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Case Report

PYRIFORM FOSSA SCHWANNOMA: A RARE CASE REPORT

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ABSTRACT

Schwannomas are benign slow-growing tumors that arise from the Schwann cells of any peripheral, cranial, or autonomic nerve sheaths. Between 25% and 45% of extracranial schwannomas arise in the head and neck region. The location of Schwannoma or neurofibroma within the larynx and hypopharynx is very uncommon. They represent 0.1% to 1.5% of all benign tumors in larynx and hypopharynx. Here we present a rare case of schwannoma of pyriform fossa in a young female patient. Presenting complaints were hoarseness of voice and difficulty in swallowing. Laryngoscopic examination revealed a smooth submucosal swelling in the left pyriform sinus. Bilateral vocal cord mobility was normal. No lymphadenopathy was detected. Contrast enhanced computerised tomography of the neck showed a well marginated hypodense lesion showing progressive enhancement with non enhancing central mass in supraglottic and pyriform sinus on left side extending superiorly from epiglottis and reaching inferiorly till glottis and right side small laryngocele. Treatment of choice is complete enucleation/excision along with preservation of function of affected nerve/site. Various approaches such as endoscopic, open transoral, and external or lateral pharyngotomy approaches can be used depending on size and location of lesion. The definitive diagnosis relies on clinical suspicion and histopathological confirmation.

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INTRODUCTION

Neurogenic tumours of the larynx are quite rare and constitute only .1 -1.5% of all benign laryngeal tumours. Schwannomas are slow growing benign well encapsulated tumour that arise from schwann cells of the nerve fibres. 25% of all schwannomas present in the head and neck region with majority occurring in the parapharyngeal space. Presentation in the larynx is a rare phenomenon. Over 130 cases of laryngeal schwannomas have been reported in the literature till date. Most common site of laryngeal schwannoma is aryepiglottic fold (80%) followed by arytenoids ventricular folds and vocal cords (20%)

A definitive diagnoses can be made histologically and complete surgical excision is curative. We describe a case of Laryngeal schwannoma in a young female which is successfully excised through a transoral microlaryngeal approach without tracheostomy

Case report

A 20 year old girl presented with a 6 month history of hoarseness of voice and difficulty in swallowing. She was non smoker. There was no associated dyspnea, odynophagia, neck swelling and significant weight loss. 90 degree Laryngoscopic examination revealed a smooth submucosal swelling in the left pyriform sinus (Figure 1). Bilateral vocal cord mobility was normal. No lymphadenopathy was detected. Contrast enhanced computerised tomography of the neck showed a well marginated hypodense lesion showing progressive enhancement with non enhancing central mass in supraglottic and pyriform sinus on left side extending superiorly from epiglottis and reaching inferiorly till glottis and right side small laryngocele (Figure 2 and Figure 3). Under general anaesthesia, transoral microlaryngoscopic excision of the lesion was done. Incision was taken over the swelling on the aryepiglottic fold, soft nodular mass was exposed, blunt dissection was done, mass was completely enucleated, diligent hemostasis achieved and margins were closely approximated.

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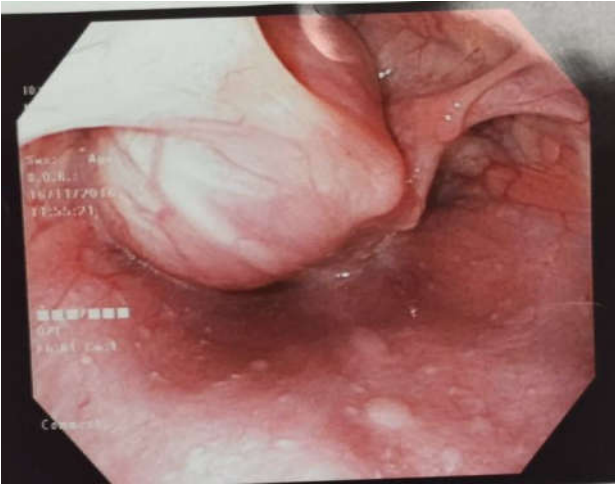


Figure 1 90 degree laryngoscopic image showing smooth submucosal swelling in the left pyriform sinus.

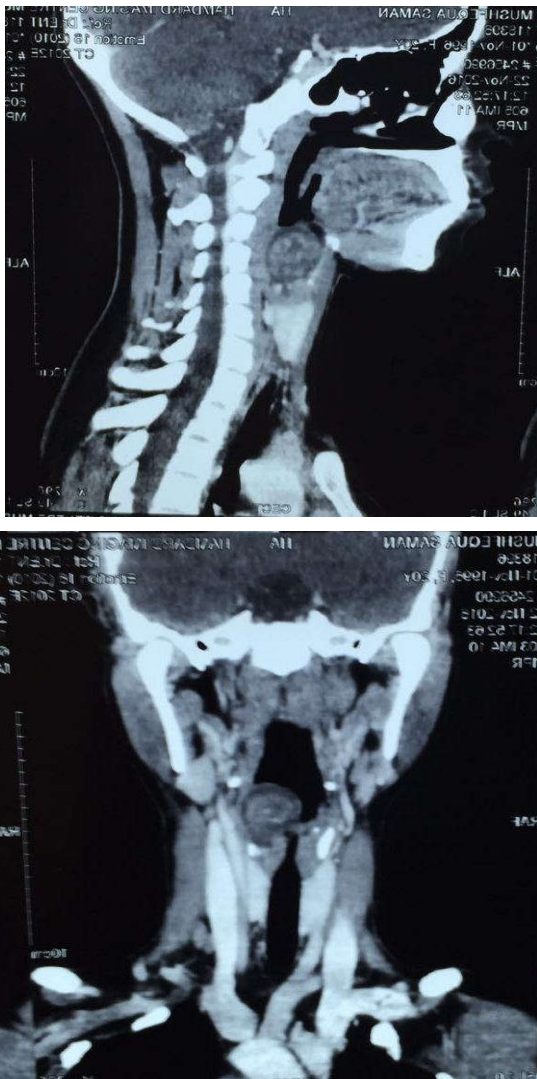


Figure 2 and Figure 3 Contrast enhanced computerised tomography of the neck showed a well margined hypodense lesion showing progressive enhancement with non enhancing central mass in supraglottic and pyriform sinus on left side extending superiorly from epiglottis and reaching inferiorly till glottis and right side small laryngocele.

Histopathology showed gray white solid areas admixed with multiple cystic areas filled with myxoid material along with areas of hemorrhage. Microscopically proliferation of spindled

shaped cells with fibrillary cytoplasm was seen with dense fibrous bands arranging the cells into nodules. Within the lesion, cellular areas were interspersed with loose myxoid and cystic areas suggestive of Schwannoma (Figure 4). Staining for S 100 protein was positive in the tumour cells. Post operative 90 degree scopy revealed restricted mobility of the left vocal cord.

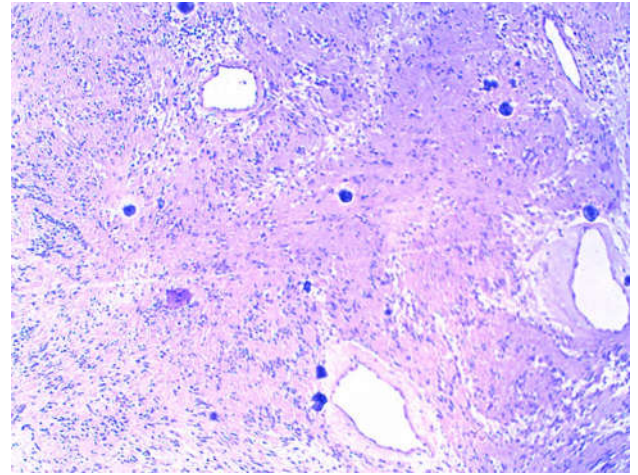


Figure 4 gray white solid areas admixed with multiple cystic areas filled with myxoid material along with areas of hemorrhage. Microscopically proliferation of spindled shaped cells with fibrillary cytoplasm was seen with dense fibrous bands arranging the cells into nodules. Within the lesion, cellular areas were interspersed with loose myxoid and cystic areas suggestive of Schwannoma

DISCUSSION

Schwannomas are benign slow-growing tumors that arise from the Schwann cells of any peripheral, cranial, or autonomic nerve sheaths. Schwannomas involving the head and neck are usually intracranial and most commonly involve the vestibular nerves. Between 25% and 45% of extracranial schwannomas arise in the head and neck region.[1]. Schwannoma was first described as a pathological entity by Verocay in 1908 who later called it neurinoma in 1910. Later the term neurilemmoma has been coined by Stout in 1935 [2]. The location of Schwannoma or neurofibroma within the larynx and hypopharynx is very uncommon. They represent 0.1% to 1.5% of all benign tumors in larynx and hypopharynx.[3] 80% are located in the aryepiglottic fold, 20% in the false or true vocal cords.[4-6] Only nine histologically confirmed hypopharyngeal schwannomas have been known.[7,8] They usually grow submucosal; with a few presenting as polypoid growth.[5]. There is a slight female preponderance.[5] The internal branch of the superior laryngeal nerve is possibly the nerve of origin.[9,10]

The presentation is generally in the form of sore throat, odynophagia, dysphagia, dyspnea, stridor, hoarseness, and sensation of a lump in the throat. Our patient presented with hoarseness and difficulty in swallowing confirming with benign hypopharyngeal masses. These salient features are due to pressure effects.[11] The differential diagnosis of the lesion included neurogenic tumors of the larynx include chondroma and adenoma.[12] Also laryngeal cyst and internal laryngocele should be taken into consideration.[5]

On the contrary, malignant hypopharyngeal neoplasms may present in addition to the above, with a recent onset of

symptoms with rapid progression, progressive weight loss and cervical swellings.

Imaging techniques such as computed tomography (CT) or magnetic resonance imaging (MRI) are also invaluable in the evaluation of schwannomas. On CT, they appear isodense, but up to 20% of these tumors may exhibit hypodense areas. On magnetic resonance imaging, a schwannoma is designated by intermediate intensity and can hardly be differentiated from muscle on T1 scans. On T2 scans, the intensity of a schwannoma increases.[13] Both also help to describe the extent of lesion and involvement of surrounding structures. Treatment of choice is complete enucleation/excision along with preservation of function of affected nerve/site. Various approaches such as endoscopic, open transoral, and external or lateral pharyngotomy approaches can be used depending on size and location of lesion. Small tumors can be excised easily by endoscopic and transoral routes while larger tumors are easily dealt using lateral pharyngotomy approaches. Here, a transoral excision was done due to its relatively small size.

Histopathologically, schwannomas have two different patterns: Antoni A and B areas. The former represents a cellular area in which sheets of spindle-shaped cells are generally arranged in a palisading fashion, called Verocay bodies. Antoni B is comprised of myxoid, loose, degenerative areas. Also, S100 protein positivity with no obvious mitotic activity is often characteristic of these cellular areas.[14] The histological pattern of this lesion with Antoni A and B as well as the S100 protein expression helped to confirm the diagnosis of schwannoma in this patient. Malignant transformation is reported in schwannoma it is very uncommon.[15]

CONCLUSION

Schwannomas of the hypopharynx are extremely rare. The preoperative diagnosis may be difficult and it is often made after the surgery. The definitive diagnosis relies on clinical suspicion and histopathological confirmation. Complete surgical excision with appropriate approaches is the treatment of head and neck schwannomas.

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