Journal of Cancer Research and Practice 3 (2016) 84–88

Contents lists available at ScienceDirect

Journal of Cancer Research and Practice

journal homepage: http://www.journals.elsevier.com/journal-of-cancer-research-and-practice

Case report

Advanced nasopharyngeal carcinoma in children

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ARTICLE INFO

Article history:
Received 22 July 2015
Accepted 23 September 2015
Available online 8 June 2016

Keywords:
Advanced nasopharyngeal carcinoma
Children
Outcome

ABSTRACT

Nasopharyngeal carcinoma (NPC) is the most familiar cancer originating in the nasopharynx, and not uncommonly found in the head and neck region in adults. Under appropriate and effective treatment, the outcome subsequent to medical intervention can be relatively good in adults with nasopharyngeal carcinoma. However, NPC is very uncommon in children younger than 10 years, and its incidence increases gradually with patient age. In general, NPC may be more aggressive in character, with frequent distant metastasis while initial diagnosis in children. We reported the case of a 12-year-old girl with advanced nasopharyngeal carcinoma, who was refractory to multiple chemotherapy regimens. We herein have provided a summary review of the literature regarding the incidence, characteristics, treatment and outcome of nasopharyngeal carcinoma in children, in the hopes of encouraging further investigations that will create additional treatment options for those who suffer NPR at any age.

1. Introduction

Nasopharyngeal carcinoma (NPC) is a tumor arising from the epithelial cells that line the surface of the nasopharynx. It is a commonly found tumor of the head and neck region in adults, and its incidence varies widely in different regions. It has been observed that high risk areas for NPC include areas such as Southern China, Hong Kong, and Taiwan. Regardless of the region, however, NPC is exceedingly rare in children. An estimated 5% of primary malignant tumors in children originate in the area of the head and neck,[1] while nasopharyngeal carcinoma constitutes about 2% of head and neck malignant tumors in children.[2] Also, NPC in children differs from its adult counterpart in the prevalence of the non-keratinizing undifferentiated subtype, a histologic pattern associated with advanced locoregional spread and frequent distant metastases. The standard therapy for children with NPC has generally followed the guidelines that have been established for adults. Here we reported a 12-year-old girl with advanced and metastatic nasopharyngeal carcinoma, and refractory to multiple treatment regimens, who ultimately expired 1 year after diagnosis.

2. Case report

A 12-year-old girl had normal developmental milestones, and further lacked any indication of abuse, alcohol consumption or contact history with smoking. She complained of right neck swelling for 1 week in May, 2014. Her associated symptoms included poor appetite, trismus, and right neck pain. The patient was treated with antibiotics initially, but the intervention was unsuccessful. We undertook further examination by way of a head and neck CT scan, which revealed a huge nasopharyngeal tumor, approximately 6 cm in size, with locally advanced extension and invasion. Furthermore, bilateral neck lymphadenopathy was also noticed (Fig. 1A, B). Under the guidance of a Karl Storz telescope, one granular mass was noted over the posterior nasopharyngeal wall, which encroached to the right fossa of Rosenmuller fossa and torus tubarius. The final histological examination confirmed the diagnosis of nasopharyngeal carcinoma, undifferentiated (WHO type 3) (Fig. 2). The patient's EBV viral load was checked and determined to be 1.09×10^3 copies/mL. Ultimately, we assessed that the patient was clinical stage T4N2M0, and stage IVA. Definitive CCRT was indicated.

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Peer review under responsibility of The Chinese Oncology Society.

http://dx.doi.org/10.1016/j.jcrpr.2016.05.007

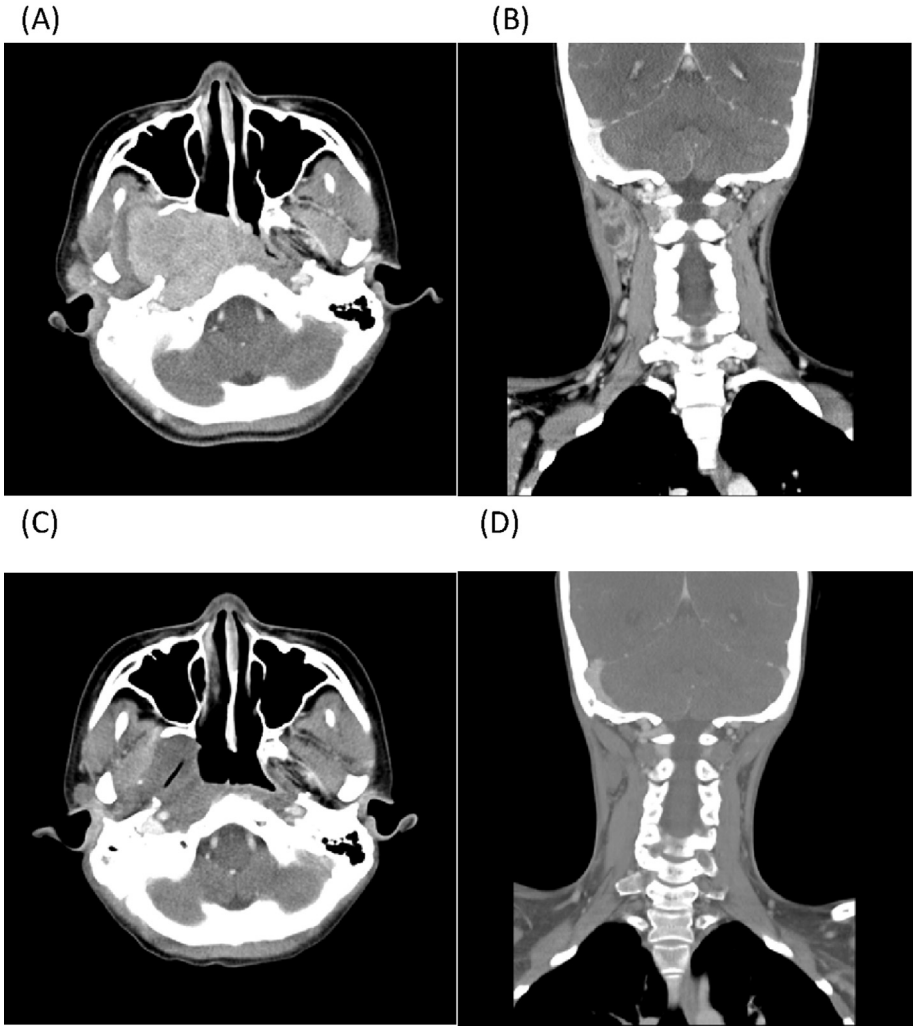

Fig. 1. Image of the Head and neck CT reveals (A) a huge nasopharyngeal tumor extending to the skull base; (B) enlarged neck lymphadenopathy. After treatment (CCRT), follow-up head and neck CT reveals shrinkage in size of the primary nasopharyngeal tumor (C), and bilateral neck lymphadenopathy (D).

After deciding on a further course of treatment, the patient received weekly Cisplatin (30 mg/m^2) and concurrent radiotherapy over the nasopharyngeal tumor and gross lymph nodes (7380 cGy/41 fractions), which was completed in August, 2014. A follow-up head and neck CT revealed shrinkage in size of the nasopharyngeal tumor and lymph nodes (Fig. 1C, D). However, multiple hepatic nodules were incidentally discovered during follow-up. Additionally, she also had unexplained simultaneous abnormal hemogram. Nasopharyngeal carcinoma with liver and bone marrow metastasis was subsequently confirmed after liver biopsy and bone marrow study (Figs. 3 and 4).

To address the patient's metastatic nasopharyngeal carcinoma, she had received multiple treatment regimens, even including target therapy with cetuximab. Although the primary nasopharyngeal tumor was still in stable disease status, her condition of distal metastases progressively deteriorated. The patient was informed as to her poor prognosis concerning ongoing efforts to treat her refractory metastatic nasopharyngeal carcinoma, and she was soon under palliative care. Later, the patient passed in June, 2015.

3. Discussion

Nasopharyngeal carcinoma (NPC) is the predominant tumor type arising in the nasopharynx. The etiology of this challenging disease is multifactorial and involves virological, environmental, and genetic components. The incidence of nasopharyngeal carcinoma demonstrates a marked geographical variation. In general, the incidence of nasopharyngeal carcinoma increases with age, peaking at around 50–59 years of age, with a early small peak found in late childhood in some populations.[3] Childhood nasopharyngeal carcinoma, less than 1% of all childhood malignant tumors, is a very rare disease.

Nasopharyngeal carcinoma commonly presents as nosebleeds, nasal congestion and obstruction, or otitis media. Given the rich lymphatic drainage of the nasopharynx, bilateral cervical lymphadenopathies are often the first sign of disease. Three histological subtypes are recognized by the World Health Organization (WHO): 1) keratinizing squamous cell carcinoma (type 1); 2) non-keratinizing squamous cell carcinoma (type 2); and 3) undifferentiated carcinoma (type 3). The type 3 nasopharyngeal carcinoma is