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Polymorphous Nasopharyngeal Adenocarcinoma: The Importance of a Correct Diagnosis

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Abstract

Polymorphous nasopharynx adenocarcinoma is a rare disease. It frequently occurs in the soft and hard palatal mucosa of the minor salivary glands. This adenocarcinoma has a slow infiltrating growing pattern with frequent perineural invasion and low metastatic potential.

We report a case of polymorphous nasopharyngeal adenocarcinoma with extension to the lateral pharyngeal space and closely proximal to the skull base and jugular vein.

Keywords: Polymorphous Nasopharyngeal Adenocarcinoma; Salivary Glands; Jugular Vein

Introduction

Polymorphous adenocarcinoma is a rare tumor that mostly affects the minor salivary glands in the palatal mucosa [1]. It was first described in 1983 [2]. Despite its infiltrative growth pattern, this tumor has a low distant metastatic potential [3]. Surgery is the gold standard treatment when possible. Radiotherapy and adjuvant chemotherapy have also been used, although there is no evidence of their benefits [4]. We found only four polymorphous nasopharyngeal adenocarcinoma medical case reports. In none of them did any of the patients complain of nasal obstruction alone, in other cases patients complained of other related symptoms. We report a case of a woman who was referred to our center with nasal obstruction that was not responding to medical treatment.

Case Report

A 44-year-old woman, who had been complaining of nasal obstruction for a year, was referred to our center for turbinoplasty. She did not present any other symptom. No disease was recorded. Her posterior nasopharyngoscopy not only showed her enlarged turbinates but also a tumor arising from the left which occupied her entire nasopharynx (Figure 1). This had been misdiagnosed before.

We performed CT (Figure 2) and MRI (Figure 3), which showed a well-defined lesion measuring approximately 40.7 mm x 53.8mm x 46.7 mm in its anteroposterior, transverse and cephalo caudal axes respectively. It presented a heterogeneous signal in different sequences, showing both solid behavior and heterogeneous enhanFigure 1: Endoscopic view. Right and left nostril.

cement, which extended towards the medial sector of the rhinopharynx. It also showed areduced diameter at that level that produced mass effect to the right in its anterior sector that contacted the fibers of the lateral pterygoid muscle but did not impress signs of invasion. Its posterior sector, which contacted the long muscle of the head in its lateral sector and the parotid space, did not show any signs of invasion as well. A small branch of the left vertebral artery was mentioned in its V3 segment, as well as the superior bulb of the ipsilateral jugular vein, that crossed the mentioned lesion from its posterior sector.



Figure 2: Computed tomography.

We then performed a lesion biopsy. Endoscopy showed a lesion arising from the left nasopharynx that occupied the entire nasopharynx but did not bleed profusely. The tumor was removed to permeabilize the nasopharynx and meliorate the patient's symptoms (Figure 4). Figure 3: Magnetic resonance imaging.

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Figure 4: Endoscopic view after partial resection.

After the surgery, the patient reported improvement of her nasal obstruction. The final pathological diagnosis was polymorphous nasopharyngeal adenocarcinoma.

We required a PET. It described a 48 mm x 30 mm x 42 mm solid formation with heterogeneous enhancement after intravenous contrast was administered. The same occupied the parapharyngeal and masticatory space on the left side, reducing the lumen of the hypermetabolic (SUV 9.8) lesion; It was also in intimate contact with the deep lobe of the ipsilateral parotid gland. We referred the patient to the oncology department with her pathological diagnosis and images. She is currently undergoing evaluation by an oncologist who is considering treatment with radiation therapy vs. partial surgical resection plus adjuvant radiation.

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Discussion

Polymorphous adenocarcinoma frequently occurs in the 5th and 6th decades of life and there is a 2:1 female predominance [4]. It is an indolent tumor that is locally invasive but metastasizes slowly. Its localization; bone involvement and size determine surgical treatment [5]. Neck dissection is not necessary when there is no evidence of lymph node involvement. Polymorphous adenocarcinoma has been reported to have a poor response to radiotherapy [6] but postoperative treatment is generally recommended [7].

The importance of rhinopharyngeal fiberoptic examination is well known. It is routinely performed when hearing loss is involved. But it should also be performed to evaluate nasal obstruction as a routine procedure.

Conclusion

We present a case report of a rare and indolent tumor that was misdiagnosed in the initial evaluation.

Regardless of the unusual case report, it seems important to emphasize the need to explore every patient's nasopharynx when they are presenting with nasal obstruction.

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