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CASE REPORTS

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## Nasopharyngeal Malignancy Presenting as Proptosis in Children

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### ABSTRACT

Nasopharyngeal malignancy is potentially devastating with a high mortality especially if there is delay in diagnosis or late presentation. Nasopharyngeal carcinoma (NPC) presenting with proptosis as a major symptom to an Ophthalmologist is uncommon especially in the paediatric age group. It is against this background we present these patients to share our experience. We retrospectively reviewed cases of nasopharyngeal cancer in children who first presented to the Ophthalmologist at the Olabisi Onabanjo University Teaching Hospital [OOUTH] Sagamu between 2008 and 2010.

Three paediatric patients presented at the eye clinic of OOUTH Sagamu. Two of the patients presented with severe uniocular proptosis and the third with bilateral proptosis. The patients also developed reduced hearing, epistaxis, severe and disturbing headache and neck swelling. There was profound vision loss in three eyes of the three patients. Their eyeballs showed restricted ocular movements. Radiological imaging showed features suggestive of nasopharyngeal cancer involving the nasopharynx, sinuses and orbit. One had Fine Needle Aspiration Cytology [FNAC] from the cervical gland confirmatory of Burkitt's lymphoma. The other two cases were worked up for biopsy from the nasopharynx but discharged against medical advice. The patient with bilateral proptosis died shortly after. The ophthalmologist may be the first to be consulted in cases of a nasopharyngeal cancer with ophthalmic manifestation. Diagnosis is usually difficult to make except with a high index of suspicion. For this reason it is imperative for the ophthalmologist to be familiar with this subject matter.

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**Key words:** Proptosis, nasopharyngeal malignancy, ocular presentation, children

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### INTRODUCTION

The nasopharynx is the nasal portion of the pharynx and is the commonest site of head and neck cancer<sup>1, 2, 3</sup>. Nasopharyngeal carcinoma [NPC] represents 35% of malignancy of Ear Nose and Throat [ENT] origin<sup>4</sup>. Nasopharyngeal cancer usually starts in the second decade and reaches a peak in the fourth decade of life<sup>4, 5, 6</sup>. It can, however, occur at any age group including children<sup>5, 7</sup>. The median age in children is 14 years<sup>8</sup>. A bimodal age incidence has classically been described, with a male preponderance.<sup>4, 6, 9-12</sup> Viral, genetic, occupational and environmental factors are important in the aetio-pathogenesis of NPC<sup>10-14</sup>.

NPC is associated with a very high mortality and a low 5 years survival<sup>11, 15</sup>.

Neck swelling due to enlarged upper deep cervical gland is an early sign and a common presenting symptom<sup>4, 9, 10, 13, 15, 16</sup>. Epistaxis, nasal obstruction with discharge, and rhinorrhea are manifestations of NPC<sup>4, 10, 15, 17</sup>. Blood-stained sputum<sup>8</sup>, tinnitus, hearing loss, headache, and cranial nerve palsies, are frequent associates. Diplopia, strabismus, visual impairment, ptosis and orbital pain are some of the ophthalmic presentations of this disease<sup>12, 19</sup>. Horner syndrome has been reported as a sign of NPC<sup>20</sup>. Optic disc oedema, optic atrophy and eyelid mass are not uncommon findings. Although NPC has been reported as one of the common causes of orbital disease,<sup>11, 21</sup> bilateral orbital extension is comparatively rare. Proptosis due to orbital invasion in NPC is a rarely reported presentation in our environment particularly in children.

The paucity of information regarding the ophthalmic manifestation of nasopharyngeal cancer in our environment necessitated this study to document our experience with NPC and

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add to literature.

### **CASE SERIES**

The records of three patients seen by the Ophthalmologist at the Olabisi Onabanjo University Teaching Hospital Sagamu with the working diagnosis of Nasopharyngeal Cancer between 2008 and 2010 were retrieved and analysed.

#### **Case 1**

A 9-year old boy presented with bilateral progressive proptosis preceded by reduced hearing. There was history of fever, epistaxis, nasal congestion, throbbing headache and rhinorrhea. His visual acuities were 6/5, 6/6 right and left eye respectively at presentation but later deteriorated due to exposure keratitis despite the application of ocular surface wetting agents. He had bilateral severe proptosis with tearing and chemosis of the conjunctiva and limitation of abduction of the eyeball. Investigation for thyroid ophthalmopathy was negative. He was also given therapeutic trial of prednisolone when a differential of pseudotumour was entertained but without improvement. Haematologist, paediatrician and ear-nose-throat (ENT) surgeon were co-opted in the management. Plain skull x-ray revealed a definite soft tissue contrast in the nasopharyngeal air column, thumb printing appearance and sutural diastasis (evidence of raised intracranial pressure). Cranial computerised tomography contrast enhancement showed nasopharyngeal mass with bilateral orbital masses. He was booked for examination under anaesthesia and biopsy of nasopharyngeal mass but was discharged against medical advice. He died shortly after leaving the hospital.

#### **Case 2**

A 15year old boy presented with right progressive proptosis and loss of vision which was preceded by catarrh. He had presented at two different hospitals before presenting in our clinic. There was history of epistaxis, blockage and secretion in the right nasal cavity, but no otological symptoms. His visual acuities were No Perception of Light (NPL) in the right and 6/5 in the left eye. He had right cervical, submandibular lymphadenitis and abdominal masses. He was co-managed by haematologist,

paediatrician and ENT surgeon. Cranial CT showed mass in the nasopharynx, nasal cavity, ethmoid, maxillary antrum and orbit. FNAC of the neck swelling confirmed Burkitt's lymphoma. He was commenced on chemotherapy. He was lost to follow up probably due to death after a third course of chemotherapy.

#### **Case 3**

A 14year old girl presented with left unilateral proptosis and bilateral loss of vision. This was preceded by a headache associated with epistaxis, nasal congestion, and hearing loss. Her visual acuities were hand movement and no light perception in the right and left eye respectively. She had bilateral proptosis which was worse in the left eye, with limitation of ocular motility and optic atrophy in both eyes. Investigation for dysthyroid orbitopathy was negative. CT showed a mass in the nasopharynx involving the sinuses, nasal cavity and the orbits. Biopsy was not done because she left the hospital against medical advice.

**Case 1: Bilateral proptosis in a 9-year old boy with nasopharyngeal caecinoma**



**Case 2: A 15year old boy who presented with right progressive proptosis and loss of vision which was preceded by catarrh**



**Case 3: A 14year old girl presented with left unilateral proptosis and bilateral loss of vision**



**DISCUSSION**

Carcinoma of the nasopharynx accounts for less than 1% of all childhood cancers, Subhabrata et al<sup>25</sup> reported that NPC is the third most common malignancy of the head and neck in children and children aged between 10 and 19 years are the most common affected age-group, with a peak age between 14-15years. About 10–20% of NPC in Nigeria occur in children<sup>7</sup> and 40-50% of nasopharyngeal malignancy in children is due to carcinoma.<sup>18</sup>

Paediatric NPC is generally not suspected clinically until late into the disease process. The three patients in this report presented late with proptosis as the initial manifestation of NPC to the ophthalmologist. Although proptosis is the most common symptom and sign of any orbital disease, it is an unusual and atypical presentation of NPC in our environment. It is usually unilateral but bilateral involvement has been reported<sup>26</sup>. One of the patients had an upper cervical and submandibular mass at presentation which was reported to be a common presentation of NPC in our environment, although this is not always the case<sup>10,12,14</sup>.

NPC frequently spreads superiorly to the base of the skull and intracranially. Cranial nerves are involved when the skull base is invaded by the tumour. Nasopharyngeal malignancies are known to cause limitation of ocular movement from cranial neuropathy which may present with diplopia in unilateral cases. Orbital involvement and ocular cranial neuropathy usually confer a poor prognosis and is worse in bilateral cases<sup>11,13,19,23,24</sup>.

All the three patients had episodes of epistaxis. NPC was documented as the third most common cause of epistaxis<sup>4,27</sup>. The three patients presented to the Ophthalmologists because of proptosis. Hsu et al also reported in his article a few cases of NPC who presented to

the Ophthalmologist first with proptosis<sup>13</sup>. The presence of epistaxis in a patient who presents to an Ophthalmologist with proptosis should draw our attention to the possibility of a NPC.

The presence of severe headache as a presenting symptom in NPC has been documented in the literature<sup>13,14,18</sup>. One of the patients also developed rhinorrhoea, Garandawa et al and Hsu et al found rhinorrhoea as the second most common presentation in NPC<sup>4,13</sup>.

Two patients had severely impaired vision due to optic atrophy while the third was due to exposure keratopathy. Similar finding was reported by, Dunmade<sup>13</sup>, Gupta et al<sup>19</sup> and Hsu et al<sup>26</sup>. It is pertinent that ENT surgeons should collaborate with Ophthalmologist in assessing these patients.

Early diagnosis and treatment directed at the underlying aetiology of proptosis may confer better prognosis but poverty and ignorance may explain why medical attention is usually sought late in our setting. Difficulty with diagnosing NPC may be resolved with detailed history and clinical examination, backed with relevant investigation where there is a high index of suspicion.

Cases 1 and 3 posed diagnostic dilemma until radiological investigation aided the diagnosis. Stambuk et al<sup>7</sup> also reported the radiographic reliability in the diagnosis of NPC in children.

Computerised Tomography [CT] is considered the gold standard radiological investigation and the best investigation for proptosis in a child<sup>28,29,30</sup>. Plain skull x-ray may be helpful in making a diagnosis of NPC in a few cases as seen in the case of the 9 year old boy in this review. CT will reveal the precise location and extent of lesion and in some cases, may aid in selecting the appropriate surgical approach and for follow up. Magnetic Resonance Imaging [MRI] is superior to CT in contrast resolution, especially in cases of tumour extension or invasion<sup>12,31</sup> but its relative scarcity in Nigeria thus limits its use.

Histology is a gold standard especially in the cases with diagnosis uncertainty. It is confirmatory of the type of malignancy and helps in deciding on the best treatment option. FNAC could also settle diagnosis where experienced is available.

Traditionally, radiotherapy is the treatment

of choice for NPC and its nodal metastasis particularly in adult. Palliative radiotherapy alone or in combination with adjuvant chemotherapy was recommended for advanced nodal and visceral metastases<sup>13, 22</sup>. Response is reportedly satisfactory in children with NPC treated with concurrent radiotherapy and adjuvant chemotherapy<sup>8, 14, 33</sup>. Surgery in the treatment of NPC is reserved for obtaining biopsy specimen for histology and for treating lymph nodes that fail to regress after irradiation or reappear after an apparently complete clinical response<sup>12</sup>.

The management of NPC is usually a multidisciplinary approach involving the ENT Surgeon, Radiologist, Pathologist, Neurosurgeon, Oncologist and Radiotherapist. The Ophthalmologist should be co-opted in the team especially when there is ophthalmic involvement.

In conclusion, Nasopharyngeal malignancy is potentially devastating with a high mortality especially if there is delay in diagnosis or late presentation. The ophthalmologist may be the first to be consulted by the patients with nasopharyngeal malignancies because neuro-ophthalmic involvement may be the only symptom and/or sign at first presentation<sup>10, 20</sup>. NPC should be considered as a differential in a child presenting with proptosis, and ophthalmologists should be familiar with this mode of presentation and its management. Difficulty with diagnosing NPC may be resolved through a thorough clinical evaluation and relevant investigative work up.

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