

Management of an Atypical Lipomatous Tumor/Well-Differentiated Liposarcoma of the Left Laterocervical Region: Report of a Rare Case

Ioannis KOMNOS^a, Maria MICHALI^a, Lentiona BASIARI^a, Georgios TSIRVES^a,
Georgios PSYCHOGIOS^a

^aDepartment of Otorhinolaryngology-Head and Neck Surgery,
University General Hospital of Ioannina, Ioannina, Greece



ABSTRACT

Liposarcomas are among the most usual cancerous growths of mesenchymal tissues and represent about 1% of head and neck sarcomas. They are extremely rare in childhood and are mostly seen between 30 and 60 years of age. The biologic behavior and histologic features of liposarcomas vary. Although these tumors grow very slowly and have a benign behavior, sometimes they grow rapidly and metastasize early, with fatal results. This case report presents a 63-year-old man with a tumor of the left side of the cervical region which has grown to a large size over four years. For an accurate diagnosis, fine needle aspiration biopsy (FNA) biopsy was performed. The cytological examination showed an adipose tumor. Surgical removal was done under local anesthesia and the pathologic examination showed a well-differentiated liposarcoma. These are usually early stage tumors, with fewer metastases than other sarcomas. Surgical abscission is the gold standard for the treatment of liposarcomas. The efficacy of postoperative radiotherapy or/and chemotherapy is controversial.

Keywords: liposarcoma, head and neck sarcoma, lipomatous tumors, well-differentiated liposarcoma, liposarcoma therapy.

INTRODUCTION

Sarcomas are rare neoplasms of mesodermal origin which account for 1% of all head and neck malignancies (1). Sarcomas of the cervical region represent 5–15% of all sarcomas in adults. Liposarcomas are among the most frequent malignant mesenchymal tumors. Head and neck lipo-

sarcomas are less common than in other body sites, comprising 2-9% of sarcomas found in the region. They are very rare in the pediatric population and most commonly occur in adults, with a peak incidence between 30 and 60 years of age. Furthermore, liposarcomas are more common in males than females. The majority of them are found in deep soft tissues of the retroperito-

Address for correspondence:

Maria Michali, MSc, Consultant

Department of Otorhinolaryngology-Head and Neck Surgery, University Hospital of Ioannina, Stavros Niarchos Avenue, 45500, Ioannina, Greece
Tel.: 00306945908292; email: maria.ch.michali@gmail.com

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neum as well as upper and lower limbs. Only a small percentage (about 2%) is located in the head and neck region, with the cheek, larynx and neck being the most usual sites implicated in this area. When they extend at the front of the neck, it is indispensable to prove that the tumor is lipomatous (2, 3).

The behavior of a tumor and its tendency for topically aggressive growth and distant metastases depends on its histologic type and staging. The size and location are also important prognostic factors. Liposarcomas usually appear as slow-growing, painless, well-circumscribed masses and their size may exceed 10 cm during presentation. In most instances, these tumors are well encapsulated (3).

Surgical resection is suggested as the fundamental treatment for managing regional expansion, excluding the cases of aggressive liposarcomas being in sites that are not accessible for excision (4, 5). Predictors of treatment failure include large tumor size, high-grade histology and positive surgical margins. Adjunct radiotherapy is suggested for these neoplasms and occasionally chemotherapy when there is a remarkable risk of systemic dissemination (4-7). $\square$

CASE PRESENTATION

A 63-year-old male came to our department with a subcutaneous painless tumor formation on the left side of the cervical region that occurred about four years prior to presentation. The tumor was asymptomatic and increased in size during the last 10 months. The patient's medical history was free. On physical examination, the patient's neck was asymmetric and it had a well-circumscribed mass of soft tissue on the left cervical region (Figure 1). The preoperative computed tomography (CT) scan of the patient's neck showed a mass with a fatty/solid consistency, extending subcutaneously above the left sternocleidomastoid muscle and measuring 6.2 cm $\times$ 5 cm $\times$ 2 cm (Figures 2a and 2b). There was no extension of the tumor into the surrounding tissues. No swollen neck or axillary lymph nodes were seen. Based on these findings, a diagnosis of lipomatous neoplasm was made. The patient underwent fine needle aspiration biopsy (FNA) under local anesthesia. Histopathologic examination revealed an adipose tissue neoplasm.



FIGURE 1. Preoperative photograph showing a large mass of the left laterocervical region



FIGURE 2a. Preoperative axial computed tomography (CT) showing a well-defined soft tissue density mass



FIGURE 2b. Preoperative coronal CT showing the tumor of the left cervical region

The decision was made to remove the mass surgically under local anesthesia. The mass had not adhered to the muscle and it was deduced with no injury sustained to the tissues around it. The incision was sutured layer by layer (Figure 3). The well-capsulated, soft, yellow tumor, which measured 8 cm $\times$ 6 cm $\times$ 2.5 cm (Figure 4), was sent for further histologic examination. Microscopically, the tumor was predominantly composed of stellate cells, many of which were darkly stained, large and exhibited pleomorphic



FIGURE 3. Intraoperative view of surgical access to tumor



FIGURE 4. The defined and soft excised tumor

nuclei. Furthermore, atypical lipoblasts were found. Immunohistochemical staining was positive for CDK4 (cyclindependent kinase 4) and MDM2 (murine double minute 2). Based on these results, an atypical lipomatous tumor/well-differentiated liposarcoma was diagnosed. After obtaining the result of histopathological examination, an abdominal ultrasound and a CT scanning of the thorax were performed, which did not detect any distant metastasis. According to the above described findings, it was a localized tumor.

The postoperative course was satisfactory. The oncologists recommended ultrasound follow-up of the cervical region three months after surgery and then every six months. They also suggested that the wide excision of the tumor was found to be adequate with no further therapy. During the 3-, 6- and 12-month postoperative follow-up there was no sign of relapse. The anatomic location, the histologic type (well-dif-

ferentiated) and a history of slowly enlarging mass without lymph or distant metastasis show the benign character of this tumor. □

DISCUSSION

Patients with liposarcoma have no clinical symptoms until the tumor grows to an enormous size, causing pressure consequences or deformation. Liposarcomas are 120 times less common than lipomas. Most of them are found in the retroperitoneum and extremities. The prevalence of head and neck liposarcomas represent only 1% of all sarcomas of this region (7, 8). Risk factors for liposarcomas include genetic background, trauma and past treatment with radiation therapy (8).

Although medical imaging methods, including CT and magnetic resonance imaging (MRI), could lead to a primary diagnosis of liposarcoma, only histologic and immunohistochemistry examinations can verify the diagnosis. In our case, the patient underwent only CT scanning because the mass was limited to the left laterocervical region. Well-differentiated liposarcomas consist of nodules of fat cells and fibrous tissue (9).

The progression of a liposarcoma depends on its histologic type. The histologic types of liposarcomas are atypical tumor/well-differentiated, liposarcoma lipomatous, pleomorphic liposarcoma, myxoid liposarcoma and de-differentiated liposarcoma. Well-differentiated liposarcoma is the most usual type (30–40% of all liposarcomas). It is locally aggressive and involves a high risk of relapse but a low risk of distant metastases. It is not easy to evaluate the outcome of all patients with head and neck liposarcoma. The histologic type is very important in prognosis. Patients with well-differentiated liposarcoma tend to recover well, with reports of local recurrence only. In our case, the patient had a slowly growing tumor for about four years with no distant metastasis, which was a sign of the benign character of his tumor (7).

The majority of patients with liposarcoma have no clinical symptoms before the neoplasm becomes large and impinges on neighboring structures. The main therapeutic method for lipomatous tumors of the head and neck region is their complete resection. In our case, the tumor was removed en block. As a result, there was no need for adjuvant therapy. In the international

literature, some authors believe that further therapy with radiation and/or chemotherapy should be avoided after a wide surgical excision, regardless of histological type of the liposarcoma (10). However, some other authors have suggested adjuvant radiotherapy, even if they had a clear fibrous capsule (11-13). In patients with distant metastases, chemotherapy is used as an adjunct to radiotherapy. Nevertheless, the decision of radiotherapy or/and chemotherapy as treatment for liposarcomas remains controversial (13). □

CONCLUSIONS

In conclusion, liposarcomas of the head and neck region are very rare tumors and sometimes, because the fibrous component may represent the majority of them, they might be misdiagnosed as other soft-tissue neoplasms.

Excision is the mainstay of treatment and the prognosis is determined by the histological type and clinical stage. Wide surgical excision of low-grade liposarcomas (well differentiated and myxoid) is usually sufficient. Even with complete resection, liposarcoma has a high local recurrence rate. Thus, long-term follow-up is required. Furthermore, it is not clear whether adjuvant radiotherapy and chemotherapy is of benefit. Our incident indicates that clinicians and pathologists keep the potential diagnosis of liposarcomas in mind when evaluating patients with tumors of the head and neck region. □

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