A case of atypical imaging of Nasopharyngeal Carcinoma in an adolescent: A diagnostic challenge

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SUMMARY

Nasopharyngeal mass in paediatric population is usually benign and malignant nasopharyngeal tumours such as carcinoma is rare. We report a case of nasopharyngeal carcinoma (NPC) in an adolescent with atypical imaging findings that mimicked a benign mass. This highlights the importance of a comprehensive review on the distinctive radiographic features of paediatric nasopharyngeal mass. We discuss the imaging features of NPC in the paediatric group, which differ from the elderly patients. By recognising these distinctive imaging features, it may help in diagnosing typical case. This case report serves as a reminder that NPC, despite being uncommon, should be considered in the differential diagnosis of a juvenile nasopharyngeal mass.

CASE REPORT

A healthy 17-year-old women presented with 6-month history of epistaxis and nasal blockage. Nasal endoscopy showed bilateral posterior nasopharyngeal lobulated mass obstructing posterior choanae with streaks of blood on its surface (Figure 1A).

Contrast enhanced computed tomography (CT) demonstrated a well-defined nasopharyngeal mass with a central 0.3 cm calcification focus (Figure 1B). It showed moderate hyperintense signal in T2-weighted (Figure 2A) and isointense signal in T1-weighted (Figure 2B) in relative to surrounding muscles in magnetic resonance imaging (MRI). No evidence of surrounding local infiltration was seen. It demonstrated only mild homogenous enhancement (Figure 3). There were bilateral enlarged level II non-necrotic cervical lymph nodes. No distant organ metastasis.

The patient underwent endoscopic biopsy of this mass under general anaesthesia. Microscopic examination showed fragments of nasopharyngeal mucosa, focally lined by stratified squamous epithelium with no keratinization seen. In terms of immunohistochemistry, these tumour cells showed cytoplasmic immunostaining for PanCK and negative for LCA, T cell and B cell. The findings were consistent with non-keratinising squamous cell carcinoma (Figure 4).

Patient was diagnosed as Stage II nasopharyngeal carcinoma (TNM staging) and subsequently completed combined

chemo-radiotherapy. One year follow up CT scan showed almost complete resolution of the tumour with a single calcification within the right paramedian posterior nasopharyngeal enhancing mucosa.

DISCUSSION

Nasopharyngeal Carcinoma (NPC) is primarily a disease of adults and its incidence among paediatric population varies with geographic location. In the United States of America, NPC accounts for less than 1% of all childhood cancers and children aged between 10-19 years old are the most common age group affected. In a study over 10-years among 1121 NPC patients conducted in Indonesia, a significant number of juvenile cases were observed.1 Patients aged 10-29 years old made up 21% of all the cases. Furthermore, the data also showed a steadily increase with age peaking at the fifth decade without a clear bimodal age distribution. The Malaysian data by Prasad et al showed similar result where the incidence rose after the age of 20-29 years and reached a plateau between 40-49 years.² Our patient aged below 20 years old and was considered in the lower risk group based on the local data.

Reports about the characteristic cross-sectional imaging features of NPC among the elderly have been well reported in comparison to children and adolescents due to its low incidence. NPC is imaged as asymmetric mass in the posterior nasopharynx in the elderly in both CT and MRI. The younger age group also shows similar findings as reported by Hilda et al, in all the nine patients in their study.³ Contrary, the mass in this case was symmetrical with some lobulated contour. Some distinguishing features such as widening of petro-clival fissure, central skull base invasion, para-pharyngeal space infiltration and post-obstructive mastoid opacification shown in the study³ were all not demonstrated in this case. In light of this fact, benign nasopharyngeal mass was made as the initial diagnosis. Yabuuchi et al., described the CT and MRI imaging features of NPC among 13 children and young adult ≤30 years old.⁴ Two imaging features had been identified which were different from older groups. Namely, primary mass was homogenous in intensity and enhancement on MR compared to heterogeneity in elderly. Its signal intensity was slightly higher than the surrounding muscles and lower to the cerebellar grey matter on T1 and T2-weighted images. Secondly, cervical lymph node metastasis without necrotic

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Fig. 1: (a) Nasal endoscopy showing lobulated posterior nasopharyngeal mass (b) Contrast enhanced computed tomography scan of neck in axial view showed mildly homogenous enhancing nasopharyngeal mass with a central calcification focus. (c) MRI Axial T2-weighted showing the mass is hyperintense (d) Coronal T1-weighted showed the mass is isointense to the surrounding muscle and did not invade the pharyngobasilar fascia. (e) MRI Axial post contrast T1- weighted showing homogenous enhancement of nasopharyngeal mass.

change was more commonly demonstrated in younger age group compared to elderly. The assumed reason was histology subtype of non-keratinizing squamous cell carcinoma (WHO type II) or undifferentiated carcinoma (WHO type III) were commoner in younger group in contrast with keratinizing squamous cell carcinoma (WHO type I) in elderly.⁴ The mass in our case showed no significant difference in radiographic findings compared with Yabuuchi et al.⁴

Lymphoma was considered one of the possible differential diagnoses for malignant mass in view of abundant lymphoid tissue at the nasopharynx. Furthermore, the homogenous signal intensity and enhancement as well as symmetrical appearance shown in this case mimicked features of malignant lymphoma. The difference in MR signal may help to differentiate NPC from lymphoma.⁴ Malignant lymphomas was moderately high compared to muscle on T2-weighted and low to intermediate on T1-weighted, as shown



Fig. 2: Sheet of malignant squamoid cells infiltrating the underlying stroma (a) H&E x10 (b) H&E x20 (c) Immunohistochemistry PanCK is positive for tumor cells x10 (d) Immunohistochemistry LCA is negative for tumor cells x20

in this case. On the other hand, carcinoma has only slightly higher signal than muscle on both T1- and T2-weighted. Based on this MR signal characteristic, lymphoma was more likely than carcinoma here. Yet, this MR signal finding should never be taken as a sole criterion, rather it should be taken as part of the overall radiographic evaluation in coming to a set of differential diagnosis. Another differential like rhabdomyosarcoma was not considered here as it is commonly seen in younger children <12 years old.

Majority of nasopharyngeal mass in paediatric population are benign and by far the most common cause is benign adenoid hypertrophy. It was likely in this case with its symmetrical and homogenous appearance without local infiltration. However, the clinical presentation of prolonged epistaxis for 6 months refuted this possibility. Juvenile angiofibroma, another benign mass commonly encountered in adolescent was unlikely in this case as there was no evidence of widening spheno-palatine fossa or enhancement pattern of a vascular mass. Other rare nasopharyngeal vascular masses such as haemangioma and vascular malformation had been taken into consideration based on the presence of intralesional calcification focus, which was presumed to be phlebolith. However, the lack of vascular enhancement and absence of reddish mass during nasal endoscopy made this differential less likely. Furthermore, follow up CT scan showed almost total resolution of the primary mass with persistent calcification focus now vividly shown within the nasopharyngeal mucosa. However, we cannot explain the reason behind its presence. Despite its benign looking radiographic features, none of the above mentioned benign differentials fit our patient.

Many studies have shown that juvenile NPC has better outcome despite being more frequently diagnosed at advanced stage. In a database conducted by Sultan et al among 129 children and adolescents <20 years old and 5885 adults, the 5-year overall survival rate was 83% in childhood and 62% in adult. However, Sultan et al., also found that juvenile NPC are at higher risk in developing second cancer than adult during long term follow up.⁵ Our patient has remained disease free after two years follow up.

CONCLUSION

We report an unusual case of nasopharyngeal carcinoma in adolescent, which mimicked a benign tumour on CT and MRI. Awareness that NPC can occur in younger patients should prompt evaluation for certain distinctive radiographic features for guidance. In case of uncertainty or atypical appearance, the need for biopsy should be done, as the ultimate histology diagnosis will most influence the subsequent management and outcome.

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