

CASE REPORT

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# Vocal fold myxoma: a case report and review of current literature

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## Abstract

**Background:** Vocal fold myxoma is a rare mesenchymal tumor with unknown exact etiology. The aim of this report is to review current literature on demographic and clinical features of vocal fold myxomas in order to increase awareness among otorhinolaryngologists on management of this rare vocal fold pathology.

**Case presentation:** We report a case of a 46-year-old female teacher presented with 1 year history of progressive dysphonia with no airway obstructive or constitutional symptoms. Flexible laryngoscopy revealed a polypoid mass at the anterior two thirds of the left vocal fold. We proceeded with endolaryngeal microsurgery, where a left vocal fold mass with prominent overlying blood vessels was incompletely excised due to poor plane, in an attempt to prevent injury to vocal ligament. The histopathological findings were consistent with myxoma. No evidence of recurrence was noted at 2 months postoperatively.

**Conclusions:** Vocal fold myxoma should be considered as a differential diagnosis of vocal fold pathology clinically resembling vocal fold polyp, especially in older male adults. Complete excision with removal of a rim of surrounding tissue is the gold-standard treatment to prevent recurrence. If diagnosis is uncertain, intraoperative frozen section can be performed to ensure margins are free from tumor. Life-long follow-up is recommended in incomplete excision of tumor.

**Keywords:** Case report, Larynx, Laryngeal myxoma, Myxoma, Vocal fold

## Background

Myxomas are rare benign neoplasms of uncertain mesenchymal cell origin that typically involve large muscles such as the heart [1]. In the head and neck region, they have been reported to occur as odontogenic tumors of maxilla and mandible [1, 2]. Myxomas originating from the larynx are rare and sites of involvement that have been reported include vocal fold, aryepiglottic fold, and epiglottis [3]. Myxomas rarely arise from vocal fold with only over twenty cases reported for the past 30 years through extensive literature search. The exact etiology of vocal fold myxoma remains unclear. It is believed that myxomas are likely to derive from primitive embryonic

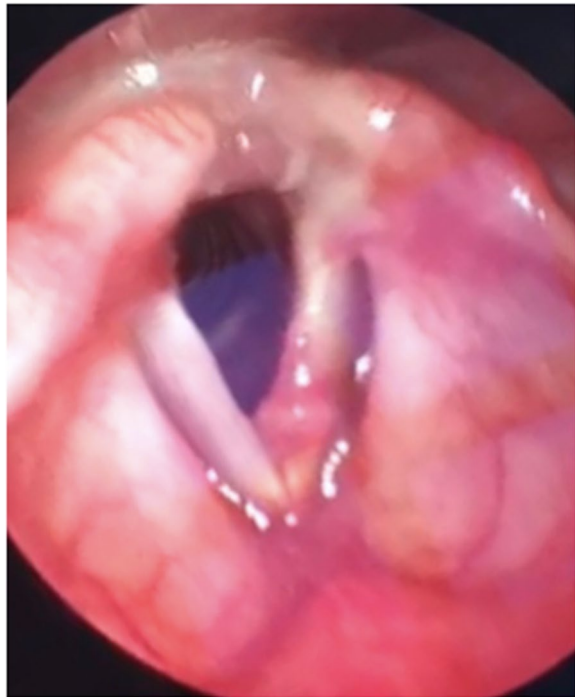
mesenchyme or fibroblasts that are capable of elaborating abundant mucopolysaccharides [1]. Herein, we describe a case of vocal fold myxoma initially diagnosed as vocal fold polyp.

## Case presentation

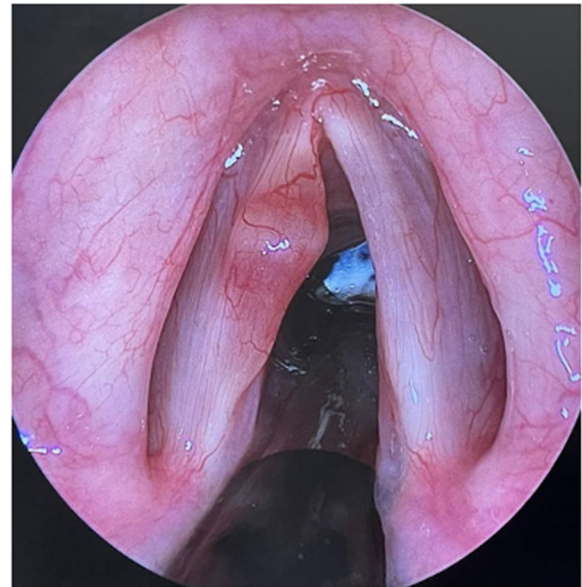
A 46-year-old female presented with 1 year history of progressive dysphonia. She worked as a teacher and had no significant medical and family history. There were no associated breathing or swallowing problems. Clinically, she appeared to be comfortable with no stridor or neck swellings. Maximum phonation time was normal (15 s), and she had overall dysphonia of grade 3 (roughness) on GRBAS scale [4]. Flexible laryngoscopy revealed an approximately 1.0 × 1.0 cm, round, pink, polypoid mass with smooth surface at the edge of anterior two thirds of the left membranous vocal fold (Fig. 1). Vocal fold mobility was normal, and there were no other abnormal

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**Fig. 1** Indirect laryngoscopy view showing polypoid mass at anterior two thirds of the left membranous vocal fold

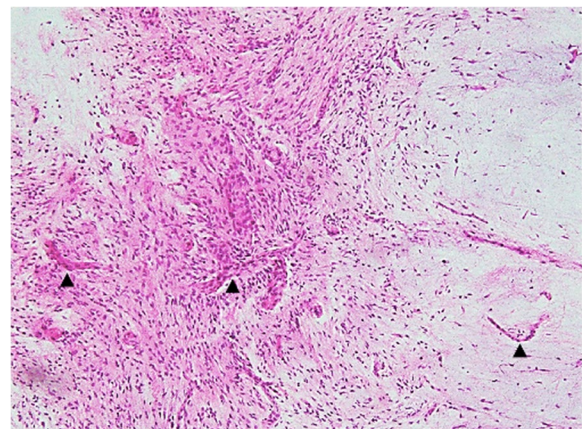


**Fig. 2** Direct laryngoscopy view showing fusiform polypoid mass at left vocal fold with prominent overlying blood vessels. Note the fullness surrounding the mass within the superficial lamina propria

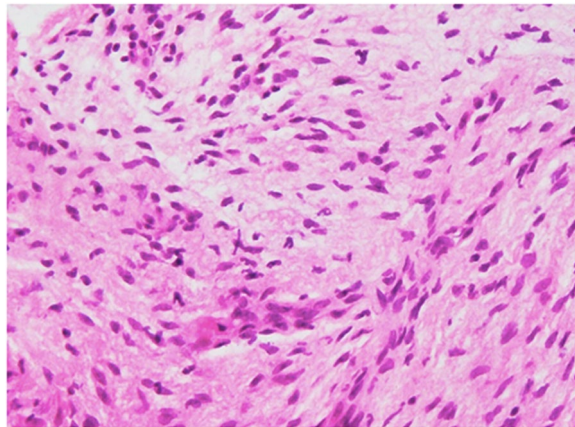
laryngeal findings. Provisional diagnosis given was left vocal fold polyp in view of her profession and laryngoscopy findings. We proceeded with endolaryngeal microsurgery using precision technique, where intraoperatively there was a  $1.0 \times 1.0$  cm mass at left vocal fold with prominent overlying blood vessels (Fig. 2). After raising medial microflap, difficulty was encountered in removing the entire mass as it extended towards vocal ligament with no clear plane, appearing as edematous tissue within superficial lamina propria. Small fragments of tissue close to vocal ligament were not removed in an attempt to preserve vocal function.

Microscopically, the lesion showed hypocellular myxoid-rich area alternating with more cellular area (Fig. 3). The myxoid-rich area showed presence of scattered bland spindle cells accompanied with presence of thin walled linear or curvilinear-like blood vessel. The cellular areas were composed of moderately cellular bland ovoid to spindle cells with eosinophilic cytoplasm and indistinct cell boundary embedded within a variably collagenized stroma. No nuclear pleomorphism, necrosis, or mitosis was detected (Fig. 4). The histopathological findings of the lesion were consistent with myxoma. Further immunohistochemical study showed the spindle cells are negative for cytokeratin AE1/AE3, epithelial membrane antigen

(EMA), desmin, actin, CD34, and S100. Full physical examination was performed to rule out heritable diseases that are associated with vocal fold myxoma such as Carney complex, Mazabraud syndrome, and Hunter's syndrome. Two months postoperatively, patient's hoarseness has improved with overall dysphonia of grade one (roughness) on GRBAS scale [4]. Laryngoscopic examination showed mild edema over the left membranous vocal fold at the operated site, and there was no residual mass visualized. The patient was



**Fig. 3** Alternating myxoid and cellular spindle shaped cells with occasional curvilinear-like blood vessels (arrow head) (hematoxylin and eosin stain, original magnification  $\times 100$ )



**Fig. 4** The spindle shaped cells showed normochromic nuclei with inconspicuous nucleoli and indistinct eosinophilic cytoplasm. No nuclear pleomorphism, mitosis, or tumor necrosis were observed (hematoxylin and eosin stain, original magnification  $\times 400$ )

informed of the difficulty encountered intraoperatively and advised for life-long follow-up. She was satisfied with the treatment given.

## Discussion

Historically, myxoma was first described by Virchow in 1871 as a group of tumors that resemble mucinous substance of umbilical cord [5]. In 1948, Stout further elucidated myxoma as non-metastasizing true mesenchymal neoplasm consisting of undifferentiated stellate cells in a loose myxoid stroma [5]. This forms the basis of current histological description, and a diagnostic criterion was also outlined.

Based on our review, vocal fold myxomas show strong male preponderance, with 20 out of 23 cases reported occurred in males (Table 1) [1–3, 6–12, 15–21]. Patients' ages ranged from 25 to 77 years old and tend to occur in older adults between 5th to 7th decades. Mode of presentation generally depends on size of tumor. Lesions measuring less than 1.5 cm commonly present with progressive dysphonia. Larger lesions measuring more than 1.5 cm are more likely to present with upper airway obstruction such as dyspnea and stridor. Two of the reported cases required emergency tracheostomy to secure airway [9, 17]. On laryngoscopic examination, vocal fold myxoma usually appears as broad-based, hypervascular, and polypoid

**Table 1** Summary of reported cases of vocal fold myxoma

Case [ref]	Year	Age	Gender	Site	Size (cm)	Main symptom	Removal	Recurrence
1 [6]	1982	30	M	Left	0.75	Dysphonia	Complete	None (12 months)
2 [6]	1982	25	F	Right	1.5	Dysphonia	Complete	None (12 months)
3 [7]	1994	64	M	Left	1.0	Dysphonia	Incomplete	None (18 months)
4 [8]	1997	57	M	Right	0.7	Dysphonia	Complete	None (6 months)
5 [9]	1997	62	M	Right	2.5	Dyspnea, stridor (requiring tracheostomy)	Complete	–
6 [10]	1999	42	M	–	–	Dyspnea, OSA	–	None (12 months)
7 [11]	2005	46	M	Right	0.8	Dysphonia	Complete	–
8 [12]	2007	53	M	Right	0.5	Dysphonia	Complete	–
9 [13]	2008	48	F	Left	–	Dysphonia	–	None (3 months)
10 [14]	2008	74	F	Right	0.4	Dysphonia, dyspnea	Complete	None (24 months)
11 [15]	2008	36	M	Right	0.7	Dysphonia	–	–
12 [16]	2013	61	M	Right	0.5	Dysphonia	Complete	None (3 monthly)
13 [1]	2014	77	M	Left	0.8	Dysphonia	–	None (3 months)
14 [17]	2014	64	M	Right	1.5	Dyspnea, stridor (requiring tracheostomy)	–	None (8 months)
15 [18]	2015	60	M	Right	1.0	Dysphonia	Incomplete	Recurrence at 27 months
16 [19]	2018	44	M	Right	–	Dysphonia	Complete	None (18 months)
17 [19]	2018	77	M	Left	–	Dysphonia	Complete	None at 3 months
18 [20]	2018	53	M	Right	0.5	Dysphonia	–	–
19 [3]	2020	69	M	Right	0.9	Dysphonia	Complete	None (1 month)
20 [2]	2020	36	M	–	–	Dysphonia	–	None (6 months)
21 [2]	2020	45	M	–	–	Dysphonia	–	None (6 months)
22 [2]	2020	72	M	–	–	Dysphonia	–	None (6 months)
23 [21]	2021	54	M	Right	1.5	Dysphonia, dyspnea, stridor	Complete	–
24 [–]	Present case	46	F	Left	1.0	Dysphonia	Incomplete	None (2 months)

[Ref] Reference; M Male; F Female, – Not reported



mass [1, 2]. It usually arises superficial to vocal ligament at variable sites of vocal fold including anterior commissure [1, 2]. Stroboscopic examination may display loss of mucosal wave [7, 8]. It is important to perform complete physical examination as myxomas can be associated with heritable disease such as Carney complex, Mazabraud syndrome, and mucopolysaccharidosis II (MPS II) or Hunter's syndrome [1]. Recurrence of tumor has only been reported once in this review.

Diagnosis of myxoma is made through histopathological examination. Histologically, laryngeal myxomas can be described as pure myxoma and myxoma of cellular variant [21]. Classic myxoma is characterized by bland spindle and stellate shaped cells embedded in hypovascular, abundant loose myxoid stroma, whereas myxoma of cellular variant is characterized by hypercellularity, more numerous collagen fibers, and increased vascularity [21]. Important histological differential diagnoses to consider are other soft tissue tumors with myxoid degeneration such as myxoid liposarcoma, myxoid leiomyosarcoma, ganglions, and rhabdomyosarcoma [22]. The absence of stromal vasculature, hemorrhage, hemosiderin-laden macrophages, and hyalinization of basement membrane can help to distinguish myxomas from vocal fold polyps [1].

The treatment of laryngeal myxoma is complete excision of the tumor with removal of a rim of surrounding normal tissue [2]. This is due to infiltrative growth pattern and presence of pseudocapsule in myxomas where incomplete excision will inevitably lead to local recurrence [2]. In doing so, intra-operative frozen section has been found to be beneficial for immediate assessment of margin status to ensure complete excision [9]. Endoscopic laryngeal microsurgery with careful excision of tumor is pertinent to preserve the integrity of vocal ligament and vocal function. An external thyrotomy approach has been described where the vocal fold myxoma was adherent to the undersurface of vocal ligament [18]. The tumor was not completely excised to prevent injury to the vocal ligament and recurrence was noted after 27 months [18]. Long-term follow-up is recommended due to its infiltrative nature, slow growth rate, and high incidence of local recurrence [13].

This case report describes a rare occurrence of myxoma arising from vocal fold with largest compilation of cases to date of over twenty cases for the past 30 years. Previous demographic findings and intraoperative techniques employed were also analyzed. The limitation of this report is that long term follow-up is not available to observe for recurrence.

## Conclusions

Vocal fold myxoma should be considered as a differential diagnosis of vocal fold pathology clinically resembling vocal fold polyp, especially in older male adults. Complete excision with removal of a rim of surrounding tissue is the gold-standard treatment to prevent recurrence. If diagnosis is uncertain, intraoperative frozen section may play a role in employing appropriate surgical technique and to ensure margins are free from tumor. Life-long follow-up is recommended in incomplete excision of tumor.

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## Authors' contributions

ZMN: design of study, data collection and interpretation manuscript drafting, final approval of manuscript. MHM: data collection and interpretation, final approval of manuscript. IHB: data collection and interpretation, final approval of manuscript. LTL: data collection and interpretation, final approval of manuscript. All authors read and approved the final manuscript.

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## Availability of data and materials

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## Declarations

## Ethics approval and consent to participate

Not applicable

## Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

## Competing interests

The authors declare that they have no competing interests.

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