

CASE REPORT

REVERSIBLE OBSTRUCTIVE SLEEP APNEA AND RIGHT HEART FAILURE DUE TO MASSIVE TONSILLAR HYPERTROPHY

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Obstructive sleep apnea is a relatively common but under diagnosed clinical entity in children. Adenotonsillar hypertrophy is the most common cause for upper airway obstruction in pediatric patients. If the obstruction to upper airways is not relieved, then the child can develop obstructive sleep apnea and its consequences. Treatment is simply to remove the obstruction thereby restoring patency of upper airways

Key Words: Obstructive sleep apnea, pulmonary hypertension, adenoid hypertrophy, tonsil hypertrophy.

INTRODUCTION

Obstructive sleep apnea (OSA) is a syndrome that mostly affects adult males of over forty years of age. It is thought to occur as a result of soft tissue laxity in the buccal and pharyngeal musculature.¹ Adenotonsillar enlargement is the most common cause of pharyngeal airway obstruction in infants and children. Often this is mild and has no sequelae. However when constant and severe obstruction is present, a condition of OSA develops which leads to disturbed sleep, snoring, behavioral abnormalities, and sometimes growth failure. Long standing OSA is known to cause hypoxia, polycythemia and cor pulmonale.² We report a case of severe tonsillar hypertrophy causing OSA and right heart failure, who improved dramatically after tonsillectomy.

CASE REPORT

Four years old AH had increasing breathlessness and abdominal distention for one year. He was also frequently ill with fever and cough for the same duration. He did not have jaundice, anorexia or contact with tuberculosis. He was an Afghan refugee and had previously been healthy.

On examination he had puffy cheeks with malar flush, central cyanosis, mild clubbing, raised jugular venous pulsations and pedal edema. He also had grossly enlarged hyperemic tonsils and no lymphadenopathy. Precordium was hyperactive with tachycardia, gallop rhythm, loud pulmonary component of 2nd heart sound and a pansystolic murmur of grade 3/6 intensity, best audible at lower left sternal edge. The abdomen was grossly distended with visible veins, moderate hepatosplenomegaly (each measuring 5 cms below costal margin) and ascites. Fine basal crepitations were audible in the chest. The initial diagnosis was congestive cardiac failure and chronic liver disease.

Laboratory investigations revealed a high hematocrit (53%), normal white cell and platelet count, normal serum electrolytes and renal functions. Liver functions were normal except a low serum albumin (total serum proteins = 5.8 gm/dl, albumin = 2.9 gm/dl, globulin = 2.9 gm/dl, A: G ratio = 1). Erythrocyte sedimentation rate was 02 mm after 1st hour and the Mantoux test was negative.

Chronic liver disease was evaluated and the results were normal for hepatitis B, collagen vascular disease, α 1-antitrypsin deficiency and cystic fibrosis (sweat chloride and delta 508). Ultrasound of abdomen showed hepatospleno-megaly and ascites. Liver biopsy did not show significant histological abnormality.

Chest X-ray revealed moderate cardio-megaly. Electrocardiogram showed right axis deviation (QRS = 120°), right atrial enlargement with "P pulmonale" in lead II and severe right ventricular hypertrophy (Figure 1).



Figure-1: Electrocardiogram showing evidence of right ventricular pressure overload

Echocardiography confirmed the right atrial & right ventricular dilatation, moderate right ventricular hypertrophy, severe tricuspid regurgitation (systolic pressure gradient being 60 mm Hg) and pulmonary hypertension.

Cardiac catheterization (Table-1) and pulmonary angiogram were done which confirmed severe pulmonary arterial hypertension and moderate degree of systemic desaturation.

During the hospital stay, AH was observed to snore very loudly and sweat significantly during sleep. The possibility of OSA was considered and an overnight oximetry was performed. It revealed moderate degree of sleep hypoxia with a lowest oxygen saturation of 48% and a mean low saturation being 78% (Figure 2), thus confirming the diagnosis of OSA.

Table – 1: Cardiac Catheterization Data

Catheterization Data	Pressures (mmHg)	Oxygen
		Saturation on room air (%)
<i>Preoperative</i>		
Right atrium	a = 38, v = 26, m = 25	30
Right ventricle	s = 118, ed = 18	22
Pulmonary artery	130 / 62, m = 90	27
	s = 118, ed = 8	56
Left Ventricle	116 / 76, m = 95	45
Aorta		
Postoperative		
Right atrium	a = 6, v = 3, m = 2	72
Right ventricle	s = 48, ed = 6	69
Pulmonary artery	47 / 12, m = 28	71
	s = 112, ed = 8	95
Left ventricle	102 / 50, m = 71	95

Aorta		
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The child underwent tonsillectomy with an uneventful post operative recovery. Follow up three months later revealed a barely audible cardiac murmur and regression of the hepatosplenomegaly. Repeat echocardiogram showed significant reduction in estimated pulmonary arterial pressures, measuring 25 mmHg. Sleep oximetry was repeated and there were no episodes of desaturation (Figure-3). A follow up cardiac catheterization was also done and the results are given in table-1. The pulmonary arterial hypertension had reduced to a significant degree with normalization of systemic hypoxemia.

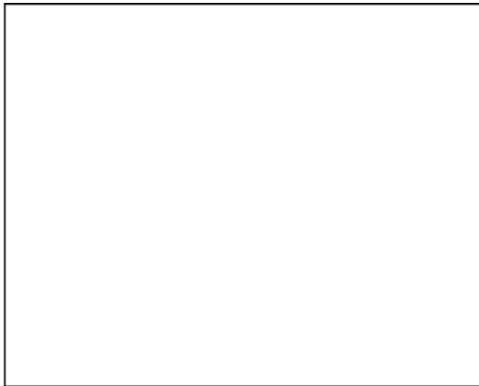


Figure-2: Pre-operative Sleep Oximetry Graph

DISCUSSION

OSA is not an uncommon entity in children but its pathophysiology is poorly understood.³ Obstruction to upper airways in children is caused by a variety of diseases, more common being adenotonsillar hypertrophy. The others include macroglossia, micrognathia, choanal atresia, Down's syndrome, mucopolysaccharidosis, craniofacial abnormalities, cleft palate repair and obesity.⁴⁻⁶

It is well known that chronic hypoxia occurring at high altitudes increases hematocrit and causes pulmonary hypertension and right ventricular hypertrophy. In OSA, the exposure to hypoxia is intermittent rather than continuous. Experimental animals like Wistar rats, when exposed to alternating periods of hypoxia and normoxia twice per minute for eight hours per day for 5 weeks, to mimic the OSA in humans, showed rising hematocrit from day 7 onwards. There was also a significant increase in the right ventricular mass.⁷ Thus intermittent nocturnal hypoxemia due to obstructive sleep apnea causes pulmonary arterial hypertension and polycythemia. The triad of loud snoring, difficulty in breathing during sleep and sleep related breathing pauses is characteristic of OSA in most children. During wakefulness such children may appear either normal or have adenoid facies with mouth breathing. The spectrum of severity has been graded over a scale of 5 by Brouillette and Waters.⁸ The grade 5 severity represents cor-pulmonale, cardiomegaly and congestive cardiac failure. Our patient had grade 5 severity of OSA.



Figure-3: Post-operative Sleep Oximetry Graph

The hepatomegaly, splenomegaly, ascites and pedal edema seen in our patient were due to the long standing right heart failure. In adults, potentially lethal complications like cardiac arrhythmias, ischemic heart disease and stroke can also occur with OSA.⁹

Gorur et al found children with OSA to have right ventricular hypertrophy when compared to controls.¹⁰ Sanner et al¹¹ have shown that right heart failure can occur in OSA independent of lung disease and is thought to be due to reduced right ventricular ejection fraction and systolic dysfunction. In our patient the right ventricular dysfunction was mild (end diastolic pressure (edp) = 18 mmHg) and pulmonary arterial pressures were grossly elevated (Table-1). The right ventricular function returned to normal (edp = 6 mmHg) and drastic reduction in pulmonary arterial pressures were seen (Table -1) after tonsillectomy.

The massive tonsillar enlargement in our patient led to OSA with its attending repetitive hypoxia, secondary pulmonary hypertension and cardiac failure. Tonsillectomy removed the upper airway obstruction and led to complete reversal of these complications in 3 months time.

In summary, this case highlights the need for increased awareness among pediatricians and general practitioners of the possibility of OSA in children. A common pediatric problem like adenotonsillar hypertrophy could possibly lead to serious complications like severe pulmonary hypertension, cardiac failure and neurobehavioral problems.¹² Emergency management would consist of tracheotomy which is certainly not a benign procedure. However definitive treatment would be of the underlying disease making tonsillectomy and adenoidectomy the mainstays of therapy in such cases.

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