CASE REPORT

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Rare sphenoid ridge intracranial mature teratoma in an adult female

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ABSTRACT

Intracranially located teratomas usually involve midline brain structures. However, they rarely occur in adults. A 26-year-old woman presented with a growing intracranial mass lesion in the left sphenoid ridge without neurological deficits. Magnetic resonance imaging revealed homogenous hyperintensities without contrast enhancement. The patient underwent gross total excision of the soft, yellowish sphenoid ridge tumor with no cystic component. The surgery was uneventful, with no intraoperative complications. Histological analysis revealed a mature teratoma. She attended regular outpatient neuroradiology follow-up appointments. The present case is an unusual example of a mature teratoma with regard to location, neuroimaging appearance, macroscopic intraoperative findings, histological tumor subtype, and patient age and sex.

Keywords: intracranial tumor, mature teratoma, sphenoid ridge

Abbreviations:
CT: computed tomography
MRI: magnetic resonance imaging

BACKGROUND

Central nervous system germ cell tumors are rare, comprising approximately 0.3%–0.6% of all primary intracranial tumors.[1,2,3,4,5] They are divided into germinomas, nongerminomatous, and mixed germ cell tumors.[3] Teratomas belong to the nongerminomatous type, which are derived from all three germ cell layers seen during organogenesis, and they are hypothesized to arise from misplacement of pluripotent germ cells.[3,4,5]

Teratomas are generally rare in adults and only account for 3% of all childhood tumors, with the majority occurring in the sacrococcygeal region and gonads, including ovarian tumors.[6,7] Histologically, these tumors are strikingly similar, regardless of their location[7]. However, teratomas

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that are intracranially located rarely occur, with only a few reported cases in adults. Even when it occurs intracranially, they preferentially involve midline brain structures.[5,7,8] Teratomas are histologically classified into mature teratomas, immature teratomas, and teratomas with malignant transformation.[4,7,9] Of these, the immature variant is the most common intracranial type.[10] It is rare for the mature variant to occur within the central nervous system in adulthood. Here, we report a rare case of a sphenoid ridge, an intracranial mature teratoma, in an adult female patient.

CASE PRESENTATION

A 26-year-old woman presented to a nearby hospital 9 years ago following head trauma. Computed tomography (CT) revealed an asymptomatic intracranial mass in the left sphenoid ridge (Figure 1a). Initially, she was managed conservatively with annual neuroimaging checkups; however, she subsequently did not attend follow-up appointments for about 5 years. She later reappeared for a follow-up appointment. A repeat magnetic resonance imaging (MRI) showed significant tumor growth.

Preoperative contrast CT showed an irregular hypodense mass in the left sphenoid ridge (Figure 1b). MRI revealed a mass with increased intensities on T1-weighted and T2-weighted images with no perilesional edema or contrast enhancement (Figure 2).

The patient underwent microsurgical resection. A frontal craniotomy was performed, and the medial Sylvian fissure was separated. The intraoperative findings consisted of a soft suckable yellowish tumor (Figure 3) with no adherence to the brain or vessels, except for the wall of the M1 segment of the left middle cerebral artery. Gross total tumor resection was accomplished. The postoperative course was uneventful.

Postoperative MRI showed complete removal (Figure 4). Histological analysis revealed that the tumor consisted of mature cartilage tissues, ductal component, stratified squamous epithelium, hair follicle, and sebaceous glands (Figure 5). According to the World Health Organization tumor classification, these findings correspond to a mature teratoma.[11] At present, the patient attends regularly scheduled outpatient neuroradiology follow-ups.

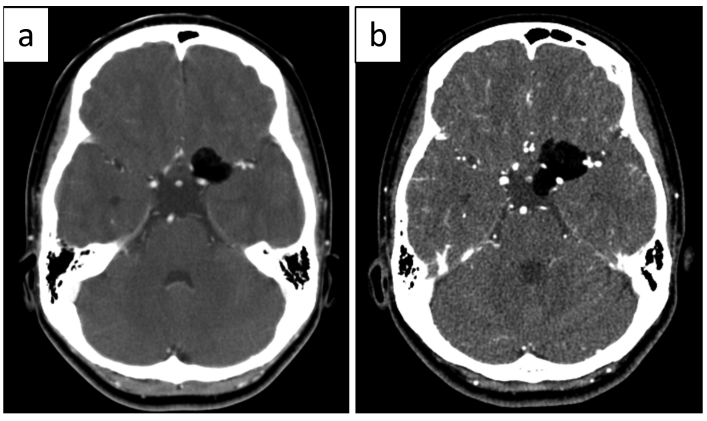

Fig. 1 Preoperative images

Fig. 1a-1b: Comparison of preoperative contrast cranial computed tomography (CT) scan (b) and initial neuroimage from 9 years prior (a) shows evidence of a size increase of the irregular hypodense mass in the left sphenoid ridge.

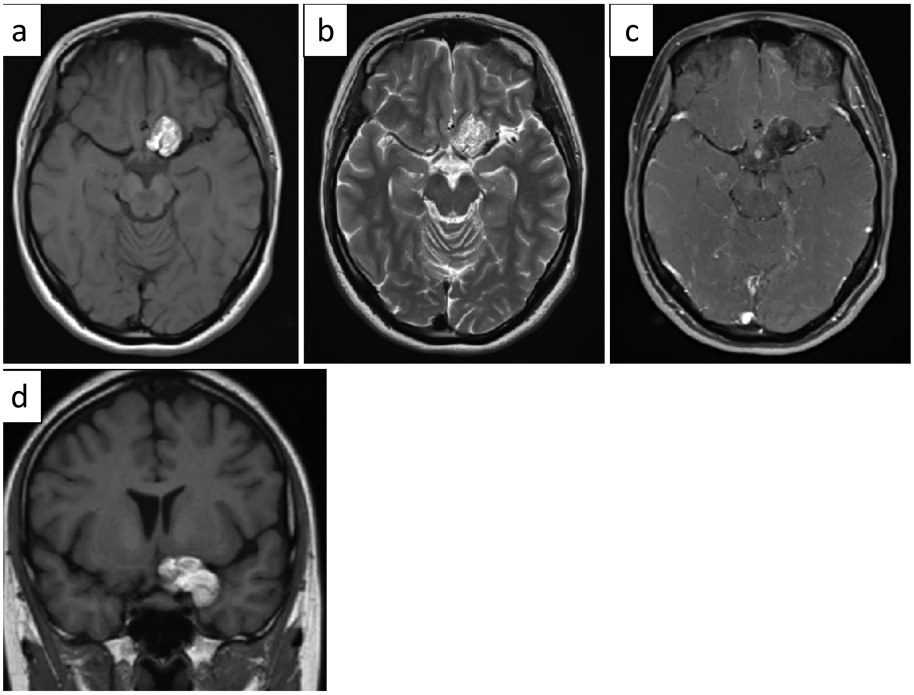

Fig. 2 Preoperative magnetic resonance imaging (MRI)
Fig. 2a and 2d: MRI showing hyperintense lesions on T1-weighted imaging in axial (a) and coronal (d) angles.
Fig. 2b: T2-weighted MRI showing a hyperintense lesion on the left sphenoid ridge.
Fig. 2c: Gadolinium MRI contrast image with fat suppression showing a non-enhanced lesion with suppression.

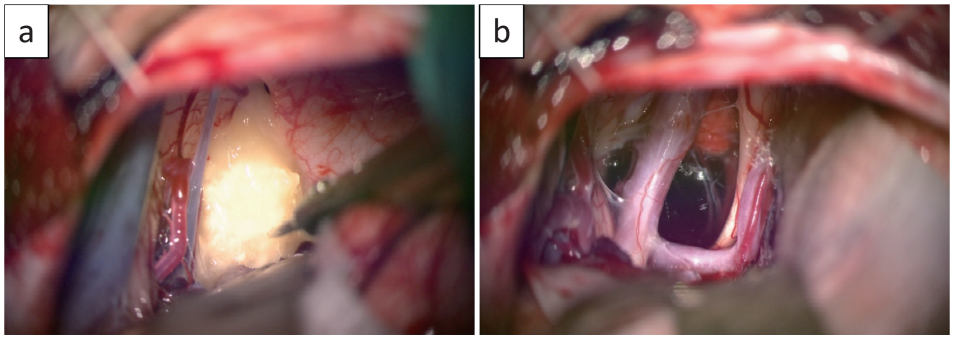

Fig. 3 Intraoperative photograph showing tumor removal
Fig. 3a: Yellowish and soft tumors were excised from the left sphenoid ridge.
Fig. 3b: View of post-tumor excision with intact surrounding vessels.

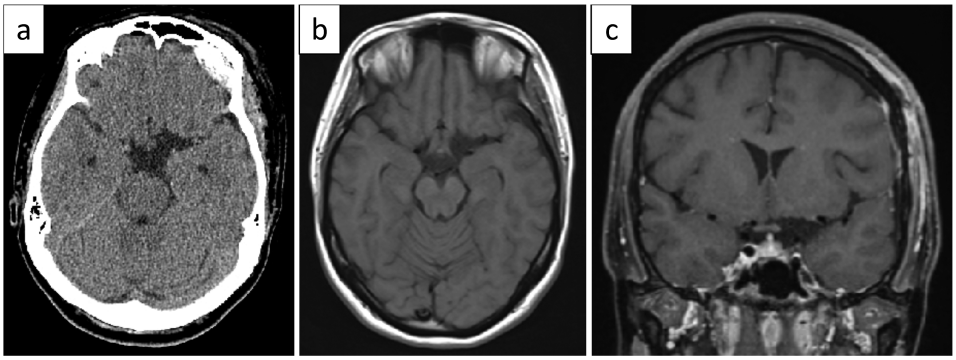

Fig. 4 Postoperative neuroimages showing complete tumor extirpation
Fig. 4a: Axial computed tomography scan.
Fig. 4b: T1-weighted axial magnetic resonance imaging (MRI).
Fig. 4c: Postcontrast T1-weighted coronal MRI.

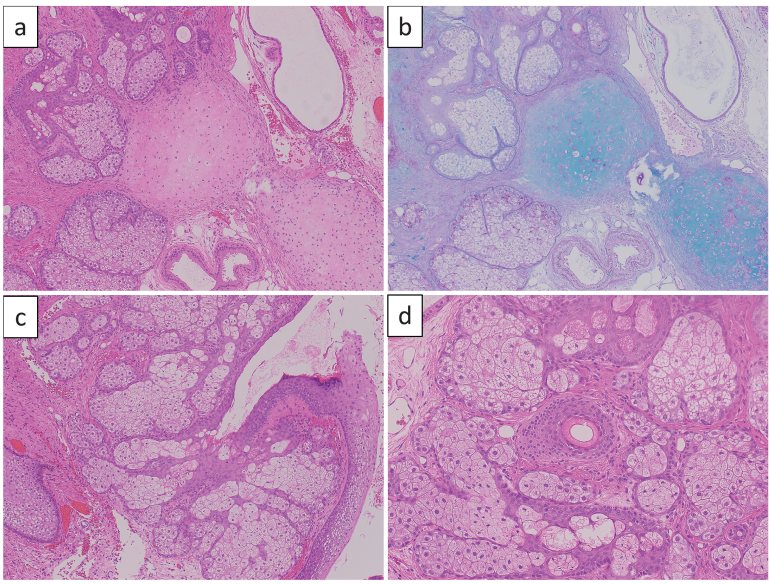

Fig. 5 Histopathology slides showing features consistent with a mature teratoma
Fig. 5a-5b: Hematoxylin and eosin staining (a) and Alcian blue/periodic acid-Schiff staining (b) showing mature cartilage tissue, ductal component, and sebaceous glands.
Fig. 5c-5d: Hematoxylin and eosin staining showing stratified squamous epithelium, hair follicle, ductal components and sebaceous glands with no malignancy (c and d).
Original magnification: 100× in a–c, 200× in d.

DISCUSSION

Intracranially located mature teratomas rarely occur in adults (incidence rate: 0.2%).[12] They usually appear as an irregular solitary mass with multilocularity and mixed signals depicting multiple tissue origins.[4] The nature of the lesion's appearance can be assessed using neurological

imaging. During CT imaging, most intracranial teratomas show the presence of fat and calcification at a minimum. Additionally, they have both cystic and solid components that contribute to an irregular outline. Solid components demonstrate variable enhancement.[13] MRI usually reveals heterogeneous intensities on T1- and T2-weighted images, followed by heterogeneous contrast enhancement. MRI signal intensity reflects the various tissue types within teratomas, including fibrous, adipose, calcification, cystic elements, hair follicles, cellular debris, and keratinocytes. This patient's neuroimaging findings did not classically fit the literature's description of a mature intracranial teratoma. It exhibited a hypodense mass, the absence of calcification, or a cystic component on preoperative CT and demonstrated homogenous intensity on T1- and T2-weighted MR images.

This case also has several other distinguishing features that are not typical in the literature. Intracranial mature teratomas are tumors that show a clear predominance (5:1) in males.[12,14] They generally occur in pediatric populations (individuals aged ≤15 years).[7,12,15] The current case occurred in an adult female patient, which is a very rare occurrence and is not in agreement with the general belief that teratomas occur in young male patients.[7,8,10,16] Teratomas are also considered partly cystic in most cases. However, in this case, the tumor was macroscopically solid without cystic components. The histological features of this tumor, which were well differentiated into mature tissue (Figure 5), diverged from the general belief that intracranial teratomas are more immature.[10,16]

Studies have shown that intracranial teratomas usually involve midline brain structures, including the third ventricle, particularly, the pineal and suprasellar/parasellar regions. Less commonly reported sites include the ventricular system, cerebellar vermis, basal ganglia, cerebellopontine angle, and cavernous sinus.[4,5,7,8] The present case appears to be unique as the teratoma was located in the sphenoid ridge, which is away from the midline. In addition to the present case, there are 5 other case reports of mature intracranial teratomas located away from the midline (Table 1). The mean patient age was 35.8 years (range, 24–67 years). Additionally, of the 6 patients, 5

Table 1 Case reports and characteristics of mature intracranial teratomas in adults located away from the midline

Reference	Patient sex/ age (y)	Symptom	Tumor location	Tumor side	Treatment	Follow-up	Recurrence
Nishigaya et al, 1994[17]	Male/67	No	Sylvian fissure	Left	No treatment, autopsy finding	NA	NA
Lee et al, 1996[18]	Male/35	Dysphasia	Middle cranial fossa	Left	Surgical resection	6 months	No
Phadke et al, 2004[19]	Male/25	Bony swelling, paresis of cranial nerves	Middle cranial fossa	Right	Surgical resection	NA	NA
Dimov et al, 2013[7]	Male/24	Headache, nausea, right-sided weakness, proptosis of the eye	Fronto-temporo-basal/orbit	Left	Surgical resection	4 years	No
Høyer et al, 2013[15]	Male/38	Generalized seizure	Frontal lobe	Left	No treatment, autopsy finding	NA	NA
Present case	Female/26	No	Sphenoid ridge	Left	Surgical resection	6 months already	No

NA: not available
y: years

were male. Four patients exhibited tumor-related symptoms. Of the 5 previously reported cases, 4 (80%) were located on the left side, similar to the present case. However, no other previous cases reported the sphenoid ridge as the tumor location. Four patients underwent total resection, while the other 2 were diagnosed based on autopsy findings. There was no evidence of recurrence during the follow-up period.

Another notable finding among the previous cases is that adult intracranial mature teratomas that are off the midline are predominantly left-sided. The left-sided pattern has not yet been described by other researchers, for which the reasons remain unknown. It is hypothesized that it may be linked with embryogenesis, as most teratomas are congenital in origin. However, left-sided intracranial mature teratomas may potentially be more symptomatic than contralateral ones due to hemispheric dominance. The clinical presentation of patients with an intracranial teratoma is nonspecific and depends on tumor size, tumor location, and signs of intracranial pressure, which may be compensated for due to slow tumor growth.[4,15] Consequently, the diagnosis of an intracranial mature teratoma would be virtually impossible, except as an unexpected finding on neurological imaging performed for another reason, such as head trauma.[15,17]

Serum tumor markers such as serum alpha-fetoprotein (AFP) and β-human chorionic gonadotropin (β-HCG) were not assayed in this patient. These can help in differentiating teratomas from other intracranial neoplasms.[4] However, some teratomas, especially the mature variant, do not usually express serum biomarkers.

Treatment strategies for teratomas remain controversial. Treatment includes radical resection whenever possible. Mature teratomas are considered chemotherapy and radiology resistant. Thus, complete tumor resection is the treatment of choice.[20] It is generally believed that any remaining portion of a diagnosed mature teratoma may also contain a small proportion of immature or malignant tissue. Mature teratomas have a lower recurrence rate after surgical resection than immature or malignant teratomas.[21] However, various studies have reported that patients with intracranial teratomas can develop other germ cell tumors at different intracranial sites.[4,22,23,24] Therefore, after the initial gross total tumor resection of mature intracranial teratomas, patients should be followed up for an extended period because of the possibility of recurrence or new occurrence.

CONCLUSIONS

The present case is an unusual example of a mature teratoma with regard to location, neuroimaging appearance, macroscopic intraoperative findings, histological tumor subtype, and patient age and sex. We believe that this case report will add to the literature on intracranial teratomas and improve clinicians' knowledge in this area.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

The patient has consented to the submission of the case report to the journal for publication.

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Not applicable.

CONFLICT OF INTEREST

None of the authors declare any potential conflicts of interest.

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