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Clinical Record

Aggressive angiomyxoma of larynx: case report and literature review

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Abstract

Objective: We report the second known case of aggressive angiomyxoma of the larynx.

Method: Case report and a review of the world literature concerning angiomyxoma of the larynx and recent advances in the immunohistochemical, cytogenic and clinical study of its female pelvic counterpart.

Results: Aggressive angiomyxoma is a rare mesenchymal tumour originally thought only to occur in the female pelvis and peritoneum, or rarely in the male genital tract. A 47-year-old man presented with a one-month history of dysphonia. He was found to have a supraglottic mass on endoscopic examination, and underwent a laryngofissure approach excision biopsy and covering tracheostomy. Histological analysis showed a characteristic proliferation of spindle cells widely separated by loose, myxoid stroma with a prominent vascular component. Aggressive angiomyxoma was diagnosed.

Conclusion: To our knowledge, this is the second report in the world literature of aggressive angiomyxoma of the larynx. Comparison with the female pelvic counterpart facilitates diagnosis, aided by recent advances, and suggests that complete surgical excision with a wide margin is the treatment of choice.

Key words: Laryngeal Neoplasm; Angiomyxoma

Introduction

Aggressive angiomyxoma is a rare mesenchymal tumour typically occurring in the female pelvis and peritoneum[1] and the male genital tract.[2] It has only once been described in the larynx.[3]

We present the case of 47-year-old, dysphonic man diagnosed as the second known laryngeal case of aggressive angiomyxoma.

Case report

A 47-year-old man presented with a one-month history of dysphonia. Although this had been preceded by subtle changes over the previous year, he had remained systemically well, with no other features of malignancy apparent.

Flexible fibre-optic laryngoscopy showed a left-sided, mucosa-covered, smooth, supraglottic mass.

The patient was subsequently admitted for microlaryngoscopy and biopsy. Endoscopic examination of the larynx proved difficult due to prominent incisors and limited cervical spine extension. Laryngoscopy confirmed the above findings; inferior to the known mass, the larynx was normal. Biopsies were taken, which unfortunately proved inadequate.

Computed tomography of the larynx confirmed a well defined tumour confined to the supraglottis (Figure 1).

Due to the above anatomical considerations, an open approach to the larynx was deemed most appropriate. The patient subsequently underwent a laryngofissure approach excision biopsy and covering tracheostomy. Intra-operatively, the left-sided supraglottic mass was completely excised.

The patient was successfully decannulated one week post-operatively and later discharged home.

Macroscopically, the mass measured $4.0 \times 2.5 \times 1.0$ cm and was well circumscribed, oval, rubbery and white. Microscopically, a bland, hypocellular, myxoid stroma was seen, with a proliferation of abnormal blood vessels. Spindle cells were seen scattered throughout the entire specimen. The nuclei were small and hyperchromatic, but there was no nuclear atypia or mitoses. Neutrophil polymorphic cells were also scattered throughout (Figure 2). Immunostaining for S-100 protein, desmin, cluster of differentiation 34 glycoprotein and smooth muscle actin was negative.

After four years of follow up the patient remained well, with no evidence of loco-regional recurrence or distant metastasis.

Discussion

Although little is currently known about laryngeal aggressive angiomyxoma, research into the immunohistochemical, cytogenic and clinical course of its female pelvic counterpart may aid our understanding. The term 'aggressive' was introduced by Steeper and Rosai in 1983[1] in their case report of this distinctive neoplasm of the female pelvis and perineum. The term refers to the tumour's locally aggressive behaviour and propensity for local recurrence, rather than its metastatic potential. To date, only two

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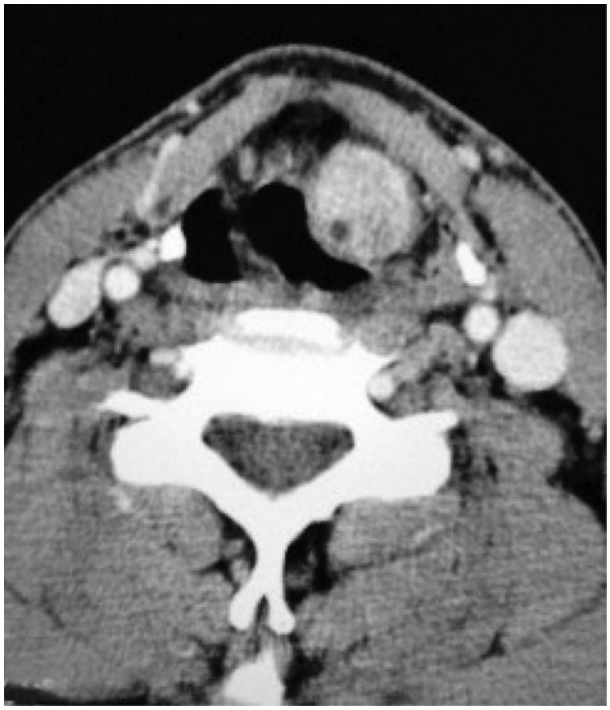

FIG. 1

Axial computed tomography scan showing left-sided, supraglottic mass.

cases of metastatic aggressive angiomyxoma have been published, both arising in the female pelvis.[4,5]

Histologically, this neoplasm is characterised by a proliferation of spindle-shaped or stellate cells widely separated by loose, myxoid stroma, in which is dispersed a prominent vascular component. Immunohistochemically, female pelvic aggressive angiomyxomas have been found to be invariably positive for vimentin, desmin, cluster of differentiation 44, 34 and K4 glycoprotein, oestrogen receptors, progesterone receptors, smooth muscle actin,[6] and cytokeratin AE1/AE3.[7] Negativity for S-100 protein is consistent,[6,7] and negativity for muscle-specific antigen variable.[6,8] In our case, the only similarity with the pelvic form was negativity for S-100 protein, as positivity for desmin, cluster of differentiation 34 glycoprotein and smooth muscle actin was not found, suggesting either little immunohistochemical similarity or greater variability in the laryngeal form.

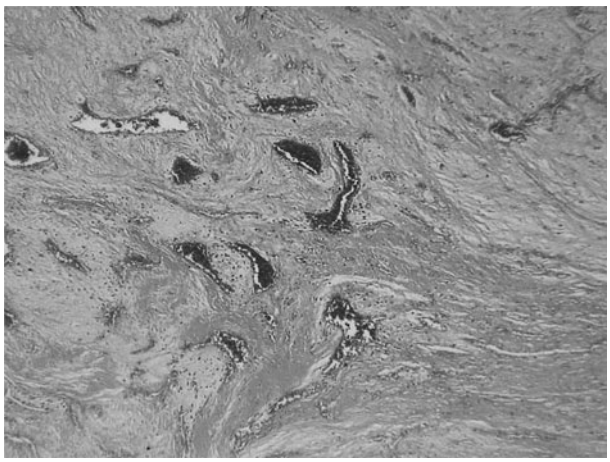

FIG. 2

Photomicrograph showing bland, hypocellular, myxoid stroma with a proliferation of abnormal blood vessels (H&E; ×100).

Chromosomal aberrations have been documented in numerous pelvic cases, identifying a non-random involvement of chromosomal band 12q15.[9–11] An additional case with 12q15 rearrangement has been described using fluorescence in situ hybridisation.[12] Such rearrangements lead to alterations of the high mobility group gene *HMGA2*.

Aggressive angiomyxoma has been classified by the World Health Organization under 'tumours of uncertain differentiation'.[13] Its tendency for local recurrence has meant that, in the female pelvic form, complete excision is favoured when possible, preferably with a wide margin. Furthermore, the first case report of a laryngeal aggressive angiomyxoma suggested that this approach should be used for the laryngeal form, given that the tumour recurred after incomplete excision.[3]

The use of chemotherapy and radiotherapy for aggressive angiomyxomas of the pelvis is not particularly effective, presumably because of the low mitotic activity demonstrated. However, due to the expression of oestrogen and progesterone receptors in the female genital tract forms, therapy with gonadotrophin-releasing hormone agonists has been applied. It has been suggested that this is useful in patients with recurrence or in whom surgical excision is not possible.[14]

- **Aggressive angiomyxoma is a rare mesenchymal tumour now known to be found in the larynx**
- **Its more common pelvic counterpart demonstrates a propensity for local recurrence, and has metastatic potential**
- **Treatment is surgical; a wide excision margin is favourable**
- **It is currently unclear whether the immunohistochemical and genetic characteristics of the laryngeal form are similar to those of its pelvic counterpart**

Given that the presented patient is only the second reported case of laryngeal aggressive angiomyxoma, the long-term prognosis remains undetermined. Although the female pelvic form was not previously thought to have metastatic potential, two case reports have now been published describing pulmonary,[4] and pulmonary and mediastinal,[5] metastases. Long-term follow up and reporting of further cases will help to determine whether the laryngeal disease bears the same metastatic traits.

Conclusion

Angiomyxoma of the larynx is an uncommon, malignant tumour of unknown aetiology and prognosis. Comparison with the female pelvic form would suggest that complete surgical excision with a wide margin is the treatment of choice. Research into the pelvic form may aid our understanding; however, it is currently unclear to what extent the laryngeal form shares the immunohistological, cytogenetic and clinical traits of its pelvic counterpart.

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