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ORIGINAL ARTICLE

# Esthesioneuroblastomas in an Asian population: Similarities and differences☆

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**Summary** *Objective:* Esthesioneuroblastoma is an uncommon tumor that is described widely among the Caucasians. In Singapore, we see predominantly Asian patients with esthesioneuroblastomas. From our experience, we note significant and interesting differences between our data on Asian patients and the published ones on the Caucasian patients.
*Methods:* A retrospective review of all patients who underwent craniofacial resection for esthesioneuroblastomas was conducted from January 1997 to January 2010. Relevant data were collected and statistical analyses were carried out to determine factors that predicted mortality or complications.
*Results:* Out of a total of 48 patients who underwent craniofacial resections, half had esthesioneuroblastomas (50%). There was a peak age distribution at the sixth decade of life and 62% of our patients were male. Both local and regional recurrence rate was 50%.
*Conclusion:* Majority of our Asian patients who underwent craniofacial resections had esthesioneuroblastomas. There is a male predilection, and we do not see a bimodal age distribution that is commonly reported.


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## 1. Introduction

Esthesioneuroblastoma is an uncommon tumor that was first reported in 1924 by Berger and Luc.[1] With a reported incidence of 3% for sinonasal neoplasms, there have been more than 1000 cases reported in the literature, mostly in the last two decades, but mainly as single case reports or case series.[2] Even large institutions have reported only relatively small case series over decades, and, as such, statistical analysis of data was limited. Moreover, most of these publications are on the Caucasian population. In our center, which sees predominantly Asian patients for craniofacial resections, we note significant and interesting differences between our data and the published ones.

## 2. Materials and methods

This was a retrospective review of all craniofacial resections performed in our center, Singapore General Hospital, from January 1997 to January 2010. Our center is the largest tertiary hospital in Singapore, which sees predominantly Asian patients comprising about 74% Chinese, 14% Malays, and 9% Indians of our Singapore population.[3] Our unique ethnic distribution allows us to analyze craniofacial resection within the Asian ethnicity. We specifically focused our data analysis on patients diagnosed with esthesioneuroblastomas.

Preoperatively, most patients will have a computed tomography (CT) scan of the paranasal sinus to assess bony invasion to paranasal sinuses or intracranial spread. A magnetic resonance imaging (MRI) scan is also obtained, unless the tumor encountered is in an early stage (Kadish Stage A), so as to assess for intracranial spread accurately and also to help distinguish between fluid within sinuses and tumor involvement. Having both modalities of imaging (CT and MRI scans) will help in more accurate assessment of staging of tumor.

All patients with esthesioneuroblastomas were then treated with a dual modality of craniofacial resection (described below) and adjuvant radiotherapy, as the tumor is known to be radiosensitive and it improves preservation of the eye.[4,5] Chemotherapy is considered for advanced, high-grade (Hyams histopathological grade) lesion, as it has been shown to improve long-term survival.[6,7]

We categorized postoperative complications into wound-related (infection, dehiscence, and flap necrosis), central nervous system (CNS) (cerebrospinal fluid leakage, meningitis, and pneumocephalus), orbital (nasolacrimal duct obstruction, diplopia, and blindness), and systemic complications. This categorization was also used by the International Collaborative Study on craniofacial resections and will help in comparing our data with the large series of data that has been published thus far.[8]

Institutional Review Board (IRB) approval was sought prior to starting this retrospective audit.

### 2.1. Surgical technique

Most patients were operated on by the same two surgeons (an otolaryngologist and a neurosurgeon) with the same open approach. Endoscopic craniofacial resection by a second surgeon was performed only on three patients. An extended lateral rhinotomy was used for the transfacial approach to access the tumor intranasally with a bicoronal incision with periosteal flap used by the neurosurgeons. After the tumor had been cleared, intraoperative frozen sections were routinely used for all patients to ensure that margins were clear prior to conclusion of the operations. Olfactory bulbs were routinely excised even if they were not clinically involved to minimize local recurrence rates. This was because this tumor was known to recur locally more than 13 years postoperatively despite clear margins of resections intraoperatively.[7] With regard to reconstruction, neurosurgeons routinely used a vascularized pericranial flap to close small defects. A temporalis fascia graft was usually used to augment the closure and a bone calvarium graft used when the defect was larger. Surgicel and Tisseel glue were then used to reinforce the closure. All nasolacrimal ducts were routinely marsupialized to prevent postoperative epiphora. A flavine-soaked pack was then used to pack the nasal cavity for 5 days postoperatively. All patients were then monitored in the neurosurgical intensive care unit (NICU) postoperatively. They were also given prophylactic intravenous amoxicillin–clavulanate for the first 5 days postoperatively.

## 3. Results

Over a period of 13 years, we had a total of 48 patients who underwent craniofacial resections. The majority of our patients had esthesioneuroblastomas (50%) (Table 1). The rest of the histological diagnoses were a heterogeneous mix of tumors. Among the patients with esthesioneuroblastomas, 22 were Chinese, one was Malay, and one was Indian. There was a peak incidence in the sixth decade of life (Fig. 1). We did not see the bimodal peak in age distribution commonly described in the second and sixth decade of life.[9] A slight male predilection (62% males, 38% females) was present. The top three commonest symptoms

**Table 1** Histological diagnosis of patients.

| Tumor | Number |
|---|---|
| Esthesioneuroblastoma | 24 (50%) |
| Squamous cell carcinoma | 3 |
| Adenocarcinoma | 3 |
| Meningioma | 3 |
| Clival chordoma | 2 |
| Sinonasal undifferentiated carcinoma | 2 |
| Adenoid cystic carcinoma | 2 |
| Nasopharyngeal carcinoma | 1 |
| Basosquamous carcinoma | 1 |
| Schwannoma | 1 |
| Inverted papilloma | 1 |
| Neuroblastoma | 1 |
| Recurrent giant cell tumor | 1 |
| Leiomyoma | 1 |
| Fibrous dysplasia maxillary sinus | 1 |
| Renal cell carcinoma metastases (adenocarcinoma) | 1 |
| Total | 48 |