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Laryngeal rhabdomyosarcoma in adults

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Abstract
Laryngeal rhabdomyosarcoma is a rare disease. Only nine of these tumours have been adequately described in world literature in the adult population. Adult patients with laryngeal rhabdomyosarcoma often present at a later stage than other laryngeal tumours, including squamous cell carcinoma. Diagnosis is made by identification of cross-striations histochemically or cytoplasmic myoglobin by immunohistological methods. We present a 66-year-old woman with pleomorphic rhabdomyosarcoma of her larynx. This is the first female in the adult age group to be presented. Surgical treatment with adjuvant radiotherapy is currently the treatment of choice for this disease. We provide a review of the literature on laryngeal rhabdomyosarcoma, including presentation, pathology and management of this rare disease.

Key words: Laryngeal neoplasms; Rhabdomyosarcoma

Introduction

Rhabdomyosarcoma is a malignant tumour of striated muscle origin. These often resemble muscle cells found in seven- to 10-day-old embryos and occur in children with a higher frequency than adults. Rhabdomyosarcomata are more rarely found in the adult population.

Malignant laryngeal tumours are usually of squamous cell origin. Sarcoma of the larynx is rare, occurring in fewer than one per cent of cases (Cady *et al.*, 1968). In one study, 214 cases of head and neck sarcoma were studied (Wanebo *et al.*, 1992) and angiosarcoma, fibrosarcoma and malignant fibrous histiocytoma were the most commonly found lesions; rhabdomyosarcoma (11/214) and embryonal rhabdomyosarcoma (four out of 214) were the least common in this group.

Rhabdomyosarcoma of the larynx is extremely rare. Sixty per cent of all rhabdomyosarcomata in the head and neck occur in the orbit, nasopharynx and the nose (Masson and Soule, 1965). Of 2500 laryngeal tumours studied between 1940 and 1965, only three of 31 sarcomas found were rhabdomyosarcoma (Cady *et al.*, 1968). Similarly, Gorenstein *et al.* (1980), reviewed 3100 cases of laryngeal tumour and found 17 sarcoma, of which three were rhabdomyosarcoma. In studies of rhabdomyosarcoma, laryngeal involvement was reported in 2.3 per cent and 3.6 per cent of cases in respective studies (Masson and Soule, 1965; Feldman, 1982).

We present a case of laryngeal rhabdomyosarcoma in an adult female.

Case report

In May of 1995, a 66-year-old woman was referred with a two-month history of hoarseness of voice and dyspnoea on exertion. In the past she had had tuberculosis treated at the age of 14. She had previously smoked 10 cigarettes a day for 30 years but had ceased smoking 16 years before presentation. Examination revealed mild stridor but no cervical lymphadenopathy. Indirect laryngoscopy showed a tumour of the left arytenoid region. Microlaryngoscopy and biopsies were performed along with debulking of the left transglottic tumour. Histological examination revealed pleomorphic rhabdomyosarcoma containing necrotic, malignant tumour with spindle cells and globoid cells.

Immunoperoxidase stains were positive for vimentin, desmin and actin. Although light or electron microscopy did not demonstrate Z bands, cytoplasmic filaments were seen in tumour cells with the electron microscope.

Magnetic resonance imaging (MRI) of the larynx revealed a low signal area of abnormal tissue on T1-weighted scans in the left side of the larynx. There was slight enhancement following gadolinium contrast. The tumour measured 2 × 3 cm. The lesion was confined to the larynx. There was no lymphadenopathy seen. The right lobe of the thyroid was prominent and extended almost to the level of the larynx superiorly. This was of unknown significance.

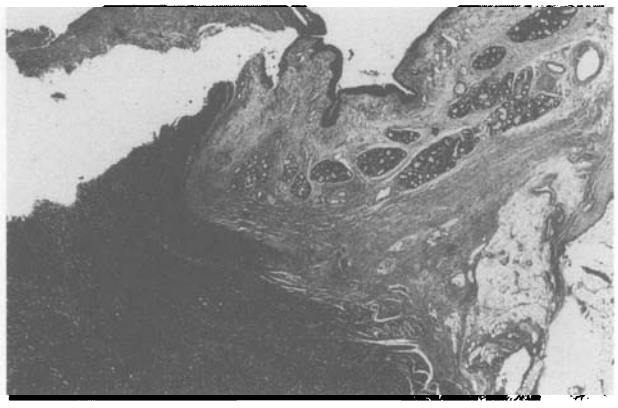

Fig. 1
Low power view of tumour showing ulceration with deep invasion of laryngeal stroma.

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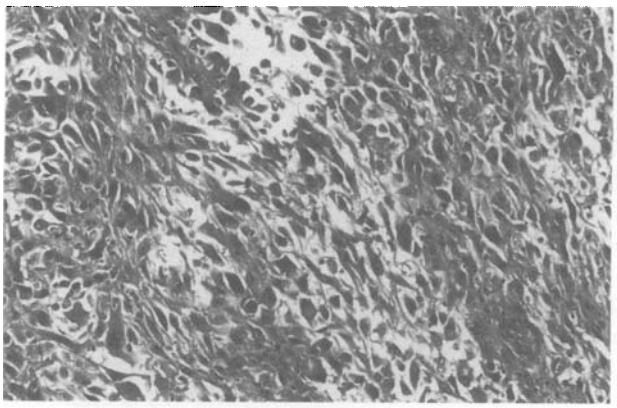

FIG. 2

High power views showing the marked degree of pleomorphism of the tumour cells and high mitotic rate. Immunocytochemistry showed positivity for vimentin, desmin, actin and negativity for cytokeratin (CAM 5.2, MNF 116), s-100, HMB 45 and LCA.

Nine days following presentation total laryngectomy was performed. The tumour was confined to the left glottis macroscopically and this was also confirmed histologically. Bilateral subtotal thyroidectomy was also performed. Two masses were palpable in the right lobe of the thyroid. These were shown to be colloid nodules on histology.

Post-operative recovery was uneventful and radiotherapy was commenced approximately four weeks following surgery. A laryngeal wedge shaped field of 9×8.5 cm received 5500 cGys in 25 fractions. The anterior oblique neck with a field size of 5×12.5 cm also received 5500 cGys in 25 fractions. After treatment for one month a smaller laryngeal field size of 5×6 cm received 1000 cGys in five fractions over a seven-day period. Following this a smaller anterior neck field of 4×4 cm received 540 cGys in three fractions over a four-day period. This made a total of 7040 cGys to the tumour bed. This high dose of radiotherapy created significant morbidity and steroids, as well as opiate analgesia, were used for palliation of these side-effects.

Following radiotherapy, stomaplasty and secondary tracheo-oesophageal fistula formation was performed. The stomaplasty was as a result of narrowing of the stoma due to radiotherapy. Follow-up after this revealed significant hypothyroidism with TSH levels greater than 700. Thyroid replacement was commenced. Follow-up after 30 months showed no recurrence of tumour and the patient is well and has good use of her speech valve.

Discussion

Rhabdomyosarcoma of the larynx is a very rare disease. Documentation of rare diseases such as this are very important for our future understanding of the nature and management of such diseases. Many rhabdomyosarcomas of the larynx have not been well documented in the past with either micrographs of histological features or complete descriptions of clinical and/or radiographical findings. As with Batsakis and Fox (1970) and Haerr *et al.* (1987), we have elected to include only well-documented cases of rhabdomyosarcoma in this review. The limited number of cases does not allow a thorough understanding of the disease.

TABLE I

ADULT CASES OF LARYNGEAL RHABDOMYOSARCOMA

Ref. No.	Age	Sex	Site	Histology	Symptoms	Duration	Treatment	Follow-up
Aleksander *et al.* (1976)	60	M	Glottic	Cross-striations seen	Acute respiratory distress Hoarseness	?	Tracheotomy Patient refused laryngectomy	?
Rodriquez and Ziskind (1970)	57	M	Right vocal fold	Rhabdomyosarcoma	Acute respiratory distress Weight loss Hoarseness	?	Laryngectomy	—
Hall-Jones (1975)	54	M	Posterior vestibular wall of larynx	Embryonal (botryoid) Cross-striations seen	Acute respiratory distress Dysphagia Hoarseness	2 years	Laryngectomy	16 months No recurrence
Haerr *et al.* (1987)	62	M	Below interaryntenoid notch	Alveolar rhabdomyosarcomas	Dysphagia Cervical lymphadenopathy	1 week	Laryngectomy Left radical neck dissection Modified right neck dissection Chemotherapy Radiotherapy	Recurrence within 1 month Neck nodes Died 3–4 months Abdominal metastases on autopsy
Winther and Lorentzen (1978)	72	M	Left vocal fold	Pleomorphic rhabdomyosarcoma Cross-striations seen	Hoarseness	1–2 months	Partial laryngectomy	Died
Frugoni and Ferlito (1976)	33	M	Left laryngeal vestibule False fold Left vocal fold	Pleomorphic rhabdomyosarcoma Cross-striations seen with PTAH stain	Hoarseness	3 months	Laryngectomy Radiotherapy	No recurrence 6 years
Srinivasan and Talvalkar (1979)	55	M	Left arytenoid Aryepiglottic fold Left piriform fossa	Pleomorphic rhabdomyosarcoma Cross-striations seen Also infiltrating SCC	Hoarseness Dysphagia	?	Laryngectomy planned	Died pre-op, secondary to acute laryngeal obstruction
Da Mosto *et al.* (1996)	69	M	Right true vocal fold	Pleomorphic rhabdomyosarcoma	Dysphonia	7 months	Laryngectomy Radiotherapy	No recurrence 2 years
Filipo*	53	M	Left false fold	Pleomorphic rhabdomyosarcoma	?	?	Laryngectomy Radiotherapy	No recurrence 8 months
Ruske (present case)	66	F	Left arytenoid region	Pleomorphic rhabdomyosarcoma	Hoarseness Shortness of breath Stridor	2 months	Laryngectomy Radiotherapy	No recurrence 1 year

*Cited Da Mosto *et al.* (1996)

Presentation of rhabdomyosarcoma in the larynx is often with more advanced symptoms than the more common squamous cell carcinoma of the larynx. Reasons for this may include the fast-growing nature of the rhabdomyosarcoma or the position of rhabdomyosarcoma in the larynx. In presented cases over the age of 20 years, half had symptoms of airway compromise such as shortness of breath, stridor or respiratory distress. Hoarseness (eight), dysphagia (three), weight loss (one), and pain (one) were other presenting symptoms.

In adults, males tended to be affected more than females. We have presented the first well-documented case of rhabdomyosarcoma of the larynx in an adult woman.

The histological appearance of rhabdomyosarcoma resembles embryological striated muscle more than adult striated muscle (Stobbe and Dargeon, 1950). Since then Horn and Enterline (1958), have suggested four subtypes for the histological classification of rhabdomyosarcoma. These include pleomorphic, embryonal, alveolar, and botryoid. Alveolar and botryoid may be subtypes of embryonal rhabdomyosarcoma. In general the younger population fall into the embryonal subtype and the older population into the pleomorphic subtype. The case presented above falls into the pleomorphic rhabdomyosarcoma classification.

Surgery–laryngectomy, is the mainstay of treatment at the present time, although radiotherapy has been used as both primary and adjuvant therapy for rhabdomyosarcoma. Success has been varied and dosages given are large, resulting in high morbidity levels. This was evident in our case. Our case received 7040 cGys and had significant radiotherapy morbidity.

Rhabdomyosarcomata of the larynx tend to be less aggressive than rhabdomyosarcomata elsewhere in the head and neck (Canalis *et al.*, 1976; Frugoni and Ferlito, 1976). Cartilaginous borders in the larynx may also restrict spread of this tumour locally and result in a better prognosis. Despite this observation the outlook for rhabdomyosarcoma of the larynx remains bleak. Survival for rhabdomyosarcoma in other sites with, or without, treatment has been quoted as 50 per cent mortality one year post-presentation and 70 per cent within two years (Masson and Soule, 1965). The 66-year-old woman presented is alive and well with no local or regional recurrence after 30 months.

Conclusion

A case of rhabdomyosarcoma of the larynx has been described. A review of adult rhabdomyosarcomata of the larynx reveals this is the first female over the age of 20 to be adequately reported with this disease. This patient's tumour was of the pleomorphic type on histological examination. Only 10 cases of adult rhabdomyosarcoma of the larynx have been adequately reported. At present it seems that a combination of surgery and radiotherapy would be the treatment of choice. Chemotherapy has been added to very advanced tumours with little success and to the paediatric population with some degree of success.

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