ejpmr, 2018,5(6), 153-155

SJIF Impact Factor 4.897

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Report

ISSN 2394-3211

EJPMR

LEIOMYOSARCOMA OF THE HEAD AND NECK: REPORT OF TWO CASES AND REVIEW OF THE LITERATURE

Khaoula Hafidi[1]*, Omar Oubry[2], Hasnae Taghzout[1], Touria Bouhafa[1], Abderahman Elmazghi[1] and Khalid Hassouni[1]

[1]Department of Radiotherapy, University Hospital Center Hassan II Fez, Morocco.
[2]Department of English Studies, Faculty of Arts and Humanities Fez Saiss, Morocco.

***Corresponding Author: Khaoula Hafidi**

Department of Radiotherapy, University Hospital Center Hassan II Fez, Morocco.

Article Received on 04/04/2018 **Article Revised on 25/04/2018** **Article Accepted on 15/05/2018**

ABSTRACT

Leiomyosarcoma is a malignant smooth-muscle tumor that is extremely rare in the head and neck region. Two cases of this uncommon tumor localized in the larynx and the nasal cavity are presented in this report. We also discuss the diagnosis and treatment of Leiomyosarcoma in this aspect.

KEYWORDS: Leiomyosarcoma; Neck; Soft tissue; Sarcoma; Neoplasm

1. INTRODUCTION

Soft-sarcomas of the head and neck region are relatively rare and represent less than 10% of all soft tissue sarcomas and less than 1% of all neoplasms of this region.[1] In fact, the majority of Leiomyosarcomas of the head and neck arise in the oral cavity, superficial soft-tissues like: scalp, paranasal sinuses, and jaws.[2] In one of the largest series of head and neck sarcomas including 352 patients, Freedman et al. reported only 4 (1.1%) cases of Leiomyosarcoma located at the neck region.[3] In this report, two new cases of Leiomyosarcomas localized in the larynx and the nasal cavity are presented, and the diagnostic difficulties and management of this exceedingly rare malignancy will be discussed.

2. CASE REPORTS

2-1. Case 1

A 23-year-old male patient was admitted for laryngeal dyspnea. On direct laryngoscopy, an invasive process of the three floors of the larynx was detected. Plus, the histopathological examination revealed a laryngeal Leiomyosarcoma grade II **(Figure 1)**, then the immunohistochemical study was conducted, and tumor cells expressed H-caldesmone and did not express desmin or cytokeratin. The computed tomography had objectified no secondary locations. Thus, the patient underwent a total laryngectomy with bilateral neck dissection, and there was no complication during the surgery.

After, the patient had an adjuvant radiotherapy with a total dose of 46 Gy in 23 sessions of 2Gy, and the evolution was marked by the appearance of secondary locations.

2-2. Case2

A 62-year-old male patient was admitted to the emergency because of an epistaxis and a left nasal obstruction; he had a biopsy as well as a resection of the tumor arising in the left nasal cavity. The histopathological examination was in favor of a Leiomyosarcoma; 5 months later, a tumor recurrence was detected in the same site by an endoscopic examination, and the computed tomography (CT) showed a tumor in the inferior horn of the left nasal cavity (figure 2), then the patient had for the second time a biopsy and a resection of the tumor with a left medial maxillectomy. Then, the histopathological examination and the immunohistochemical staining both revealed a Leiomyosarcoma, and the limits of resection were healthy. The patient had an adjuvant radiotherapy with a total dose of 66 Gy with Intensity Modulated Radiation Therapy (IMRT), and he showed good loco regional control.

Figures

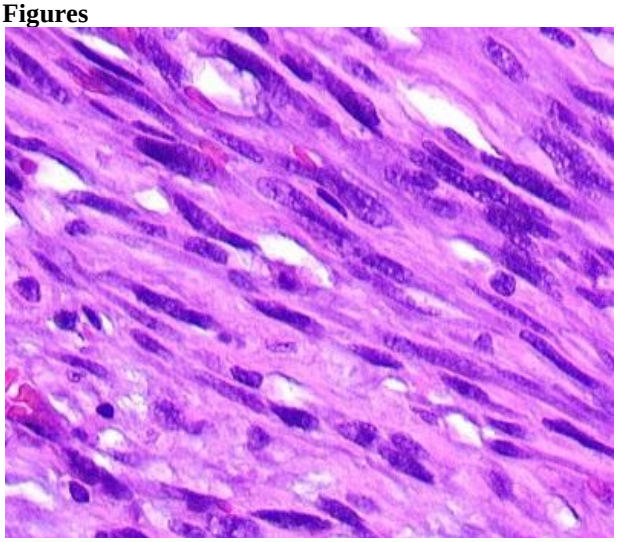

Figure 1: Atypical spindled mesenchymal cells with ''cigar'' shaped nuclei in laryngeal grade II of leiomyosarcoma (haematoxylin-eosin, 400×).

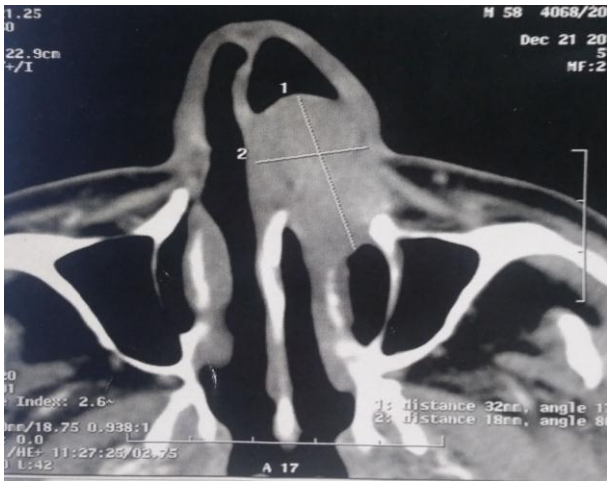

Figure 2: Axial section of computed tomography that shows the thickening at the nasal cavity.

3. DISCUSSION

The symptoms of Leiomyosarcoma involving the head and neck region depend on the site and size of the tumor. Hoarseness, stridor, dyspnea and dysphagia are the most common complaints of patients with laryngeal tumors. While the initial symptom of the patient with laryngeal Leiomyosarcoma - in this report - was only dyspnea, the main symptom of the second patient with Leiomyosarcoma of nasal cavity was the obstruction due to mechanic compression. The existence of few smooth muscles in the head and neck region is the potential reason for the rarity of Leiomyosarcoma. Since blood vessels are the only structures in the larynx and nasal cavity with smooth-muscle tissue, it has been speculated that it develops from smooth muscle in the tunica media of the vessel walls.[4] Aberrant mesenchymal differentiation and metastasis are the other possible modes of origin.[5]

The differential diagnosis of this rare tumor may be problematic; hence, the diagnosis should be supported by immunohistochemical and ultra-structural investigations. Histologically, the tumor is characterized by prominent interlacing bundles and fascicles of elongated spindle cells with: elongated with ‘cigar-shaped’ or ‘blunt-ended’ nuclei, prominent nucleoli, and abundant eosinophilic cytoplasm. The mitotic rate is usually high and atypical mitotic figures are present.[2] The Leiomyosarcoma is positive for muscle specific actin, and vimentin, and it is negative for S-100 protein myoglobin and epithelial membrane antigen on immunostaining.[6] The electron microscopy is very helpful, and the surgical resection remains the principal treatment method of the soft-tissue sarcoma. Because the adjacent pseudocapsule is commonly infiltrated by the tumor cells and satellite lesions are often found at some distance from the main lesion, the margin of the excision should be at least 1 cm in all directions when surgery is used alone. If a combination of surgery and radiation therapy is performed, that margin can be reduced to approximately 0.5 cm.[7] Even the extent and the adequacy of excision both determine the survival and local recurrence, because of the proximity of the adjacent neurovascular structures or vertebral column**;** en bloc resection and achieving these margins at all tumor planes is almost impossible in the head and neck region. During the surgical removal of the lesion in the presented case with nasal cavity localization the limits of resection were healthy. The most aggressive surgical therapy of the laryngeal Leiomyosarcoma is ‘total laryngectomy’, but partial laryngectomy procedures can also be preferred in some selected cases.[2,4,8] Total laryngectomy was performed in the present case in order to achieve more proper surgical margins, and a disease free survival rate, keeping in mind that the nature of the tumor may have satellite or skip lesions. Unless there is a lymphadenopathy, neck dissection is usually not essential because of the poor metastatic lymph node rate.

Moreover, there are no randomized trials for head and neck soft tissue sarcomas; the effectiveness of adjuvant radiation in these sarcomas has been clearly demonstrated through three prospective randomized trials that have compared surgery alone with surgery and radiation.[9–10] Preoperative or postoperative choice of external-beam irradiation is still questionable for soft-tissue sarcomas, since there are no supportive data. Some of the potential advantages of preoperative External-Beam Radiation Therapy (EBRT) include decreased intraoperative seeding of tumor cells, and tumor shrinkage that might facilitate surgery later. Suit et al. evaluated the relationship between tumor size and the sequencing of radiation, and they showed that preoperative radiation was superior to postoperative radiation in terms of local control for patients with tumors greater than 15 cm.[10] However, others have shown no difference.[11] The adjuvant radiotherapy was carried out to the base of the skull where the tumor was microscopically positive. Although recurrence was

expected in this site due to the poor radio response rate of the Leiomyosarcoma and positive surgical margins, relapse occurred outside the radiation field where the surgical margins were negative. This evidence emphasizes the role of adjuvant radiotherapy in reducing the risk of recurrence of the head and neck Leiomyosarcomas that are treated surgically, even without tumor positive margins. For this purpose, radiation therapy may be necessary after surgery of these sarcomas.

CONCLUSION

Leiomyosarcomas are malignant tumors of smooth muscles accounting for 5–6% of all soft tissue sarcomas. They are mostly seen in the gastrointestinal tractus, the uterus, and the retroperitoneum. Soft tissue sarcomas in head and neck region are rare and total surgical excision with safe margins and adjuvant radiotherapy are the mainstay of treatment.

Competing interests

The authors declare no competing interests.

Authors' contributions

All authors read and approved the final version of the manuscript.

REFERENCES

1. Eeles RA, Fisher C, A'Hern RP, Robinson M, Rhys-Evans P, Henk JM, et al. Head and neck sarcomas: prognostic factors and implications for treatment. Brit J Cancer, 1993; 68: 2017.
2. Marioni G, Bertino G, Mariuzzi L, Bergamin-Bracale AM, Lombardo M, Beltrami CA. Laryngeal leiomyosarcoma. J Laryngol Otol, 2000; 114: 398–401.
3. Freedman AM, Reiman HM, Woods JE. Soft-tissue sarcomas of the head and neck. Am J Surg, 1989; 158: 367–72.
4. Freije JE, Gluckman JL, Biddinger PW,Wiot G. Muscle tumors in the parapharyngeal space. Head Neck, 1992; 14: 49–54.
5. Chen JM, Novick WH, Logan CA. Leiomyosarcoma of the larynx. J Otolaryngol, 1991; 20: 345–8.
6. Chae SW, Geon C, Choong SC, Jong OC. Leiomyosarcoma of the larynx. J Otolaryngol, 1999; 28: 293–5.
7. Eilber FR, Eckardt J. Surgical management of soft tissue sarcomas. Semin Oncol, 1997; 24: 526–33.
8. Yoshizaki T, Horikawa T, Nonomura A, Furukawa M. Leiomyosarcoma of the larynx. J Laryngol Otol, 1999; 113: 167–9.
9. Pisters PW, Harrison LB, Leung DH, Woodruff JM, Casper ES, Brennan MF. Long term results of a prospective randomized trial of adjuvant brachytherapy in soft tissue sarcoma. J Clin Oncol, 1996; 14: 859–68.
10. Suit HD, Mankin HJ, Wood WC, Gebhardt MC, Harmon DC, Rosenberg A, et al. Treatment of the patient with stage M0 soft tissue sarcoma. J Clin Oncol, 1988; 6: 854–62.
11. Cheng E, Dusenbery K, Winters M, Thompson R. Soft tissue sarcomas: preoperative versus postoperative radiotherapy. J Surg Oncol, 1996; 61: 90–9.