A Rare Tumor in the Neck of a Child: Plexiform Neurofibroma

ABSTRACT

Plexiform neurofibroma represents an uncommon variant of neurofibromatosis type 1, constituting only 5%–30% of all cases. Plexiform neurofibroma is usually diagnosed during childhood and arises from multiple nerves, manifesting as bulging and deforming masses that can also involve connective tissue and skin folds. We report a case of a two-year-old girl who presented with worsening stridor since birth and later exhibited progressively increasing left neck swelling at the age of 10 months old. Ultrasound and magnetic resonance imaging (MRI) showed a lobulated solid mass in the left deep neck space extending to the midline and having a mass effect on the airway with involvement of the supraglottic region. Tracheostomy was done, and a biopsy of the supraglottic lesion revealed a plexiform neurofibroma. The patient was conservatively managed after a discussion with her parents concerning the associated potential of operative morbidity. The patient's parents had learned about tracheostomy care, and the patient was scheduled for yearly MRI surveillance. MRI was performed again three months after the initial diagnosis and showed stable lesion. Plexiform neurofibroma is a slow-growing tumor. A treatment decision must consider the benefits of surgery and the morbidity of the progressing disease. Hence, airway management is crucial prior to the final decision of such cases