CASE REPORT

Laryngeal Sarcoma: A Case Report

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ABSTRACT

Laryngeal sarcoma is a rare malignant tumour and the diagnosis requires comprehensive histopathological and immunohistochemical examinations. Due to its rarity, there is paucity of information on its behaviour and management. We report a case of an elderly male presented with one month history of hoarseness. Laryngoscopic examination showed a large exophytic mass at anterior commissure causing partial airway obstruction. Tracheostomy, direct laryngoscopy and excision of tumour with adequate margin were performed. The histopathological examination revealed myofibroblastic sarcoma. Computed tomography of the neck showed no regional or distant metastasis. Post-operatively, he was decannulated and there was no clinical evidence of recurrence. Management of LS is highly individualised. Complete tumour excision with clear margins is the primary treatment. Neck dissection is not required if there is no nodal metastasis. Radiotherapy is advocated in selected cases to achieve loco-regional control.
Malaysian Journal of Medicine and Health Sciences (2023) 19(SUPP19):13-15. doi:10.47836/mjmhs.19.s19.3

Keywords: Larynx, Laryngeal cancer, Laryngeal sarcoma, Laryngoscopy, Sarcoma

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INTRODUCTION

Laryngeal sarcoma (LS) is a rare malignant tumour of the larynx and the diagnosis requires histopathological with immunohistochemical examination. Due to its rarity and presence of various histological subtypes, there is limited information on clinical features, histology and standard treatment in the literature. Herein, we report a case of laryngeal sarcoma without clinical evidence of recurrence post-surgery.

CASE REPORT

A 58-year-old Indian male presented with one month history of hoarseness. He denies difficulty breathing, reduced effort tolerance or dysphagia. Clinically, there was no stridor and no palpable cervical lymph nodes. Flexible laryngoscopy examination showed a large exophytic mass at anterior commissure, partially obstructing airway (Fig.1). Bilateral vocal folds were mobile. Subsequently, tracheostomy under local anaesthesia, direct laryngoscopy and endoscopic excision of tumour with adequate margin were performed (Fig.2). Intraoperatively, there was an exophytic mass arising from anterior half of right vocal fold, sparing anterior commissure, ventricle and subglottis. The histopathological examination (HPE) was reported as spindle cells arranged in fascicles and in some area forming bundles. The spindle cells display pleomorphic round to fusiform vesicular nuclei with prominent nucleoli and abundant eosinophilic cytoplasm. Immunohistochemical staining tests were positive for SMA and S100, and negative for desmin, CD34, CKAE1/3 and Caldesmon (Fig.3). The HPE confirmed the diagnosis of myofibroblastic sarcoma. Post-surgery,

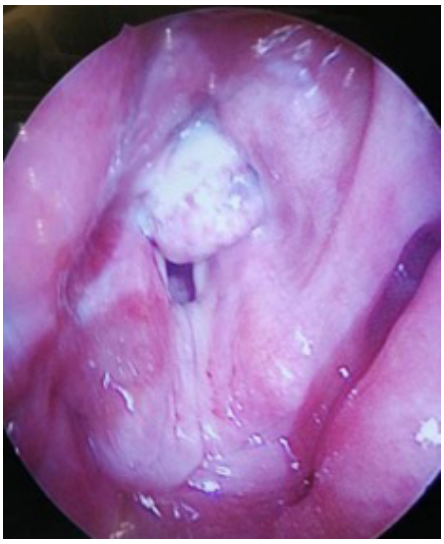

Figure 1: Flexible laryngoscopy view showing a large exophytic mass at anterior commissure

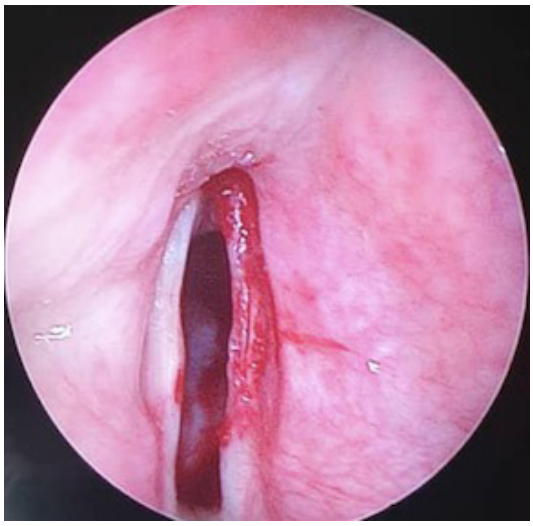

Figure 2: Direct laryngoscopy view showing complete tumour excision

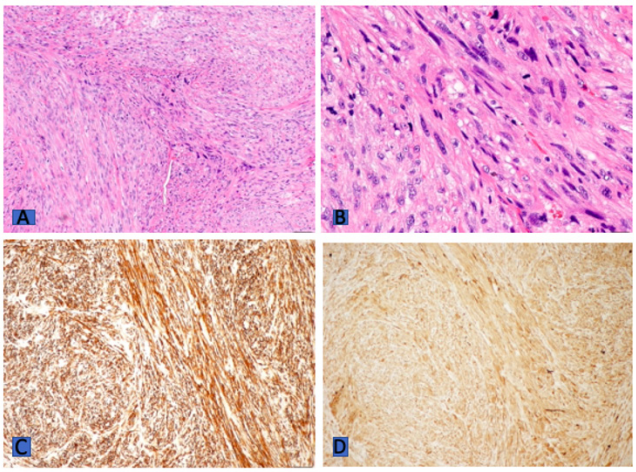

Figure 3: Micrographs revealing the histological appearance of the tumour. (A and B) Hematoxylin and eosin stain revealing spindle cells arranged in fascicles. The cells display hyperchromatic nuclei, conspicuous nucleoli and abundant eosinophilic cytoplasm (A,x100: B,x400). (C) Spindle cells positive for smooth muscle actin (SMA). (D) Spindle cells positive for S-100 protein

patient was well and decannulated after two months. There was no clinical evidence of recurrence observed at three years follow-up.

DISCUSSION

Sarcoma is an aggressive form of malignant soft tissue tumour that usually originate from upper or lower limbs (1). In head and neck region, malignant tumours are commonly of epithelial origin whereby squamous cell carcinoma (SCC) is the most common histology found (1). Soft tissue sarcomas are rare in head and neck region where it has been reported to occur in less than 5% of cases (2). In head and neck region, the common sites of tumour origin are neck, face, forehead and sinuses, usually presenting as painless submucosal or subcutaneous mass (3). Sarcoma arising from larynx is extremely rare and accounts for less than 1% of all laryngeal malignant tumors (2). Because of the rare occurrence, there is lack of information on the behaviour, management and outcome of laryngeal sarcoma (LS).

The aetiology of LS remains unclear thus far. However, the incidence have been found to be higher in older age males based on previous reported cases (2). Clinical presentation of LS is similar to laryngeal SCC. Hoarseness is usually the presenting symptom, followed by upper airway obstruction symptoms such as dyspnoea, reduced effort tolerance and stridor, as the tumour progressively increase in size. Patients may complain of dysphagia when the tumour is large enough to involve hypopharynx (4). The laryngoscopic appearance is usually described as submucosal polypoidal or lobulated mass and rarely ulcerative, as opposed to laryngeal SCC where the appearance is commonly described as ulcerative (3, 4). LS is also more localised and less likely to infiltrate in contrast to laryngeal SCC (3). The majority of LS originate from vocal folds, although it can arise from any laryngeal subsites (4). However, involvement of contralateral vocal fold is uncommon (3).

Definitive diagnosis requires histopathological examination (HPE) and LS is known to have various histological subtypes. More than half of LS are fibrosarcomas followed by chondrosarcomas (2). Other rare subtypes are spindle cell sarcoma, osteosarcomas, liposarcomas, undifferentiated pleomorphic sarcomas, synovial sarcomas, rhabdomyosarcomas, leiomyosarcoma, undifferentiated epithelial sarcoma and myofibrosarcomas (2). Sarcomas are distinguished based on immunohistochemical (IHC) markers such as desmin, vimentin and alpha-smooth muscle actin (2). It is important to ensure core of the tumour is obtained rather than superficial part of tumour to allow complete HPE and extensive IHC tests (3). Electron microscopy can aid diagnosis in ambiguous cases (2). Apart from that, sarcomas have a wide spectrum of clinical activity from slow-growing, locally and regionally aggressive, and potential for distant metastasis (3). For example, chondrosarcoma has slow growth potential as compared to undifferentiated epithelial sarcoma that has a high metastatic ability (2).

Contrast-enhanced computed tomography (CECT) or Magnetic Resonance Imaging (MRI) of the neck are the imaging modalities of choice to assess size, extension and presence of cervical metastasis (2). However, there is no specific radiological features in LS (5). MRI has superior soft tissue delineation and commonly purported for preoperative planning (5). It is also used to assess tumour recurrence (2). At present, there is no valid standardised staging for LS. However, some authors used American Joint Committee on Cancer (AJCC) for staging (3).

It is important to note that treatment for LS is highly varied

and individualised, and multidisciplinary team approach is vital to optimize management of LS. Currently, there is no definitive guideline on the management of LS. Most studies supported surgical excision and radiotherapy in selected cases. Complete tumour excision with clear margins is the mainstay of treatment (5). Larynx preservation is preferable and possible as most cases are diagnosed early and remain operable for a considerably longer period after diagnosis as compared to squamous cell carcinoma (SCC) (2, 3). The treatment of LS depends on histological type, location and size (3). The type of surgery recommended depends on the size of tumour where options include endoscopic approach, partial laryngectomy or total laryngectomy (2). In advanced cases, a more aggressive surgical approach is required in order to ensure margins are free from tumour. Large infiltrating tumours may require vertical partial laryngectomy (laryngofissure) or total laryngectomy (2). Specifically for chondrosarcoma subtype, a non-aggressive surgery can be opted because of the nature of the tumour which is usually pedunculated, well-differentiated, less likely to infiltrate surrounding structures and metastasize later (2, 3). Chondrosarcoma generally has better prognosis (2).

Cervical metastases are uncommon in LS except at advanced stage (3). Studies have shown that 88-90% of head and neck soft tissue sarcomas do not have cervical metastases (2). Neck dissection is performed when there is confirmed cervical lymph node metastasis (5). Therefore, elective neck dissection is generally not required (3).

Role of radiotherapy and chemotherapy is controversial. Sarcomas are generally deemed to be resistant to radiotherapy (2, 3). However, some authors cited that radiotherapy can be used as an adjunct to surgery to achieve loco-regional control (2, 5). It is indicated for high-grade tumour, positive surgical margins, large tumour (more than 5 cm) and recurrent tumors (3). It is also recommended when surgery is not possible in marginal excision (2). Liu et. al reported one patient who received a full course of radiotherapy without surgery due to small lesion and old age at diagnosis with good prognosis reported (3).

Chemotherapy can be offered for aggressive histological subtypes such as rhabdomyosarcoma (2). Chemotherapy combination of vincristin, doxorubicin, cyclophosphamide/iphosphamide, mesna, and etoposide has been used (2). However, it has been found that effect of chemotherapy did not influence the survival (2).

There is lack of evidence on patterns of recurrence, however, late recurrence can occur in slow-growing tumour. Hence, it is recommended to follow-up patients every three to six months for at least five years (1). Apart from laryngoscopic examination during follow-up, MRI surveillance once a year can also be opted to monitor recurrence (2). No survival data from a large series is available at present due to rarity of LS (2). In a review of ten cases, the five-year overall survival (OS) and disease-specific survival (DSS) were 76% and 90%, and ten-year OS and DSS were 57% and 90% (3). Two patients had local recurrence and distant metastasis to lung after eight months and 20 months after surgery (3).

CONCLUSION

Laryngeal sarcoma is rare and there is paucity of data on behaviour, management and outcome. Diagnosis requires comprehensive histopathological and immunohistochemical examination. Multidisciplinary approach is important in management. Complete tumour excision with clear margins with laryngeal preservation is the cornerstone of treatment. Elective neck dissection is not necessary when there is no evidence of nodal metastasis. Radiotherapy is advocated in selected cases to achieve loco-regional control.

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