Mimicking Malignancy in Adolescents

Case Report

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ABSTRACT

Nasopharyngeal masses in young males can be benign or malignant. As the treatment regimen for these masses differs considerably, it is up most important to differentiate benign from malignant masses before deciding in treatment of patients and also for minimizing morbidity from unnecessary interventions. We hereby present a case with nasopharyngeal carcinoma (NPC) mimicking juvenile nasopharyngeal angiofibroma (JNA). This case initially presented as JNA and to reveal NPC on final histopathology report. As JNA was the initial provisional diagnosis an imaging was done prior to any tissue biopsy. It might had safe the patient's life as a biopsy might be catastrophic. The aim of our case report is to highlight the importance of complete clinical examination and preoperative imaging in differentiating and ideal management of nasopharyngeal masses.

Key Words: Carcinoma; juvenile nasopharyngeal angiofibroma; nasopharynx.

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INTRODUCTION

Nasopharyngeal carcinoma (NPC) is an epithelial malignant tumour of the nasopharynx. NPC is the fourth most common cancer among Malaysians (5.2% of all cancers)[1]. It is most common among Chinese but constitutes only 0.7% cancers worldwide. Children and adolescent NPC is very rare worldwide as compared to adult[2]. Advanced regional disease and high prevalence of metastasis are common in children[3]. It is usually diagnosed late due to trivial presentation of painless neck lumps, blood stained saliva or nasal secretion and unilateral mild ear block. In view of late presentation, its survival outcome is poor.

Juvenile nasopharyngeal angiofibroma (JNA) are rare benign tumours presenting in male adolescents as nasal obstruction and epitaxis. Although these tumours are benign, they are highly vascular and locally aggressive. Biopsy or surgical resection prior to imaging, angiography or embolization may be hazardous. The definitive treatment is surgical resection.

We present a case of adolescent male with a nasopharyngeal mass that presented with physical findings suggesting of JNA. Considering this diagnosis, biopsy was not done due to its potential of torrential bleeding. Radiographic assessment done and results were reviewed before a biopsy was taken. Subsequently histopathological report revealed NPC.

CASE REPORT

A 14-year-old teenager presented with a six months history of recurrent episodes of epitaxis, left side nasal obstruction and neck swelling. It was associated with reduce hearing over the same side. He denied other constitutional symptoms such as sudden loss of weight or appetite. On neck examination, there was an isolated firm swelling over left lateral neck (level III) measuring 3x4 cm which is not tender, fixed and has no skin changes. External nose and oral cavity examinations showed normal findings. A rigid nasoendoscopy had revealed a mass obstructing the chaona. Irregular in margins and bleed on contact (Picture 1). Pure tone audiometry revealed left sided mild to moderate conductive hearing loss with Type B tympanometry. Subsequently a contrast-enhanced computed tomography (CT) scan was obtained which showed an ill-defined heterogenously enhancing soft-tissue mass with the epicentre in the left nasopharynx 6.0 x 7.4 x 7.0 (AP x W x CC) with erosion of the left pterygoid plate, posterior aspect medial and lateral left maxillary sinus wall with minimal extension of the soft tissue mass into left maxillary sinus (Figure 1). There is also extension into the left pterygomaxillary fissure and destruction of bilateral sphenoid sinus floor, posterior ethmoidal air cell floor and left mastoid bone (Figure 2). Multiple enlarged matted lymph nodes in all levels of bilateral cervical region, with the largest measuring 3.0 x 4.0 cm (AP x W) at the left cervical level II region (Figure 3). Some of these

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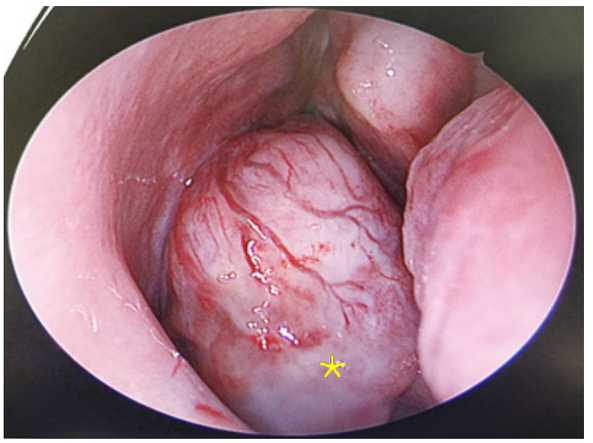
have necrotic centre within. Based on radiological features we decided to have a biopsy taken for histopathology as it favours more towards NPC with no further imaging such as magnetic resonance imaging was done on the patient. Subsequently, histopathological examination revealed non-keratinising squamous cell carcinoma. The patient was referred for chemoradiation. His currently is under frequent surveillance follow ups post chemoradiation. No signs or symptoms residual disease. The patient had provided appropriate informed consent for publication of the case and the images.



Picture 1: Endoscopic view of the left nasal cavity with appearance of vascular mass occupying the nasal floor (yellow asterisk).

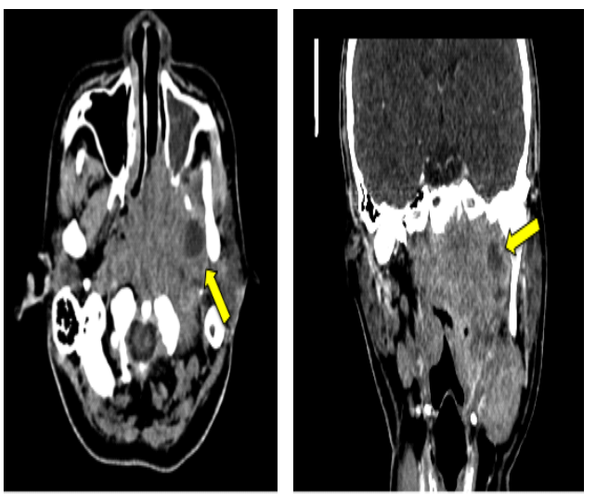


Figure 1 (a) Figure 1 (b)

Fig. 1 (a) axial cut and (b) sagittal cut contrast enhanced CT neck demonstrates heterogeneously enhancing soft tissue mass with the epicentre in the left nasopharynx and extensive local infiltration to the posterior nasal space, left parapharyngeal space prevertebal muscle and infratemporal space. cystic component is seen within the mass (arrow).

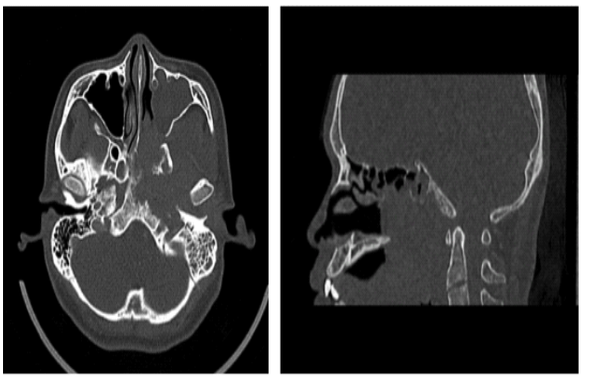


Figure 2 (a) Figure 2 (b)

Fig. 2 (a) and (b) both bone window studies demonstrate destruction and erosion of the adjacent bones by the enhancing mass with involvement of the left sided paranasal bones, clivus and base of skull.

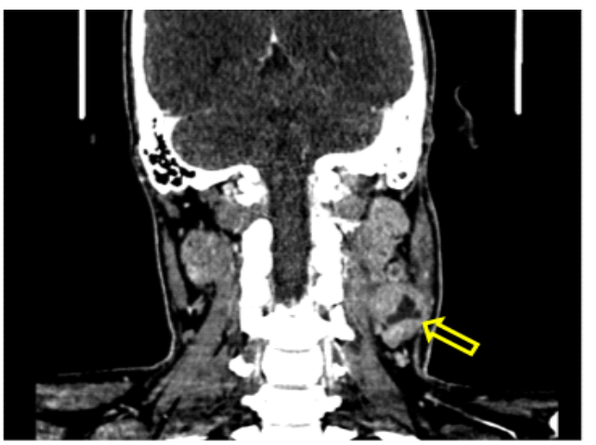


Fig. 3: Sagittal view of the CECT neck shows multiplr matted enlarged bilateral cervical lymph nodes with some of the nodes have central necrosis (open arrow).

DISCUSSION

Nasopharyngeal carcinoma (NPC) is an epithelial neoplasm arising in the nasopharynx. NPC displays a distinct racial and geographic distribution, which is reflective of its multifactorial etiology. This kind of tumour is rare in children but has a predilection for adolescents as per this indextient. Malignant tumours of the nasopharynx are rare and are detected for one to three percent of all malignancies in children [4] and NPC only accounts for 20-50% of nasopharyngeal malignancies. Children between 10 to 19 years old are the most common age group affected by NPC. If this is to be compare to the adult population, NPC accounts for almost all nasopharyngeal malignancies. The diagnosis of NPC in children is often delayed and the patient will only present as a locally advanced disease. In addition to the non-specificity of the symptoms, not considering the entity for the differential

diagnosis might be the other reason for the delayed diagnosis. Childhood NPC is believed to have a better diagnosis than the adult counterparts. The most common symptom of NPC is a painless cervical mass which is bilateral in half of the cases. The possibility of nasopharyngeal malignancies in any child with progressive nasal obstruction, unilateral otitis media and cervical lymphadenopathy when sign and symptoms persists for more than three weeks despite treatment NPC should be considered. On computed tomography, NPC appears as a heterogeneously enhancing mass arising from the nasopharyngeal mucosa or submucosal space with skull base erosion and invasion into the surrounding structures due to its malignant nature. Cervical lymphadenopathy is frequently evident and is the main presentation for most of the patients. As in the case mentioned above, the patient has a heterogeneously enhancing soft tissue density arising from the left nasopharynx with multiple enlarged bilateral lymph nodes.

Juvenile nasopharyngeal angiofibroma (JNA) is a unique and rare benign vascular tumour of nasopharynx exclusively seen in adolescent males. This tumour is rare as it accounts less than one percent of head and neck tumours worldwide. A higher incident can be seen in South East Asia as compared to Europe. The reason for this is still not completely understood but the knowledge about JNA is increasing [5]. The pathognomonic radiological feature of JNA is the anterior bowing of the posterior maxillary wall (Holmen-Miller sign) on computerised tomography. Other features may include a mass originating at the sphenopalatine foramen with widening and erosion of the pterygomaxillary fossa, sphenoid sinus, and infratemporal fossa [6]. Biopsy is absolutely contraindicated because of the risk of massive haemorrhage. Further confirmation of JNA diagnosis is usually provided by angiography, which also doubles as treatment with embolization and gives information on specific blood supply of the tumour indirectly reducing potentially life threatening events preoperatively. Thus, the diagnosis of JNA is based on clinical and radiological examination to ensure precise diagnosis and accurate management.

The case as mentioned above is presented to highlight the salient imaging features that should not be overlooked for evaluation and diagnosis of JNA and NPC and the importance of detailed clinical examination. Because 90% of JNAs present with involvement of the pterygopalatine fossa and very typically homogeneously enhance, isolated heterogeneous masses arising from the nasopharyngeal mucosa without involvement of the sphenopalatine foramen and features like muscle invasion and the presence of cervical lymphadenopathy should prompt consideration for entities, such as NPC.

CONCLUSION

Strong clinical suspicion with the help of preoperative imaging of patients with presumed JNA should be carefully reviewed to ensure accurate diagnosis and proper management. Furthermore, an accurate correlation between clinical and radiological findings is necessary for the best diagnosis. Uncommon features seen on imaging, such as the lack of involvement of the pterygopalatine fossa, the presence of soft-tissue invasion, and cervical lymphadenopathy should alert the physician to malignant etiologies such as NPC and every clinician should be aware that NPC can occur in children and therefore should deliberately look for associated secondary diagnostic signs. The earlier a diagnosis is made the better the prognosis will be.

CONFLICT OF INTEREST

There is no conflict of interest.

REFERENCES

1. Al-Sarraf M, LeBlanc, M., Giri PG, *et al.* Chemoradiotherapy versus radiotherapy in patients with advanced nasopharyngeal cancer: phase III randomized intergroup study 0099. J Clin Oncol 1998; 16: 1310-1317.
2. Liu W, Tang Y, Gao L, Huang X, Luo J, Zhang S, Wang K, Qu Y, Xiao J, Xu G, *et al*. Nasopharyngeal carcinoma in children and adolescents – a single institution experience of 158 patients. Radiant Oncol 2014 Dec 5;9:274.
3. Downing NL, Wolden S, Wong P, Petrik DW, Hara W, Le QT. Comparison of treatment results between adult and juvenile nasopharyngeal carcinoma. Int J Radiant Oncol Biol Phys 2009 Nov 15;75(4):1064-1070.
4. Ayan I, Kaytan E, Ayan N. Childhood nasopharyngeal carcinoma: from biology to treatment. Lancet Oncol. 2003;4(1):13-21.
5. Nicolai P, Castelnuovo P. Benign tumors of the sinonasal tract. In: Flint PW, Haunghey BH, Lund VJ, Niparko JK, Robbins KT, Thomas JR, Lesperance MM, editors. Cummings otolaryngology head and neck surgery. 6th ed. Philadelphia (PA): Elsevier Saunders; 2015. P. 744-746.
6. Blount A, Riley KA, Woodworth BA. Juvenile nasopharyngeal angiofibroma. Otolaryngol Clin 2011 Aug;44(4):989-1004.