DÜZCE TIP DERGİSİ
DUZCE MEDICAL JOURNAL

OLGU SUNUMU / CASE REPORT

Esthesioneuroblastoma with Unusual Metastasis to T10 Vertebra: Case Report and Review of the Literature

T10 Vertebraya Metastaz Yapmış Estezyonöroblastom: Vaka Sunumu Ve Literatür Taraması

[1] Süleyman YILMAZ

[1] Elif KARALI

[1] Ender GÜÇLÜ

[1] Özcan ÖZTÜRK

[2] Ümran YILDIRIM

[1] Düzce Üniversitesi Düzce Tıp Fakültesi KBB AD.

[2] Düzce Üniversitesi Düzce Tıp Fakültesi Patoloji AD.

Submitted/Başvuru tarihi:
05. 11. 2009
Accepted/Kabul tarihi:
01. 01. 2011
Registration/Kayıt no:
09 11 73

Corresponding Address /Yazışma Adresi:

Dr. Süleyman Yılmaz
Düzce Üniversitesi Düzce Tıp Fakültesi KBB Anabilim Dalı
Düzce/Türkiye

e-posta:
dryilmazsuleyman@yahoo.com

e-ISSN 1307- 671X
www.tipdergi.duzce.edu.tr
duzcetipdergisi@duzce.edu.tr

ABSTRACT

Esthesioneuroblastoma is a rare malignant tumor orginating from the olfactory epithelium. A 52–year-old man admitted to our clinic with a 3 months history of progressive nasal obstruction, epistaxis and mass on the nasal radix. On rhinoscopy, a polypoid mass was seen in the both nasal cavity and we performed an intranasal biopsy with local anesthesia. On histopathologic analysis the tumor was identifed as an esthesioneuroblastoma. The tumor was classified as Kadish stage B. The mass was excised via bilateral endoscopic endonasal resection and lateral rhinotomy approach. Radiotherapy was performed postoperatively. During the follow up, submandibuler lymph node metastasis occured ten months after surgery. The patient underwent bilateral type III modified radical neck dissection followed by radiotherapy to the neck. Distant metastasis developed on the T 10 vertebra 12 months after the initial treatment. Although he had distant metastasis he was free of local recurrence 13 th month after surgery. In conclusion because of locoregional recurrences are common in esthesioneuroblastomas, patients must be followed carefully but also we have to remember that distant metastasis may appear without local recurrence.

Keywords: Esthesioneuroblastoma, endonasal resection, distant metastasis, local recurrence.

ÖZET

Estezyonöroblastom olfaktuar epitelden köken alan nadir görülen malign bir tümördür. 52 yaşındaki erkek hasta kliniğimize 3 aydır devam eden ilerleyici burun tıkanıklığı, burun kanaması ve nazal radikste kitle ile başvurdu. Rinoskopik muayenede her iki nazal kavitede polipoid kitle görüldü ve lokal anestezi ile biyopsi yapıldı. Histopatolojik analiz sonucunda estezyonöroblastom tanısı konuldu. Tümör Kadish evre B olarak değerlendirildi. Endoskopik endonazal ve lateral rinotomi ile yaklaşımları ile kitle eksizyonu yapıldı. Postoperatif radyoterapi uygulandı. Takiplerinde cerrahiden 10 ay sonra submandibuler lenf nodu metastazı saptandı ve hastaya bilateral tip 3 modifiye radikal boyun diseksiyonu yapıldı ve sonrasında radyoterapi uygulandı. Cerrahiden 12 ay sonra ise T10 vertebrada uzak metastaz saptandı. Hastanın uzak metastazı olduğu halde primer bölgede rekürrens saptanmadı. Sonuç olarak estezyonöroblastomalarda lokal nüksler sık olduğu için hastaların dikkatli bir şekilde takip edilmeleri gereklidir ve aynı zamanda lokal rekürrens olmadan da uzak metastaz olabileceği de akılda tutulmalıdır.

Anahtar kelimeler: Estezyonöroblastom, endonazal rezeksiyon, uzak metastaz, lokal rekürrens.

INTRODUCTION

Esthesioneuroblastoma is a rare malignant tumor orginating from the olfactory epithelium. Malignant tumors of the nasal cavity are rare. Esthesioneuroblastoma account for only 6 % of these neoplasms (1). These tumors have a bimodal age distribution occuring on the second and sixth decades of life, but it can be seen in all age groups (2). Esthesioneuroblastomas are locally aggressive and can metastasize by lymphatic and hematogenous routes. The cervical lymph nodes are the most common site of metastasis. It can spread submucosally in all directions, involving the paranasal sinuses, nasal cavities and cross the cribriform plate involving brain. Because of non specific symptoms such as nasal obstruction, epistaxis and headache diagnosis is frequently delayed. In this article we reported a delayed case who attended our clinic after the mass had involved on the nasal radix.

CASE REPORT

A 52–year-old man admitted to our clinic with a 3 months history of progressive nasal obstruction, epistaxis and enlarging mass on the nasal radix. On rhinoscopy, a polypoid mass was seen in the both nasal cavity. Examination revealed a 2 x 3-cm, painless mass on the radix. There was no palpabl mass on the neck at the time of the diagnosis. His vision and eye movements in both eyes were normal. A computed tomography demonstrated a mass that filled both nasal cavities extending into the left anterior and posterior ethmoid cells, bilateral frontal sinuses and invading the left lamina papyracea and no intracranial extension was seen (Figure I). The maxillary and sphenoid sinuses were also free of tumor. We performed an intranasal biopsy under local anesthesia. On histopathologic analysis the tumor was identifed as an esthesioneuroblastoma. These findings were confirmed by immunohistochemistry, tumor cells were strongly positive stained with NSE, chromogranin (Figure II), synaptophysin and CD99 (Figure III) and weakly positive stained with neurofilament. Besides it was not stained with S-100 protein, keratin, desmin, HMB45. The tumor was classified as Kadish stage B. The mass was excised via bilateral endoscopic endonasal resection and lateral rhinotomy approach. The tumor was peeled away completely from the cribriform plate and left lamina papyracea with endoscopic resection. Radiotherapy was performed postoperatively. At follow up, bilateral submandibuler lymph node metastasis occured ten months after surgery. The patient underwent bilateral type III modified radical neck dissection followed by radiotherapy on the neck. Distant metastasis developed on the T 10 vertebra 12 months after initial treatment (Figure IV). He was free of local recurrence at follow up with nasal endoscopy 13 months after surgery. The rarity of the distant metastasis of esthesioneuroblastoma to vertebra without local recurrence is the reason of presenting this case.

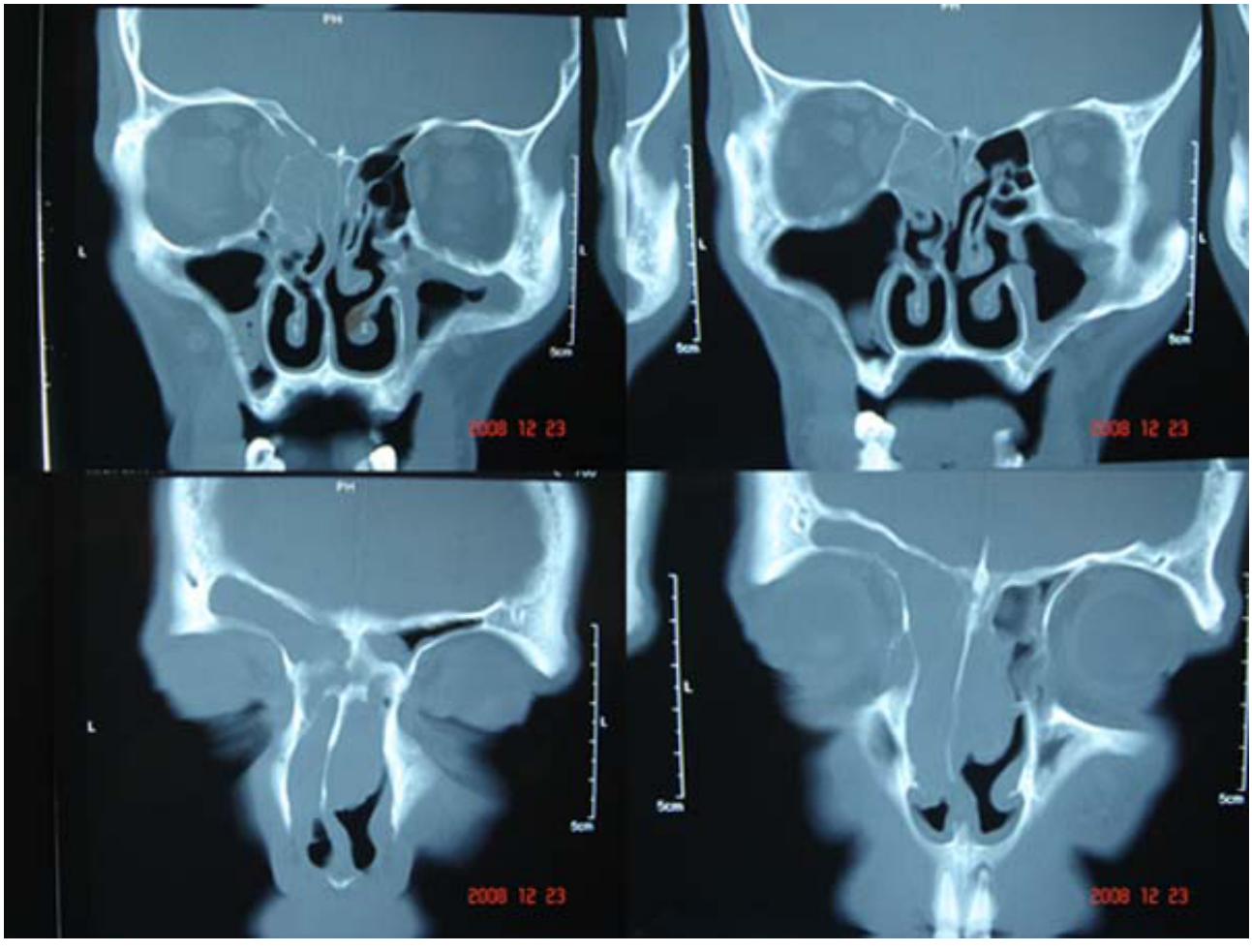

Figure I: A computed tomography shows a mass in the cribriform area extending into the left anterior and posterior ethmoid cells, bilateral frontal sinuses and invading left lamina papyracea.

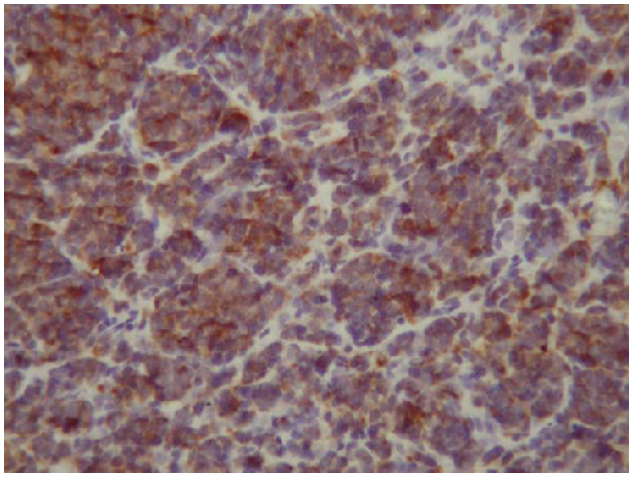

Figure II: This photomocrograph showed positive stained tumor with chromogranin (X 200).

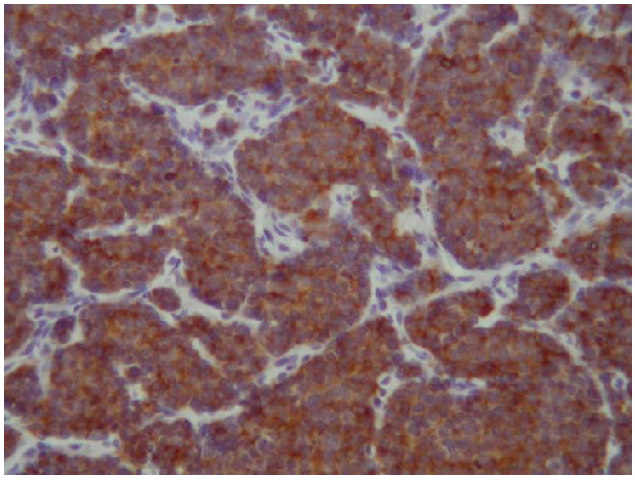

Figure III: Diffuse membranous CD 99 staining demonstreated in tumor cells.

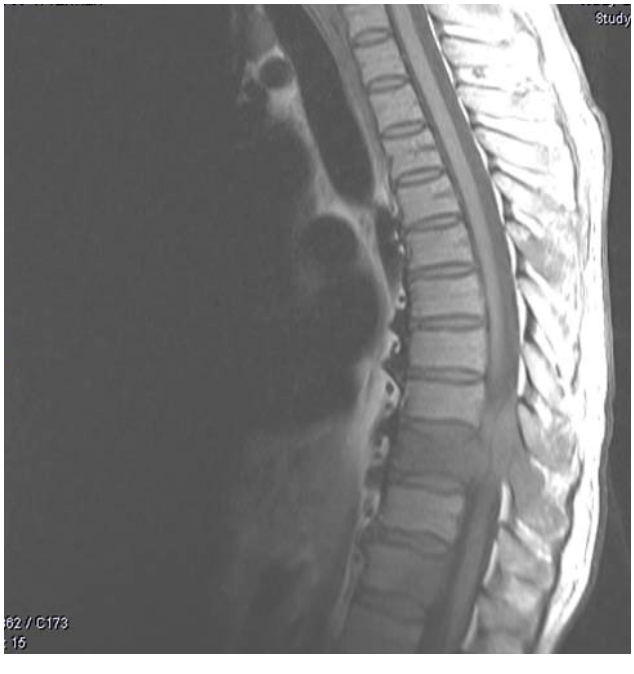

Figure IV: MRI shows distant metastasis on the T 10 vertebra.

DISCUSSION

Esthesioneuroblastoma is a rare malignant tumor of nasal cavity arising from the olfactory neuroepithelium located in the nasal septum, cribriform plate, the middle and superior turbinates(3). The most common symptoms of esthesioneuroblastoma are unilateral nasal obstruction and epistaxis. Less commonly, patients experience anosmia, headache and may have orbital symptoms such as proptosis, diplopia and excessive lacrimation if the mass extends to the orbita. In our case the patient complained of bilateral nasal obstruction, epistaxis and the enlarging mass on the nasal radix. The typical histologic appearance of an olfactory neuroblastoma (esthesioneuroblastoma) includes the presence of irregular nests of small hyperchromatic cells separated by stroma, diffuse sheets of cells with a prominent background of capillaries and little intervening stroma (4). The cells are small round blue cells with hyperchromatic nuclei and a high nuclear-cytoplasmic ratio. Occasional nuclear molding was seen.

Esthesioneuroblastomas are staged clinically with kadish system. The kadish system is based on the spread of the tumor (2). According to this system, stage A tumors are confined to the nasal cavity, stage B lesions involve the sinuses, and stage C masses involve the middle cranial fossa and the retrobulbar orbit (5). In a report by Diaz at al., all the recurrence occured in patients with kadish stage C tumors and the kadish staging system has been demonstrated to be of prognostic value for recurrence and survival (6). Nevertheless, the high incidence of local recurrence is directly related to inadequate resection margins (7). The most frequent recurrence is local. In our case no local recurrence was observed at follow up to 13 months after surgery.

Craniofacial resection combined with radiotherapy is considered to be the gold standard treatment in the management of these tumors (8). In recent years, many authors have begun treating esthesioneuroblastoma with a minimally invasive approach, obtaining good results (7-9). Surgery alone seems to be ineffective for local control, and many authors suggest the use of postoperative radiotherapy to reduce the risk of local recurrence. Serious complications were reported after craniofacial resection. These complications could potentially be avoided using an endoscopic approach. Other advantages of the endoscopic treatment are the short duration of surgery, short hospitalization and a better quality of life without sometimes aesthetic damage (9). In our case we performed bilateral endoscopic endonasal resection

and lateral rhinotomy approach. The patient underwent postoperative radiotherapy.

The incidence of cervical metastasis varies from 10 % to 33 % at the time of the diagnosis (10). Neck metastasis can occur early in the disease or many years later. Neck dissection is indicated only in the presence of nodes, elective dissection appears to be unnecessary. In our case, there was no palpabl mass on the neck at the time of diagnosis so elective neck dissection was not performed. At the follow up, neck metastasis occured 10 months after initial treatment. The patient underwent bilateral type III modified radical neck dissection followed by radiotherapy on the neck. The incidence of distant metastasis occur in 12 % to 25 % of patients, in whom lung, brain and bone are the areas most commonly involved (11, 12). In our case distant metastasis developed on the T 10 vertebra 12 months after initial treatment without local recurrence.

In conclusion because of locoregional recurrences are common in esthesioneuroblastomas, patients must be followed carefully but also we have to remember that distant metastasis may appear without local recurrence.

REFERENCES

1. Svane-Knudsen V, Jorgensen KE, Hansen O et al. Cancer of the nasal cavity and paranasal sinuses:a series of 115 patients. Rhinology 36:12-14, 1998.
2. Bradley PJ, Jones NS, Robertson I. Diagnosis and management of esthesioneuroblastoma. Curr Opin Otolaryngol head Neck Surg 11:112-118, 2003.
3. Diaz EM, Johnigan RH III, Pero C et al. Olfactory neuroblastoma:the 22 –year experience at one comprehensive cancer center. Head Neck 27:138-149, 2005.
4. Bellizzi AM, Bourne TD, Mills SE et al. The cytologic features of sinonasal undifferentiated carcinoma and olfactory neuroblastoma. Am J Clin Pathol. 129: 367-76, 2008.
5. Kadish S, Goodman M, Wang CC. Olfactory neuroblastoma. A clinical analysis of 17 cases. Cancer 37:1571-6, 1976.
6. Lund VJ, Howard D, Wei W et al. Olfactory neuroblastoma:past, presentand future Laryngoscope 113:502-507, 2003.
7. Walch C, Stammberger H, Andrehuber W et al. The minimally invasive approach to olfactory neuroblastoma:combined endoscopic and stereotactic treatment. Laryngoscope 110:635-640, 2000.
8. MoritaA, Ebersold olsen KD, Lewis JE et al. Esthesioneuroblastoma: prognosis and management. Neurosurgery 32:706-715, 1993.
9. Unger F, Haselberger K, Walch C et al. Combined endoscopic surgery and radiosurgery as treatment modality for olfactory neuroblastoma. Acta Neurochir 147:595-602, 2005.
10. Rinaldo A, Ferlito A, Shaha AR et al. esthesioneuroblastoma and sevical lymph node metastases:Clinical therapeutic implications.Acta Otolaryngol 122:215-21, 2002.
11. Eden BW, Debo RF, Larner JM et al. Esthesioneuroblastoma: long term out come and pattern of failure the University of Virginia experience . Cancer 73:2556-2562, 1994.
12. Resto VA, Eisele DW, Forastiere A et al. Esthesioneuroblastoma: the Johns Hopkins experience. Head Neck 22:550-558, 2000.