro-navigation helps the surgeon to exactly visualise the extent of the section and do it safely. The medical fraternity can precisely direct the excision to achieve a good radio surgical target while minimising the risk of surgical morbidity and maximizing the rate of local control [5]

MATERIAL AND METHOD:

This retrospective study carried out at the apex tertiary care centre of the armed forces medical setup in India, retrospectively analysed the use of Gamma knife radiosurgery as a primary or adjunctive form of therapy for Vestibular Schwannoma, offered to patients over a period of 10 years at this centre and compared the same with available national and international statistics. The radiosurgery was carried out using a Leksell Gamma knife Elekta instruments, Stockholm, Sweden, model 4 C, installed in 2007. The gamma knife was planned with safety tolerance limit of dose to brain stem, cranial nerve& Cochlea not to exceed 12 Gy, 20 Gy & 5 Gy respectively. The prescribed tumor margin dose was more than 12 Gy if possible. A retrospective chart analysis of 247 patients who underwent a radio surgical procedure as a primary or adjunctive therapy was carried out.

RESULT:

Of the 763 cases who received Gamma knife radiosurgery at this centre till Sep 2018, only 247 cases with Vestibular Schwannoma underwent the procedure, either as a primary therapy or as adjuvant therapy to post-operative residual tumour. The tumor volume was ranging being between a minimum of 0.18 cc to a maximum of 36.5 cc .

Tumor Volume	Number of Cases
0-10	230
10-15	14
15+	3
Grand Total	247

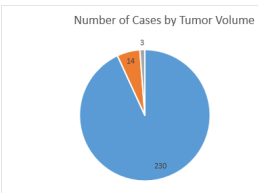

Fig 1.

Gender	Number of Cases
Female	109
Male	138
Grand Total	247

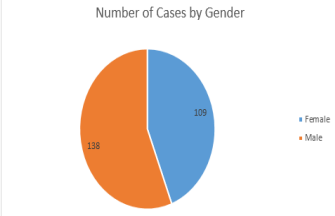

Fig 2.

Age Group	Number of Cases
0-20	5
20-30	38
30-40	45
40-50	51
50-60	64
60-70	30
70-80	12
80 +	2
Grand Total	247

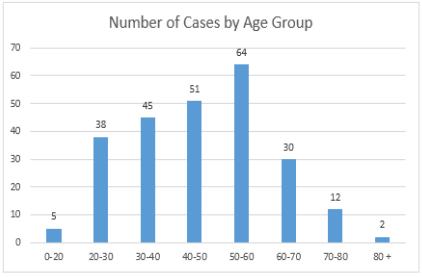

Fig 3.

Male is to female ratio was 1:1.3. 93% of the tumours knifed were between the volume range of less than 10 cc and the average dose of radiation was 12 Grey. Maximum prevalence was noted in the age group of 40 to 60 years. The follow-up has ranged from 6 months to 10 years. There has been no loss to follow up.

All cases, except one, which received gamma knife radiosurgery not only exhibited tumour control (defined as unchanged or reduced tumour volume in follow up) but a few also showed subtle improvement in status viz recovery of lower cranial nerve deficits. The clinical improvement, if any, in cases where gamma knife surgery was offered as adjunctive postoperative therapy for residual lesion, was ignored to offset any confounding factor generated by spontaneous recovery over a period of time from iatrogenic neurological paresis caused by surgery. No radiation induced adverse effects were noted in this particular series.

All the cases of Vestibular Schwannomas were of sporadic type except for a case of 30 years old lady who was knifed bilaterally in the same sitting with a tumour volume of 6.8cc on left & 2.6 cc on right.

Tumour progression was noted in only one case which was a case of 36 years old serving soldier with an initial tumour volume of 2.4 cc who was administered a dose of 12 Grey at 50% isodose with a tumour coverage of 96% on 17 sep 2011. (Fig 4a & 4b)

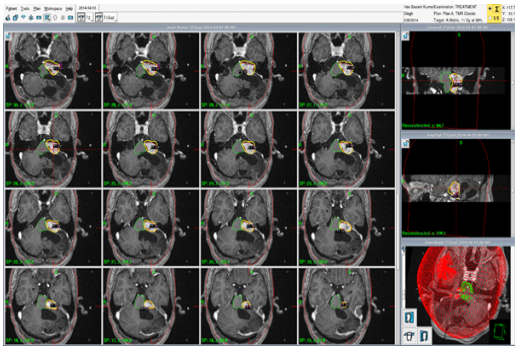

Fig 4a.Initial GKS Protocol

He showed tumour progression and had to be re-knifed on 03 Apr 2014 with 11.5 Grey dose, 50% isodose.

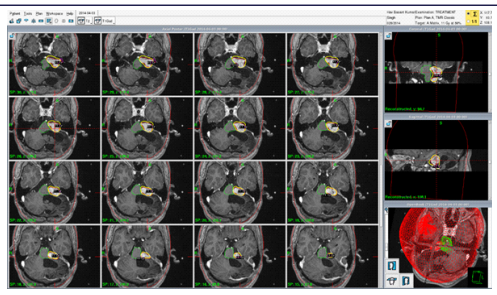

Fig 4b. Re-do GKS protocol on tumour progression

A 52 years old lady with a tumour volume of 36.5 cc, who was a high risk surgical candidate and was unwilling for fractionated radiotherapy, was offered gamma knife in two stages. Sectorial knifing was done for her on 24 Feb 2018 and 16 Aug 2018. The patient is under follow up.(Fig 5a & Fig 5b)

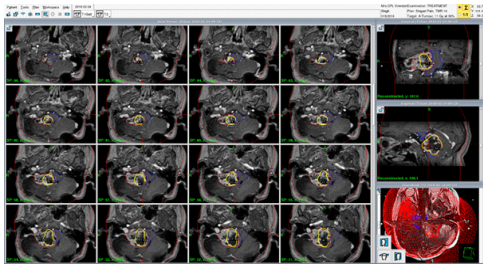

Fig 5a Knifing of medial sector- First sitting

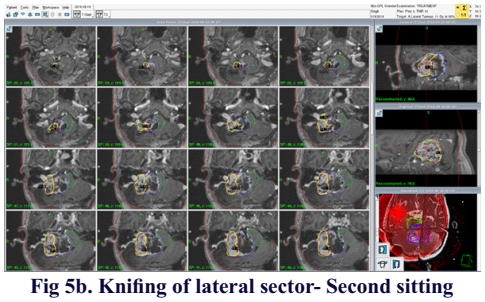

Fig 5b. Knifing of lateral sector- Second sitting

DISCUSSION:

Progression rates following stereotactic radiosurgery (SRS) are similar to recurrence and progression rates seen following surgery and fractionated radiotherapy respectively [6,7,8] Most tumours with single dimension less than 3cm and tumour volume around 10 cc can be effectively managed with GKS. Tumour volume more than 15 cc respond poorly to GKS.[9]

None of the cases in our series showed progression except one. To our surprise the tumour that progressed was not in the volume range of >15 cc as noted for high propensity for progression in some series[9].It fell in the group of less than 10 cc which have been conventionally considered to be the most responsive to GKS, reinforcing the importance of innate tumour pathology in growth progression. Progressive tumors are treated with either microsurgery or radiosurgery.[10]. We treated our only case that progressed despite GKS, with repeat GKS & with good result.

Tumors < 3cm in diameter can be treated initially with Gamma knife, fractionated radiation therapy (range 50–54 Gy) or proton therapy.[6] There are no direct comparisons among these modalities and outcomes with respect to tumor control and hearing preservation have been similar.[6,7,8]

We in our series have treated 3 cases with volume more than 3 cc with GKS with good outcome. The largest tumour treated in our series was more than 53 cc in volume. This was knifed in two sittings in sectorial fashion and is in regular follow up.

An analysis of international results show that majority of these lesions respond well to SRS with 32% showing regression and 59% showing senescence. Around 9 % of these lesions show progression despite SRS.

We in our series have seen a tumour progression rate of 0.4 % only. Most series are for lesions smaller than 2.5 cm. A few series for tumours >3 cm have shown more incidence of progression, facial nerve paresis. An SRS renders the tumours to have increased post op morbidity than usual in case they do need to undergo surgery.[5] However in our series we treated our only post GKS progression with repeat GKS so we have nothing to comment on these lines.

CONCLUSION:

- There may be a controversy regarding the modality of action on these tumors by radiosurgery – however- evidently, there seems to be no controversy about the efficacy of the same.
- Conventional Surgery is associated with a significantly higher morbidity as compared to radiosurgery
- Radiosurgical treatment is strongly recommended for small tumors < 10 cc or recurrent tumor after surgery with minimal or no brainstem compression.
- Fractionated radiosurgery is recommended for unresectable large tumors.
- Large symptomatic tumors with significant mass effect on the brainstem however may need to be surgically debulked.
- In a benign disease, tumor control and quality of life indices are probably more significant than eradication and morbidity.
- The overall quality of life appears to be significantly better with GKS as a primary and is an effective adjunct as a secondary modality of therapy.

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