CASE REPORT

Rare case of mortality after hemorrhagic property of esthesioneuroblastoma

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ABSTRACT

Esthesioneuroblastoma is a tumor arising from olfactory neuroepithelium with an incidence of four per million. This case presents a rare nasal cavity neoplasm with hemorrhagic properties that may lead to significant sequelae. We present a 69-year-old male patient who presented with worsening altered mental status over the past 6-7 months. His Glasgow coma scale was a nine, showing a large mass centered at the cribriform plate with extension intracranially, sinonasal cavity, and bilateral orbits on diagnostics. Bifrontal craniotomy was performed one day after admission with resection of the tumor in the cranial fossa and intranasally, and decompression of the brainstem, frontal lobes, and third ventricle. After surgery, the patient was managed in the intensive care unit but continued to deteriorate. He was confirmed to have no meaningful brain activity and eventually deceased seven days after admission. Depending on the tumor stage, esthesioneuroblastoma can have a favorable prognosis with proper therapies. Different surgical techniques for ENB lead to the question of which therapeutic modality is the best because of the tumor's gross hemorrhagic properties. With adjunctive radiation therapy and/or chemotherapy, more research can bring light to proper surgical techniques.

KEYWORDS: esthesioneuroblastoma; olfactory neuroblastoma; postoperative hemorrhage; frontal craniotomy; Hyams grading system; modified Kadish staging system

INTRODUCTION

Esthesioneuroblastoma (ENB), also known as olfactory neuroblastoma, is a rare tumor that arises from the olfactory neuroepithelium located commonly in the cribriform plate, roof of the nose, upper portion of the nasal cavity, and superior nasal concha which then can invade the skull and cranial structures [1]. ENB was first described by Berger and Luv in 1924 accounting for 3-6% of nasal cavity neoplasms [2]. The incidence of ENB is 0.4 per million with a mean age of 53, range of 35-70 years of age [3]. This case provides light on hemorrhagic properties of a rare nasal cavity neoplasm that can conclude in mortality after resection. Mortality seen in our patient brings in the question if different surgical approaches should be warranted. Our patient presented in an atypical presentation for esthesioneuroblastoma which ultimately ended in his demise.

CASE PRESENTATION

We present a 69-year-old patient that presented to the emergency department with worsening altered mental status over the past 6-7 months. He had a past medical history of bilateral cataracts and benign prostatic hyperplasia. He was accompanied by his friend, who provided all the information. He is not able to perform activities of daily living. He has lost 8.16 kilograms unintentionally during this time frame. The patient did not present with a headache, abdominal pain, chest pain, sinus drainage, or a recent fall/trauma. Vital signs showed a blood pressure of 160/101 mmHg with a mean arterial pressure (MAP) of 120.7 mmHg and 98% oxygen saturation on room air. Physical examination showed him to be alert and oriented to his name only, with a Glasgow Coma Scale of 9 (eyes 2, verbal 2, motor 5). Additional neurological assessment was difficult to attain with the status of the patient. He was drooling and urinating on himself. Cardiovascular and abdominal examinations were unremarkable. Initial lab work showed no abnormalities in his complete blood count, coagulation panel, complete metabolic panel, ammonia, and lipase levels that were ordered. Previous significant radiological findings include a fusiform aneurysmal dilation of the mid-descending thoracic aorta measuring 3.4 x 3.1 cm on a low-dose lung computed tomography (CT) scan in December 2022. In the emergency department, the patient received a CT head or brain without contrast which showed a large mass centered at the cribriform plate with intracranial extension, extension

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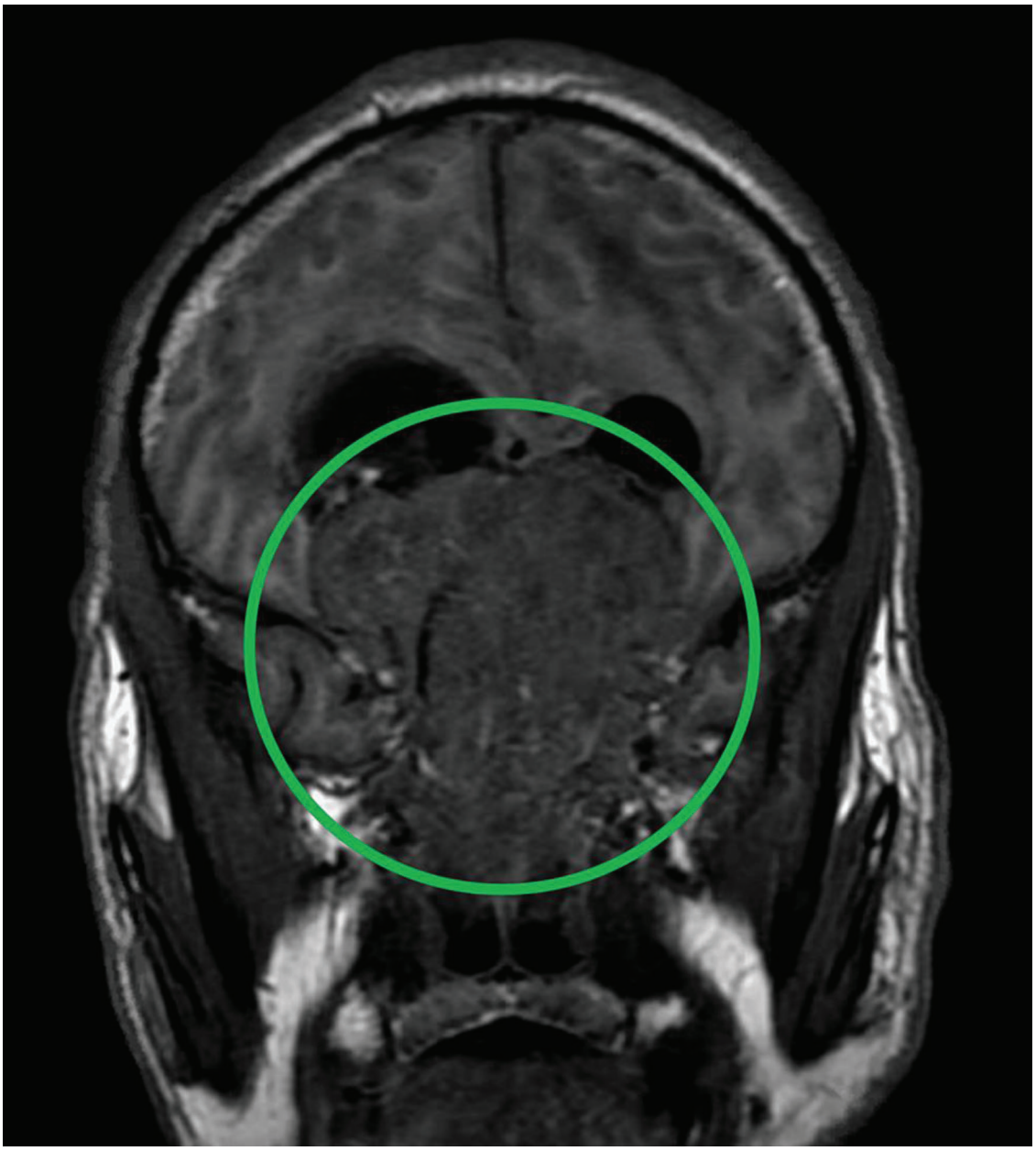

Fig. 1. Coronal view T1 magnetic resonance imaging with contrast showing a large mass aiding in the suspicion of ENB.

into the sinonasal cavity with osseous destruction, and minimal extension in the left greater than right orbits. This gave our patient a stage 3 on the Kadish scale as his tumor burden was beyond the sinus cavities and into the cranium. Magnetic resonance imaging (MRI) of the brain was then performed, confirming these findings, and raising suspicion of ENB. These findings can be seen in Figure 1. Also noted was that the mass had intracranial peripheral cystic components with vasogenic edema causing a 1.5 cm right-to-left midline shift. The patient was then admitted to the neurological intensive care unit (ICU) for further evaluation, and consultation with Neurosurgery was requested.

The patient was assessed and medically managed in the neurological ICU, and the decision for a bifrontal craniotomy was performed one day after admission. There was resection of the anterior cranial fossa tumor, decompression of the brainstem, frontal lobes, and third ventricle, and partial resection of the intranasal tumor component during the surgery. During the operation the tumor was observed to be extensive as it enveloped the cavernous sinus and constricted the carotid arteries. There was extensive hemorrhaging during the operation but it was controllable with cautery. Findings of the mass was reported as a large, extremely vascular, and extremely fibrous structure with the frozen section showing small blue cell malignant tumor. A lumbar drain was placed at the end of the procedure. The resected mass sent for pathological examination was positive on immunohistochemistry for synaptophysin, chromogranin, and CD56. S100 showed focal peripheral positive cells and SSTR2A was strongly and diffusely positive. This confirmed the mass to be a Hyams I/II low-grade ENB. The patient returned to the Neurological ICU slightly hypertensive at

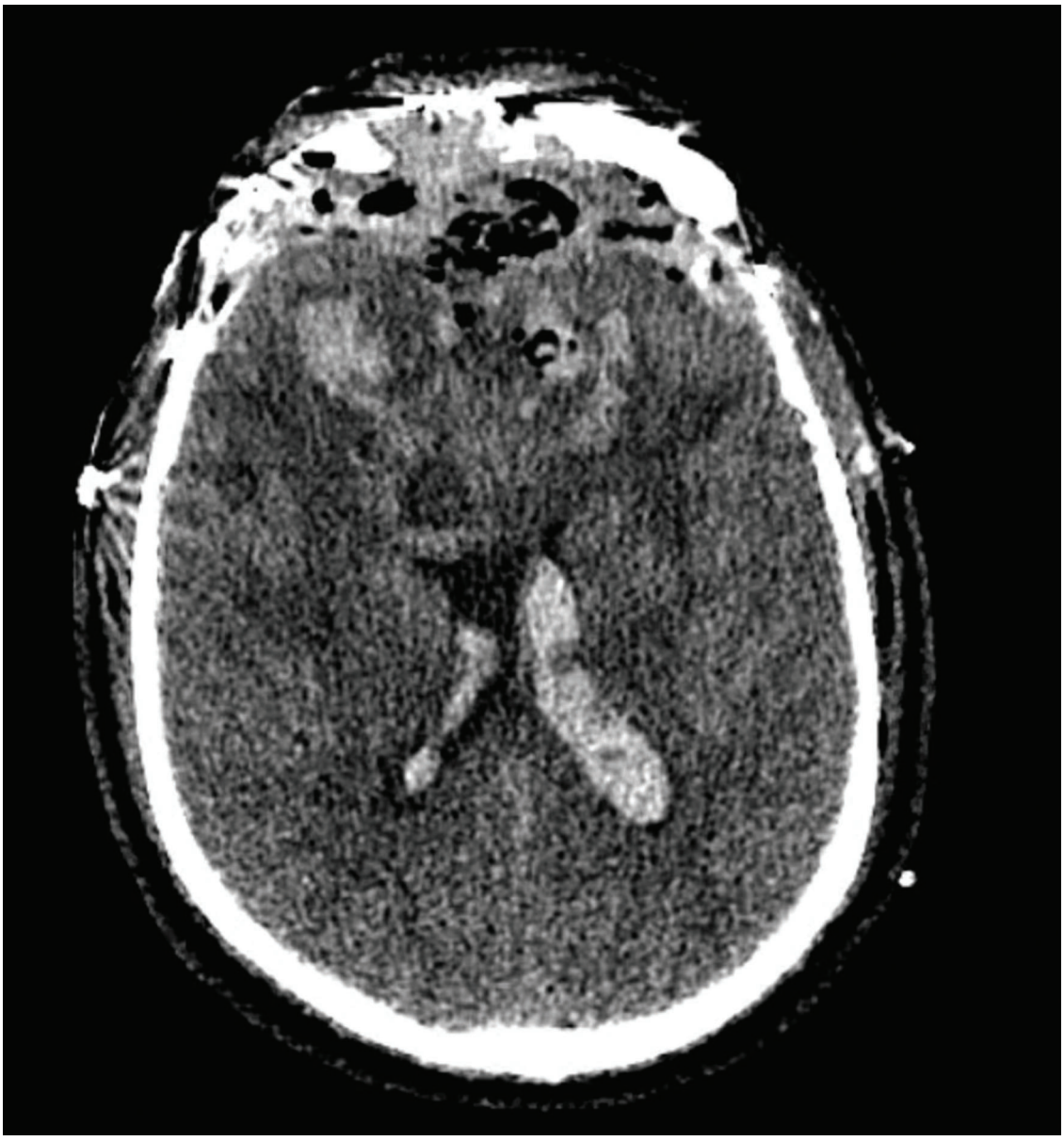

Fig. 2. CT head axial view w/o contrast showing post-operative high-density hemorrhage within the posterior and right basal frontal lobes which then extended into the suprasellar cistern and lateral, third, and fourth ventricles.

137/79 mmHg w/ MAP of 97.3 mmHg and was placed on a ventilator. He showed a rise in his lactic acid of 4.2 but this down trended during the next few days. Post-operative CT head or brain without contrast showed an improved mass effect, but newly formed fluid and hematoma accumulation was demonstrated within the subdural space. There was high-density hemorrhage within the posterior and right basal frontal lobes, extending into the suprasellar cistern and lateral, third, and fourth ventricles the following day seen in Figure 2.

The patient did not recover post-surgery and continued to deteriorate. He would not open his eyes or respond to painful or verbal stimulation on physical examination. Patient seemed to not be breathing over the vent. Corneal reflexes were not noted. Due to the patient's poor prognosis, palliative care was consulted. The patient was confirmed to have no meaningful brain activity, additionally shown with electroencephalogram. He was weaned off his ventilator setting, deceasing seven days after admission to the hospital.

DISCUSSION

ENB may have different clinical manifestations depending on the mass effect of the tumor. Common presenting symptoms include nasal obstruction, rhinorrhea, and sinus pain. Other manifestations that can be observed include proptosis, blindness, unilateral otalgia, retrobulbar pain, and unintentional weight loss [4]. Common imaging studies include CT and MRI for olfactory neuroblastoma. Still, they can use other modalities such as positron emission tomography/computed tomography and speckle reduction imaging [5]. For our patient, the pathology that was sent was confirmed to be a Hyams I/II low-grade ENB. Microscopically, olfactory neuroblastoma will show focal

ulceration and rich vascular supply depicting gross hemorrhagic properties. With these hemorrhagic properties, it is essential to be delicate with these tumors. If a biopsy is applicable, it must be done in the operating room to help control bleeding. The Hyams grading system grades tumors I to IV based on several pathological features such as mitotic activity and necrosis [6]. The tumor showed moderate pleomorphism and moderate mitotic activity, but no necrosis and consequently, low grade of the tumor. Literature to date shows a lack of controlled trials leading to a wide range of management for these tumors. Management comes from a multidisciplinary approach and depends on the Kadish stage criteria [7].

Surgical management primarily involves craniofacial resection through coronal scalp incision and bifrontal craniotomy [7]. With the invasiveness of this technique, it is found that there is an endoscopic approach to help patients cosmetically and minimize complications [7]. The invasiveness of the bifrontal craniotomy that our patient endured may make you wonder if his prognosis would have been better with an adjacent endovascular approach. Since our patient could not leave the ICU due to bleeding, he could not receive radiation therapy. Data indicates that combined surgery and radiotherapy can affect overall survival for patients [8]. Adjunctive chemotherapy has shown promising results for high-grade lesions defined as Hyams grade III or IV but shows little evidence to lower grade tumors described as Hyams grade I or II [1]. With no evidence of specific chemotherapy treatment, the standard follows regimens for neuroblastomas, small-cell lung carcinoma, and primitive neuroectodermal tumors [1].

With the dissemination of the disease, the most common place is the cervical lymph nodes. However, that incidence at the presentation of the disease is shown to be less than 10 percent of the patients [9]. Like many other tumors, the prognosis worsens once there is metastasis to lymph nodes. With the rare incidence of ENB and the favorable prognosis of the tumor when there are no lethal complications and appropriate treatment, distant metastasis can be prevented. This leads to a lack of research to treat this later state of progression. Further research can be attributed to the lack of knowledge of the distant spread of cancer and new therapeutic options to limit any lethal complications that can occur with surgery on this tumor.

CONCLUSION

ENB is a rare tumor and, depending on the stage of the tumor, can have a favorable prognosis with proper therapeutic pathways. Different therapeutic modalities may be used to treat this tumor depending on the Hyams grading scale. Adjunctive radiation therapy has been proven to improve the prognosis for patients, with chemotherapy being more beneficial with more high-grade lesions. With the different surgical techniques used for ENB, more research is needed to understand the most effective therapeutic modality for these patients because of the hemorrhagic properties the tumors present with.

Conflict of Interest

The authors declare that they have no competing interests.

Informed Consent

Written informed consent was obtained from the son of the patient for publication of this case report and accompanying images.

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