e - ISSN - 2349 - 8005

INTERNATIONAL JOURNAL OF ADVANCES IN CASE REPORTS

Journal homepage: www.mcmed.us/journal/ijacr

PAEDIATRIC PHARYNGEAL NEUROENDOCRINE TUMOUR: A RARE ENTITY

Vaibhav Pandey[1], Prabhat Kumar[2], Shweta Singh[3], Preeti Tiwari[4]*

[1]Senior Resident, Department of Paediatric Surgery, Institute of Medical Sciences, Banaras Hindu University, Varanasi, U.P., India.
[2]Consultant, Department of Paediatrics, Popular Hospital, D.L.W. Road, Near Flyover, Kakarmata, Varanasi, U.P., India.
[3]Assistant Professor, Department of Paediatrics, Heritage Institute of Medical sciences, Ramnagar Mohansarai Bypass Road, Varanasi, U.P., India.
[4]Senior Resident, Trauma Centre, Institute of Medical Sciences, Banaras Hindu University, Varanasi, U.P., India.

Corresponding Author:- **Preeti Tiwari**
E-mail: drtiwaripreeti@gmail.com

Article Info
Received 15/01/2015
Revised 27/02/2015
Accepted 23/03/2015

Key words:
Neuroendocrine tumour, Nasopharyngeal carcinoma, Pharyngeal tumour.

ABSTRACT
Neuroendocrine tumours of the head and neck are rare. They include carcinoid tumors, atypical carcinoid, neuroendocrine carcinomas and large cell neuroendocrine small cell carcinomas [1]. Form large cell is exceptionally disclosed in the upper aerodigestive tract entity, with a frequency of 0.98% [2]. Elloumihas et al reported first case of nasopharyngeal large cell neuroendocrine tumour. To best of our knowledge this is first case of paediatric nasopharyngeal large cell neuroendocrine tumour.

CASE REPORT

A 3-year-old girl presented in emergency with severe respiratory distress (tachypnea, nasal flaring, grunting, retractions, and cyanosis). The child developed above symptoms over last 3 months. Physical examination revealed the following conditions: heart rate 156/min and respiratory rate 45/min with a saturation of 84 on pulse oximetry.

Physical examination revealed whole of nasopharaynx and oropharynx filled by a large tumor. Nasal and oral suction was performed. Rest of examination was normal. Chest radiograph showed diffuse parenchymal infiltrates. Arterial blood gas analysis showed respiratory acidosis and child was in impending respiratory failure. After resuscitation with intravenous fluid and antibiotics, child was taken for emergency tracheostomy. Tracheostomy and a biopsy from the lesion were performed. Histological examination of the biopsy specimen showed a nasopharyngeal proliferation large cells tablecloths, round or cuboid. The cytoplasm was abundant and slightly eosinophilic. The nuclei were round or oval, regular, large nucleoli and rather little monomorphic. Mitoses were quite numerous. The stroma was scarce and highly vascular, endocrine type (Fig 1).

CT scan revealed a voluminous process, expansive, centred on the nasopharynx, invading the skull base and deep spaces of the face (Fig. 2). The child improved symptomatically. The staging with chest radiography, abdominal ultrasound were normal. The patient received three cycles of chemotherapy with cisplatin and cyclophosphamide. Child did not responded to treatment and developed progressive disease and died after 6 months.

DISCUSSION

Neuroendocrine tumors of the head and neck are very rare. These include commonly the carcinoid tumors and rarer variants like atypical carcinoid, neuroendocrine carcinomas and large cell neuroendocrine small cell carcinomas [1]. Hui et al. reported the first case of large cell neuroendocrine carcinoma of the head and neck region arising from the parotid gland [5]. Since then, five cases have been described in the parotid level. Few series have been published since then with neuroendocrine tumour involving the head and neck region [6-7]. Kusafuka et al. reported eight observations neuroendocrine large cell carcinoma of the head and neck, four laryngeal three oropharynphryngeal and hypopharyngeal [4]. To our knowledge this is first case of neuroendocrine tumour involving the nasopharaynx and oropharynx in paediatric age group.

Male preponderance and older age group involvement has been reported in the published series [3-4]. The presentation with respiratory distress is not reported till now and might be explained by the aggressive nature of the tumour. The clinical presentation is aggressive and rapidly progressive, with almost constant nodal involvement [4]. In our case the disease was rapidly progressive with extensive involvement of whole pharynx in a short duration. In contrast neuroendocrine small cell carcinomas, no manifestation of paraneoplastic syndrome has been reported with large cell tumours [3-4]. Our case also did not showed any features of paraneoplastic syndromes. Histological examination shows a trabecular architecture, pseudo-rosettes pattern, or sometimes a palisade arrangement. The cells are large with a low ratio nucleocytoplasmic. The mitotic activity is high (greater than 10 mitoses per ten fields). The tumor is often invasive, with extensive foci of necrosis [6-8]

The rarity of these tumors poses a challenge for therapeutic options and treatment protocol [9]. In the lungs, as in the head and neck, these tumors are supported as neuroendocrine small cell carcinomas, chemotherapy and radiotherapy association rather than surgery [9-11]. Salts of platinum and etoposide are the most used molecules. The large cell neuroendocrine tumors are poor prognosis tumors.

CONCLUSION

The neuroendocrine carcinoma of the head and neck is a rare entity in children. The clinical presentation is not specific. These tumours are aggressive with rapid progression and their therapeutic management is not codified.

Conflict of interest: None.

Figure 1: Patient with tracheostomy

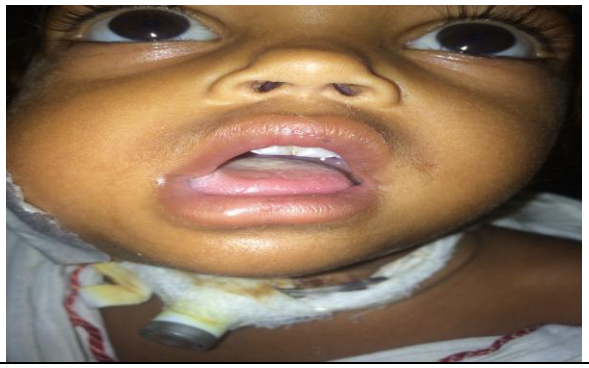


Figure 2: Proliferation of epithelioid cells tablecloths showing a large eosinophilic cytoplasm and vesicular nuclei (hematoxylin-eosin × 400)

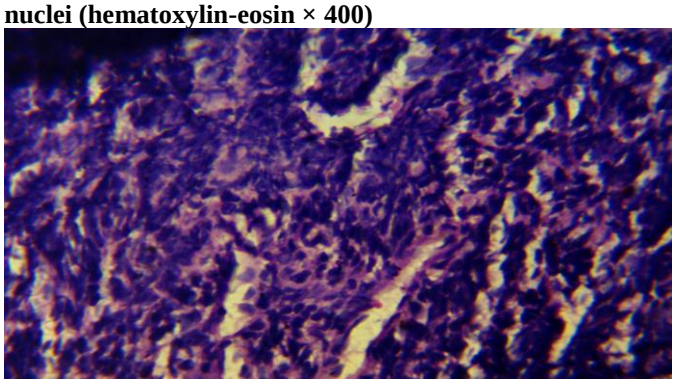
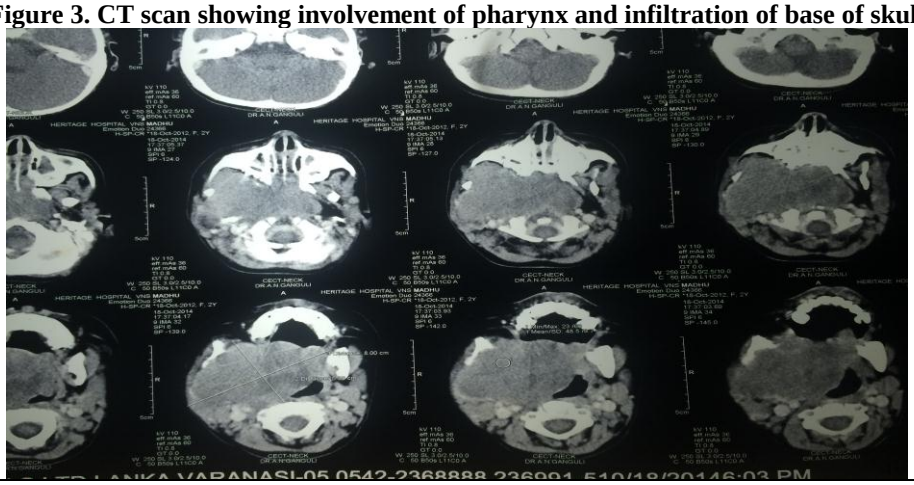


Figure 3. CT scan showing involvement of pharynx and infiltration of base of skull



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