Case Report

JUVENILE LARYNGEAL PAPILLOMATOSIS – A CASE REPORT

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ABSTRACT

We report a rare case of juvenile laryngeal papillomatosis in a 7 year old male child. The child was born through normal vaginal delivery and had recurrent episodes of dyspnoea, stridor and noisy breathing during sleep since an year. Laryngoscopy revealed multiple papillary lesions and histopathological confirmation of squamous papilloma was given. Virological studies demonstrated HPV. Mothers HPV status was not available.

KEYWORDS: JLP, HPV, child

Introduction:

Juvenile laryngeal papillomatosis (JLP) is a rare, benign and recurrent condition secondary to HPV infection with a varying clinical course. [1,2] The Viral aetiology for papilloma was proposed as early as 1923 by Ullman and HPV was identified by electron microscopy in 1949. A maternal history of genital warts, secondary to both HPV low risk types 6 & 11 have been identified as the strongest risk factor.[3]

CASE REPORT :

Our case is a 7 year old male child, who presented with progressive difficulty in breathing, and noisy respiration during sleep. Maternal history revealed normal vaginal delivery and history of genital warts could not be elicited. Clinical examination of the child revealed multiple florid laryngeal papillomatosis, more on the right side of larynx compared to left side. Due to progressive severity of symptoms, surgery was advised and papillomas were excised and specimen was sent for histopathology.

Histopathology showed multiple small papillomas (Fig 1). The papillomas were lined by stratified squamous epithelium with a fibrovascular core (Fig 2). No dysplasia of lining epithelium was noted. A diagnosis of Juvenile laryngeal papillomatosis was given.

An HPV study was positive for HPV 11 and regular follow-up was advised due the recurrent nature of JLP.

DISCUSSION:

JLP is a rare disease caused by infection with non-oncogenic HPV types 6 & 11 in young children. The child acquires the infection at birth or perinatally, from the infected maternal genital tract. Cases onset until the age of 14 is classified as JLP, and onset at older ages by HPV 6 & 11 is probably acquired by sexual contact and is classified as adult onset respiratory paillomatosis. The overall prevalence of JLP is 3.6 to 4.3 per 100,000 children and mostly seen in children younger than 4 years old. [4,5]

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Diagnosis is usually delayed as young patients are not cooperative in laryngoscopy. Severity of the disease can be assessed based on modified version of staging assessment, which was developed by Derkey. Although, JLP is benign, it is known to grow rapidly, need repeated surgery to keep airway patent; and in some cases, 100 surgeries may be needed by the age of 10 years. Surgical intervention frequency is reduced with the use of non surgical treatments like interferon, cidovir, celecoxib, photodynamic therapy, etc. Tumour excision is usually with a pedestal laryngoscope and other auxiliary treatments such as CO2 laser, cryotherapy, electric cauterization, etc are also done. Photodynamic therapy is known to particularly reduce recurrence as it is known to remove the virus without any systemic effects. [4-7]

Hajek first suggested the transmission from mother to child at birth. Later on, many investigators showed several cases of mothers of children with JLP frequently giving history of genital wart. The maternal HPV infection seems to be contracted intrapartum by contact of foetus to the infected maternal genital tract. Caesarean section is not known to fully protect against JLP suggesting that infection may also be transferred during the perinatal period. [5]

HPV 6 & 11 types are known to infect the epithelium, enter base cells via micro wounds and later infect squamous cells of larynx and other areas. Once inside the nucleus, they can proliferate as HPV virons and can persist in free state outside the chromosome. These HPV virons can be found both in papilloma tissue as well as normal tissue. Studies have shown the lesion to more HPV Ag positivity, if the age of infection inset is much lesser. [3]

JLP is often misdiagnosed as asthma and patient can present with severe airway obstruction and many cases of morbidity has been reported secondary to delayed diagnosis. Timely diagnosis is very important and general practitioners should proceed with caution in any child with shortness of breath, particularly when there is associated hoarseness of voice. [8]

Immunization of pregnant women or expecting mothers with quadrivalent HPV vaccine has been proven to be effective in preventing infectious diseases in both women and their children. Risk of malignant transformation particularly with HPV 11 variant has been reported in 2-4% of the cases. Spontaneous remission is known to occur rarely during puberty[3,7,9]

CONCLUSION :

Even though JLP is rare, the disease can present a enormous challenge for the young patients, their family and their treating physicians.

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FIGURES :

Fig 1: Multiple papillomas on slide section

Fig 2: Squamous papilloma with fibrovascular core – H&E 10x

Fig 1:

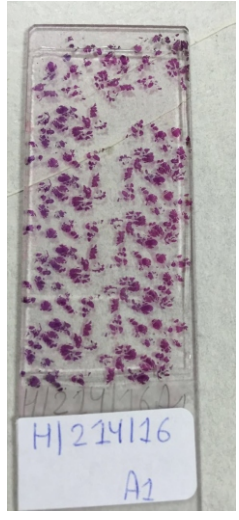
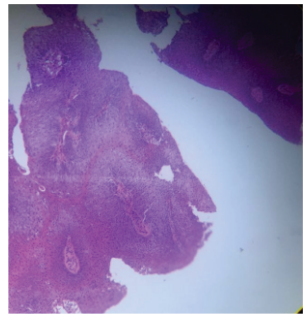


Fig 2:



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