Case Report

Hidden Diagnosis in the Subglottic Larynx: Schwannoma Mimicking as Bronchial Asthma

Fayaz A Sofi, Mohmad Hussain Mir, Farhana S Bagdadi, Khalid Mehmood

Department of Internal Medicine, Sheri Kashmir Institute of Medical Sciences, Soura, Srinagar, Jammu and Kashmir, India

Abstract

Schwannomas are rare neurogenic tumors derived from the schwann cells. Their laryngeal location is uncommon and the diagnosis is difficult. We report a case of a 17-year-old female who presented with symptoms of intermittent breathlessness mimicking acute attacks of bronchial asthma and resulted in delayed diagnosis, which lead to upper airway obstruction and an emergency tracheostomy. Computed tomography (CT) of neck revealed a soft-tissue mass within the subglottic region. Surgical excision of the growth was done by laryngofissure technique. Histopathological examination of the tumor revealed it to be a schawanoma. The subglottic occurrence of schwannoma is very rare. Case history and detailed clinical examination are important and will guide for relevant investigations and help to reach at a definitive diagnosis.

Keywords: Neurogenic tumors, Schwannoma, Subglottic

Address for correspondence: Dr. Farhana Siraj Bagdadi, Hamambal, Khanyar, Srinagar – 190 002, Jammu and Kashmir, India.
E-mail: bagdadifarhana6@gmail.com

Introduction

Schwannoma is a neurogenic tumor arising from nerve sheaths of peripheral and cranial nerves, where it originates from Schwann cells which are embedded in the neurilemal sheath as a multinucleated syncytial network.[1] Schwannoma involving the larynx is rare[2] and diagnosis is difficult. Resulting symptoms include sore throat, odynophagia, dysphagia, dyspnea, stridor, etc., and are related to the mass effect of a slow-growing tumor in the larynx. Final diagnosis is based on the histological demonstration of characteristic spindle cells and positive immunohistochemistry for S100 protein. Surgical excision is the treatment of choice.

We present a case of a young female with symptoms mimicking acute bronchial asthma. The symptoms used to worsen on crying or laughing. Symptoms were even attributed to hysteria in secondary care hospital. This resulted in delayed diagnosis which lead to upper airway obstruction and an emergency tracheostomy. It was on computed tomography (CT) of neck that a soft-tissue mass was detected in the subglottic region. Mass was excised and patient is doing well at present.

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Case Report

A 17-year-old female was referred to Department of Internal Medicine of a Tertiary Care Hospital with history of increased breathlessness of 2 months duration. She was following a district hospital where she was labeled as a case of bronchial asthma. Despite adequate treatment she was not doing well. She had no history of atopy or any family history of bronchial asthma or any other allergic disorder. She was a nonsmoker and was not rearing any pets. Her physical examination revealed no stigmata of chronic allergic disorder like nasal polyps, allergic shiners, etc. Her chest examination revealed no wheeze. She gave a typical history of worsening of breathlessness on crying or laughing for last 2 months. Her X-ray chest was normal and her arterial blood gas analysis during the acute events showed high p_{co2} of the order of 75, 82, 67 mmHg. Her po_2 was in the range of 82–87 mmHg and saturation was 90–97%. Her pulmonary function tests revealed Forced expiratory

volume in 1st second (FEV_1) 62.7% of predicted, Forced vital capacity (FVC) 89.0% of predicted with FEV_1/FVC ratio of 70.4%, which was suggestive of obstructive pattern. Her postbronchodilatation FEV_1 increased to 65.9% with an increase of 3.2% only, which suggested fixed obstruction. Her bronchial asthma was ruled out because asthma is characterized by reversible obstruction. Meanwhile, during hospitalization, she developed acute upper airway obstruction with stridor, and emergency tracheostomy was done.

Noncontrast computed tomography (NCCT) neck was done which showed a soft-tissue mass within the subglottic region on the right side measuring 1.6 × 1.2 × 2.5 cm with evidence of surrounding pneumomediastinum. Her virtual bronchoscopy showed a subglottic mass filling almost whole of trachea leaving a small space on left side [Figure 1]. Surgical excision of the growth by laryngofissure technique was performed under general anesthesia. The mass was found to arise from the right side of the subglottis extending to under surface of vocal cords and into the tracheal lumen. The tumor was pale yellow, globular with smooth surface. Tumor was removed in toto and measured 2 × 2 cm. Histopathological examination of the tumor revealed it to be a schwannoma [Figure 2]. The immunohistochemistry for S100 protein was positive. The postoperative period was uneventful and the patient was decannulated on 12th postoperative day. Presently, patient is on our follow-up and is doing well.

Discussion

Schwannoma is a tumor derived from the schwann cells. Approximately 25–35% of all reported schwannomas occur in the head and neck region. However, schwannomas involving the larynx are rare.[2] Almost all laryngeal schwannomas present insidiously and arise from the aryepigllotic fold or arytenoids.[3] Schwannomas may arise from schwann cells of peripheral, cranial, or sympathetic nerves. They involve males and females equally and can occur at any age.[4] Most common nerve of origin of laryngeal schwannoma is the internal branch of superior laryngeal nerve.[5] Symptoms are related to the mass effect of a slow-growing tumor in the larynx and include sore throat, odynophagia, dysphagia, dyspnea, stridor, etc. Our patient had developed a ball valve mechanism of the growth and that is why used to get increased breathlessness on laughing and crying and on two occasions was labeled as hysterical in the emergency department of a secondary care hospital. Bronchial asthma was ruled out in view of fixed obstruction as bronchial asthma is characteristically a reversible airway disease. Imaging can help in diagnosis. On noncontrast computer tomography scan of neck, small schwannomas are seen as homogenous enhancing mass.[2] On Magnetic Resonance Imaging (MRI) studies, T_1-weighted imaging of a schwannoma shows variable intensity and it is well enhanced after gadalonium injection.[6] However, CT and MRI appearances are not diagnostic.

The diagnosis can be achieved by endolaryngeal fine needle aspiration or incision biopsy, although it may be difficult to distinguish between schwannoma and neurofibroma.[7] The ultimate diagnosis is based on the histologic demonstration of characteristic spindle cells with typical nuclear palisading and positive immunohistochemistry for S100 protein , but negative for α-smooth muscle actin, CD34 and vimentin.[8] Surgical removal is the treatment of choice. Various surgical approaches for the removal of the laryngeal schwannoma have been mentioned in the literature, varying from endoscopic removal, laryngofissure, and anterior and posterior pharyngotomies to lasers.[7] The prognosis is good if surgical excision is complete. Malignant transformation is rare. Radiation should not be employed as schawanomas are highly radio resistant tumors.

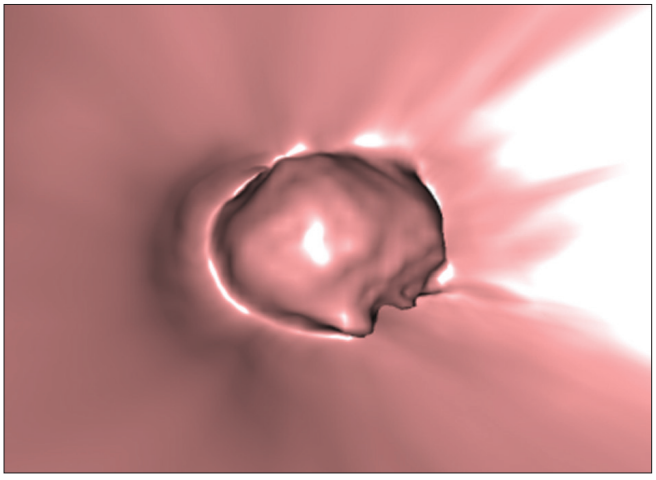

Figure 1: Photograph of virtual bronchoscopy showing growth in the subglottic region filling almost whole of tracheal lumen

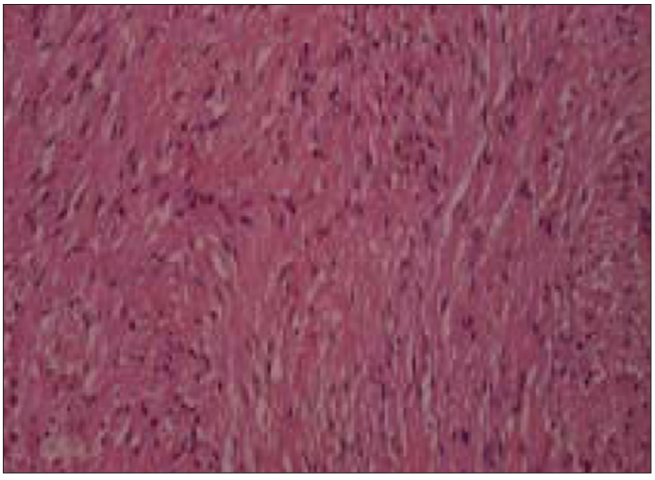

Figure 2: Histopathological examination of specimen showing spindle cells characteristic of schwannoma

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