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Case Report

# Nasopharyngeal carcinoma in childhood

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Nasopharyngeal carcinoma (NPC) is a rare tumor which originates from the nasopharyngeal epithelium. It accounts for <1% of childhood malignancy cases, whereas almost all adult nasopharyngeal cancers are carcinomas.[1,2] In children, nasopharyngeal malignancies are mostly rhabdomyosarcomas and only 20-30% are carcinomas or lymphomas.[2] Approximately one third of the undifferentiated type NPC is diagnosed in adolescents or young adults.[1]

The highest incidence of this disease occurs in Guangdong Province of Southern China, where the risk may be as high as 30-50/100,000 persons/year.[3] In Indonesia, the incidence is approximately 4.7/100,000 persons/year.[4]

A report from Sutomo Hospital, Surabaya in 1997 stated that the patients' ages were between 12-75 years and the male to female ratio was 2.1:1. Lutan *et al* reported that this incidence at the ear, nose, and throat (ENT) department in Pirngadi Hospital, Medan during 1970-1979 was 39.6% (170 out of 429 carcinoma cases).[5] Lubis reported four cases of NPC from the year 1999-2004 at the child health department in Adam Malik Hospital, Medan.[6]

Yet the etiology of NPC remains undetermined, current epidemiologic and experimental data suggest that there are at least three major etiological factors, i.e. viral, genetic, and environmental.[3] Antibody titers towards Epstein-Barr virus (EBV) antigens are elevated in NPC patients regardless of their ethnic and geographic origin. The EBV genome has been demonstrated by nucleic acid hybridization in biopsies from NPC. The majority of malignant nasopharyngeal tumor (80-90%) originates from the epithelium and should be considered as variants of squamous cell carcinoma.[1,3]

According to the World Health Organization (WHO), NPC is classified into three histologic types, i.e. squamous cell carcinoma, non-keratinizing carcinoma, and undifferentiated carcinoma. [1-3]

Generally, the keratinizing squamous cell type NPC is relatively radioresistant, though less aggressive in behavior. The undifferentiated NPC is more radiosensitive and aggressive, frequently with an advanced local regional spread.[7] Most childhood and adolescence cases are of the undifferentiated type, with a few non-keratinizing type, while the squamous cell type are found more in older population. The non-keratinizing and undifferentiated types are associated with elevated EBV titers, however the squamous cell type is not.[1]

Clinically, significant cervical lymphadenopathies exist in most patients at diagnosis. Other presenting signs and symptoms are related to the local spread of the tumor, trismus, epistaxis, chronic otitis media, hearing loss, and cranial nerve palsies. Me-

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tastasis spread may result in bone pain or symptoms related to organ dysfunction at sites of visceral metastasis.[2]

The extent of the tumor at diagnosis is conveniently described by the tumor, node, and metastasis (TNM) classification of the American Joint Committee on Cancer (AJCC) 1988/1992. [7]

Standard care of treatment for NPC over the years has been external beam radiation therapy. Data accumulated over the last decade shows dramatic responses using chemotherapy (called neo-adjuvant therapy) to shrink the tumor prior to radiation, anticipating that the smaller size is more susceptible to curative radiation. Radiation, which starts three weeks after chemotherapy ends, is given daily for approximately seven to eight weeks using a high energy liner accelerator beam.[8]

## Case Report

### Case I

A 12-year-old boy was admitted to the pediatric hemato-oncology division, referred from the ENT department at Adam Malik Hospital, with complaints of swelling on the right and left neck, headache, nasal obstruction, epistaxis, tinnitus, and decreased eyesight (**Figure 1**). This condition began two years ago and during the last seven months he had difficulty swallowing and chewing. One year ago, he was treated in Banda Aceh Hospital as sinusitis and was suggested to be operated, but his parents refused. Two months later, at a private hospital, after endoscopy (**Figure 2**), the diagnosis was NPC with blockage of the Eustachian tube. Aspiration biopsy from the regional side found granulation tissue. The patient was then referred to Adam Malik Hospital, where CT imaging and aspiration biopsy was done. Findings of lymphadenopathy in the right and left neck, nasopharyngeal tumor with extension to posterior cavum nasale; sphenoid sinus, para pharyngeal space, right maxillary sinus, right oropharynx, fossa infratemporalis, middle cranial fossa, and right extra conal lateral retro bulbar with bone destruction of basis cranii; cervical lymphadenopathy; and metastases revealed from CT imaging of the neck, nasopharynx, and aspiration biopsy from nasopharyngeal site, respectively, proved undifferentiated carcinoma. This patient was diagnosed with NPC Stage IV.

We gave chemotherapy and radiotherapy, with Cisplatin 100 mg/m$^2$/iv per day, Bleomycin 15 mg/iv, bolus, 16 mg/m$^2$/iv per day in 5 day continuous infusion and 5-Flourourasil 650mg/m$^2$/iv per day in 5 day continuous infusion every 4 weeks for two courses then followed by radiotherapy 3500 rad for 3.5 weeks.

### Case II

A 12-year-old boy was admitted to Adam Malik Hospital, with the chief complaint of swelling of the neck, experienced 3 months ago (**Figure 4**). He also had difficulty swallowing, nasal obstruction, epistaxis, hearing was decreased since 2 months ago. He was consulted to the pediatric hemato-oncology division by the ENT department after aspiration biopsy (**Figure 5**).

The biopsy showed the spread of epithelial cells with larger nuclei, coarse pleiomorphic chromatic form, and eosinophilic cytoplasm. From this finding, we concluded NPC with metastases.

On CT scan of the head, we found that the infratentorial cerebellum and ventricle IV were nor-

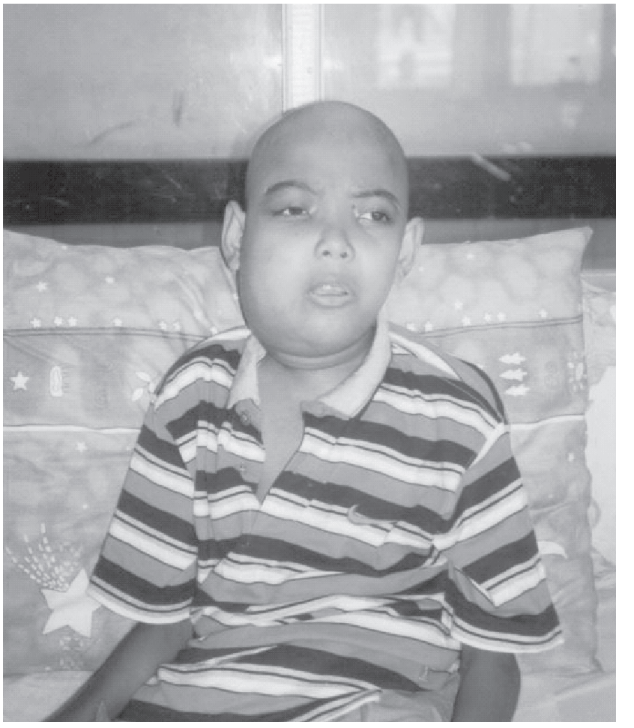

**Figure 1.** Before treatment

mal and the supratentorial did not show hyperdense/hypodense lesion. There was neither mass effect nor midline shift. Ventricular system and cortical sulci were normal. Destruction of basis cranii was not found and right nasopharynx was thickening. We diagnosed this patient with NPC Stage IV.

This patient was treated with chemotherapy and radiotherapy (**Figure 6**). The second case received the same chemotherapy as the first. However, he did not come for radiotherapy.

## Discussion

NPC in children rarely attracts medical attention before it spreads to regional lymph nodes or at a later stage.[2] Enlargement and extension of the tumor in the nasopharynx may result in symptoms of nasal obstruction, changes in hearing usually associated with blockage of the Eustachian tube, direct extension into the ear, and cranial nerve palsies. The diagnosis of NPC was made after "a voe biopsy" of the nasopharynx was done.

Both cases of nasopharyngeal carcinoma have the similar clinical presentations, and diagnosis of NPC was found after 2 years in the first case and 5 months in the second case. The presence of lymph node enlargement is the reason for them to seek medical attention.

Treatment of NPC depends on how far the cancer has spread. For stages 0, I and II, standard treatment is radiation therapy aimed at the nasopharyngeal tumor and nearby lymph nodes of the neck. Although the spread to lymph nodes is not detected at these stages, it is considered preventive (prophylactic) radiation. For Stage III and IV nasopharyngeal cancer, patients receive radiotherapy of the nasopharyngeal mass and neck lymph nodes as well as chemotherapy. Combination therapy helps reduce the risk of recurrence in large cancers that have not spread to lymph nodes.[9]

Both of our cases were diagnosed as NPC stage IV and we planned to treat them with chemotherapy followed by radiotherapy.

The prognosis in children is better than in adults, yet early diagnosis and general condition helps encourage the success of the treatment.

In conclusion, both cases of nasopharyngeal carcinoma were reported in Indonesian boys of 12 years

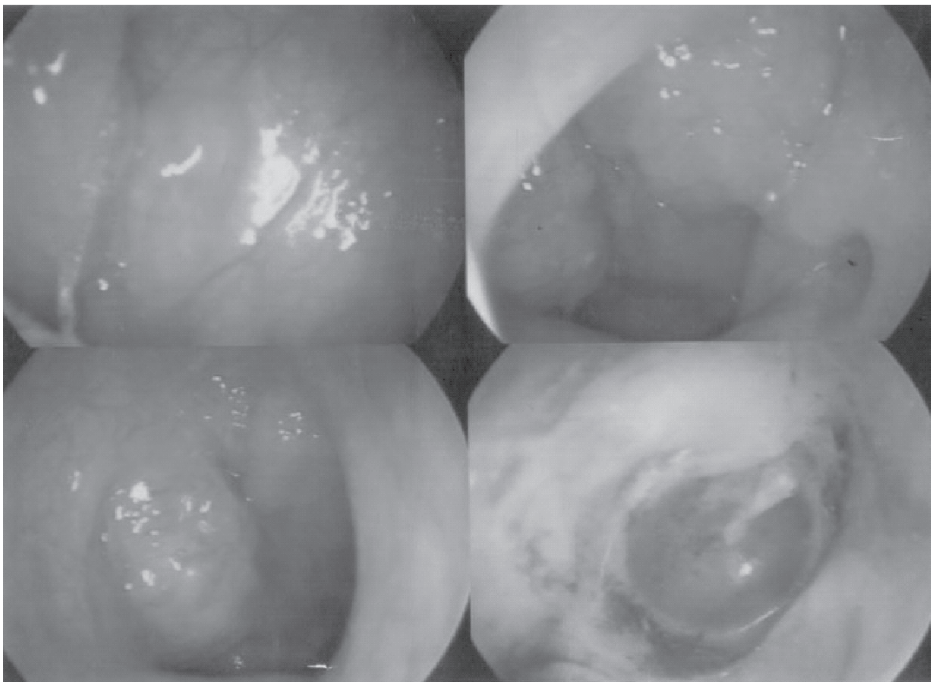

**Figure 2.** Endoscopy from nasopharynx

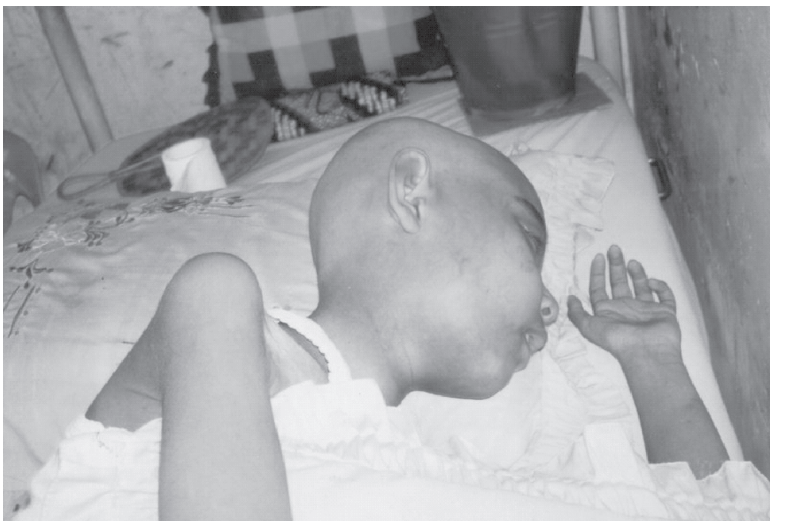

**FIGURE 3.** AFTER TREATMENT

**FIGURE 4.** BEFORE TREATMENT

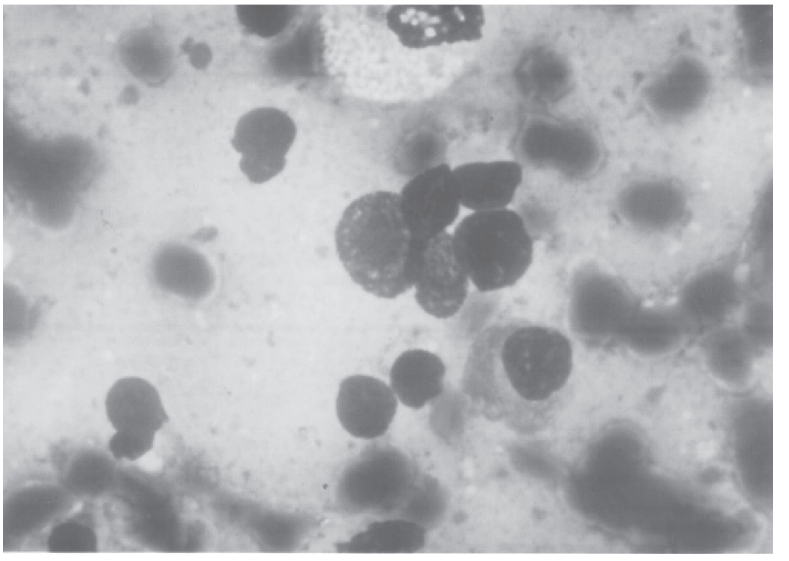

**FIGURE 5.** ASPIRATION BIOPSY FROM NASOPHARYNGEAL CARCINOMA

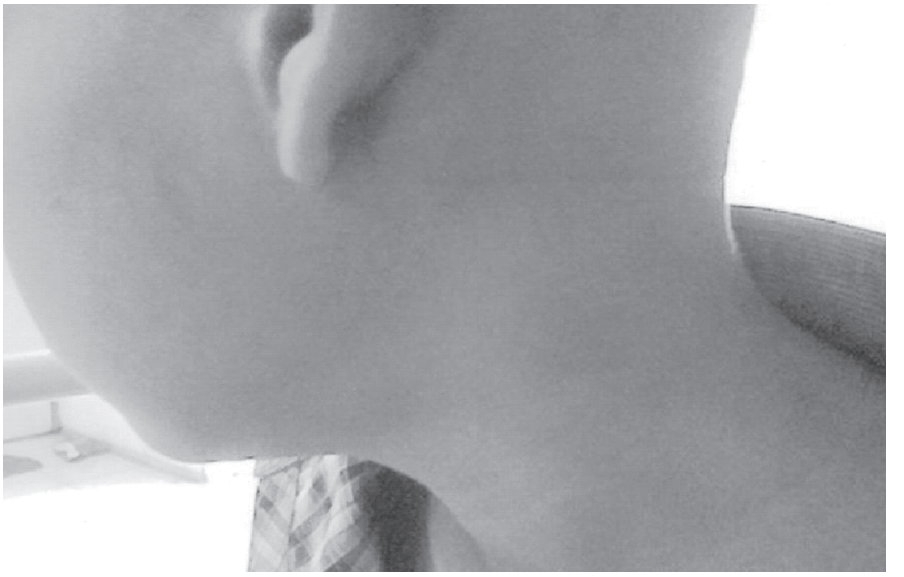

**Figure 6.** After treatment

old. The enlargement and extension of the tumor resulted in symptoms of nasal obstruction, epistaxis, and decrease of hearing. A biopsy and CT of head and neck revealed NPC. The first case was treated with chemotherapy and radiotherapy and the second one was treated with chemotherapy alone. Diminished tumor size and improvement in symptoms occurred in both cases.

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