Received / Geliş tarihi: November / Kasım 20, 2006 Accepted after revision / Düzelti sonrası kabul tarihi: June / Haziran 5, 2007 Published online / Online yayın tarihi: September / Eylül 14, 2007 doi:10.2399/tao.06.022

OLGU BILDIRISI / CASE REPORT



Extramedullary plasmacytoma of the nasopharynx: two-handed transnasal endoscopic surgery

E. Serbetci, G.A. Sengör

Nazofarenksin ekstramedüller plazmasitomu: transnazal endoskopik iki-el yaklaşımı

Ekstramedüller plazmasitom, sıklıkla nazofarenks ve paranazal sinüslerin submukozal lenf dokularından kaynaklanan, nadir görülen malign bir tümördür. Bizim hastamızın düzgün yüzeyli, polipoid, saplı ve 18x11x13 mm boyutlarında submukozal bir kitlesi mevcuttu. Bu kitle sağ torus tubarius mukozasından kaynaklanmaktaydı. İki-el tekniği yardımıyla, endoskopik görüntüleme altında, sapı nazofarenkste bağlanarak kitle çıkartıldı. Histopatolojik incelemesi sonucunda ekstramedüller plazmasitom olduğu belirlendi. Hastanın sistemik araştırmasında lokal veya sistemik yayılıma ait bulguya rastlanmadı. Plazmasitomlarda tercih edilen tedavi yöntemi radyoterapi olmasına rağmen, tek başına cerrahi tedavi uyguladığımız hastamızda, 33 ay boyunca rekürrens gelişmedi. Bu tümöre uyguladığımız transnazal endoskopik cerrahi, nazofarenks lezvonlarının tedavisi icin uygulanan minimal invazif cerrahi ve ileri endoskopik tekniklere bir örnek teşkil etmektedir.

Anahtar Sözcükler: Nazofarenks, ekstramedüller plazmasitom, iki-elle endoskopik cerrahi.

Abstract

Extramedullary plasmacytoma is a rare malignant tumor that frequently involves submucosal lymphoid tissue of the nasopharynx and paranasal sinuses. Our patient had a smooth surfaced, polypoid, pedicled, submucosal mass with the dimensions of 18x11x13 mm. It originated from the mucosa of the right torus tubarius. Applying the two-handed technique, the tumor was excised under endoscopic vision by ligating its pedicle in the nasopharynx. The histopathologic examination revealed extramedullary plasmacytoma. There was no local or systemic disease on systemic investigation. Although the preferred treatment method in plasmacytomas is radiotherapy, with surgery alone, we did not observe any recurrence in 33 months of follow-up. The transnasal endoscopic approach for this tumor is a sample for minimally invasive surgery and advanced endoscopic techniques for the treatment of nasopharyngeal lesions.

Key Words: Nasopharynx, extramedullary plasmacytoma, twohanded endoscopic surgery.

Türk Otolarengoloji Arsivi, 2008; 46(1): 45-48

Turk Arch Otolaryngol, 2008; 46(1): 45-48

Erhun Şerbetci, MD; Gani Atilla Şengör, MD Nisantası ENT Group, İstanbul

Introduction

As surgeons gain experience in endoscopic surgery, more techniques will be developed for endonasal tumors at different locations, and the surgery will be more comfortable for both patients and doctors. With the advances in radiology, a threedimensional resection plan can be devised, and the surgeon can clearly determine whether the tumor is removable by the transnasal endoscopic route.¹

Both extramedullary and medullary plasmacytomas are localized forms of plasma cell neoplasms and 80-90% of extramedullary plasmacytomas (EMP) are localized in the head and neck.^{2,3} They arise from the submucosal lymphoid tissue of the nasal cavity, and 40% occur in the paranasal sinuses, 20% in the nasopharynx, and 18% in the oropharvnx^{2,4,5} Most of the patients are over 40 years old and 4:1male predominance is observed.^{3,4} а Radiotherapy is considered the primary form of management, but there are several reports on surgical management with or without radiotherapy.^{2,4,6} Regional lymph node metastasis occurs in 20 to 25% of patients.^{3,4} As the local recurrence rate is 6–10%³ and the risk of disseminated disease (multiple myeloma) is 9.5–50%,⁵⁸ long-term follow-up is necessary.

In this report, we present a case of nasopharyngeal EP, which was managed with surgery alone, using the transnasal two-handed endoscopic approach.

Case Report

A 31-year-old man presented with bilateral nasal obstruction and with a history of two recent episodes of nasal bleeding. On endoscopic examination, he had a smooth surfaced, polypoid, pedicled, submucosal mass located in the right lateral wall of the nasopharynx. Its pedicle was based on the right torus tubarius (Figure 1). Computed tomography showed an 18x11x13 mm mass with soft tissue density and peripheral contrast enhancement, protruding into the lumen of the nasopharynx. Excisional biopsy under general anesthesia was planned. He underwent a transnasal endoscopic procedure using the two-handed technique, in which the assistant holds the endoscope. Using Blakesley forceps, the tumor was ligated by its pedicle with 3/0 polyglycolic acid to prevent potential bleeding from a likely vascular tumor. On ligation, the tumor turned purple (Figure 2). Its pedicle was cut with Zurich nasal scissors and cauterized using monopolar cautery after it was totally excised (Figure 3). The histopathologic examination of the specimen revealed an EMP. To rule out disseminated disease, a postoperative systemic investigation was performed. The panendoscopy of the head and neck area was normal. No monoclonal gamma globulin peak was evident. Urine protein electrophoresis was also normal. Quantitative immunoglobulins were within normal limits. A bone marrow biopsy and skeletal survey were found negative by the hematologist. The surgical borders of the specimen were free of tumor and radiotherapy was not used. No local or systemic recurrence has been observed during 33 months of follow-up (Figure 4).

Discussion

The transnasal endoscopic approach for removing juvenile nasopharyngeal angiofibromas and for adenoidectomy has been the subject of many reports.^{9,10} With advances in radiology, we can obtain sufficient information about the dimensions and spread of the tumor to choose the type of surgery.

EMPs may pursue one of several clinical courses:³ a) localized disease may be apparently cured by surgery and/or radiation; b) local disease can recur but may eventually be eradicated; c) local disease may persist and cause death by uncontrolled growth; d) disseminated disease (multiple myeloma) may develop.

The treatment of EMPs relies mostly on the radiosensitivity of the tumor. Wax et al. demonstrated 75% locoregional control using radiotherapy,⁷ whereas Kotner and Wang reported 16 patients treated with radiation therapy with a 5-year survival of 69% and a disease-free survival of 63%.¹¹ For patients with the diagnosis of multiple myeloma, chemotherapy for the systemic disease often takes precedence.⁵ Susnerwala et al. reviewed which lesions best respond to chemotherapy, in addition to primary radiation therapy.¹² Using criteria originally intended for the grading of multiple myelo-



Figure 1. Endoscopic view of the plasmacytoma in the right lateral wall of the nasopharynx. [Color figure can be viewed in the online issue, which is available at www.turkarchotolaryngol.org]



Figure 2. The color of the mass after ligation. [Color figure can be viewed in the online issue, which is available at www.turkarchotolaryngol.org]



Figure 3. The mass was separated from its pedicle and the pedicle was cauterized with monopolar cautery. [Color figure can be viewed in the online issue, which is available at www.turkarchotolaryngol.org]

mas, tumors were classified as low, intermediate, and high grade.¹³ EMPs with a low-grade histology were controlled by primary external beam radiation in 83% of cases. Lesions considered to be intermediate to high grade had only a 17% control rate with primary external beam radiation. There was a significant difference in local control but not overall survival. These authors recommended the use of adjuvant chemotherapy in patients diagnosed with



Figure 4. Endoscopic view in the postoperative 33rd month. [Color figure can be viewed in the online issue, which is available at www.turkarchotolaryngol.org]

intermediate- and high grade EMP.¹² In the head and neck region, in areas like the nasopharynx, which is adjacent to vital structures, the primary treatment modality for EMPs is radiotherapy, but there are reports of successful treatment with surgery alone.^{5,6,14} Complete resection of nasopharyngeal EMPs in either a primary therapeutic or salvage modality are associated with high morbidity rates. In the literature most of the nasopharyngeal EMPs were extensive or needed extended surgical approaches like transpalatal or anterior skull base approaches.⁵ Surgical therapy as the primary treatment modality may be used in limited diseases. Additional reported treatment options include stereotactic radiosurgery and cryoablation.¹⁵ Longterm follow up is critical to detecting any local recurrences and identifying patients who convert to the disseminated form of multiple myeloma.

In our case, the location and morphological features of the tumor facilitated endoscopic removal. Since the surgical borders were tumor-free histopathologically, we did not apply radiotherapy, and the patient has shown no local recurrence or systemic disease.

With the two-handed technique described by May et al.¹⁶ in which the assistant holds the endoscope, we used both hands for ligating the tumor pedicle in the nasopharynx. If the lesion needs to be managed using bimanual interventional techniques, the two-handed endoscopic technique is suggested as the best approach compared to traditional open approaches or radiotherapy.

In experienced hands, the excision of limited tumors from areas like the nasopharynx becomes possible using minimally invasive techniques. The technical decision concerning the surgical approach should be made after a detailed investigation of the tumor extent and the anatomy.

References

- 1. Rice DH. Endonasal approaches for sinonasal and nasopharyngeal tumors. *Otolaryngol Clin North Am* 2001; 34: 1087-93.
- Korolkowa O, Osuch-Wojkiewicz E, Deptala A, Suleiman W. Extramedullary plasmacytoma of the head and neck. *Otolaryngol Pol* 2004; 58: 1009-12.

- Mills SE, Fechner RE. The nose, paranasal sinuses and nasopharynx. In: Diagnostic surgical pathology. Sternberg SS, editor. 3rd ed. Philadelphia: Lippincott Williams&Wilkins; 1999. p. 907.
- Yavaş Ö, Altundağ K, Sungur A. Extramedullary plasmacytoma of nasopharynx and larynx: synchronous presentation. *Am J Hematol* 2004; 75: 264-5.
- **5. Wein RO, Popat SR, Doerr TD, Dutcher PO.** Plasma cell tumors of the skull base: four case reports and literature review. *Skull Base* 2002; 12: 77-86.
- Miller FR, Lavertu P, Wanamaker JR, Bonafede J, Wood BG. Plasmacytomas of the head and neck. *Otolaryngol Head Neck Surg* 1998; 119: 614-8.
- Wax MK, Yun KJ, Omar RA. Extramedullary plasmacytomas of the head and neck. Otolaryngol Head and Neck Surg 1993; 109: 877-85.
- **8. Kapadia SB, Desai U, Cheng VS.** Extramedullary plasmacytoma of the head and neck. A clinicopathologic study of 20 cases. *Medicine (Baltimore)* 1982; 61: 317-29.
- Durr DG. Endoscopic electrosurgical adenoidectomy: technique and outcomes. J Otolaryngol 2004; 33: 82-7.
- Scholtz AW, Appenroth E, Kamen-Jolly K, Scholtz LU, Thumfart WF. Juvenile nasopharyngeal angiofibroma: management and therapy. *Laryngoscope* 2001; 111: 681-7.
- 11. Kotner LM, Wang CC. Plasmacytoma of the upper air and food passages. *Cancer* 1972; 30: 414-8.
- 12. Susnerwala SS, Shanks JH, Banerjee SS, Scarffe JH,Farrington WT, Slevin NJ. Extramedullary plasmacytoma of the head and neck region: clinicopathological correlation in 25 cases. *Br J Cancer* 1997; 75: 921-7.
- Bartl R, Frisch B, Fateh-Moghadam A, Kettner G, Jaeger K, Sommerfeld W. Histologic classification and staging of multiple myeloma. A retrospective and prospective study of 674 cases. *Am J Clin Path* 1987; 87: 342-55.
- 14. Baumann I, Ruck P, Dammann F, Plinkert PK. Locally recurring extramedullary plasmacytoma of the upper aerodigestive tract. *Laryngorhinootologie* 2000; 79: 213-20.
- 15. Corsetti RL, Ready NE, Wanebo HJ. Cryotherapy for a plasmacytoma of the nasopharynx: a newer approach to a difficult surgical access site. *Surg Rounds* 2000; 707-12.
- 16. May M, Hoffmann DF, Sobol SM. Video endoscopic sinus surgery: a two-handed technique. *Laryngoscope* 1990; 100: 430-2.

Conflict of interest statement: *No conflicts declared.*

Correspondence: Gani Atilla Şengör, MD Valikonağı Cad, 107E, Daire 1 Nişantaşı 34363 İSTANBUL Phone: +90 212 296 91 54 Fax: +90 212 231 27 04 e-mail: dr_atilla@yahoo.com