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Giant retropharyngeal liposarcoma

A 64-year-old man presented at our Otolaryngology (ENT) department with a history of progressive dysphagia spanning a few months, and swallowing cough. Thirteen years earlier, he had undergone surgery for partial removal of a retro-hypopharyngeal liposarcoma with successive selective radiotherapy. Definitive histology reported low-grade liposarcoma. The recurrence of the disease had been documented by magnetic resonance imaging (MR), and the residual mass had been monitored with imaging follow-up over a number of years, although the patient did not exhibit any kind of symptom until a few months ago. Fiberoptic laryngoscopy identified a considerable hypopharyngeal mass.

At the last MR control, a fatty mass had been found, with few septa and foci inside; the mass, with capsulate aspect, was localized in the retro-hypopharynx region, compressed on the axial plane (Fig. 1) between prevertebral muscles and the esophago-tracheal axis and extending down, beyond the thoracic inlet, extensively occupying the cranial portion of the posterior mediastinum (Fig. 2).

Surgical excision of the liposarcoma was performed using a cervical U-shaped incision. After separation and medialization of thyroid lobes, a wide mass was found situated between the prevertebral plane and the esophago-tracheal axis in the upper posterior mediastinum. Isolation of the liposarcoma easily proceeded almost completely with digital dissection; in fact, the slow-growing behaviour of the neoplasm had allowed the formation of a thick fibrous cap all around it. The mass, totally separated from the surrounding tissues, was removed en bloc from the thoracic inlet and the mediastinum (Fig. 3). The definitive histological result was well-differentiated 'lipoma-like' liposarcoma. An MR carried out 1 month after surgery showed regular post-surgical outcomes; no signs of persistent disease were evident. Based on radiological control, definitive histology and past therapy, further postoperative chemotherapy or radiotherapy were not considered. However, annual radiological and clinical follow-up have been recommended.

Basing on Stout's classification, there are four main histological variants of liposarcoma: well-differentiated, myxoid, round cell and pleomorphic.[9,10] Well-differentiated liposarcoma is the most common subtype, accounting for 45% of all liposarcomas.[3] It is divided into two histological subtypes: 'lipoma-like', which has occasional lipoblasts and minor areas of pleomorphism, and 'sclerosing', which has more regions of fibrosis[6]. These well-differentiated tumours have the most favourable clinical course compared with the other subtypes of liposarcoma and rarely metastasize,[1] but, like the other subtypes, well-differentiated liposarcomas have the propensity for local recurrences.[3] The most common sites

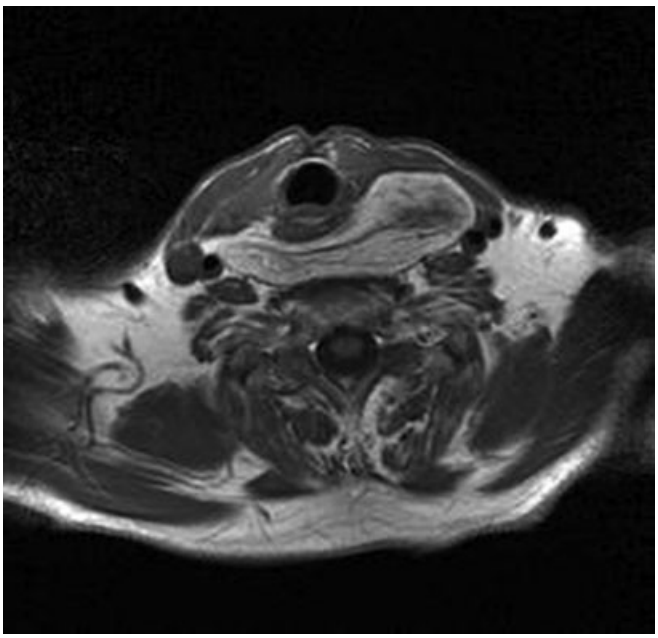

Fig. 1. T1-weighted magnetic resonance imaging on axial plane, showing the typical hourglass shape of retropharyngeal liposarcomas.

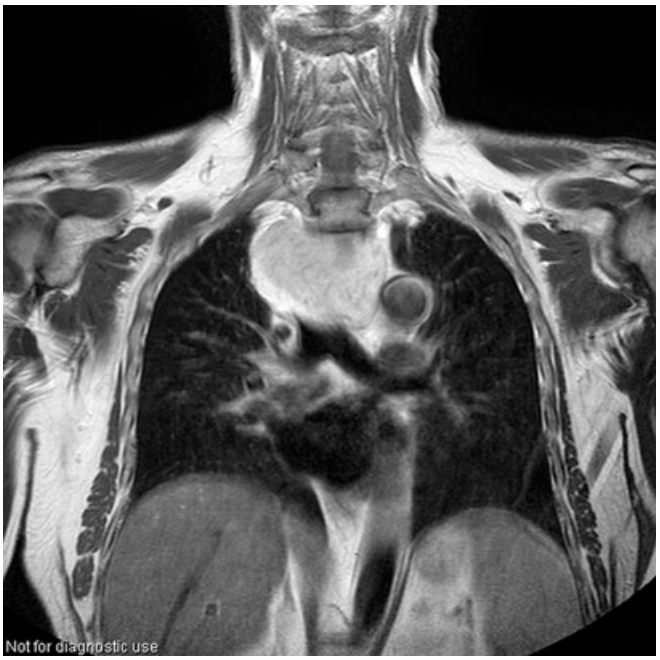

Fig. 2. Post-contrast T1-weighted magnetic resonance imaging on sagittal plane: the extension of retropharyngeal liposarcomas to the posterior upper mediastinum can be seen.

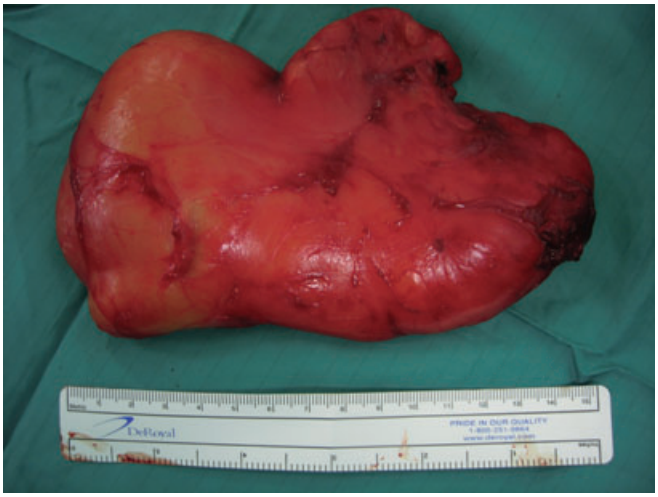

Fig. 3. Macroscopic appearance of retropharyngeal liposarcomas.

of occurrence of head and neck liposarcomas are the cheek (14%), anterior neck (13%), larynx and pyriform sinus (13%), orbit (8%), and supraglottis (7%).[1]

Based on a MEDLINE and PUBMED search, our case is the ninth report of a retropharyngeal localization in the international literature[1–8] (Table 1). A literature review of retropharyngeal liposarcomas (RLS) is resumed. The particular anatomic localization where the RLS arises makes the progressive growth of that neoplasm frequently silent: the neoplasm starts to be symptomatic when generally reaching large dimensions and extending into the mediastinum (including our case).[2,3,5,6] The mean age of first presentation in the patients is 59.9(+/–16 standard deviation). A slight male predilection is seen (5 male/4 female), according with the literature on liposarcomas of other sites (unlike lipomas, which predominate in females).[1] Common presenting symptoms of reported cases are dysphagia (5/9 cases), dysphonia (3/9), neck swelling (3/9), obstructive sleep apnea (3/9), swallowing cough (1/9), and globus pharyngeus (1/9). The hypothesis of a malignant transformation from a lipoma has been suggested by some authors.[1,6]

Computed tomography (CT) and MR imaging of well-differentiated liposarcoma typically demonstrate a predominantly fatty mass, well circumscribed, with no signs of local invasion and no associated lymphoadenopathies. A distortion of the larynx and pharynx, pushed anteriorly, is always found. The hourglass shape in the axial plane has to be considered strongly typical, and has been reported most (8/9) of the cases reviewed. Irregular thickened linear or nodular septa are often present. The CT scan typically demonstrates a low-density signal, in some cases of inhomogeneous composition.[4] MR can show a mass with a fat-intensity signal, and a mixture of hyperintense and intermediately intense signals in T2-weighted images.[4,8] In T1, the mass appears intermediately intense, enhanced after gadolinium administration.[1,4,8] Typically, the amount of radiologically identifiable fat at MR is variable, but a well-differentiated liposarcoma will generally demonstrate at least 75% of adipose tissue.[1,3,5] Essentially, CT and MR also tend to reflect the degree of tumour differentiation, that is, the more differentiated the tumour, the more the image appearance will approach that of adipose tissue.[3]

Table 1 Review of the literature

	Age, sex	Mediastinal extension	Approach	Hystology	Postoperative radiotherapy	Recurrence after first attempt	Notes
Present case	64-year-old male	Yes	Cervicotomic	Well differentiated, lipoma like	After first operation	13 years	None
F. M. Stoller *et al.*[1]	42-year-old female	No	Cervicotomic	Myxoid	Yes (and chemotherapy with oral MTX and 5FU)	No	None
B. Yueh *et al.*[2]	88-year-old female	Yes	Cervicotomic	Well differentiated, lipoma like	After second attempt	1 year	Dedifferentiation into a malignant fibrous histiocytoma. Postoperative chemotherapy
F. Ballestros *et al.*[3]	64-year-old male	Yes	Cervicotomic	Well differentiated, n.o.s.	No	No	Previous multiple subcutaneous lipoma removal
R. Gundelach *et al.*[4]	62-year-old male	No	Cervicotomic	Well differentiated, sclerosing sub-type	Yes	No	None
R. Hermans *et al.*[5]	40-year-old male	Yes	Cervicotomic	Sclerosing, myxoid	No	No	None
I. B. A. Menown *et al.*[6]	69-year-old female	Yes	Cervicotomic	Well differentiated, n.o.s.	No	18 month	Dedifferentiation from lipoma
M.E. Prince *et al.*[7]	39-year-old female	No	Cervicotomic	Well differentiated, n.o.s.	No	No	None
H. Ozawa *et al.*[8]	56-year-old male	Yes	Trans-oral	Myxoid	After second operation	1 year	

5FU, fluorouracil; MTX, metotrexate; n.o.s., not otherwise specified.

At the present time, surgery is considered the treatment of choice for RLS. Although 4/9 cases reported (including our case)[2,3,5] had a mediastinal extension, an exclusive cervicotomic approach has been performed in seven out of eight cases, for the absence of infiltrates and easy dissectability from surrounding tissue. According to the behaviour of liposarcomas from other sites, of the nine retropharyngeal liposarcomas reported, 4/9 recurred (including our case)[1,6,8]. To date, the postoperative treatment of head and neck liposarcoma has been based on the results achieved with tumours of the extremities of the trunk, and has not changed significantly in recent years.[1] The effectiveness of postoperative radiotherapy in well-differentiated liposarcomas has not yet been demonstrated.[1,6] In general, chemotherapy has not be demonstrated to be effective in the treatment of soft-tissue liposarcomas,[6] and adjuvant chemotherapy could represent an option only in cases where distant metastases or local aggressive behaviour have been documented.

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