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TO ASSESS THE QUALITY OF LIFE IN SKULL BASE TUMORS – ESTHESIONEUROBLASTOMA

Otorhinolaryngology

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ABSTRACT

Introduction: Esthesioneuroblastoma (ENB) is a rare malignant neoplasm, comprising about 3% of all intranasal neoplasms. Most of these patients present in locally advanced stages and require multimodality treatment. Multimodality approach with a risk-adapted strategy is required to achieve good control rates.

Objective : This article outlines the various newer details of quality of life assessment in esthesioneuroblastoma and The importance of early diagnosis of esthesioneuroblastoma.

Methodology :A Retrospective study of 2 years duration with a sample size of 14 cases. Pre-Treatment and 1½ year follow-up questionnaire was given to all the patients who were included in the study.

Results: Recorded data shows poor Quality of Life (QOL) soon after diagnosis. Same worsened if the presenting stage was advanced.

Conclusion: Early detection and radio-imaging, with diagnostic nasal examination (DNE) helps in improving the QOL in patients with esthesioneuroblastoma.

KEYWORDS

Esthioneuroblastom, Quality Of Life, Diagnostic nasal endoscopy

Introduction:

Esthesioneuroblastoma (ENB) is a rare malignant neoplasm arising from the olfactory neuroepithelium. ENB was first described by Berger *et al* in 1924 [1]

It constitutes only 3% of all malignant intranasal neoplasm [2]

Sites involved - nasal septum, Olfactory fossa, turbinates - extends to skull base

Manifests across all ages with peaks in the second and sixth decades of life [3]

Patients present in locally advanced stages and require multimodality treatment which includes surgery, chemotherapy and radiotherapy [4]

Multimodality approach with a risk-adapted strategy is required to achieve good control rates while minimizing treatment related toxicity [5]

Clinically presents as unilateral nasal obstruction, epistaxis, change in sense of smell, headache, facial distortion, visual change and neck swelling

Intracranial extension commonly causes proptosis (infiltration into superior orbital fissure) and Cranial nerve palsies (infiltration into skull base foramen and brain) Hyams histopathological grading [6]

Grade	Lobular architecture preservation	Mitotic index	Nuclear polymorphism	Fibrillary matrix	Rosettes	Necrosis
I	+	Zero	None	Prominent	HW rosettes	None
II	+	Low	Low	Present	HW rosettes	None
III	+/-	Moderate	Moderate	Low	FW rosettes	Rare
IV	+/-	High	High	Absent	None	Frequent

Modified Kadish staging [7]

Stage A: Tumour limited to the nasal fossa
Stage B: Tumour extension into the paranasal sinuses
Stage C: Tumour extension beyond the paranasal sinuses and nasal cavity
Stage D: Distant metastasis

Aims & Objectives: Study and compare quality of life in Esthesioneuroblastoma patients

Methodology:

A Retrospective study (open – consecutive cases - convenient sampling)

Duration - 2 years (Aug 2015 to Aug 2017)

Sample size of 14 cases (n=14)

Institutional Review Board /Ethical committee clearance obtained

Written Informed consent was obtained from the patients and their attenders

Inclusion Criteria

- Histopathology confirmed esthesioneuroblastoma
- No previous intervention
- Age > 12 years

EORTC QLQ C30 Pre-Treatment and 1½ year follow-up questionnaire was given to all the patients who were included in the study

All patients underwent diagnostic nasal endoscopy and radio-diagnostics

Specialty advise taken and treated accordingly (Neuro-, Plastic-, Vascular- surgery and psychology)

Statistical tool used was paired t-test

Statistical analysis was obtained using IBM SPSS v22 for Windows

p value < 0.05 was considered statistically significant

Observation – results:

n=14.

Clinical staging and histo-pathological grading were done

To R/o observer related bias – department secretary collected data.

Followed up for 1½ years, monthly once for first 6 months, once every 2 months for next 6 months, once every 3 months for last 6 month, Patients were on age-dependent treatment regime / care / diet

Demographics and statistical data tabulated

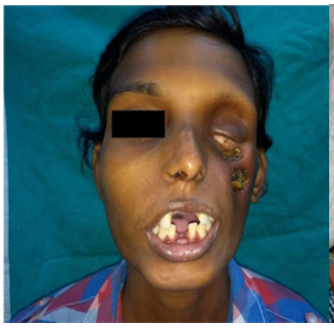

Figure 1A.Pre op following radiotherapy.

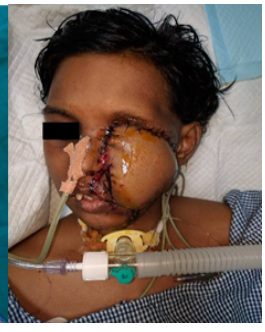

Figure 1B. Post op image of patient after orbital enucleation and flap closure

Table 1 statistical data

Paired Samples Test									
Data compared		Paired Differences					t	df	Sig. (2-tailed)
		Mean	Std. Deviation	Std. Error Mean	95% C.I. of the Difference				
					Lower	Upper			
Pair 1	PT1PRETX - PT1POSTTX	0.7	1.44198	0.26327	0.16156	1.23844	2.659	29	0.013
Pair 2	PT2_PRETX - PT2_POSTTX	1.13333	1.81437	0.33126	0.45583	1.81083	3.421	29	0.002
Pair 3	PT3_PRETX - PT3_POSTTX	0.13333	1.19578	0.21832	-0.31318	0.57984	0.611	29	**0.546**
Pair 4	PT4_PRETX - PT4_POSTTX	0.7456	1.18447	0.20966	0.17338	1.24954	2.67	29	0.011
Pair 5	PT5_PRETX - PT5_POSTTX	1.18799	1.19445	0.25767	0.18556	1.25544	3.221	29	0.001
Pair 6	PT6_PRETX - PT6_POSTTX	0.74457	1.33178	0.25627	0.16036	1.23964	2.943	29	0.003
Pair 7	PT7_PRETX - PT7_POSTTX	1.12933	1.82347	0.32226	0.45643	1.87983	3.001	29	0.002
Pair 8	PT8_PRETX - PT8_POSTTX	0.13553	1.18878	0.20732	-0.22317	0.51384	0.711	29	**0.676**
Pair 9	PT9_PRETX - PT9_POSTTX	0.8376	1.09947	0.21976	0.18009	1.25224	2.87	29	0.011
Pair 10	PT10_PRETX - PT10_POSTTX	1.18977	1.18885	0.27467	0.19956	1.30084	3.027	29	0.001
Pair 11	PT11_PRETX - PT11_POSTTX	0.88347	1.33178	0.23317	0.13456	1.21554	2.718	29	0.013
Pair 12	PT12_PRETX - PT12_POSTTX	1.20333	1.79937	0.29126	0.64583	1.77083	3.452	29	0.012
Pair 13	PT13_PRETX - PT13_POSTTX	0.13473	1.19578	0.21832	-0.31318	0.57984	0.611	29	**0.512**
Pair 14	PT14_PRETX - PT14_POSTTX	0.87356	1.09947	0.19966	0.19958	1.28854	2.997	29	0.001

Discussion:

- **Age and sample size:** n = 14. Age range was 17 – 26 years. Mean age - 21.66 (± 4.51). A Study by Wade et al (1982) which included 5 patients showed a bimodal distribution
- **Sex:** This study had a Female: Male ratio of 3:1. Similar study showed a ratio of F:M=3:2
- The Commonest clinical denominator was presentation with epistaxis
- **Metastasis -** 4 patients developed skull base metastasis
  - Advised chemotherapy – intermediate response
  - 1 candidate succumbed to the disease due to intracranial extension
  - A study of 5 recurrent esthesio-neuroblastoma by Smith et al (1996) showed intermediate response to chemotherapy.

- **Neck node status:**
  - 10 patients had N0 neck
  - 4 patients had significant lymphadenopathy in Level V and Level II with the largest nodal size of 3.4 cms
  - Study by Johns et al (2001) documented that node positive skull base lesions had poor prognosis

- According to Modified Kadish staging, 12 patients were of stage C and 2 patients were of stage D. Wade et al (1982) documented 3 cases with metastasis stating that higher the staging, poorer was the prognosis
- The histopathology showed Hyams Grade 3 for 11 patients and grade 4 for 3 patients
- **EORTC – QLQ C30 (version 3)** Quality-of-Life questionnaire scores were computed and comparison between pre- and post-treatment.
  - The Mean pre-treatment score was 2.87 (± 0.263) and post-treatment score was 2.14 (± 0.57)
  - Paired t-test provided an overall p-value of < 0.05 which showed that QOL improvement was statistically significant
  - Individually comparing in 3 patients, (p > 0.05) showed no statistically significant improvement – possibly due to skull base metastasis / psychological issues / restriction of daily activities (psychiatric assessment / counselling)

Limitations:

- Retrospective study
- Small sample size
- Rare case scenario
- Other demographic data comparison not done

Conclusion

- Better QOL noted in patients presenting at early stages
- QOL dipped as soon as diagnosis was presented / developed metastasis
- Attender care and comfort level had a significant bearing on patient's quality-of-life

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