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

T10 Vertebra metastaz yapmış esthesyonöroblastom: Vaka sunumu ve literatür taraması

ABSTRACT
Esthesioneuroblastoma is a rare malignant tumor originating 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 identified 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, submandibular lymph node metastasis occurred 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 T10 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

Anahtar kelimeler: Estezyonöroblastom, endonasal reseksiyon, uzak metastaz, lokal rekürrens.

INTRODUCTION
Esthesioneuroblastoma is a rare malignant tumor originating 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 occurring 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 palpable 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 identified 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 submandibular lymph node metastasis occurred 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.
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 et al., all the recurrence occurred 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.

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

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

Figure IV: MRI shows distant metastasis on the T 10 vertebra.
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 palpable mass on the neck at the time of diagnosis so elective neck dissection was not performed. At the follow up, neck metastasis occurred 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