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Adult Laryngeal Rhabdomyosarcoma: is aggressive treatment justified in all cases? A case report and review of the literature

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Abstract

Background: Adult laryngeal rhabdomyosarcomas are rare tumours commonly treated by laryngectomy.

Case report: We present a case of subglottic laryngeal rhabdomyosarcoma in an elderly woman, treated by endoscopic resection.

Conclusion: Despite the fact that this tumour is traditionally treated aggressively, this approach is not supported by the literature. Due to the varying biological behaviour of this tumour in adults, we believe that conservative surgical procedures or combination therapies should be preferred, rather than total laryngectomy.

Key words: Rhabdomyosarcoma; Larynx; Adult

Introduction

Rhabdomyosarcomas represent 40 per cent of all head and neck sarcomas.[1] Although they are the most common soft tissue sarcomas of childhood (with an annual incidence of 4–7 per million children), they rarely appear in adults (being the third most common sarcoma in this age group).[2–4] The majority of head and neck rhabdomyosarcomas occur in the orbit, nasopharynx, nasal cavity and paranasal sinuses.[4] Laryngeal involvement occurs in only 3 per cent of head and neck rhabdomyosarcomas.[5]

We present a case of subglottic laryngeal rhabdomyosarcoma in an elderly woman, which was treated by endoscopic resection.

The histological classification of rhabdomyosarcomas is controversial. Most authors accept the system proposed by Horn and Enterline.[6] According to this system, there are three major histological rhabdomyosarcoma subtypes: embryonal, alveolar and pleomorphic.

Case report

An 83-year-old woman presented to our department with a three-month history of progressive dyspnoea. She reported that a subglottic lesion had been removed under general anaesthesia seven years previously, and had been reported as a haemangioma.

On physical examination, the patient had tachypnoea, stridor and intercostal recession.

Fibre-optic laryngoscopy showed a large, exophytic mass arising from the subglottis.

The patient underwent an emergency tracheostomy under local anaesthesia; however, due to a severe arrhythmia during surgery, biopsies were not performed at the time.

Subsequent high resolution computed tomography (CT) scanning showed a large, subglottic mass with 95 per cent obstruction of the lumen. Chest radiography and CT scans of the thorax and abdomen were normal.

Direct microlaryngoscopy was performed three months later, as the patient suffered a stroke three weeks following her tracheostomy. It was possible to fully excise the lesion macroscopically, endoscopically as well as through the tracheostomy.

The patient recovered well and there were no post-operative complications.

Histopathological examination revealed a laryngeal pleomorphic rhabdomyosarcoma. Grossly, the tumour mass was whitish and firm with areas of haemorrhage, and measured $4 \times 2.5 \times 1$ cm. Microscopically, the tumour was diffuse and cellular without any distinct morphological pattern. Ulceration was found on the surface epithelium. In the biopsy specimen, there was a variation in the cellularity, with alternating hyper- and hypocellular areas; the latter were associated with a myxoid stroma. The cellular components consisted of round cells (resembling lymphocytes) with dark nuclei, and also larger oval- to spindle-shaped cells with eccentric, eosinophilic cytoplasm. Mitotic figures and necrosis were easily assessed. Immunoperoxidase stains were positive for desmin, vimentin and myoglobulin and negative for S100 protein, cytokeratin (CAM 5.2), smooth muscle antigen, Leucocyte common antigen, carcinoembryonic antigen and cluster of differentiation 68 glycoprotein (Figure 1).

Due to the patient's age and physical condition, no further treatment was pursued. The patient was followed up regularly every three months, and eventually died 16 months after her operation from a second stroke, free of disease.

Discussion

Rhabdomyosarcoma is the most common soft tissue neoplasm of the head and neck in children, and usually appears during the first decade of life.[2] It is more rarely found in adults (being the third most common soft tissue sarcoma in this age group). Although the most common site of presentation is the head and neck region (approximately 35 per cent), laryngeal rhabdomyosarcoma is extremely rare, and institutional experience is thus quite limited, with only a few published reports.[4] Males tend to be affected more than females.[7] Only two female cases have

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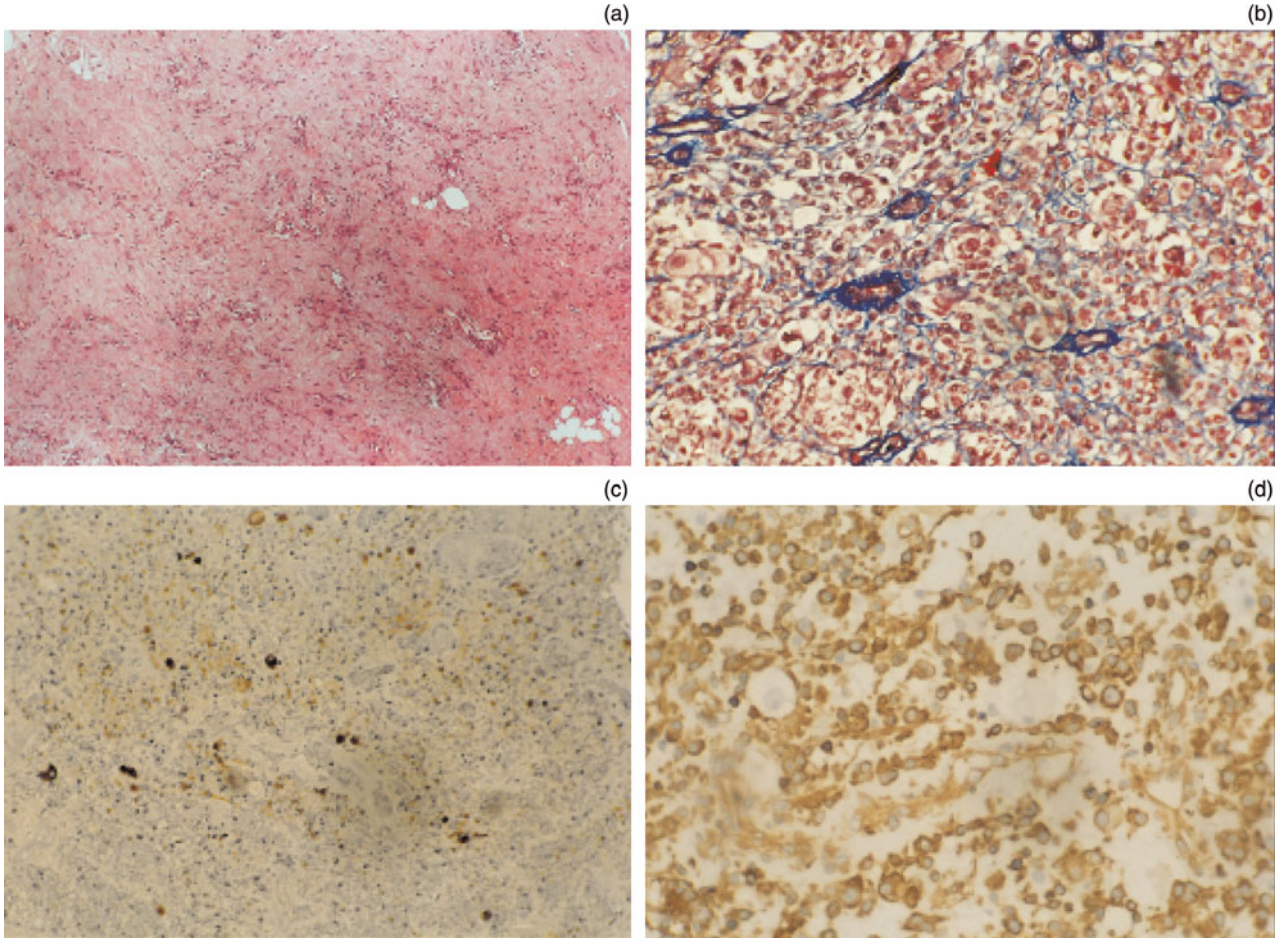

FIG. 1

Photomicrographs: (a) H&E, ×4; (b) Masson trichrome staining, ×10; (c) desmin immunoreactivity staining, ×20; (d) vimentin immunohistochemical staining, ×20.

been reported in the literature, including the present case (see Table 1).

The most widely accepted classification for rhabdomyosarcomas is that of Horn and Enterline.[1,6] This system classifies rhabdomyosarcomas into three basic histological types: embryonal, alveolar and pleomorphic. Botryoid rhabdomyosarcoma is considered to be a subtype of embryonal rhabdomyosarcoma.[1,26] Newer, rare histological types such as spindle cell and sclerosing rhabdomyosarcoma have been reported in both children and adults.[26] Embryonal rhabdomyosarcoma tends to appear mainly in infants and young children, while the alveolar type is more common in young adults.[27–30] Pleomorphic rhabdomyosarcoma is usually found in older adults, although an overlap often occurs.[13,31]

The differential diagnosis of rhabdomyosarcoma includes lymphoma, fibrosarcoma, melanoma, neuroblastoma, retinoblastoma, haemangioendothelioma and granular cell myoblastoma.[1,24,26] Special stains and immunohistochemical markers are necessary to establish a histological diagnosis.[1,23]

The symptomatology of laryngeal rhabdomyosarcomas is usually quite similar to that of other laryngeal tumours.[7,23] The anatomical position of such tumours, along with their rapid growth, usually result in a more advanced clinical appearance than the more commonly observed laryngeal squamous cell carcinoma.[23] The most common signs and symptoms of laryngeal rhabdomyosarcoma include hoarseness of voice, dyspnoea, stridor and dysphagia, while pain and weight loss are relatively rare, usually indicating advanced disease.[7,23,24]

Computed tomography and MRI usually provide valuable information about tumour size and location, nerve or vascular invasion, and bony erosion.[7,24,26] Computed tomography is often preferred when information about laryngeal cartilage erosion is needed.

Surgical excision appears to be the treatment of choice for laryngeal rhabdomyosarcoma.[1] Of the reported cases in adults, only two patients were not treated surgically. Emergency tracheostomy was performed in some cases because the airway was at grave risk by the time of admission. Most of the remaining cases underwent total laryngectomy. However, according to Da Mosto *et al.*, conservative laryngectomy should be employed if the tumour can be adequately removed.[1] Conservative procedures such as partial laryngectomy, cordectomy or tumour debulking are supported by some authors when the tumour is resectable, mainly because these laryngeal lesions tend to be less aggressive than other head and neck rhabdomyosarcomas, and it is believed that the laryngeal cartilages may act as anatomical barriers to tumour expansion.[12,23,24,32]

In our patient, emergency tracheostomy was performed in order to secure the patient's airway, and the tumour was macroscopically removed at a second stage. No other therapeutic procedure was involved, and the patient survived for 16 months following her operation, subsequently dying from an unrelated cause with no evidence of recurrence. Her survival time is comparable to that of other patients treated with nonconservative methods. Although some authors suggest that a radical excision is preferable in such lesions, a large number of surgical cases would be

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TABLE I

REPORTED ADULT CASES OF LARYNGEAL RHABDOMYOSARCOMA

Case no	Study	Pt sex, age (yrs)	Histological type	Site	Treatment	Follow up	Pt status by end of follow up
1	Filipo & Crifo[8]	M, 53	Pleomorphic	False vocal fold, laryngeal sinus	Total laryngectomy, RT	8 mths	Alive & disease-free
2	Rodriquez & Ziskind[9]	M, 57	Pleomorphic	R true vocal fold	Total laryngectomy	NS	NS
3	Grouls & Bechtelsheimer[10]	M, 67	Pleomorphic	L vocal fold	RT	3 wks	Dead (cardiogenic shock)
4	Hall-Jones[11]	M, 54	Embryonal (botryoid)	Post vestibular wall of larynx	Laryngectomy	2 yrs	Alive & disease-free for 16 mths
5	Frugoni & Ferlito[12]	M, 33	Pleomorphic	L true & false vocal folds	Total laryngectomy, RT	6 yrs	Alive & disease-free
6	Aleksandar *et al.*[13]	M, 60	Cross-striations seen; exact type NS	Glottic	Tracheostomy, pt refused laryngectomy	NS	
7	Lamendola & Buonocore[14]	M, 61	Pleomorphic	R true vocal fold	Total laryngectomy, bilat neck dissection	1 yr	Alive & disease-free
8	Marasso *et al.*[15]	M, 65	Pleomorphic	R false vocal fold, subglottic extension	RT, CT	NS	
9	Winter & Lorentzen[16]	M, 72	Pleomorphic	L true vocal fold	Partial laryngectomy	1 yr	Alive & disease-free
10	Franz[17]	M, 57	Pleomorphic	L true vocal fold	RT	2 yrs	Alive & disease-free
11	Srinivasan & Talvalkar[18]	M, 55	Pleomorphic	L arytenoids, L aryepiglottic fold & pyriform fossa	Laryngectomy planned	Died pre-op (airway obstruction)	Dead
12	Kleinsasser & Glanz[19]	M, 45	Mixed type (pleomorphic-embryonal)	Left vocal fold	Cordectomy	7 yrs	Alive & disease-free
13	Haerr *et al.*[20]	M, 62	Alveolar	Interarytenoid notch	Total laryngectomy, RT, CT	5 mths	Dead
14	De Agostino *et al.*[21]	M, 70	Pleomorphic	L vocal cord, ant commissure	Total laryngectomy	NS	
15	Jahnke & Vogl[22]	M, 45	Pleomorphic	R vocal fold	Total pharyngectomy, cervical oesophageal-tracheal resection, R thyroidectomy, CT, RT	NS	
16	Da Mosto *et al.*[1]	M, 69	Pleomorphic	R true vocal fold, subglottic extension	Total laryngectomy, thyroidectomy, RT	2 yrs	Alive & disease-free
17	Ruske *et al.*[23]	F, 66	Pleomorphic	L arytenoid region	Laryngectomy, RT	30 mths	Alive & disease-free
18	Akyol *et al.*[24]	M, 68	Pleomorphic	Transglottic tumour, invasion of extralaryngeal structures	Total laryngectomy, R modified neck dissection, RT	8 mths	Dead
19	Abali *et al.*[25]	M, 45	NS	Subglottic (metastatic invasion)	RT (palliative)	Approx 22–24 mths	Alive
20	Dikbas *et al.*[7]	M, 28	Embryonal	R ventricle, R vocal fold, extralaryngeal & subglottic extension	Tracheostomy, CT, RT	22 mths	Alive
21	Present case	F, 83	Pleomorphic	Subglottic	Emergency tracheostomy, endoscopic tumour excision	16 mths	Dead (disease-free)

No = number; pt = patient; yrs = years; mths = months; wks = weeks; M = male; F = female; R = right; L = left; post = posterior; ant = anterior; RT = radiotherapy; CT = chemotherapy; bilat = bilateral; NS = not specified; pre-op = pre-operatively; approx = approximately

needed in order to determine whether prognosis is related to the extent of tumour removal. Neck dissection is not usually suggested, as rhabdomyosarcomas do not generally metastasise to cervical lymph nodes.[1,4] Cervical lymph node infiltration has been reported in only one case (case 14, Table I). Distant metastasis of laryngeal rhabdomyosarcoma has not been reported.[1]

The role of radiotherapy in the management of laryngeal rhabdomyosarcoma remains controversial. Radiotherapy has been used as primary and adjuvant therapy in several cases.[23] The results were variable and difficult to interpret, as radiotherapy often followed radical excision of the tumour. Radiotherapy was adopted as the only therapeutic procedure in two reported cases. Grouls and Bechtelsheimer reported that their patient died of cardiogenic shock three weeks later, while Franz's patient was alive and disease-free after two years.[10,17]

Chemotherapy has been involved in four cases of laryngeal rhabdomyosarcoma (see Table I). A number of chemotherapeutic agents appear to be effective against rhabdomyosarcoma, including vincristine, cyclophosphamide and actinomycin D.[2] It has been suggested that rhabdomyosarcoma is the only tissue sarcoma for which chemotherapy may have a significant adjunctive role when used alongside both surgery and radiotherapy.[20,33] Triple therapy seems to have encouraging results in embryonal and botryoid laryngeal rhabdomyosarcomas.[1] The efficacy of chemotherapy can be augmented by concurrent use of several agents (i.e. combination chemotherapy).[2]

Conclusion

Rhabdomyosarcomas are soft tissue neoplasms which rarely involve the adult larynx. It is not uncommon for airway obstruction to occur quite early in such patients, requiring immediate airway intervention. Despite the fact that these tumours are traditionally treated aggressively, this approach is not supported by the literature, which remains scarce. Due to the variable biological behaviour of this tumour in adults, we believe that conservative surgical procedures or combination therapy should be considered, rather than total laryngectomy, depending on the tumour size and location and the patient's age and physical condition.

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