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CASE SERIES AND REPORTS

Solitary fibrous tumour of the supraglottic larynx

Tumore fibroso solitario della laringe sopraglottica

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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 invovlement; 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

RIASSUNTO

Il tumore fibroso solitario (SFT) è una neoplasia rara, benigna, di origine mesenchimale che generalmente origina nella pleura ma che raramente può coinvolgere altre sedi al di fuori degli spazi sierosi (mediastino, polmone, fegato, tiroide); il coinvolgimento laringeo è molto raro con solo pochi casi riportati in letteratura. Riportiamo un caso di SFT in una paziente di 41 anni con coinvolgimento della laringe sopraglottica. La sintomatologia è comparsa con disfonia e modesta odinofagia da 2 anni; L'esame fibrolaringoscopico ha evidenziato una massa sottomucosa con coinvolgimento della sovraglottide di sinistra e della parete mediale del seno piriforme. L'RMN rappresenta l'esame principale per escludere altre diagnosi (schwannoma, paragangliome ed emangioma) e per una corretta stadiazione mentre l'immunoistochimica e l'analisi citomorfologica (bcl-2 e CD34 positiva nel 90% dei casi) è la base per una diagnosi definitiva. La chirurgia (endoscopica o cielo aperto) è la prima scelta di trattamento e l'obbiettivo è un bilancio tra la radicalità oncologica e la funzione d'organo; nel caso riportato l'approccio è stato a cielo aperto per il volume della massa tumorale. La prognosi è buona e solo in alcuni casi (specialmente nei SFT pleurici) il comportamento biologico del tumore può essere di tipo maligno.

PAROLE CHIAVE: *Tumore fibroso solitario • Patologia laringea • Tumore benigno laringeo*

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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 laryngel 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.

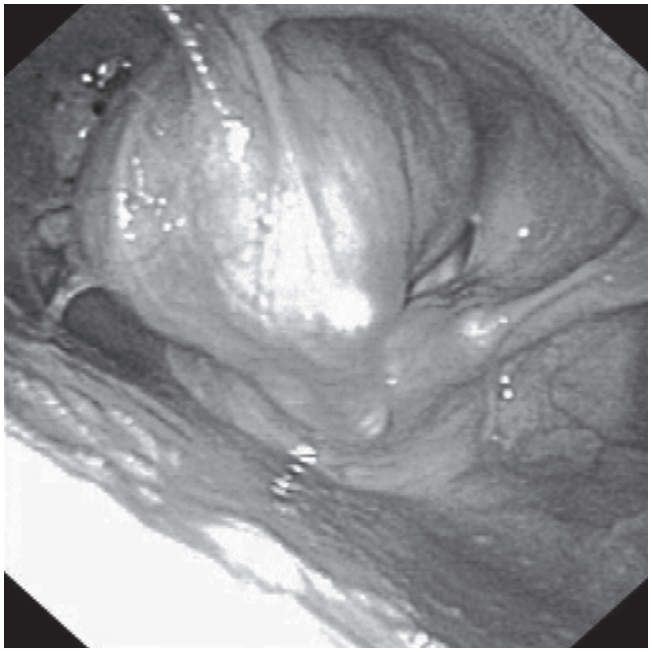

Fig. 1. Fibre-optic hypopharyngeal-laryngeal examination showing a sub-mucosal mass involving the left supraglottis and medial wall of the pyriform sinus.

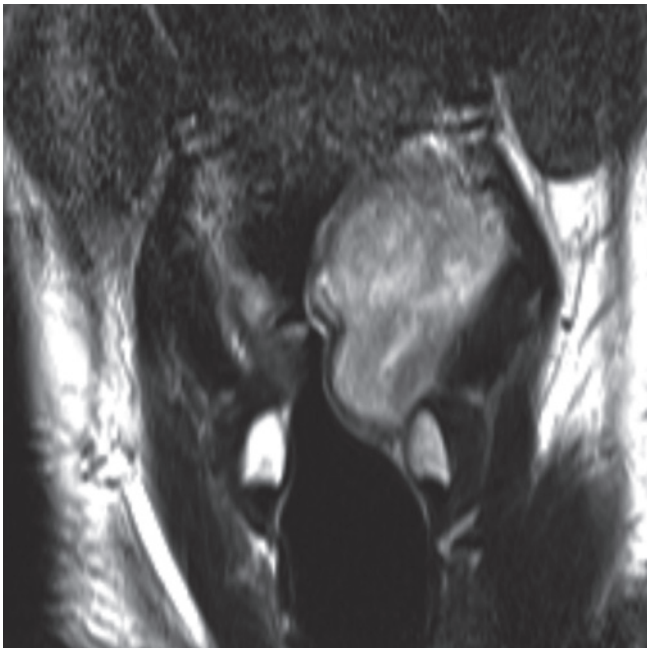

Fig. 2. MRI coronal view showing a solid mass (37x22 mm) arising from the left paraglottic space at the level of the ventricle reaching caudally the conus elasticus; cricoid cartilage is remodelled in its superior and medial aspects.

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 centred at the level of the ventricle, with caudal spread reaching 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 thyroaritenoid 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

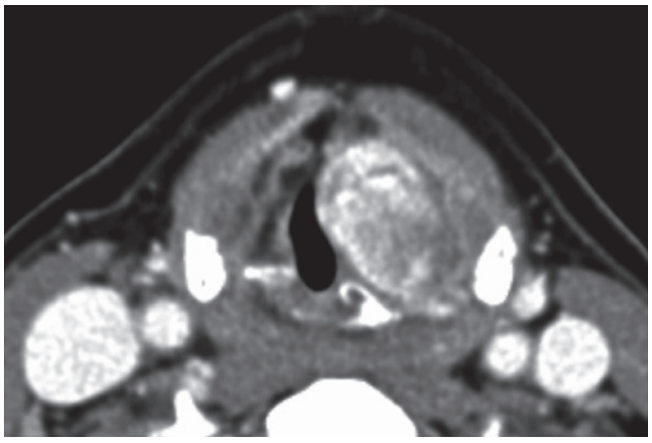

Fig. 3. CT-scan with contrast axial view showing low enhancement in both arterial and venous phases. The mass widens the thyroarytenoid space without cartilage infiltration.

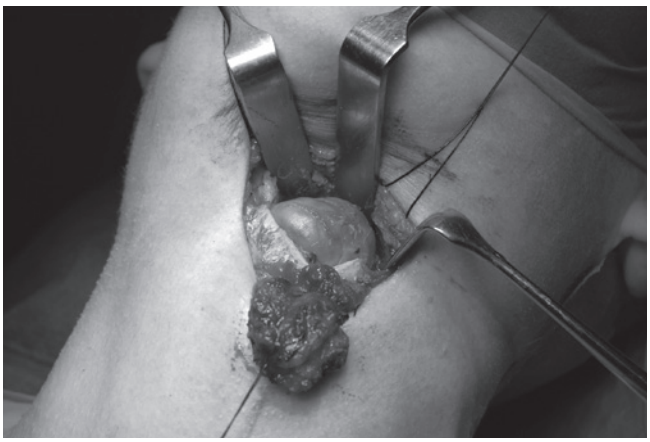

Fig. 4. Intra-operative view showing the surgical approach to the larynx. A 5-cm transverse cervical neck incision at the level of crico-thyroid membrane is performed, and the left thyroid lamina is identified and its superior half is removed in order to expose the neoplasm, occupying the left paraglottic space down to the superior aspect of the cricoid.

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 [12]. Although the mediastinum is the most frequently affected site, other localisation such as lung, urogenital tract, and orbit have been described [13 14]. For these reasons, SFT has been divided in "extrapleural" and "extrathoracic" subgroups [16].

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% [17]. "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 phonatory 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 [17] 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 [18].

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).

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Author	**Present case**	**Safneck [3]**	**Benlyazid [4]**	**Alobid [5]**	**Alobid [6]**	**Fan [8]**
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	Lateral pharyngectomy	Vertical hemilaryngectomy	Laser resection	Lateral pharyngectomy	Partial laryngectomy
Outcome (months)	NED (24) alive	NED (12) alive	NED (14) dead	NED (18) alive	NED(36) alive	NED(6) alive

M: male; F: female; VC: vocal cord; n/r: not reported; NED: not evidence of disease

Table I. Literature review concerning SFT of the larynx (follows).

	Case 7	Case 8	Case 9	Case10	Case 11
Author	**Stomeo [10]**	**Chang [19]**	**Thomson [9]**	**Morvan [11]**	**Dotto [7]**
Age	73	34	49	52	38
Gender	M	M	M	F	M
Location	Supraglottic/ commisure	Supraglottic	Glottic/subglottic	Supraglottic	False VC
Clinical presentation	Foreign body sensation/swallowing disorders	Foreign body sensation	Difficult breathing	Dysphonia/dyspnea	Cough/deepening voice
Symptoms duration (months)	3	6	24	Several months	12
Radiologic findings	Mass	Mass	Mass	Mass	Mass
Endoscopic findings	Submucosal swelling	Smooth submucosal mass	Smooth avascular mass	Bulky avascular mass	Submucosal mass
Tumour size (cm)	1.0	4.0	2.3	5.0	5.1
Treatment	Laser resection	Supraglottic partial laryngectomy	Biopsy	Lateral pharyngectomy	Laser resection
Outcome (months)	NED (24) alive	n/r	NED (12) alive	n/r	n/r

M: male; F: female; VC: vocal cord; n/r: not reported; NED: not evidence of disease

haemorrhages and nuclear atypias, although a benign evolution is seen in 50% of SFTs showing these features [16].

At MR, the lesion typically shows on T1-weighted sequences a signal that is isointense to muscle and variable on T2-weighted, with some areas with a slight enhancement and other areas with iso-hypointensity. These findings are probably due to the different histological arrangement of the tumour, which is rich in collagen and fibroblasts mixed with other areas where these components are less represented [19].

In the present case, CT with contrast revealed a low enhancement during the arterial phase, which became intense and heterogeneous in the interstitial phase, probably in view of the low presence of blood vessels. The axial CT scan showed a well-defined neoplasm occupying 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, haemangioma is typically localised in the subglottic area, whereas in adult patients the supraglottis is more frequently 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 enhancement, while on T2 WI shows a hyperintense signal compared to muscle tissue and often shows poorly defined margins; phlebitis (small round calcifications of the venous vessels) are typically present. Schwannoma generally arise from the superior laryngeal nerve and tend to dislocate the larynx without involving the laryngeal intrinsic musculature 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 after gadolinium injection while in T2 WI appear hyperintense. Paraganglioma is a highly vascularised, non-encapsulated lesion that rarely involves the supraglottis, showing on MRI T1WI a typical "salt and pepper" appearance due to signal voids combined with high signal 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 tumour 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-optimal laryngeal exposure and for bulky lesions invading the paraglottic space, with transglottic or extralaryngeal extension. Tracheotomy can be planned for both types of

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 cranio-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.

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