

Myxoma-rare laryngeal presentation.

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Citation

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Abstract

Objective: We report a rare case of laryngeal myxoma. **Case report:** Laryngeal myxomas are rare benign tumors of mesenchymal origin, to our knowledge only six have been reported in the English-language literature. We present the case of a woman seen in the ENT outpatient clinic with a lifelong history of dysphonia. On examination she was found to have bilateral swollen vocal cords. This was thought to represent Reinke's oedema. Pathological analysis after routine microlaryngoscopy showed this to be a vocal cord myxoma. **Conclusion:** We recommend excision of this lesion and long term follow-up.

INTRODUCTION

We present the case of a 48-year-old lady who was seen in the ENT outpatient clinic with a lifelong history of dysphonia. Indirect laryngoscopy showed bilateral swollen vocal cords with a polypoidal lesion on the left vocal cord, an appearance thought to be typical of Reinke's oedema. Microlaryngoscopy was carried out, and the polyp removed. Pathological analysis showed this to be a myxoma. Most myxomas in the head and neck arise from the mandible¹. Laryngeal myxomas are benign tumors of mesenchymal origin, and are very rare, to date six have been reported in the English-language literature.

CASE REPORT

We submit a case of a 48-year-old lady who presented to the ENT outpatient department with a life long history of dysphonia. She described having a deep, husky voice since childhood. Her mother's explanation to her as a child had been that her vocal cords had not developed properly. Recently her voice had got much worse and this was especially affecting her role as a business manager. She found her voice had become deeper and at times had a breathy quality. She was also a smoker and had a history of hyperthyroidism. Flexible nasal endoscopy was carried out and this showed bilateral vocal cord swelling and a polypoidal lesion on the left vocal cord. The appearance was in keeping with a severe case of Reinke's oedema. She was given general advice on voice care, advised to stop smoking and micro-laryngeal surgery was arranged within four weeks.

Intra-operative findings were of oedema of the right vocal

cord and large polyp arising from the left vocal cord (figure 1).

Figure 1

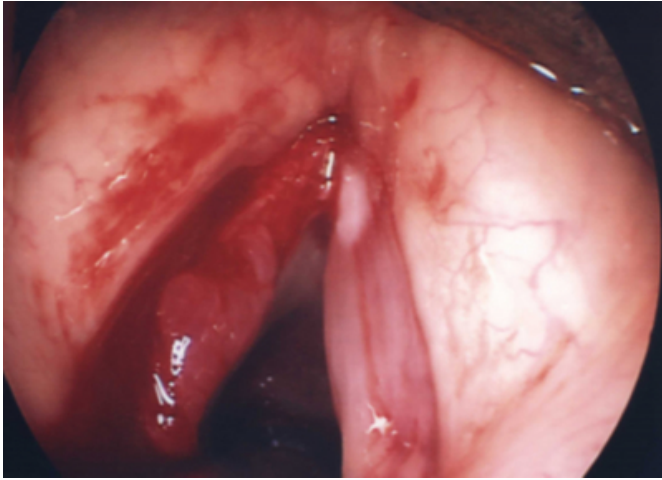
Figure 1



The right vocal cord was aspirated and the left vocal cord polyp removed with standard microlaryngoscopy instrumentation (figure 2).

Figure 2

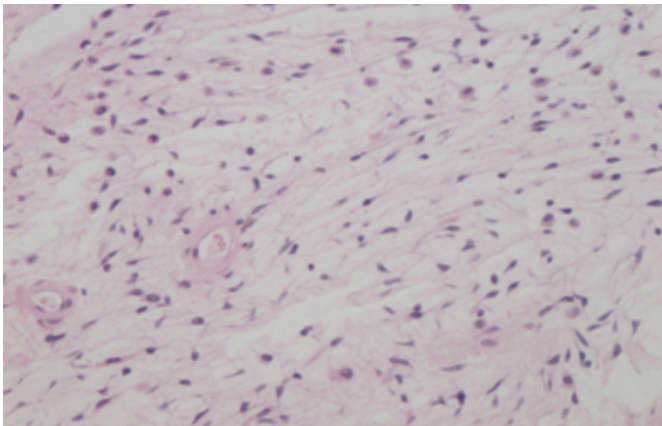
Figure 2



Histological examination of the polypoid lesion revealed hyperplastic surface squamous epithelium with a variably cellular lesion in the submucosa composed of spindle cells arranged in loose myxomatous stroma (figure 3).

Figure 3

Figure 3 Photomicrograph showing the presence of spindle cells arranged in loose myxomatous stroma the appearances of which are in keeping with a myxoma.



The spindle cells showed a mild degree of atypia but no mitotic figures or evidence of necrosis was identified. Immunohistochemistry revealed the spindle cells to express CD34 but to be negative for S100 and smooth muscle actin. Only very occasional cells were noted to express the proliferation marker Ki67. These appearances were interpreted as being in keeping with a myxoma of the vocal cord.

At three-month clinic review, her voice had much improved and there was no evidence of recurrence. At her last review to date (3 year follow up), there has been no evidence of

recurrence, and her voice is now normal.

DISCUSSION

Myxomas are benign mesenchymal tumors that occur mostly in subcutaneous soft tissue, intramuscular tissue or heart. From our literature search only six have been reported in the English language literature. In the head and neck region the most common areas affected are the mandible and maxilla¹. It has been reported these benign tumours in the maxilla can be so invasive that it results in destruction and deformation of the facial skeleton². However these lesions are rare in the larynx. Myxoma of the larynx is frequently misdiagnosed as a vocal polyp. A correct diagnosis of myxoma relies on strict pathological criteria, and is a rare diagnosis in contrast to myxomatous degeneration in laryngeal polyps, which can occur frequently³.

Dual location has also been reported in the oropharynx, and hypopharynx and it is suggested that patients undergo screening to exclude cardiac lesions which are more common presentations of this type of tumor⁴. Of the six reported cases in the larynx, four arose from the glottis with dysphonia being the most common complaint. Supraglottic myxomas tend to present later than those in vocal folds because they can grow larger before they become symptomatic, whereas vocal fold myxomas are detected earlier because they can cause disturbance of vocal fold vibrations. A large vocal cord myxoma, however, has been reported to cause airway obstruction that required tracheostomy⁵.

Within the larynx, the main differential diagnosis is between myxoma, myxoid degeneration of a laryngeal polyp and prominent Reinke's oedema. Low-grade myxoid liposarcoma and chondrosarcomas can also mimic myxoma. Immunohistochemical staining such as S100 protein (which is negative for myxoma, but positive for lipoblasts and chondroblasts) can be used as can immunoreactivity of smooth muscle⁶. Myxomas are connective tissue tumours composed of multinucleate stellate cells suspended in an oedematous mucopolysaccharide-rich stroma. They also infiltrate surrounding tissue and are characterized by a slow growth rate, thus they have a high incidence of local recurrence. Therefore long-term follow-up is required.

Laryngeal myxomas can usually be excised using micro-laryngeal techniques; however an external approach may be necessary in some cases. As previously mentioned the most important aspect is securing an airway, which may even necessitate tracheostomy. To prevent recurrence, the

literature suggests myxoma should be surgically excised with surrounding normal tissue¹⁻³. Intra-operative frozen section can also be useful for immediate assessment of margin status⁵. In our case we did not suspect this diagnosis beforehand therefore, no particular margin was taken. The patient has also had no recurrence thus far, and her voice remains normal.

SUMMARY

Laryngeal myxomas are benign tumours of mesenchymal origin and are very rare. Only six have been reported in the English-language literature. In the head and neck areas the commonest site is the mandible. Surgically excision is advocated with surrounding normal tissue as infiltration of the adjacent tissue can occur. They also have a high incidence of local recurrence with a slow growth rate, therefore long-term follow-up is recommended.

References

1. Nakamura A, Iguchi H, Kusuki M, Yamane H, Matsuda M, Osako S. Laryngeal myxoma. *Acta Otolaryngol.* 2007 Jun 18;1-3
2. Deron PB, Nikolovski N, den Hollander JC, Spoelstra HA, Knecht PP. Myxoma of the maxilla: a case with extremely aggressive biologic behavior. *Head Neck.* 1996;18: 459-64.
3. Hadley J, Gardiner Q, Dilkes M, Boyle M. Myxoma of the larynx: A case report and a review of the literature. *Journal of Laryngol Otol.* 1994 ;108: 811-812.
4. Ayache S, Chatelain D, Tramier B, Strunski V. Oropharyngeal and hypopharyngeal myxoma: case report and literature review. *Journal of Laryngol Otol* 2007 May;121(5) e5.
5. Kim KM, Kim SC, Jeong HJ, Kie JH. Myxoma: life-threatening benign nonepithelial tumor of the larynx. *Yonsei Med J.* 1997; 38:187-89.
6. Idrees MT, Hessler R, Terris D, Mixson C, Wang BY. Unusual Polypoid Laryngeal Myxoma. *Mt Sinai J Med.* 2005 Jul; 72: 282-4.

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