



## Case Report

## Olfactory neuroblastoma resection through endoscopic endonasal approach: A rare case report

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## ABSTRACT

**Background:** Olfactory neuroblastoma (ON) is a rare malignant tumor originating from the nasal cavity and/or skull base.

**Case presentation:** A 53-year-old female complained of a runny nose mixed with blood on the left nasal cavity since 1 year ago. Histopathology result showed malignancy and immunohistochemistry showed ON supported by CT-Scan result. The patient underwent endoscopic endonasal approach resection with anterior septectomy followed by debulking with cautery and cutting instruments. The patient received 35 times of radiotherapy combined with 6 times chemotherapy subsequently.

**Discussion:** Firstly, reported successful management of ON in Indonesia.

**Conclusion:** An ON case of an Indonesian female was successfully resected without complication through a join operation of endoscopic endonasal approach four-hand technique with a neurosurgeon.

## 1. Introduction

Olfactory neuroblastoma (ON) is a rare malignant tumor originating from the nose and/or skull base, 3–6% of sinonasal tumors. The standard surgical management for years is an anterior craniofacial approach including bifrontal craniotomy and lateral rhinotomy (LR) or Weber-Ferguson incision to get tumor resection. In this decade, therapeutic modalities have changed. The endoscopic endonasal approach is an alternative standard for the surgical management of ON [1,2]. The advantages of endoscopic procedures compared to transfacial and transcranial approaches are shorter surgery time and hospitalization, no external incision, minimal brain parenchymal injury, no neurovascular structural manipulation, and no oncologic outcome difference even if piecemeal resection is needed [3]. Based on the reports, we are interested in reporting cases of ON in Indonesian women who have successfully carried out the endoscopic endonasal approach reported using the SCARE 2020 Guideline [4].

## 2. Case presentation

A 53-year-old Indonesian female complaint of blood-stained discharge from her left nasal cavity since 1 year along, almost every

day, and could stop spontaneously, with left nasal obstruction since 2 months ago and left side headache. There was no complaint of the eye, ear, and throat area and no lump in the neck. There was no numbness in the facial area and no anosmia was found. Nasal examination revealed a spacious nasal cavity, no visible mass, and secretion on both sides. No enlarged lymph node was found in the neck. The patient was a housewife. Patient had undergone biopsy surgery in the same area one year ago. Histopathology result of left nasal cavity mass biopsy revealed a malignant round cell tumor differential diagnosed with ON, neuroendocrine, and undifferentiated carcinoma. Immunohistochemistry examination concluded an ON.

Computerized tomography scan (CT-scan) with contrast showed enhanced solid lesion with the size of  $1.8 \times 1.2 \times 2.1$  cm olfactory region with left side predominant, extended to the anterior part of both nasal cavities (Fig. 1). Preoperative nasoendoscopy (NE) showed a mass on the olfactory region in both nasal cavities (Fig. 2). Magnetic resonance imaging (MRI) examination with contrast showed an image ON with other descriptions that corresponded with CT scan result. It was found a dura metastasis on vertex convexity (Fig. 3). Diagnosis ON stage 3C based on Kadish criteria [5].

Tumor resection was performed through a join operation of endonasal endoscopic approach four-hand technique with a neurosurgeon,

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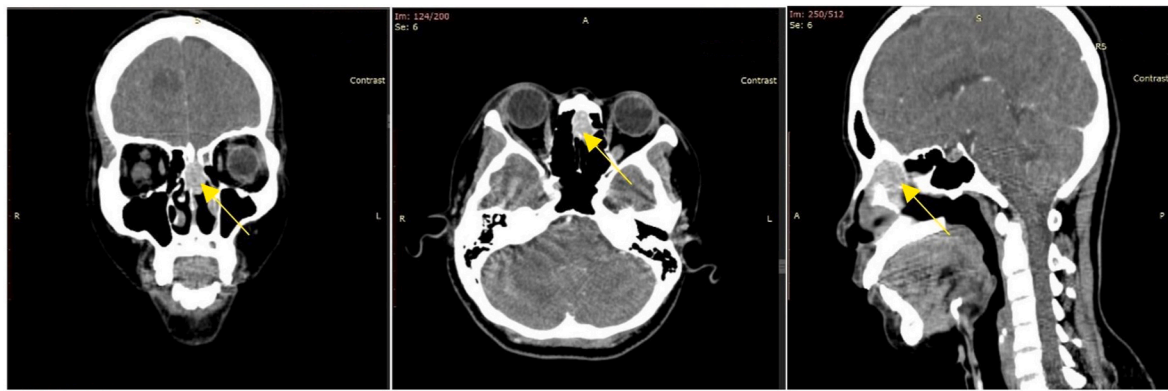
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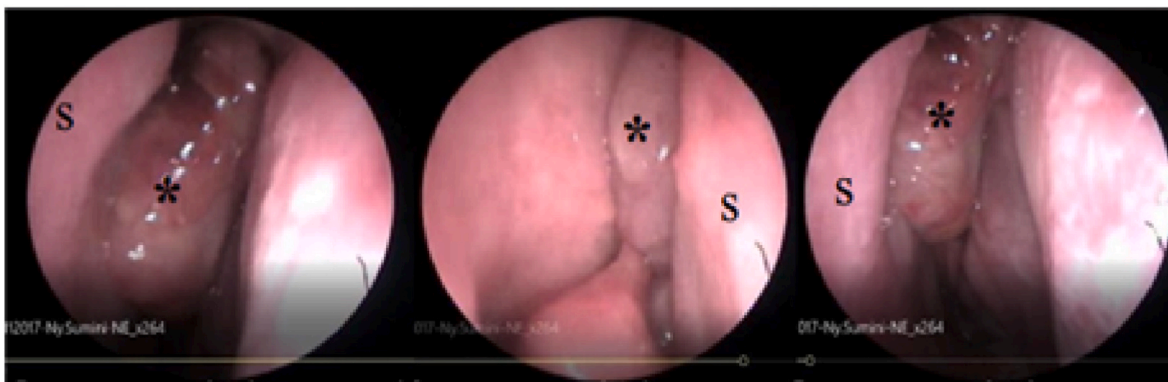
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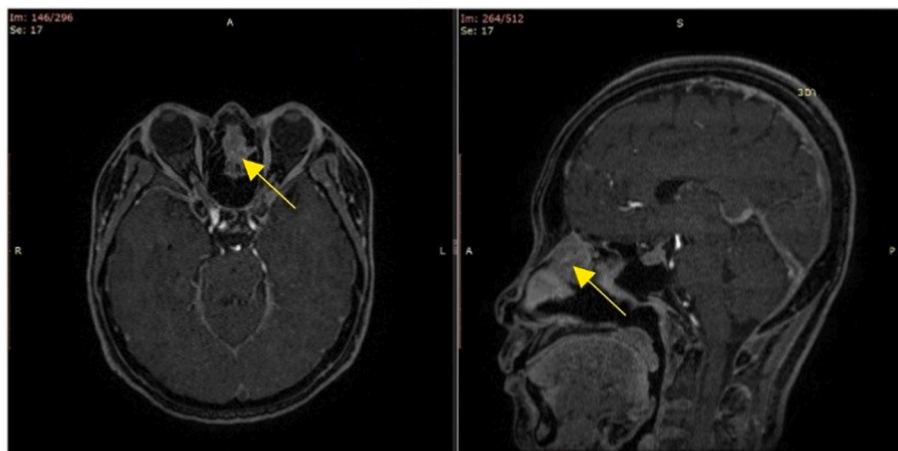
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**Fig. 1.** CT-scan paranasal sinuses of coronal, axial, and sagittal views. Mass in the olfactory region left side dominant that destructed nasal cavity and extended to the left and right nasal cavity anterior side (yellow arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)



**Fig. 2.** Nasoendoscopy found the mass in the upper right and left nasal cavity (\*), septum nasal (S).

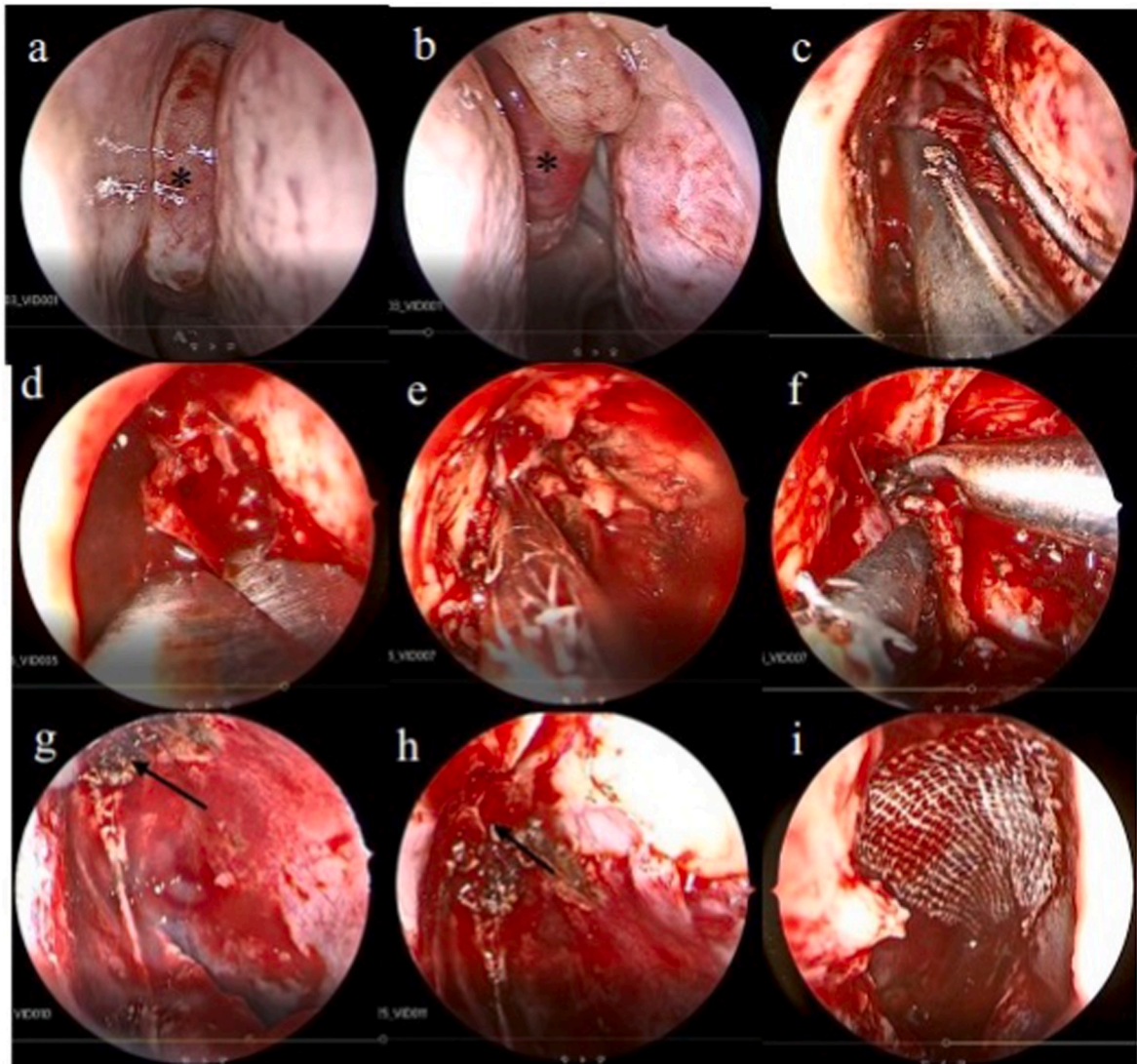


**Fig. 3.** Head MRI with contrast axial, sagittal, and coronal views. Mass in the olfactory region with left side dominant destructed left-right nasal cavities anterior side and extended to left ethmoidal sinus (yellow arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

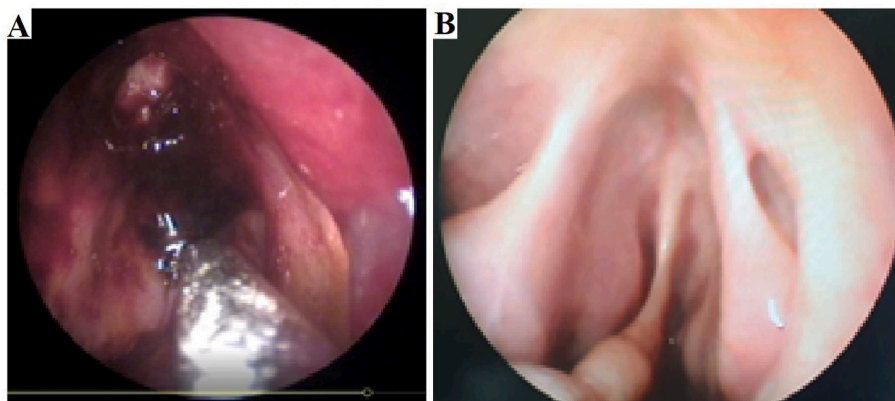
began with anterior septectomy, and continued with tumor debulking until it reached the cribriform plate. The bone of the cribriform plate was intact. Bleeding was controlled with suction and cauterization. Blood stop® was applied to the surgical area. Blood loss was about 100 ml and tumor specimens were sent for the histopathology examination. Postoperative histopathology result concluded an ON (Fig. 4).

Postoperative management included supine position 20° head elevation, nose-blowing, leaning forward, and straining were not

allowed. The patient was advised to open her mouth whilst sneezing or coughing. The patient was discharged on the following day. Nasal saline irrigation was administered for a month postoperatively. One week postoperative showed nasal crusting on the surgical site. Optimal and safe debridement was performed (Fig. 5a). Fifteen months postoperatively showed no recurrence. Smell ability was permanently lost (Fig. 5b). The patient had received chemotherapy six times and radiotherapy 35 times.



**Fig. 4.** Tumor resection with the endoscopic showed endonasal approach. (a–b) Tumor in the superior intranasal (\*). (c–d) Septectomy. (e–f) Tumor was covered resection processing. (g–h) Condition after tumor removal. It shows the cribriform plate area (arrow). Most of the septum was removed. (i) Cribriform plate was covered with Blood Stop®.



**Fig. 5.** Nasoendoscopy (a) One week after surgery. (b) Fifteen months after surgery. Mucosa was healed, no recurrent tumor was seen.

### 3. Discussion

ON is a rare malignant neoplasm derived from the superior portion of

the nasal cavity [6,7]. Imaging is crucial for the diagnosis and staging. A combination of CT-scan and MRI is usually required. A fine-cut CT scan (1.5 mm thick cut) is the best initial imaging for ON shows a

homogeneous soft mass of the nasal cavity with moderate and uniform enhancement. The primary advantage of CT scans is to determine bone involvement superior to MRI. CT scan help to evaluate bone erosion of the cribriform plate, fovea ethmoidalis, and lamina papyracea. MRI on ON is the gold standard for evaluating detailed extensions and staging. Dumbbell shape tumor extending into the cribriform plate is a rare finding [6,8].

Over the past two decades, endoscopic approaches have been popular as a treatment approach for ON [6]. Stammberger et al. first published in the English literature their experience with endoscopic treatment of nasal malignancies, including ON [9]. Surgeries were carried out under the dual supervision of a rhinologist and a neurosurgeon [1]. The advantages of endoscopic procedures, compared with transfacial and transcranial approaches, are the shorter duration of operation time, no external incision, minimal brain parenchymal injury, no manipulation of neurovascular structures, no need for blood transfusions, and no difference in oncological results even if piecemeal resection is necessary [3,10]. The operation time required for the endoscopic technique is shorter than those in patients treated with traditional craniofacial resection (CFR) techniques with craniotomy [8]. Complete endoscopic resection can be achieved in Kadish A, B. Kadish C resection with intracranial extension can be performed with endoscopic approach plus bifrontal craniotomy [11].

Transethmoid and transcribriform approaches can be done unilaterally and bilaterally dependent on lesions [12]. In general, septectomy was performed to facilitate a binostril 3-handed to 4-handed approach. Wide paranasal sinus dissection was performed involving the maxillary, ethmoid, sphenoid, and frontal sinuses. An endoscopic modified Lothrop procedure and wide sphenoid drill-out were performed to obtain anterior and posterior boundaries, respectively, for the skull base resection [1].

Surgical resection followed by postoperative radiation provides the best end-stage results in the life level and decreases the incidence of local recurrence. Most of the institutions adopt surgical resection followed by postoperative radiotherapy as a standard therapy approach. Postoperative radiation showed improved local control of the disease. Adjuvant radiotherapy (ART) is given in all cases in the form of intensity-modulated radiotherapy (IMRT). The radiation dose used is usually in the range 55–65 Gy given in 2 Gy per dose, which is divided into 30–33 doses [11,13,14]. The use of chemotherapy in ON is still controversial regarding the optimal indication and best drugs to be used. Chemotherapeutic agents were cisplatin or cisplatin plus etoposide [1, 14].

ON has a good survival outcome, with an average of 5-year survival rates reported between 57 and 93%. According to the staging system by Kadish, there is a decrease in life expectancy along with an increase in clinical staging. ON has a high recurrence rate and requires long-term follow-up. The highest risk is locoregional recurrence with most frequently occurring between 5 and 10 years. Long-term follow-up to 10–15 years is recommended. The risk of recurrence is higher in those with poor prognostic factors such as high Hyams degrees, advanced Kadish stage, intracranial extension, positive resection limits, presence of cervical lymph node metastases (35%), and distant metastases (10%). Additional negative prognostic indicators include female gender, age <20 or > 50 years at early diagnosis, extensive intracranial involvement, and high proliferation index [1,6,14].

Recurrence of diseases involving the dura layers is common [8,15]. Uncommonly, ON may present as an intracranial mass in the frontal lobe with involvement of the cribriform plate, even more rarely, it may appear as an intracranial tumor without an intranasal component [14].

#### 4. Conclusion

A 53-year-old Indonesian female was diagnosed with ON successfully performed a join operation of endoscopic endonasal approach four-hand technique with the neurosurgeon. Fifteen months follow-up showed no

recurrence. A surgical resection followed by chemoradiation would become the standard of management for ON.

#### Ethical approval

Not applicable.

#### Sources of funding

None.

#### Author contribution

All authors contributed toward data analysis, drafting and revising the paper, gave final approval of the version to be published and agree to be accountable for all aspects of the work.

#### Registration of research studies

Name of the registry:

Unique identifying number or registration ID:

Hyperlink to your specific registration (must be publicly accessible and will be checked):

#### Guarantor

Budi Sutikno.

#### Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Provenance and peer-review

Not commissioned, externally peer reviewed.

#### Consent

We have requested the patient's consent to publish this case report for educational purposes.

#### Declaration of competing interest

The author declare that they have no conflict of interest.

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