CASE REPORT

Dedifferentiated liposarcoma of the pyriform sinus

Giovanna Giordano a,*, Luigi Corcione a, Letizia Gnetti a, Giuseppe Mercante b, Teore Ferri b

a Department of Pathology and Laboratory of Medicine, Parma University, Viale A. Gramsci, 14, 43100 Parma, Italy
b Department of Otolaryngology, Parma University, Viale A. Gramsci, 14, 43100 Parma, Italy

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Summary

The authors presented a second reported case of dedifferentiated liposarcoma of the pyriform sinus in a 50-year old male, with a history of heavy smoking, airway obstruction and dysphagia. A computed tomography (CT) of the neck and thorax revealed a retroesophageal lobulated heterogenous mass. A laryngoscopic examination disclosed sessile polypoid mass in the right pyriform sinus. Microscopically, the neoplasm consisted of areas of well-differentiated liposarcoma and a non-lipogenic component (dedifferentiated component). These two parts were co-mingled, giving a mosaic pattern. Well-differentiated liposarcoma was characterized by adipocytes with a great variation in size, and multivacuolated lipoblasts. Immunohistochemical studies showed positive reactivity with vimentin and smooth muscle actin in non-lipogenic component. This neoplasm shows histological and immunohistochemical features of dedifferentiated liposarcoma with myogenic differentiation.

KEYWORDS

Liposarcoma; Dedifferentiated liposarcoma; Larynx; Pyriform sinus

Introduction

In the present paper, we describe clinical and pathological features of an extremely rare subtype laryngeal liposarcoma. This subtype, named dedifferentiated liposarcoma, is located in the pyriform sinus. Moreover, we review the literature in order to clarify the clinical and pathological features of this rare neoplasm.

Material and methods

A 50-year old male was admitted to the hospital with a history of heavy smoking, airway obstruction and dysphagia. The symptoms had been present for a period of six months.

Esophagostroscopy showed an extrinsic lesion, which had reduced the esophageal lumen by compression (Fig. 1a, asterix: neoplasm).

A computed tomography (CT) of the neck and thorax revealed a retroesophageal lobulated heterogenous mass (Fig. 1b, asterix: neoplasm).

A laryngoscopic examination disclosed sessile polypoid mass in the right pyriform sinus.

* Corresponding author. Tel.: +39 0521 290391; fax: +39 0521 292710.
E-mail address: giovanna.giordano@unipr.it (G. Giordano).
The lesion was firm and circumscribed and readily excised.

No further therapy was administered to the patient, who, six months after surgery, is well, without any evidence of recurrence or metastases.

The specimen was examined histologically and immunohistochemically.

Results

Grossly, the neoplasm was polypoid mass, measuring 5 cm (Fig. 2a) and on section showed white-grey areas and adipose tissue (Fig. 2b).

Microscopically, the neoplasm consisted of areas of well-differentiated liposarcoma and a non-lipogenic component (dedifferentiated component). These two parts were co-mingled, giving a mosaic pattern (Fig. 3a). Non-lipogenic component was also present below squamous epithelium of the mucosa, where numerous vessels were also seen (Fig. 3b).

At high-power view, well-differentiated liposarcoma was characterized by adipocytes with a great variation in size, atypical spindled cells and multivacuolated lipoblasts (Fig. 3c).

Non-lipogenic zones were characterized by fusiform or spindle cells, that varied little in size and shape, had scanty cytoplasm, indistinct cell border and were separated by collagen fibers, arranged in a parallel fashion. Nuclear atypia was light. Occasional mitoses can be observed in these non-lipogenic areas of neoplasm (Fig. 3d). No mitotic figures, instead, were seen in the areas of well-differentiated liposarcoma.

Immunohistochemical studies showed positive reactivity with vimentin and smooth muscle actin in non-lipogenic component (Fig. 4a). Lipogenic component was positive for S-100 protein.

Final pathological diagnosis, suggested by the histologic appearance and immunohistochemical profile, was low-grade dedifferentiated liposarcoma.
Microscopically, the neoplasm consisted of areas of well-differentiated liposarcoma and a non-lipogenic component ((a) hematoxylin–eosin, ×40). Non-lipogenic component was also present below squamous epithelium of the mucosa where numerous vessels were seen ((b) hematoxylin–eosin, ×40). At high-power view, well-differentiated liposarcoma was characterized by multivacuolated lipoblasts ((c) hematoxylin–eosin, ×400; arrows: lipoblast). In dedifferentiated component there were spindle cells, with light nuclear atypia and occasional mitoses ((d) hematoxylin–eosin, ×400; arrows: a mitosis).

On immunohistochemical analysis non-lipogenic areas showed positivity for vimentin ((a) ×100) and for smooth muscle actin ((b) ×100).
Discussion

In relation with other laryngeal neoplasms, sarcoma are uncommon tumors. These lesions, in fact, account for less than 1% of all laryngeal malignancies.1

Laryngeal liposarcoma is very rare, with fewer than 50 cases reported in literature.2–6

Histologically, most laryngeal liposarcomas are of the biologically favorable types, representing well-differentiated liposarcoma (lipoma-like) or mixoid liposarcoma.7,8

Liposarcomas of the larynx involve the supraglottic, glottic region and hypopharynx and are more frequent in males.7,8 Smoking has been suggested as an environmental factor in the development of this neoplasm.7

Prognosis of laryngeal liposarcoma is considered better than non-laryngeal liposarcomas, except for pleomorphic and round cell subtypes, which have high-metastatic potential.9

Our review suggested that dedifferentiated liposarcoma are an extremely rare subtype of laryngeal liposarcoma, with only four previous cases reported3,10–12 (Table 1). The first laryngeal dedifferentiated liposarcoma was reported by Tobey in 1979 as a malignant mixed mesenchymal tumor (malignant mesenchymoma), because the neoplasm, on ultrastructural analysis, was characterized by different mesenchymal elements, with the features of lipoblasts, fibroblasts, skeletal and smooth muscle cells.10 Since 1979 only three other cases of laryngeal dedifferentiated liposarcoma have been reported in the International Literature3,11,12 and only one has been seen in the pyriform sinus3 (Table 1).

Thus, our case represents the second case of dedifferentiated liposarcoma of the pyriform sinus.

In conclusion, we believe that, although dedifferentiated liposarcoma of larynx is rare, its diagnosis is possible, when the features of dedifferentiated liposarcoma, suggested by Evans13 and by Elgar and Goldblum,14 are borne in mind.

This neoplasm, in fact, characteristically, according to these authors, shows two parts: a lipogenic and non-lipogenic component. The lipogenic component consists of well-differentiated liposarcoma, while non-lipogenic component has the features of malignant fibrous histiocytoma, mixofibrosarcoma, fibrosarcoma and rhabdomyosarcoma, hemangiopericytoma-like, angiosarcomatous, osteosarcomatous, neural-like, menigioma-like differentiation or leiomyosarcomatous differentiation.12–15

Moreover, non-lipogenic component can show a low-grade and high-grade of dedifferentiation.14 Low-grade dedifferentiation, as observed in our case, is characterized by only focal pleomorphism, light nuclear atypia and rare mitoses. Conversely, high-grade dedifferentiation shows more pleomorphism and more severe nuclear atypia. Although low- and high-grade of dedifferentiation are different on histological examination, in a clinicopathological analysis of 20 retroperitoneal liposarcomas, Elgar and Goldblum have established that there is no significant difference in clinical outcome and prognosis between these subtypes of neoplasms.12 The dedifferentiated liposarcomas should be differentiated from benign lipogenic tumors, such as lipoma,16 fibrolipoma17 spindle cell lipoma18 and from spindle cell liposarcoma.19

Table 1 Laryngeal dedifferentiated liposarcoma: review of literature

<table>
<thead>
<tr>
<th>Authors</th>
<th>No. case</th>
<th>Age (Yrs)</th>
<th>Sex</th>
<th>Site</th>
<th>Treatment</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tobey</td>
<td>1</td>
<td>61 M</td>
<td></td>
<td>Right vocal cord</td>
<td>Partial laryngectomy, with cervical esophagectomy and chem</td>
<td>Local recurrences (3-10 m) Died of disease D</td>
</tr>
<tr>
<td>Steiger et al.11</td>
<td>1</td>
<td>64 M</td>
<td></td>
<td>Vallecra epiglottica</td>
<td>Resection of neoplasm</td>
<td>Alive without evidence of disease 18 m D</td>
</tr>
<tr>
<td>McCormick et al.12</td>
<td>1</td>
<td>62 M</td>
<td></td>
<td>NR</td>
<td>Partial laryngectomy, resection of neoplasm</td>
<td>Local recurrence 23 yrs later, few m later further recurrence D</td>
</tr>
<tr>
<td>Gonzales-Lois et al.3</td>
<td>1</td>
<td>69 M</td>
<td></td>
<td>Pyriform sinus</td>
<td>Partial laryngectomy, resection of neoplasm</td>
<td>Alive without evidence of disease 6 m D</td>
</tr>
<tr>
<td>Present Case</td>
<td>1</td>
<td>50 M</td>
<td></td>
<td>Pyriform sinus</td>
<td>Resection of neoplasm</td>
<td>Alive without evidence of disease 6 m D</td>
</tr>
</tbody>
</table>

Chem: Chemotherapy; m: months; Yrs: years.
The lipoma on histological examination is characterized by mature uniform adipocytes without evidence of cellular atypia. Microscopically the fibrolipoma is composed by benign fibrous connective tissue and mature adipose tissue, lacking of nuclear atypia. On microscopical examination spindle cells lipoma is a neoplasm with mature adipocytes mixed with collagen-forming spindle cells. Spindle cell liposarcoma is characterized by spindle cell proliferation, organized in fascicles and whors with variable myxoid change and by adipocyte component with features of atypical lipoma. On immunohistochemical analysis spindle cell component in this subtype of liposarcoma shows positivity for vimentin, supporting the immature mesenchymal nature of these elements. The dedifferentiated liposarcoma presents instead a spindle cells elements, who are positive for specific mesenchymal phenotypes, such as fibrous histiocytoma, rhabdomyosarcoma and smooth muscle cells. In our case, spindle cell component showed a myogenic differentiation with reactivity for smooth muscle actin.

In the larynx, on the other hand, because of the rarity of dedifferentiated liposarcoma, it is not possible to establish a prognostic difference between the low- and high-grade of differentiation. Thus, further studies are necessary to clarify prognostic significance of the low-grade and high-grade dedifferentiated liposarcoma in this site.

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References