Intraosseous schwannoma in the clivus mimicking chordoma treated with endoscopic endonasal surgery: A case report

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INTRODUCTION

Schwannomas comprise 8.6% of central nervous tumors in the Unites States,[19] and these tumors are the third most common benign brain tumors. Although neurosurgeons encounter them frequently, intraosseous schwannomas are extremely rare, accounting for <0.1–0.2% of all primary bone tumors.[12] Because most schwannomas originate from peripheral nerves and are associated with sensory nerves,[1,12] the vestibular nerve is the most frequent origin of intracranial schwannomas, and the clivus is not usually invaded by these tumors.

Here, we present the first case, to the best of our knowledge, of a schwannoma located in the clivus that was treated with endoscopic endonasal surgery (EES). We discuss the
preoperative radiological diagnosis, origin, and treatment of this tumor.

CASE REPORT

A 62-year-old man presented with a 5-year history of gradually worsening hoarseness. He was referred to our department, because magnetic resonance imaging (MRI) revealed a mass lesion in his clivus. His medical history included diabetes mellitus and hyperlipidemia, and he had no remarkable family history. Neurological examination revealed dysphagia and atrophy of the left tongue, trapezius muscle, and sternocleidomastoid muscle. There were no abnormal laboratory findings, including blood counts or alkaline phosphatase levels. Laryngoscopy revealed paresis of the left vocal cord. MRI showed that the tumor was of moderately high intensity, accompanied by cystic components on T2-weighted images and heterogeneous enhancement mainly in the peripheral area with gadolinium (Gd) on T1-weighted images [Figures 1a and b]. The tumor was mainly located in the clivus and it extended down to the left jugular tubercle and occipital condyle, partially protruding toward the medulla oblongata from the hypoglossal canal [Figure 1b]. A computed tomography (CT) scan revealed an osteolytic lesion that destructed the hypoglossal canal and expanded the clivus while preserving the bony cortex [Figures 1c-e].

Our preoperative diagnosis was chordoma, schwannoma, or chondrosarcoma; however, the diagnosis was inconclusive due to the atypical MRI findings.

To define the histological diagnosis, treat the patient's symptoms, and decompress his brain stem, EES was performed using intraoperative MRI. A rapid pathological diagnosis was planned to determine the radicality of resection depending on the tumor malignancy. Nerve integrity monitoring (NIM, Medtronic, Minneapolis, MN, USA) was used to localize the lower cranial nerves (IX, XI, and XII). After dissection of the nasal septal mucosa and wide sphenoidotomy with drilling of the anterior wall, septum, and inferior wall of the sphenoid sinus, the upper part of the tumor was exposed. The tumor seemed to have no typical feature of chordoma or chondrosarcoma, rather of schwannoma. Rapid pathology of a specimen from this part of the tumor revealed no malignant findings and features of typical schwannoma. Considering the intraoperative findings and pathological result, not radical but maximum safe resection was intended. Subsequently, a left transmaxillary-transpterygoid approach with dissection of the nasopharyngeal mucosa was performed to expand the surgical field laterally and caudally. The left Vidian nerve and artery were cauterized and cut, and the contents of the left pterygopalatine fossa were lateralized to facilitate resection of the left medial pterygoid plate. The nasopharyngeal mucosa was incised in an inverted U-shape, dissected, and caudally

Figure 1: Preoperative magnetic resonance imaging. Axial T2-weighted (a) and T1-weighted images with gadolinium enhancement (b) images show that the tumor is isointense with cystic components and is enhanced mainly in the peripheral area. The tumor is mainly in the clivus, and the small part of the tumor protrudes toward the brainstem around the left hypoglossal canal (the left hypoglossal canal: b,c, and e, white arrow). Axial (c and d) and coronal (e) bone window computed tomography images show destruction of the hypoglossal canal and jugular foramen (d and e, white dot arrow). The clival bone itself is expanded due to the tumor invasion (c and d), and the bony cortex is preserved in most parts (d, arrowhead).
reflected with a monopolar electrocautery to expose the lower clivus. The bone overlying the left paraclival carotid artery was removed, and the fibrocartilaginous tissue attached to the internal carotid artery (ICA) and Eustachian tube around the foramen lacerum was disconnected to enable mobility of these structures [Figure 2a]. The tumor in the jugular tubercle and occipital condyle behind the ICA and Eustachian tube were resected. The cortical bone in the clivus was thinned, and the cancellous bone was replaced by the tumor [Figure 2b]. The tumor was tanned and gray-yellowish, soft and fragile in most parts, and easily dissected from the clival dura. The left cranial nerve IX, X, or XI was observed at the rostral margin of the tumor around the jugular foramen during tumor removal [Figure 2c]. After resecting most of the tumor, we performed an intraoperative MRI scan, which showed a residual tumor lateral to the left occipital condyle and a small intradural tumor adjacent to the medulla oblongata. These tumors were additionally excised, except for the most lateral part. Subsequently, the arachnoid membrane appeared to be bulging from the dural defect and cerebrospinal fluid leak was repaired with Gelfoam (Pfizer, New York, NY, USA) and DuraGen (Integra LifeSciences, Princeton, NJ, USA). Finally, the surgical field was covered with the nasopharyngeal mucosa and harvested sphenoid sinus mucosal flap [Video 1].

The histopathological findings revealed typical schwannoma, which showed spindle cells arranged in a palisading pattern with Verocay bodies (Antoni A) and a small number of tumor cells dispersed in edematous stroma (Antoni B). The tumor cells were positive for S100 protein, and the prevalence of Ki-67 positive cells was <1%. Based on the histology, preoperative MRI, and intraoperative findings, the tumor was considered an intraosseous schwannoma in the clivus.

Postoperative MRI showed subtotal resection of the tumor with successful decompression of the brain stem with a small residual tumor lateral to the occipital condyle [Figures 3a and b]. Nine days after the surgery, the patient was discharged without complications, including new neurological deficits. The preoperative IX, X, XI, and XII cranial nerve palsies remained unchanged. The residual tumor showed no growth as observed through MRI 1 year after the surgery.

DISCUSSION

Intraosseous schwannomas are rare, because there are very few sensory nerves in bone,[7,22] and several cases of intraosseous schwannoma have been reported. However, intraosseous schwannomas in the clivus have not been reported, and preoperative radiological diagnosis is extremely difficult, because it mimics clival chordoma. In the current case, the lesion was successfully resected using EES.

Origin of the tumor

Intraosseous schwannomas frequently occur in the mandible, sacrum, long bone, and vertebrae.[7,22] Seventeen cases of intraosseous schwannomas of the skull have been previously reported [Table 1]. The sites affected by the tumor were discharged without complications, including new neurological deficits. The preoperative IX, X, XI, and XII cranial nerve palsies remained unchanged. The residual tumor showed no growth as observed through MRI 1 year after the surgery.

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the petrous bone in eight cases,\textsuperscript{8,11,20,23,29,30} frontal bone in three,\textsuperscript{1,4,9,10} occipital bone in two,\textsuperscript{4,17} parietal bone in one,\textsuperscript{7} frontoparietal bone in one,\textsuperscript{14} fronto-orbital bone in one,\textsuperscript{26} and sphenoorbital bone in one.\textsuperscript{6} To the best of our knowledge, schwannomas involving the clivus have not yet been reported. The origin of the tumor is presumed on the basis of the tumor location on imaging and neurological and intraoperative findings.\textsuperscript{6,14,25,30} Three mechanisms of schwannomas involving the bone are proposed as follows: extraosseous tumors erode bone secondarily; tumors arise in the center of bones; and tumors arise and grow in the nutrient canal, which forms a dumbbell-shaped configuration.\textsuperscript{19} Intraosseous schwannomas are commonly defined when all parts of the tumor are located within the bone. However, the term “intraosseous schwannomas” is not precisely defined,\textsuperscript{34} and several authors defined tumors mainly located in intraosseous region as intraosseous schwannomas, even if the tumors were associated with extraosseous invasion.\textsuperscript{13,21,24} In our case, the tumor was mainly located in the clivus; therefore, it was defined as an intraosseous schwanna, although a small part of the tumor was in the extraosseous region. In addition, the tumor protruded toward the medulla oblongata along with the hypoglossal nerve and expanded the hypoglossal canal as observed through preoperative MRI; therefore, it appeared to originate from the hypoglossal nerve. Arnold’s nerve, which is a branch of the vagal nerve, was also considered an origin of the tumor, because this nerve runs in the petrous bone; however, this nerve did not appear to be the origin of the tumor in our case based on the site of intradural tumor extension along the hypoglossal nerve. Thus, the origin of the tumor was not clearly defined, but the hypoglossal nerve was the most plausible origin in our patient.

**Radiological diagnosis**

Schwannomas rarely arise and extend into the clivus, and if they arise there, they mimic chordoma, which frequently arises in the clivus, as observed in our patient. The radiological differential diagnoses of clivus lesions include chondrosarcoma, fibrous dysplasia, multiple myeloma, and metastatic tumors in addition to chordoma. Chondrosarcoma frequently arises along the petro-occipital fissure and petrous bone,\textsuperscript{2,15} and most of the tumors are located laterally in the posterior cranial fossa. Fibrous dysplasia typically shows a ground-glass, sclerotic, or cystic appearance on CT images.\textsuperscript{13} Multiple myeloma and metastatic tumors usually show erosion of the cortical bone on CT images.\textsuperscript{18} Skull base chordoma is typically located in the center of the clivus and presents with extensive bone destruction, including the cortex on CT scans.\textsuperscript{24} T2-weighted MR images are of high intensity due to the high protein content of tumors,\textsuperscript{16} which are separated into multilobulated lesions.\textsuperscript{15} In our patient, the preoperative differential diagnosis of chordoma or schwannoma was difficult, because most of the tumor was in the clivus, which is a nontypical location. Radiological features of intraosseous schwannomas usually show lytic and expansile bone lesions without cortical erosion on CT images.\textsuperscript{12,22} On MRI, schwannomas often appear as nonspecific findings, hypo-iso signals on T1-weighted images, iso-hypersignals on T2-weighted images, and moderate Gd contrast enhancement of the solid component of the tumors. Cystic components or hemorrhages are frequently found in tumors.\textsuperscript{15} However, when compared to chordoma, schwannomas generally have a lower signal on T2-weighted images, expand, and erode bone without cortex destruction (observed on CT images) and contain cystic components or hemorrhages. MRI and CT findings in our patient were more typical of schwannomas than of chordoma due to these characteristics of intraosseous schwannomas. Although intraosseous schwannomas in the clivus are extremely rare, if findings of CT images and T2-weighted images are similar to those of our present case, schwannomas should be included in differential diagnosis of the tumor in the clivus.

**Treatment and prognosis**

In our patient, intraosseous schwannomas in the clivus were successfully resected using an endoscopic endonasal transmaxillary-transpterygoid approach without any complications. Most of the reported cases of intraosseous schwannomas in the skull have been treated through craniotomy, except for one case that was treated with radiation as the first line of treatment [Table 1]; therefore, to the best of our knowledge, the present case is the first one involving treatment with EES. The tumor was resected with EES, because it was located in the clivus. EES is generally less invasive than craniotomy, and with the advancement of EES in recent years, tumors in and around the clivus can be radically resected using expanded EES without injuring the cranial nerve.\textsuperscript{32} For tumors extending to the petrous bone,
### Table 1: Reported cases of intraosseous schwannomas of the skull.

| Authors                  | Age/Sex | Site                        | Bony findings on CT | Intensity of tumor on T2-weighted MRI | Treatment | Follow-up (mo) | Recurrence (Y/N) |
|--------------------------|---------|-----------------------------|---------------------|--------------------------------------|-----------|----------------|------------------|
| Solodnik et al. (1986)   | 59/M    | Petrous apex                | Erosion, Expansion  | NR                                   | TCS       | NR             | NR               |
| Schiffer et al. (1991)   | 3/M     | Fronto-orbital bone         | Erosion             | NR                                   | TCS       | 12             | N                |
| Horn et al. (1995)       | 46/F    | Petrous apex                | Erosion, Expansion  | Iso                                  | TCS       | 36             | N                |
| Celli et al. (1998)      | 3/M     | Occipital bone              | Erosion             | NR                                   | TCS       | NR             | N                |
| Erşahin et al. (2000)    | 14/M    | Frontal bone                | Erosion, Expansion  | Hyper                               | TCS       | 24             | N                |
| El-Bahy (2004)           | 40/M    | Spheno-orbital bone         | Erosion, Expansion  | Hyper                               | TCS       | NR             | N                |
| Goyal et al. (2008)      | 11/M    | Frontal bone                | Erosion, Expansion  | Hyper                               | TCS       | NR             | N                |
| Goiney et al. (2011)     | 48/F    | Petrous apex                | Erosion, Expansion  | Iso                                 | TCS       | NR             | NR               |
| Parikh et al. (2013)     | 26/F    | Petrous apex                | Erosion             | Iso                                 | SRT       | 12             | NR               |
| Amita et al. (2014)      | 41/F    | Frontal bone                | NR                  | Hyper                               | TCS       | 3              | N                |
| Tamura et al. (2015)     | 47/M    | Petrous apex                | Erosion, Expansion  | Hyper                               | TCS       | NR             | N                |
| Kawai et al. (2016)      | 24/F    | Frontoparietal bone         | Erosion, Intact cortex | Hyper                           | TCS       | 60             | N                |
| Mathieu et al. (2018)    | 7/M     | Occipital bone              | Erosion, Intact cortex | Hyper                           | TCS       | 2              | N                |
| Sato et al. (2019)       | 35/F    | Petrous apex                | Erosion, Expansion  | Iso-hyper                           | Biopsy    | 84             | N                |
| Rozman et al. (2019)     | 68/F    | Petrous apex                | Erosion, Expansion  | Iso-hyper                           | TCS       | 22             | Y                |
| Our case                 | 62/M    | Clivus                      | Erosion, Expansion  | Iso-hyper                           | EES       | 12             | N                |

mo: Month, N: No, Y: Yes, M: Male, F: Female, TCS: Transcranial surgery, SRT: Stereotactic radiation therapy, RT: Radiation therapy, EES: Endoscopic endonasal surgery, NR: Not reported, Iso: Isointensity, Iso-hyper: Iso-hyperintensity

In addition, the nasopharynx route was selected to reach the tumor invading the lower clivus. In this case, the schwannoma was mainly located in the clivus and it extended from the petrous bone around the foramen lacerum to the jugular tubercle and occipital condyle, extending laterally behind the ICA close to the sigmoid sinus and internal jugular vein. Using the transmaxillary-transpterygoid approach, we created a surgical corridor to the lateral area of the lower clivus. Using the translacerum approach, we mobilized the paracral and petrous segments of the ICA around the foramen lacerum and Eustachian tube while preserving the Eustachian tube by removing the fibrocartilaginous component. The tumor was subtotally resected using a combination of these approaches.
Only one previously reported case with a high proliferation index (Ki-67 index, 10%) showed recurrence after surgery, and stereotactic radiation therapy was performed. No malignant intraosseous schwannomas of the skull have been reported. If the residual tumor lateral to the occipital condyle progresses in future, stereotactic radiation therapy will be planned in our case.

CONCLUSION

Here, we report a case of intraosseous schwannoma in the clivus mimicking clival chordoma. The tumor was subtotally resected without any complications with EES using the transmaxillary-transpterygoid and translacerum approaches. Although an intraosseous schwannoma in the clivus is extremely rare, it should be included as one of the differential diagnoses of clivus lesions if the lesion is expanding the bone while preserving the cortex on CT images or is not hyperintense on T2-weighted MR images.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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