CASE REPORT

Extraskkeletal osteosarcoma of the larynx: an extremely unusual tumour

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SUMMARY

Osteosarcoma of the larynx is probably the rarest mesenchymal tumour of the larynx, with only 16 cases reported so far. The majority of them occur in males between the sixth and eighth decades of life. Patients usually present with non-specific symptoms such as dysphonia and upper airway compromise. The most common site of distant metastasis is the lung. Clinically, the tumour follows an aggressive course and is associated with high mortality. The case we present is unusual as it occurred at a young age (38 years) as compared with the norm and it did not arise from the endolarynx, unlike many of the other cases. This was the only known case where a Pearson near-total laryngectomy was performed whereby the patient's natural speech mechanism was preserved. This surgery was possible because the contralateral half of the larynx was clearly disease free and the interarytenoid region was uninvolved. The patient underwent postoperative adjuvant external beam radiotherapy beginning 4 weeks after surgery. The patient is doing well after 15 months of follow-up and shows no signs of recurrence.

BACKGROUND

Mesenchymal tumours of the larynx are extremely rare and constitute less than 2% of laryngeal tumours.^{1 2} Chondrosarcoma and fibrosarcoma are the commonest laryngeal sarcomas. With only 16 reported cases, osteosarcoma of the larynx is probably the rarest mesenchymal tumour of the larynx.^{1 2} This is the first reported case in India.

CASE PRESENTATION

A 38-year-old man with a 3-pack year history of smoking presented with a 2-month history of hoarseness of voice. There was no history of radiation exposure. His medical and family history was unremarkable. On physical examination, a hard mobile mass approximately 4×3 cm was found on the right side of the neck which moved with deglutition.

INVESTIGATIONS

A fiberoptic laryngoscopy revealed a bulge on the lateral wall of the right pyriform fossa, which appeared obliterated. The left vocal cord mobility was normal, the right was sluggish. There was no apparent mucosal abnormality. The subglottis and trachea appeared normal.

A contrast-enhanced CT (CECT) scan demonstrated an expansile lytic lesion measuring about 4.5 cm in the axial plane ([figure 1](#)) and about 5 cm in the sagittal plane ([figure 2](#)) involving the thyroid cartilage with resultant mass effect over the

Figure 1 CT of the neck (axial views) showing Expansile, lytic lesion involving the thyroid cartilage. Mass effect over laryngeal passage, supraglottis and glottic region with narrowing of the laryngeal passage. Right pyriform fossa appears effaced.

laryngeal passage, and glottic and supraglottic region with effacement of the right pyriform fossa. However, no definite loss of fat plane was observed between the lesion and underlying muscles and soft tissue. Compression effect was also noted over the right vocal cord. The epiglottis, hypopharynx, the visualised part of the thyroid gland and bilateral carotid vessels appeared normal. The hyoid bone

Figure 2 CT of the neck (sagittal view) showing Expansile, lytic lesion involving the thyroid cartilage. Mass effect over laryngeal passage, supraglottis and glottic region with narrowing of the laryngeal passage. Right pyriform fossa appears effaced.

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Figure 3 The tumour has been excised. Note the preserved left arytenoid and posterior two-thirds of the left vocal cord.

and other laryngeal cartilages showed no signs of erosion. CECT of the thorax and the abdomen showed no pulmonary or liver metastasis. There was no cervical lymphadenopathy on imaging or on clinical examination.

DIFFERENTIAL DIAGNOSIS

- ▶ Laryngeal soft tissue sarcoma
- ▶ Laryngeal carcinoma

TREATMENT

A near-total laryngectomy was performed on this patient. The operative procedure was performed under general anaesthesia. The patient was nasotracheally intubated. His skin was opened by a transverse incision, incorporating the scar of a previous incisional biopsy. The strap muscles were divided and the laryngeal skeleton was exposed. The patient was tracheostomised at the third tracheal ring. Normally, laryngeal entry is initially obtained by a transvallecular incision, which allows for the inspection of the disease, but in this case it was already known through fiberoptic laryngoscopy that the disease did not involve the endolarynx and the contralateral larynx was clear of the disease. The thyroid cartilage was divided preserving the posterior lamina at the insertion of pharyngeal muscles, without disturbing the inferior cricothyroid joint to prevent damage to the recurrent laryngeal nerve. The tumour was removed. Contralateral arytenoids, two-thirds of the vocal cord and recurrent laryngeal nerve were preserved ([figure 3](#)).

Creation of shunt

The first tracheal cartilage was excised in the midline by removing a wedge. A 12 French catheter was placed from the

Figure 4 Creation of the shunt. A Foley's 12F catheter passed through the shunt from the tracheostome to the neopharynx.

Figure 5 Mass on the lateral aspect of the larynx (right side). Cut section of the mass shows haemorrhagic, granular whitish areas and bony tissue. Mass is well demarcated from the surrounding tissue by a bony capsule. The mass did not involve the hyoid bone grossly. There was no apparent invasion of the mucosa of endolarynx.

interarytenoid region to the tracheostome. A shunt was sutured over the catheter ([figure 4](#)).

Mucosal edges were brought together at the level of true vocal cord. The thyroarytenoid was sutured to the interarytenoids. This creates a shunt that remains closed during swallowing and opens during phonation.

A level 2 to level 4 lymph node clearance was performed.

The postoperative course was uneventful. The patient received an oral diet on the third postoperative day. The shunt catheter was removed on day 7. The nasogastric tube was removed on day 10. Full diet was established on day 14. Voice training and vocal rehabilitation began after day 14.

Gross examination of the resected near-total specimen showed a mass of 6×4 cm involving the soft tissue plane on the lateral aspect of the larynx ([figures 5 and 6](#)). The mass was covered with intact fascia and was present on the right hemiglottis on the laryngeal aspect with no mucosal involvement. On serial sectioning, the mass was seen centred in the soft tissue plane near the hyoid bone without involving it. It was sharply demarcated from the surrounding tissue by a bony shell. The cut surface of the mass showed many haemorrhagic areas,

Figure 6 Mass on the lateral aspect of the larynx (right side). Cut section of the mass shows haemorrhagic, granular whitish areas and bony tissue. Mass is well demarcated from the surrounding tissue by a bony capsule. The mass did not involve the hyoid bone grossly. There was no apparent invasion of the mucosa of endolarynx.

Figure 7 Microscopic section: telengectatic, osteoblastic, chondroblastic and giant cell rich areas between osteoid tissue. There was no evidence of malignant or dysplastic epithelial cells.

granular whitish areas and bony tissue. A total of 28 lymph nodes were isolated from the left selective neck dissection specimen, a total of 10 from the central group of nodes and a total of 11 were isolated from the right selective neck dissection specimen. The specimen also included part of the parotid and two lymph nodes.

On microscopy, the section's variety of areas included telengectatic, osteoblastic, chondroblastic and giant cell rich areas with telengectatic forming the majority. In many areas, the tumour cells showed neoplastic chondroid tissue of poor differentiation and well-differentiated foci (figures 7–9). There was no evidence of malignant or dysplastic epithelial cells. A bony and soft tissue pseudocapsule limited the tumour with no extension beyond its confinement. Immunohistochemically, the tumour cells were positive for vimentin; however, these cells were negative for cytokeratin (AE1/AE3, CAM5.2) and epithelial membrane antigen. These features were consistent with a diagnosis of extraskeletal osteosarcoma. The diagnosis was extraskeletal osteosarcoma arising from the soft tissue involving the larynx.

The patient was advised to undergo postoperative adjuvant external beam radiotherapy beginning 4 weeks after surgery.

Figure 8 Microscopic section: telengectatic, osteoblastic, chondroblastic and giant cell rich areas between osteoid tissue. There was no evidence of malignant or dysplastic epithelial cells.

Figure 9 Microscopic section: telengectatic, osteoblastic, chondroblastic and giant cell rich areas between osteoid tissue. There was no evidence of malignant or dysplastic epithelial cells. The arrow shows cystic spaces.

OUTCOME AND FOLLOW-UP

The patient is doing well on 15 months of follow-up and shows no signs of recurrence or metastasis.

DISCUSSION

Extraskeletal osteosarcoma is an extremely rare tumour with fewer than 300 cases being reported in the literature.³ The important features of extraskeletal osteosarcoma are summarised in box 1.

Box 1 A brief review of extraskeletal osteosarcoma

- **Introduction**^{14 15}
 - Typically occurs in older adults: during fifth and sixth decades of life.
 - Males are affected more frequently than females: 1.9:1.
 - Predisposing factors: prior therapeutic irradiation at the site of the tumour and trauma.
 - Delayed diagnosis and poor prognosis (cause-specific survival rate at 5 years <25%).
- **Histopathology**^{14 15}
 - Large, 5–10 cm in size.
 - Well-circumscribed, grossly heterogeneous with areas of haemorrhage and/or necrosis.
 - Highly cellular, cytologically pleomorphic, mitotically active sarcomas: with neoplastic osteoid or bone.
 - Osteoblastic is commonest, followed by fibroblastic, chondroid, telengectatic, small cell and well-differentiated variants.
- **Immunohistochemistry**^{14 15}
 - Uniformly positive for vimentin
 - Smooth muscle actin (68%)
 - Desmin (25%)
 - S-100 protein (20%)
 - Epithelial membrane antigen (52%)
 - Keratin (8%)
- **Differential diagnosis**^{14 15}
 - Spindle cell sarcoma
 - Metastasis from a primary osteosarcoma
 - Carcinosarcoma
 - Malignant tumours with osseous metaplasia and sarcomas.

Only 16 cases of laryngeal osteosarcoma (LOS) have been reported in the literature.^{2–13} The majority of them occur in males between the sixth and eighth decades of life. LOS has been associated with previous radiotherapy in two cases.^{4 13} Patients usually present with non-specific symptoms such as dysphonia and upper airway compromise. Most symptoms display as polypoid soft tissue tumours of the endolarynx. LOS of the laryngeal skeleton most commonly arises from the cricoid cartilage. One case was accompanied by invasion of the anterior laryngeal cartilage, which was probably due to a previous thyrotoxicosis.⁵ The most common site of distant metastasis is the lung. Clinically, the tumour follows an aggressive course and is associated with high mortality.

Histologically, dysplastic stellate or spindled cells are seen; they produce a variable osteoid component, ranging from a delicate eosinophilic latticework pattern to a denser, well-formed osteoid matrix.

The general treatment guidelines of head and neck sarcomas are wide surgical excision wherever possible. Postoperative radiotherapy has been known to reduce recurrence in some cases. The role of chemotherapy has been inconsistent.

Owing to a very low number of reported cases, exact treatment guidelines are difficult to formulate. Complete surgical resection is indicated and total laryngectomy is the most commonly performed surgery and is indicated wherever possible. Postoperative radiotherapy could possibly reduce recurrence. The role of chemotherapy is controversial; however, one case received postoperative ifosfamide/epirubicin-based chemotherapy.

Learning points

- ▶ Head and neck sarcomas account for <1% of head and neck neoplasms, and they present with significant mortality and morbidity.
- ▶ Owing to the rarity of head and neck sarcomas in general and laryngeal osteosarcoma in particular, there is a paucity of management guidelines.
- ▶ Surgical excision is the treatment of choice, wherever possible.
- ▶ As the contralateral larynx was clearly uninvolved, the contralateral arytenoids, two-thirds of the vocal cord and recurrent laryngeal nerve could be preserved enabling preservation of the voice.
- ▶ Radiotherapy remains an important adjunct to reduce recurrence in high-grade tumours, tumours with positive surgical margins and tumours >5 cm in size.

The present case presented at a relatively younger age (38) than earlier reported cases of LOS. Our patient had a 3-pack year history of smoking. The tumour did not arise from the endolarynx, unlike many of the previously known cases. This was the only known case where a Pearson near-total laryngectomy was performed. This surgery helps in preserving the natural speech of the patient. This surgery was possible because the contralateral half of the larynx was clearly disease free and the interarytenoid region was uninvolved.

Contributors NRB wrote the manuscript and acted as a guarantor. GAK wrote and revised portions of the manuscript. RM assisted in drafting and revising the manuscript. RB was the operating surgeon. He also revised the article.

Competing interests None.

Patient consent Obtained.

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