Case series and reports

Solitary fibrous tumour of the supraglottic larynx

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SUMMARY

Solitary fibrous tumour (SFT) is a rare, benign, mesenchymal neoplasm that usually arises in the pleura, but rarely involves other sites outside the serosal space (mediastinum, lung, liver, thyroid gland); larynx involvement is very rare with only sporadic cases reported in the literature. We report a case of SFT in a 41-year-old woman with supraglottic laryngeal involvement; symptoms included dysphonia and mild odynophagia lasting 2 years, and fibre-optic laryngeal evaluation showed a sub-mucosal mass involving the left supraglottis and medial wall of the pyriform sinus. MRI represents the gold standard tool for differential diagnosis (with schwannoma, paraganglioma and haemangioma) and correct staging, while immunohistochemical and cytomorphologic analysis (bcl-2 and CD34 positivity in 90% of cases) is needed for definitive diagnosis. Surgery is the main treatment (endoscopic and open conservative technique), and its goal is a balance between safe oncological resection and good preservation of laryngeal functions; in this particular case an open laryngeal approach was scheduled due to the size of the tumour. Prognosis is good and in only a few cases (especially in pleural SFT) does the biological behaviour take a malignant course.

KEY WORDS: Solitary fibrous tumour • Larynx disease • Benign larynx neoplasm

Introduction

Solitary fibrous tumour (SFT), first described in 1931 by Klemperer and Rabin 1, is a benign mesenchymal neoplasm with a predilection for male gender (M/F 6:1) that usually arises in the pleura as a well-defined mass. It rarely involves other sites outside serosal space such as the mediastinum, lung, liver, thyroid gland, orbit and upper aero-digestive tract. SFT belongs to a tumour category group fraught with diagnostic uncertainty due to the association with haemangiopericytoma 2. Its diagnosis is not straightforward and is generally based on cytomorphologic, immunohistochemical and radiologic findings. Extra-pleural localization, upper aero-digestive tract and especially the larynx are rarely involved with only a few cases reported in the literature 3-11 19. Surgery represents the gold standard for treatment and is associated with good prognosis. Here, we report the rare case of a supraglottic laryngeal SFT in a female patient that was treated with a conservative open laryngeal approach.

Case report

A 41-year-old non-drinker and non-smoker woman was referred to our department for progressive dysphonia and mild odynophagia lasting 2 years. She denied any symptoms related to airway obstruction. Her clinical history was negative.
Physical examination did not reveal any neck masses and nothing relevant was observed in the oral cavity and oropharynx. Flexible fibre-optic hypopharyngeal-laryngeal examination showed a sub-mucosal mass involving the left supraglottis and medial wall of the pyriform sinus, covered by intact mucosa. The left vocal fold was normal in appearance and mobility (Fig. 1).

Magnetic resonance (MR) revealed a solid mass (37x22 mm) arising from the left paraglottic space at the level of the ventricle reaching caudally the conus elasticus. The cricoid cartilage appeared remodelled in its superior and medial aspect. In T2-weighted sequences, the lesion presented non-homogeneous contrast enhancement. No pathologic neck nodes were present (Fig. 2). Axial contrast enhanced computed tomography (CT) revealed low enhancement in both arterial and venous phases. The mass widened the thyroarytenoid space without cartilage infiltration (Fig. 3).

Surgery via an external approach was scheduled. Through a 5-cm transverse cervical neck incision performed along a skin crease at the level of crico-thyroid membrane, the left thyroid lamina was identified and its superior half was removed in order to fully expose the neoplasm occupying the left paraglottic space down to the superior aspect of the cricoid. The tumour shape was rounded.
and sharp with a tense-elastic consistence, covered by a homogeneous greyish-coloured avascular capsule. The mass was completely removed with previous identification and preservation of the recurrent laryngeal nerve. The cartilaginous defect was covered by infra-hyoid muscles (Fig. 4). A temporary tracheostomy was performed and removed after 2 days. Histopathologic examination revealed a solid neoplasm characterised by the presence of spindle cells in a hyalinised stroma; some myxoid spots were also present. Immunohistochemical analysis revealed positivity for CD34 and low positivity for Bcl-2.

Discussion

Solitary fibrous tumour (SFT), also known as benign localised mesothelioma, submesothelioma and subserosal fibroma, is a very uncommon benign neoplasm that arises from pleura and other serosal membranes. Although the mediastinum is the most frequently affected site, other localisation such as lung, urogenital tract, and orbit have been described. For these reasons, SFT has been divided in “extrapleural” and “extrathoracic” subgroups.

In general, “extrathoracic” SFT has a more indolent course, with a very low rate of malignant transformation and development of distant metastases in 6-10% of cases. In contrast, pleural lesions have a recurrence rate of 9-19% and are associated with a distant metastasis rate of up to 19%. “Extrathoracic” SFT are more commonly symptomatic and when located in the larynx, and they are always associated with long-standing unspecific symptoms like progressive hoarseness, foreign body sensation and phonaory changes. Laryngeal SFT is usually located in the supraglottis, where it appears at endoscopy as a swelling covered by normal mucosa; pure glottic localisations are extremely rare and only sporadic cases of larynx involvement are reported in English literature (Table I).

The lesion is hypothesised to originate from mesenchymal tissue, and in particular from myofibroblastic cells. From a histopathologic standpoint, SFT is characterised by the presence of spindle cells with headlong nuclei arranged in unspecific pattern with a collagenous background, the so-called “patternless-pattern”. Immunohistochemical analysis reveals positivity to vimentin, and in about 50% of cases for CD99 and Bcl-2 protein. Staining for cytokeratin, smooth muscle actin, desmin, S-100 protein and CEA is always negative; CD34, which is involved in proliferation of myofibroblastic cells, is positive in 90-95% of cases.

SFT is commonly characterised by a slow growth, without invasion of surrounding tissues, associated with mild and vague symptoms, even though cases with malignant transformation, local invasion, recurrence and distant metastases have been reported. This aggressive behaviour is more typically observed in large volume lesions, with a high number of mitoses/field, presence of necrosis or

| Table I. Literature review concerning SFT of the larynx (continues). |
|------------------|--------|--------|---------|---------|---------|---------|
| **Author**  | **Case 1** | **Case 2** | **Case 3** | **Case 4** | **Case 5** | **Case 6** |
| **Age**   | 41     | 13     | 60      | 29      | 71      | 65      |
| **Gender** | F      | M      | M       | M       | F       | F       |
| **Location** | Supraglottic/false VC | epiglottis | Ventricular fold | False VC | Epiglottis | Supraglottis |
| **Clinical presentation** | Dyspnoea/ dysphonia | Foreign body sensation | Laryngeal dyspnea | Hoarseness/foreign body sensation | Foreign body sensation | Hoarseness |
| **Symptoms duration (months)** | 24     | 1.5    | 20      | 6       | 6       | 12      |
| **Radiologic findings** | mass   | mass   | mass    | mass    | mass    | Polypoid mass |
| **Endoscopic findings** | Submucosal avascular sharp mass | Bulky, pedunculated mass | Occupying space mass | Bulky avascular mass | Smooth mass | n/r |
| **Tumour size (cm)** | 3.7    | 2.2    | 2.5     | 2.5     | 3.4     | 3.0     |
| **Treatment** | Open lateral thyrothomy, NED (24) alive | Lateral pharyngectomy, NED (12) alive | Vertical hemilaryngectomy, NED (14) dead | Laser resection, NED (18) alive | Lateral pharyngectomy, NED(36) alive | Partial laryngectomy, NED(6) alive |
| **Outcome (months)** | NED (24) | alive | NED (12) | alive | NED (14) | dead |

M: male; F: female; VC: vocal cord; n/r: not reported; NED: not evidence of disease
haemorrhages and nuclear atypias, although a benign evo-
lution is seen in 50% of SFTs showing these features. At MR, the lesion typically shows on T1-weighted se-
quences a signal that is isointense to muscle and variable on T2-weighted, with some areas with a slight enhance-
ment and other areas with iso-hypointensity. These find-
ings are probably due to the different histological ar-
rangement of the tumour, which is rich in collagen and
fibroblasts mixed with other areas where these compo-
nents are less represented.

In the present case, CT with contrast revealed a low en-
hancement during the arterial phase, which became in-
tense and heterogeneous in the interstitial phase, prob-
ably in view of the low presence of blood vessels. The
axial CT scan showed a well-defined neoplasm occupy-
ing the entire left paraglottic space with enlargement of
the tyro-arytenoid space, but with no signs of cartilage
infiltration.

Based on MR and CT features, differential diagnosis
includes other benign lesions such as haemangioma,
 schwannoma and paraganglioma. In children, hae-
mangioma is typically localised in the subglottic area,
whereas in adult patients the supraglottis is more fre-
quently affected; on MRI T1 weighted-images (WI) the
lesion shows a signal that is isointense to muscle (with
up to 30% of lesions showing high signal foci due to
haemorrhage), with a diffuse heterogeneous enhance-
ment, while on T2 WI shows a hyperintense signal com-
pared to muscle tissue and often shows poorly defined
margins; phlebitis (small round calcifications of the ve-
 nous vessels) are typically present. Schwannoma gen-
erally arise from the superior laryngeal nerve and tend
to dislocate the larynx without involving the laryngeal
intrinsic muscle or supraglottic folds. On CT, the
disease presents as well defined, hypodense submucosal
mass without signs of infiltrative or destructive growth;
in MRI, the lesion is isointense to slightly hyperintense
in T1WI with strong, inhomogeneous enhancement af-
ter gadolinium injection while in T2 WI appear hyper-
intense. Paraganglioma is a highly vascularised, non-
encapsulated lesion that rarely involves the supraglottis,
showing on MRI T1WI a typical “salt and pepper” ap-
pearance due to signal voids combined with high sig-
nal foci secondary to haemorrhages within the tumour,
while on T2-weighted sequences the signal of the mass
is superior to that of muscle.

Surgery is the mainstay of treatment and can be performed
with an endoscopic or open approach in relation to tu-
mour extension. An endoscopic technique is indicated for
patients with good laryngeal exposure and for lesions not
involving the pharyngeal constrictor muscle or without
significant extralaryngeal extension. Otherwise, an open
surgical technique is indicated for patients with sub-opti-
mal laryngeal exposure and for bulky lesions invading
the paraglottic space, with translaryngeal or extralaryngeal
extension. Tracheotomy can be planned for both types of

<table>
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<tr>
<th>Case 7</th>
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<td>Smooth avascular mass</td>
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<td>biopsy</td>
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<td>Outcome (months)</td>
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<td>n/r</td>
<td>NED (12) alive</td>
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</tbody>
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M: male; F: female; VC: vocal cord; n/r: not reported; NED: not evidence of disease
techniques and depends on the size and localisation of the mass and anatomical configuration of the upper aerodigestive tract of the patient.

The patient herein underwent an open approach with partial resection of the thyroid lamina and en-bloc resection of the tumour, combined with a temporary tracheotomy and nasal feeding tube (NFT) positioning. The indications for this approach were the considerable volume of the lesion and its crano-caudal transglottic extension into the paraglottic space reaching the superior border of the cricoid cartilage. In this case, an endoscopic technique was considered unsafe because of potential damage to the inferior laryngeal nerve, since the posterior extension of the tumour was close to the crico-arytenoid joint.

The post-operative course was uneventful, and the tracheotomy and NFT were removed on the 2nd and 4th post-operative days, respectively, and the patient was discharged the day after; endoscopic post-operative evaluation performed at 6 months showed normal motility of the vocal cords and complete healing of the laryngeal mucosa.

At 2-year follow-up, the patient is free of disease and clinically without any signs of recurrence; long-term clinical follow-up is required for possible rare risk of recurrence. In case of a large primary tumour (size > 10 cm), there is a possible association with a more aggressive behaviour of the tumour with metastatic spread within 6 months, although no cases of larynx involvement are described 16.

In conclusion, SFT is a rare benign mesenchymal tumour that very rarely occurs in the larynx. MR represents the first imaging tool, and the main goal of surgery is to ensure complete resection with no impact on upper aerodigestive tract function in terms of phonation and swallowing.

References