Obstructive sleep apnea with pulmonary hypertension and cor-pulmonale in an 11-year-old Nigerian boy with sino-nasal non-hodgkin lymphoma

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ABSTRACT
Children are predominantly nasal breathers, therefore obstruction of the nasal passage presents early with difficulty in breathing; however, with advance in age they soon adapt to mouth breathing. Chronic upper airway obstruction may result in cardiovascular complications such as pulmonary hypertension, right ventricular heart failure, and also renal disease. Therefore, we report the case of an 11-year-old Nigerian boy who had upper airway obstruction complicated with sleep apnea, pulmonary hypertension and cor-pulmonale resulting from sino-nasal (nasopharyngeal) non-Hodgkin lymphoma which is rare in African children

Key words: Cor-pulmonale, obstructive sleep apnea, pulmonary hypertension, sino-nasal non-Hodgkin lymphoma

INTRODUCTION
Sleep is a physiological process, which is mentally refreshing and is required for growth and development. From the age of 6 months, two stages are recognized - rapid eye movement (REM) and non-REM sleep.[1]

Early childhood is a period of predominantly nasal breathing therefore obstruction of the nasal passage will manifest early with difficulty with breathing, especially during sleep. During sleep especially in REM the tone of the pharyngeal dilators which maintain patency of upper airway is reduced increasing upper airway resistance[8] hence during inspiration, negative pressure is created with sub-atmospheric intraluminal pressure and tendency to collapse of pharyngeal walls; however, during wakeful state maintenance of patency is achieved through voluntary muscle contraction. Sleep related breathing disorders range from snoring to obstructive sleep apnoea/hypoventilation syndrome.[2] Common cause of childhood obstructive sleep apnoea is adeno-tonsilar hypertrophy often due to chronic infective and/or allergic rhino-sinusitis.[9] However, we report a case of an 11-year-old-boy who rather had sino-nasal non-Hodgkin lymphoma as his cause of obstructive sleep apnea, which was complicated with pulmonary hypertension and cor-pulmonale.

CASE REPORT
An 11-year-old boy presented with difficulty with sleeps characterized with excessive night wakes and snoring for a month; this had been progressive with mouth breathing and worsening inability to sleep. There was no fever or cough, but there were complaints of ear discharge and hearing loss. He had several antibiotics without any improvement. On examination, he had an adenoid facie [Figure 1]; the throat and ear examinations were not remarkable; however, nasal endoscopy reveal reddish masses in both nostrils; he had tachycardia of 120/min; blood pressure of 100/70 mmHg (right arm), jugular venous pulse was elevated at 8 cm of water with
displaced apex at the six left intercostal space at the anterior axillary line; he had 1\textsuperscript{st} and 2\textsuperscript{nd} heart sounds with loud P2 and diastolic murmur at the left upper sternal margin and a pansystolic murmur at left mid-sternal margin. Computed tomography (CT) of nasal and paranasal spaces reveal bilateral masses involving both maxillary cavities, nasal cavities and nasopharyngeal space [Figure 2]; chest X-ray showed cardiomegaly [Figure 3]; electrocardiogram showed sinus tachycardia with right ventricular hypertrophy; while echocardiogram showed right ventricular hypertrophy with pulmonary and tricuspid regurgitations [Figures 4]; full blood count and bone marrow biopsy results were normal while tissue biopsy of the mass showed large malignant lymphoid cells [Figure 5]. Based on the tumor site, the diagnosis of sino-nasal non-Hodgkin’s lymphoma was made.

The patient was placed on cyclophosphamid, doxorubicin, vincristine, and prednisolone; patients obstructive symptoms improved after the first course of chemotherapy and the cardiomegaly, pulmonary and tricuspid regurgitation normalized with further courses of chemotherapy.

**DISCUSSION**

Common childhood head and neck tumors vary, with different centers reporting different patterns. While lymphomas were more common in most centers,

\[4,5\] in Kano retinoblastoma was the most common,

\[6\] but non-Hodgkin lymphoma as a cause of obstructive sleep apnea is a rare event, which makes our case unique.

Non-Hodgkin lymphoma in children may present with nonspecific symptoms of fever, weight loss with lymphadenopathy, but these were absent in the index case. In Burkitt lymphoma, the lymph nodes are not commonly involved rather jaw tissues, orbital, and abdominal sites are commonly affected.

Nasal-type non-Hodgkin lymphoma are mostly of the NK/T-cell subtype and is more common among Asians, Central and South Americans which accounts for 5-10% of all non-Hodgkin lymphoma.\[8\] It is uncommon in United States, Europe, the Middle East, and Africa and the median age at presentation is 52 years;\[8\] furthermore, it is rarely reported in children, its occurrence in our case is therefore an uncommon event.
Absence of identifiable lymph node involvement delayed the diagnosis in this case. While endemic Burkitt’s lymphoma is predominantly extra-nodal; the true incidence of isolated extra-nodal non-Burkitt, non-Hodgkin’s lymphoma in Africans is not known. Most staging systems in children combine nodal and extra-nodal tumors as a single group; while in adults, efforts have been made to separate nodal from extra-nodal groups because their prognosis and treatment approach differ. [9]

This case lacked symptoms and signs such as cough, wheezing or shortness of breath; and complaints suggestive of high grade lymphoma such as pain, cranial nerve manifestations, nonhealing ulcer were absent except for facial swelling. [10] Furthermore, the chest X-ray only showed cardiomegaly and there were no significant peri-hilar opacities; though, a chest CT would have given a better picture of the extent of intra-thoracic involvement, but we were unable to do so because of the clinical state of the patient; however, cardiomegaly resolved remarkably by the fourth course of chemotherapy with complete resolution of obstructive symptoms.

Association with cor-pulmonale was not surprising in the index case because enlargement of the pharyngeal and tubal tonsils (Waldeyer’s ring) may have been responsible for the obstructive symptoms and the chronic ear discharge respectively-chronic hypoxia due to airway obstruction may results in pulmonary hypertension due to pulmonary arterial vasoconstriction and vascular remodeling. The right ventricle pumps against high pressure load resulting in right ventricular hypertrophy (cor-pulmonale) and finally right side heart failure. Other complication of chronic upper airway obstruction is systemic hypertension, which was absent in this case.

Our setting is resource limited with large population of oncology cases therefore nonavailability, affordability, and in some cases inadequate diagnostic tools and supportive services make the practice of pediatric oncology difficult; our inability to carry out immune-typing is a limitation and has denied us the opportunity to determine if this was NK/T-cell phenotype, which is mostly associated with nasal type non-Hodgkin lymphoma; be that as it may, patient responded to chemotherapy fashioned for B-cell lymphoma which makes it a possibility.

**CONCLUSION**

Nasal type non-Hodgkin lymphoma is a rare tumor among Africans and also in childhood and our case primarily presented with clinical features of obstructive apnea and cor-pulmonale. Furthermore, he also responded to chemotherapy fashion for B-cell non-Hodgkin lymphoma despite our inability to do immune-phenotyping.

**REFERENCES**


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