Esthesioneuroblastoma located in the thoracic extradural space: Case report

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\textbf{ABSTRACT}

\textbf{OBJECTIVE:} Esthesioneuroblastoma accounted for only 6% of the malignant nasal cavity neoplasms (ENB) is a rare tumor which originates from the olfactory epithelium. ENB’s are locally aggressive and can metastasize by lymphatic and hematogenous routes. A patient with the mass on the nasal dorsum was reported in this article.

\textbf{CASE HISTORY:} A 52–year-old man admitted to the hospital with a 3 months history of progressive nasal obstruction, epistaxis and mass on the nasal dorsum. On rhinoscopy, a polypoid mass was seen in the both nasal cavity and intranasal biopsy with local anesthesia was performed. Histopathologic diagnosis of the tumor was Kadish stage B esthesioneuroblastoma. Tumor was excised by using bilateral endoscopic endonasal resection and lateral rhinotomy approach and paranasal radiotherapy performed postoperatively. Ten months after surgery, neck metastasis was occurred and patient was underwent neck dissection. Twenten months after initial treatment, distant metastasis was identified on the T 10 vertebra and following the cranial and spinal radiotherapy to the neck he was free of local recurrence at follow up 13 months after surgery.

\textbf{CONCLUSION:} It has been known that the metastasis of the ENB to the spinal cord is an uncommon event, and it occurs often years after initial diagnosis. MRI scan is helpful for making the diagnosis, and surgery is the treatment of choice for obtaining diagnostic tissue and debulking the tumor. Radiotherapy is also a mainstay of postoperative treatment.

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1. Introduction

Esthesioneuroblastoma (ENB) accounted for only 6% of the malignant nasal cavity neoplasms (ENB) is a rare tumor which originates from the olfactory epithelium [1]. Although these tumors have a bimodal age distribution occurring second and sixth decades of life, it can be seen all age groups [2]. ENBs 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 and involving brain. Because of non specific symptoms such as nasal obstruction, epistaxis, headache, diagnosis frequently delayed. In this article we reported a case of diagnosis delayed patient attended our clinic after occur the mass on the nasal dorsum.

2. Case report

A 52–year-old man admitted to the Ear-Nose-Troath clinic with history of progressive nasal obstruction, and epistaxis during 3 months. On rhinoscopy, a painless polypoid mass in 3 × 3 cm diameters was seen in the both nasal cavity and the nasal dorsum without palpable mass on the neck. His vision and eye movements were normal in both eyes. A paranasal computed tomography (CT) scan confirmed this mass filled both nasal cavity. Although this mass extended into the bilateral ethmoid and frontal sinuses and invaded left lamina paprisea, its intracranial extension was not seen on CT images. Intranasal biopsi with local anesthesia was performed to the mass, and histopathological diagnosis was ENB with kadish stage B. The mass was removed near total through the bilateral endoscopic endonasal approach and lateral rhinotomy approach. Whole paranasal radiotherapy was performed postoperatively. Uneventfully, neck metastasis of residual mass was identified ten months after surgery, and whole neck radiotherapy protocol was performed to the patient. After radiotherapy, he was

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underwent to the extensive neck dissection surgery. Histopathological diagnosis was same.

Two months after the second surgery, he was admitted to the hospital with acute paraplegia. Sensory exam was unfeeling to light touch and pin prick of lower dermatomas to T12 with areflexi. Babinski reflex was plantar extensor in right leg and flexor in left leg. Anal tonusite was mild. His complete blood chemistry was within normal limits. An emergency thoracal magnetic resonance imaging (MRI) with gadolinium showed an enhancing extradural tumor which almost completely filled the spinal canal and thoracal 10 (T10) vertebrae body. Other organ metastasis was not found radiologically (Fig. 1). The patient was immediately taken to operation room. Total laminectomy was performed to the T10 vertebrae. Then, large extradural mass was removed subtotally. The histopathological diagnosis was ENB (Fig. 2). One week after laminectomy, T9 and T10 anterior corpectomy, T10-11 disectomy and T9-T10-T11 cage-plaque-screw-link stabilization through the right toracotomy was performed to the patient to remove the tumoral mass gross totally and to protect him from the progressive kyphotic deformity. The postoperative period was uneventful, and his neurological examination was better than preoperative period. He was discharged to home on 14 th day postoperatively. He was died of local recurrence at follow up 13 months after the last surgery.

3. Discussion

ENB is a rare malignant tumor of nasal cavity, and it arises from the olfactory neuroepithelium located in the nasal septum, cribiform plate and the middle and superior turbinates [3]. Commonly, ENB causes unilateral nasal obstruction and epistaxis. It also causes anosmia, headache, proptosis, diplopia and excessive lacrimation when tumor extends to the orbita. In present case, the patient complained of bilateral nasal obstruction, epistaxis due to the tumor located in the both nasal cavity and the nasal dorsum.

ENBs are staged clinically with kedisham system 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 [4].
Diaz et al. suggested that all the tumoral recurrences occur in patients with Kadish stage C tumors [5]. On the other hand, many other authors reported in literature that the most frequent tumoral recurrence is local; and this high incidence of local recurrence is directly related to in adequate tumor resection margins [6]. It has been demonstrated in literature that surgery alone seems to be ineffective for local control of ENB, and many authors suggest postoperative radiotherapy to reduce the risk of local recurrence of the tumor. In the adjuvant or neo-adjuvant platinum-based chemotherapy in addition to radiotherapy has been often chosen for local control of recurrence or distant metastasis [13]. Although craniofascial resection combined with radiotherapy is considered the gold standard treatment in management of ENB today, some serious complications are represented after craniofascial resection in recent literature [7,8]. To avoid from these complications, recent reports have suggested to treat the ENB with minimally invasive surgery such as endoscopic approaches which could have some advantages such as short duration of surgery, short hospitalization and a better quality of life without anesthetic damage [6-8]. In present case, before the radiotherapy administration, bilateral endoscopic endonasal resection and lateral rhinotomy approach which could give enough surgical corridor to the surgeon to remove the tumor from the nasal dorsum was preferred and performed to the patient. Eleven months after surgery, no local recurrence in nasal cavity and nasal dorsum was observed at follow up exams.

Neck metastasis can occur early in the disease or many years later. Neck dissection is indicated only in the presence of nodes, and elective dissection appears to be unnecessary. The incidence of distant metastasis (such as lung, brain, bone etc.) occur in 12%–25% of patients [10]. Additionally, metastasis to the spinal cord from ENB which is an uncommon event presents often years after initial diagnosis. The incidence of cervical metastasis of the ENB varies from 10% to 33% at the time of the diagnosis [9]. But thoracic metastasis of the ENB is vary rare. In literature nearly 30 patients with vertebral drop metastases has been documented today. ENB is a locally aggressive tumors and its recurrence ratio is reported 10% to 60%. On the other hand, despite aggressive treatment it commonly could metastase to the cervical lymph nodes or lungs beyond 1 month to 10 years in literature [11,12]. Drop metastasis to spine is classified by Cedars-Sinai (See Table 1) [13]. In addition to Mori and Zhang et al. described cauda equina metastases of the ENB treated with surgical resection and radiation therapy [12,14]. In present case, distant metastasis to the T10 vertebra 12 months after initial treatment and it could be diagnosed.

In conclusion, practitioners should suspect the drop metastases in their patients with ENB who suffers from the back and/or radiculopathic pain.

Table 1
Cedars-Sinai classification system for metastatic esthesioneuroblastoma to the spine.

| Stage | Anatomic location of lesion |
|-------|-----------------------------|
| CS0-leptomeningeal disease | Involvement of the CSF and leptomeninges (arachnoid and pia mater) |
| CS1a-one level disease | Involves one level of the spine or spinal cord: cervical, thoracic, lumbar, or sacral |
| CS1b-one level disease and leptomeningeal disease | Involves one level of the spine or spinal cord: cervical, thoracic, lumbar, or sacral; with involvement of the CSF and leptomeninges (arachnoid and pia mater) |
| CS2a-two level disease | Involves two levels of the spine or spinal cord: cervical, thoracic, lumbar, or sacral |
| CS2b-two level disease and leptomeningeal disease | Involves two levels of the spine or spinal cord: cervical, thoracic, lumbar, or sacral; with involvement of the CSF and leptomeninges (arachnoid and pia mater) |
| CS3a-three level disease | Involves three levels of the spine or spinal cord: cervical, thoracic, lumbar, or sacral |
| CS3b-three level disease and leptomeningeal disease | Involves three levels of the spine or spinal cord: cervical, thoracic, lumbar, or sacral; with involvement of the CSF and leptomeninges (arachnoid and pia mater) |
| CS4a-four level disease | Involves four levels of the spine or spinal cord: cervical, thoracic, lumbar, or sacral |
| CS4b-four level disease and leptomeningeal disease | Involves four levels of the spine or spinal cord: cervical, thoracic, lumbar, or sacral; with involvement of the CSF and leptomeninges (arachnoid and pia mater) |
4. Conclusion

Spinal metastasis has been shown to occur in many different tumors, but intradural and cauda equina metastases are rare. Because of locoregional recurrences are common, patients must be followed carefully for locoregional and distant metastasis.

Conflict of interest

None.

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