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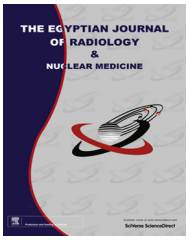

ORIGINAL ARTICLE

Nasopharyngeal carcinoma: Imaging features of unusual cancer in children

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KEYWORDS

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Abstract *Background:* Nasopharyngeal carcinoma (NPC) is a disease of elderly population while the benign adenoidal hypertrophy is the most common cause of a mass in the nasopharynx in pediatrics. NPC is rare in this age group, unfortunately, these tumors tend to be locally advanced by the time they are diagnosed. Our objective is to emphasize that, although rare, NPC does occur in children and can be diagnosed reliably when certain key radiographic features are recognized.
Material and methods: From January 2008 to May 2014, 50 pediatric patients presented to our hospital with pathologically proven NPC. The initial radiological studies (CT of 35 patients and MRI of all 50 patients) are retrospectively assessed regarding the nasopharyngeal masses and cervical lymph nodes.
Results: All 50 patients had a nasopharyngeal mass. Intracranial extension was detected in 38 cases, 15 of them invaded the central skull base, the other 23 extending along the nerves. The perineural spread along V3 was the commonest in 16 cases followed by V2 in the other 7 cases. Almost all the nasopharyngeal masses showed restricted diffusion with average ADC: 0.7×10^{-3} mm^2/sec. Enlarged lymph nodes presented in 47/50 of the cases.
Conclusion: Pediatric NPC is generally not suspected clinically until late into the disease process. Awareness that NPC can occur in children should prompt careful evaluation for distinctive radiographic features. Earlier diagnosis may then direct the patient to timely appropriate therapy when these key radiographic features are present and recognized.

1. Introduction

Nasopharyngeal carcinoma (NPC) is a disease of elderly people while the benign adenoidal hypertrophy is by far the commonest nasopharyngeal mass in pediatrics. Even when the malignancy attacks the nasopharynx in children, the pathological type mostly is lymphoma. NPC is rare in this age group (1). Unfortunately, these tumors tend to be locally advanced by the time they are diagnosed. Because they are rare, they may not be high on the list of differential diagnoses in children who present with a nasopharyngeal mass. The clinical presentation of the nasopharyngeal carcinoma is usually nonspecific. Also the delayed diagnosis reflects on the management and prognosis (2,3).

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Early detection and accurate staging are imperative for optimal treatment planning, helping to improve clinical outcome and survival rate (4).

The radiological features of the nasopharyngeal carcinoma in the adults are frequently addressed in many studies, yet to our knowledge the imaging features of the nasopharyngeal carcinoma in the pediatrics were only in few studies (1,5,6). Our objective is to emphasize that, although rare, NPC does occur in children and can be diagnosed reliably when certain key radiographic features are recognized.

According to WHO classification, NPC is pathologically divided into three categories: keratinizing squamous cell carcinoma (WHO type I), nonkeratinizing squamous cell carcinoma (WHO type II), and undifferentiated carcinoma (WHO type III). Most of the cases are undifferentiated carcinoma (WHO type 3) in the advanced stage (7).

NPC is caused by the interaction of genetic susceptibility, environmental factors (e.g., exposure to chemical carcinogens), and infection with Epstein–Barr virus. High antibody titers to Epstein–Barr virus antigens are useful diagnostic markers, and there are many tests to detect both IgG and IgA titers (8).

Children with NPC generally present with neck mass and/or nasal symptoms such as obstruction, bleeding and discharge or fever of unknown origin (1).

CT has been used for staging NPC for long time, especially for better visualization of skull base invasion and tumor involvement with lytic or sclerotic lesions, but it has now largely been replaced by MRI for primary and nodal staging. However, CT is still used for radiotherapy planning and with PET using 18F-FDG. PET/CT is of value in NPC staging, but its main advantage is detection of distant metastasis (4,9).

A lower ADC value is associated with poorly differentiated or undifferentiated tumor, larger tumor volume and presence of metastatic cervical lymph nodes (10).

2. Patients and methods

We retrospectively reviewed 50 pediatric patients (18 year old or younger) with pathologically proven nasopharyngeal carcinoma that were referred to our institution between January 2008 and May 2014. The age range was 12–17 years (median 15 years). There were 30 male and 20 female patients. Children were examined at initial presentation and before any therapy. Presenting clinical presentation was neck mass (cervical lymph node) in 25 patients, sore throat in 20 patients and trismus in 5 cases. Pathological diagnosis was performed using the specimens obtained by endoscopic biopsy in all cases, and there were 39 squamous cell carcinomas and 11 undifferentiated carcinomas.

2.1. CT imaging technique

CT for 35 cases was performed on (Siemens Somatom Sensation 64, multislice machine). IV contrast material was used in all cases. Acquisitions were started from the ventricular level down to the superior mediastinum with 5 mm-thick slice at 5 mm interval. CT images of the soft tissue and bone window setting were obtained in each study.

2.2. MR imaging technique

Conventional MR imaging was performed on 1.5T MR Unit (Siemens MRI Magnetom Espree) with a protocol that included coronal, sagittal & axial non-contrast T1WI, axial T2WI, coronal T2WI, as well as post contrast enhanced axial, coronal, and sagittal T1WI. DWI was acquired by using b values of 0, 500, and 1000 s/mm^2 applied in the X, Y, and Z directions. Processing of ADC maps was performed automatically on the MR scanners. The ADC was measured by manually placing ROIs 50–100 mm^2 in size within the tumor regions on the ADC map.

Each case was assessed regarding the following:

- The size, signal behavior (compared to the muscles), enhancement pattern of the nasopharyngeal mass.
- The skull base of invasion, the intracranial extension and perineural spread.
- The behavior on the DWI and the ADC values.
- The presence of the cervical lymph nodes enlargement, their side, levels, size of the largest and ADC value.

Staging of NPC was done depending on the 7th edition of American Joint Committee for Cancer (AJCC); International Union against Cancer (UICC) (11,12) which is as follows:

2.3. T-classification

T1: nasopharynx, oropharynx or nasal cavity.
T2: parapharyngeal extension.
T3: bony structures and/or paranasal sinuses.
T4: intracranial extension and/or cranial nerves, hypopharynx, orbit or infratemporal fossa/masticator space.

2.4. N-classification

N0: none.
N1: unilateral cervical and/or unilateral or bilateral retropharyngeal node(s), $\leqslant$6 cm in greatest dimension, above the supraclavicular fossa.
N2: bilateral cervical node(s), $\leqslant$6 cm in greatest dimension, above the supraclavicular fossa.
N3a: $>$6 cm.
N3b: in supraclavicular fossa.

2.5. M-classification

M0: no distant metastasis.
M1: distant metastasis.

3. Results

3.1. Regarding the nasopharyngeal masses

The lesions include the entire nasopharynx in 30/50 cases, the lateral wall in 15/50 cases and posterior superior wall in 5/50