

Recurrent respiratory papillomatosis mimicking retropharyngeal abscess in a Nigerian child

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Summary

Recurrent respiratory papillomatosis (RRP) is not an uncommon disease of the aero-digestive tract, usually involving points of narrowing. Involvement of the oropharynx, particularly when florid, may be easily confused with a retropharyngeal abscess.

Study design: Case report of a 1½ year-old child with florid pharyngeal RRP.

Results: The clinical presentation and features on radiography of this patient mimicked retropharyngeal abscess. The patient successfully had a preoperative tracheostomy and excision of the RRP.

Conclusion: There was a diagnostic dilemma in this patient because of the identical features of the two conditions. A high index of suspicion and painstaking, careful clinical and radiological evaluation is the key to diagnosis and prompt definitive management.

Keywords: Recurrent respiratory papillomatosis, Oropharynx, Retropharyngeal abscess.

Résumé

Introduction: Papillomatose respiratoire périodique est une maladie aerotube digestive tres courante qui habituellement implique les points de rétrécissements, impliquant l'oropharynx quand le florid en particulier pourrait être facilement confondu avec l'abcès rétropharyngéal.

Plan d'étude: Il s'agit d'un cas d'un enfant âge de 1½ ans atteint du florid pharyngéal RRP.

Resultats: La présentation clinique et des traits sur la radiographie de cet enfant ont démontré l'abcès rétropharyngéal. Le patient a eu la trachéotomie préopératoire et l'excision de la RRP couronnée de succès.

Conclusion: On a remarqué un dilemme diagnostique chez ce patient à cause des traits identiques des deux conditions. L'indice de soupçon plus élevé et une évaluation clinique et radiologique assidue et soigneuse et la clé du diagnostic et la prise en charge rapide et définitive.

Introduction

Airway obstruction in a child is a serious condition. The onus is on the clinician to deduce the site of the obstruction and the possible etiology. Recurrent respiratory papillomatosis (RRP) is a condition usually affecting infants and young children.¹ The initial term for this entity, "laryngeal" papillomatosis, was coined because of the erroneous belief that it was primarily a laryngeal condition.²

It is however well recognised that the whole of the aerodigestive tract may be involved diffusely or a single site may be affected. Laryngeal involvement is however the most common form of clinical presentation.^{3,4} The localization of this neoplasm is not random, showing a preference for regions

of juxtaposition of ciliated and squamous epithelia.⁵ Such sites include the limen vestibuli, nasopharyngeal surface of the soft palate and the carina.

We hereby present a clinical presentation of RRP mimicking an acute retropharyngeal abscess in a Nigerian child.

Case report

The patient was an eighteen-month-old female child who presented with a month history of paroxysms of cough, noisy and difficulty with breathing and cough. There was

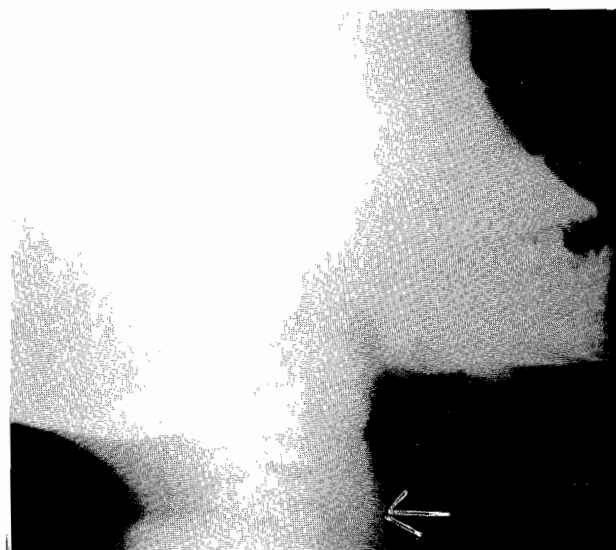


Fig. 1 Lateral view of soft tissue radiograph of the patient showing increased pre-vertebral soft tissue shadow with irregular margin (white arrows) and loss of cervical lordosis.

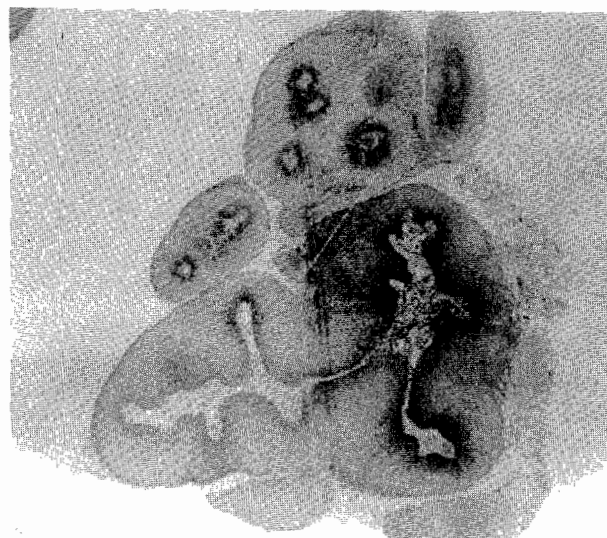


Fig. 2 Photomicrograph showing papillary tissue fragment lined by benign squamous epithelial cells enclosing fibrous stroma (Hematoxylin & Eosin x 35)

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associated bilateral mucopurulent rhinorrhea of two weeks duration. Symptoms were of insidious onset and progressively worsened till presentation.

Examination revealed a sweating, dyspneic and febrile child (Temperature 37.8°C). There was scanty bilateral mucopurulent nasal discharge. Proper visualization of the oropharynx was difficult due to pooling of saliva.

Soft-tissue X-ray of the neck (Figure 1) revealed diffuse soft tissue fullness in the prevertebral space. Haematocrit, electrolyte and urea values were within normal limits and retroviral screening was non-reactive. Based on this, a pre-operative diagnosis of retropharyngeal abscess was made. The child was promptly taken to theatre for emergency tracheostomy, incision and drainage of the abscess.

However after tracheostomy and under general anaesthesia, a proper direct pharyngoscopy revealed multiple, sessile and exuberant papillomatous growths 'carpeting' the tonsils, posterior oropharyngeal wall, hypopharynx and filling the laryngeal inlet and left vocal cord. The nasal cavities, nasopharynx, subglottis and esophageal inlet were not involved. Surgical clearance was done using Remington-Hobbs tonsil stripping and Paterson's Laryngeal biopsy forceps. The specimen was sent for histology.

Histological examination (Figure 2) confirmed the clinical diagnosis of squamous papilloma. The patient was decannulated on the 4th day and discharged to the outpatient clinic on the 9th day post-surgery. The child has remained disease free one-year post surgery.

Discussion

This case report illustrates the bizarre way RRP may mimic other causes of acute upper airway obstructive disease condition in a child. Its etiology is now known to be due to the human Papilloma virus (HPV) with confirmatory evidence from electron-microscopic evaluation and immunohistochemistry studies.^{6,7} HPV serotypes 6 and 11 are the specific ones implicated as confirmed by DNA hybridization (Southern blot) technique and polymerase chain reaction.^{1,8,9} RRP had been confused with, and treated as bronchial asthma, as was the case in the peripheral hospitals our patient had earlier presented at, prior to specialist intervention.¹⁰

HPV infection is thought to be acquired via vertical (mother to child) transmission during childbirth in a mother with genital warts.¹¹ It is expected that other siblings should develop the same disease condition if delivered via the same route; however, this is most times not the case.⁸

Although the entire aerodigestive tract is at risk, predominant pharyngeal involvement is generally uncommon. To cause further diagnostic confusion in the presented case, the X-ray of the neck soft tissue revealed features suggestive of retropharyngeal abscess, which however on a closer look reveals, a serrated anterior border of the increased prevertebral soft tissue shadow. Acute retropharyngeal abscess is a common disease of suckling infants, commonly following suppurative retropharyngeal lymph nodes.¹² The child classically presents acutely ill, pyretic and with difficulty in breathing and feeding. There is neck stiffness, and throat examination reveals fullness in the posterior pharyngeal wall, usually with pooling of saliva. Sudden death from

laryngeal edema and asphyxia, or spontaneous rupture and aspiration are grave sequelae. The soft tissue neck X-ray usually assists in diagnosis of this condition. It is characterized by revealing pre-vertebral soft-tissue fullness pushing the air column anteriorly and compromising it. The diagnosis of RRP in this patient however was made intraoperatively.

A high index of suspicion of this challenging disease condition is the key to diagnosis, and prompt careful management is rewarded with good patient response. This is because its propensity for recurrence is unparalleled.^{2,13} In fact, the site of recurrence has been shown to be hitherto grossly normal surrounding mucosa adjacent to the papillomatous site.¹³ Recurrence rate is variable, from as short as 2 weeks to as long as 20 years after surgical removal.⁹ The post operative clinical course in the case presented is worth noting in that inspite of the florid disease the child has remained without recurrence for one year now. In addition preoperative tracheostomy is known to further encourage recurrence though it was for a short duration in our patient.^{3,5,14} Recurrences are characteristically multiple and sessile.

Several attempts at preventing recurrence had been tried. These are adjuvant to primary microdebrider¹⁴ or CO₂ laser vaporization,^{2,4,14} surgical options currently not available in our center. Interferon, isotretinoin and methotrexate are a few of the recently investigated agents, yielding substantial clinical response.^{9,14} In a recent study, photodynamic therapy has been shown to have beneficial effect.¹⁵ The importance of close follow-up cannot be overemphasized especially in monitoring recurrence of this disease.

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