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Case Report

An Unusual Presentation of Hearing Impairment in an Adult with Pineal Region Germinoma: A Case Report

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Abstract
Tumors of the pineal region typically present with symptoms and signs of mass effect and increased intracranial pressure. However, although rare and can be overlooked, hearing impairment is a potential clinical finding in these cases. The authors describe a 24-year-old male who presented to the emergency room complaining of bilateral hearing impairment. Brain computed tomography showed a pineal region tumor. Histopathological examination demonstrated features consistent with germinoma. This case reports a rare presentation rarely seen in the literature and in practice as evident by the conducted literature review. Therefore, we highlight the importance of considering hearing impairment as a presenting symptom of pineal region tumors since prompt recognition and intervention, as demonstrated in this case, can lead to successful outcomes.

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Introduction

The presentation of pineal region tumors typically includes signs and symptoms of obstructive hydrocephalus due to the compression on the cerebral aqueduct. Additionally, Parinaud's syndrome, which is attributed to the compression of the superior colliculi of the midbrain tectum, is frequently observed [1]. While hearing impairment is not a typical feature of pineal region tumors, it can be due to the compression of the tumor on the dense auditory pathways in the region. In this case, the authors present a patient with a pineal region germinoma in an adult patient who presented with hearing impairment as a chief complaint. This case highlights the importance of including bilateral hearing impairment as a rare but possible presenting symptom in pineal region lesions. The CARE Checklist has been completed by the authors for this case report, attached as supplementary material (for all online suppl. material, see https://doi.org/10.1159/000533518).

Case Presentation

A 24-year-old, right-handed male with no prior known medical illness presented to the emergency room with a chief complaint of sudden-onset bilateral decreased hearing that had started 1 month earlier, associated with bilateral blurry vision, dizziness, and fatigue of 1 week duration. No history of recent traumas, headaches, seizures, loss of consciousness, sensory/motor deficits, nausea, vomiting, fever, night sweats, anorexia, or unintentional weight loss. On examination, the patient was fully conscious, alert, oriented, and ambulating freely. Pupils were reactive bilaterally and equally, measuring 3 mm. Extraocular muscles examination was bilaterally normal. No signs of Parinaud's syndrome or any motor/sensory abnormalities. The cranial nerve and coordination examinations were unremarkable. He reported a previous visit to another institution where he was managed as a case of otitis media without improvement. Brain computed tomography (CT) demonstrated a large mass located in the pineal region, resulting in obstructive hydrocephalus (Fig. 1). Cerebrospinal fluid analysis results showed positive beta-human chorionic gonadotropin (Beta-hCG) and placental alkaline phosphatase. A right ventriculoperitoneal shunt was inserted.

A contrast-enhanced brain magnetic resonance imaging confirmed the earlier CT findings of the tumor. The mass measured 3.5 × 2.6 × 4.0 cm (AP × TS × CC; anteroposteriorly, transversely, and coronal, respectively). Additionally, another smaller lesion was noted in the genu of the corpus callosum and septum pellucidum, measuring 2.5 × 1.4 × 1.7 cm (Fig. 2a–c).

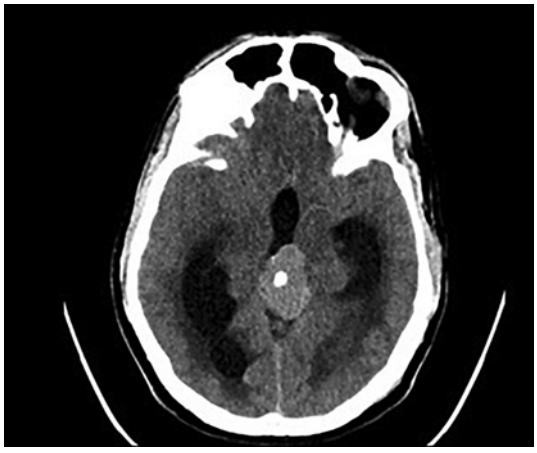

Fig. 1. Axial non-contrasted brain CT showing a hyperdense well-defined pineal region mass extended to the posterior third ventricle with a central calcification.

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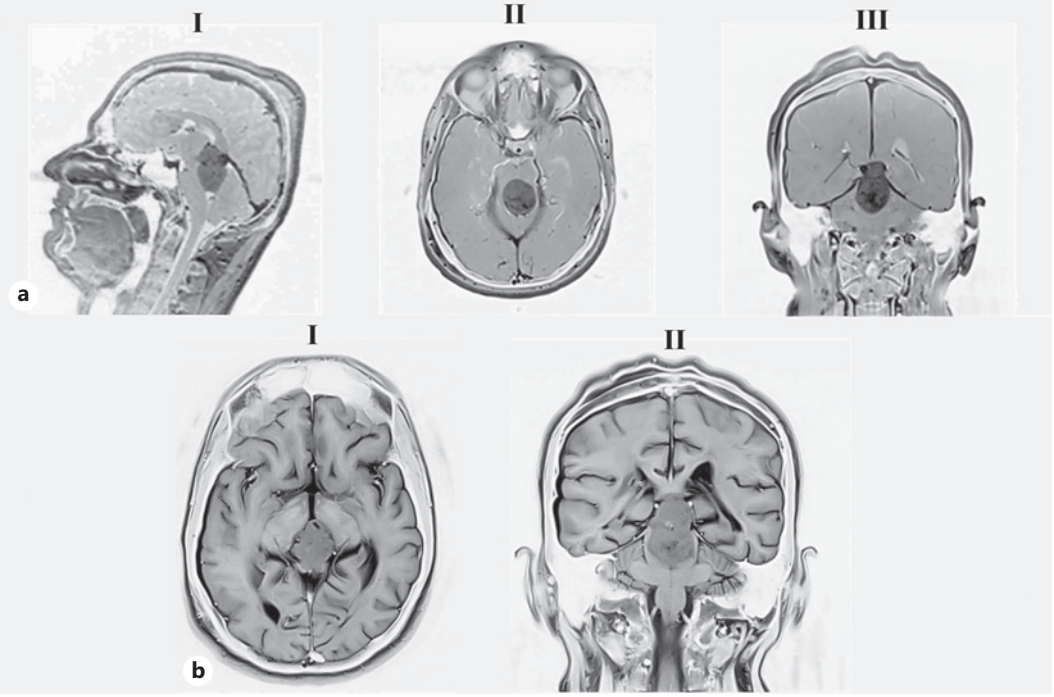

Fig. 2. MRI brain; **a** T1-WI demonstrating a pineal region lesion (I: sagittal, II: axial, III: coronal) that after gadolinium administration showing as a well-defined and circumscribed vividly enhancing mass engulfing the pineal gland, causing narrowing of the cerebral aqueduct and causing mild ventricular dilatation without transependymal permeation, mass effect on the thalamus, midbrain, pons, and effacing the ambient and quadrigeminal cisterns. **b** T2-WI showing a well-defined and circumscribed isointense lesion engulfing the pineal gland (I: axial, II: coronal), causing narrowing of the cerebral aqueduct and causing mild ventricular dilatation without transependymal permeation, mass effect on the thalamus, midbrain, pons, and effacing the ambient and quadrigeminal cisterns. MRI, magnetic resonance imaging.

Workups showed no evidence of metastases. The patient underwent a left paramedian supracerebellar infratentorial approach. With the guidance of neuro-navigation, the tumor was exposed and internally debulked. Then the tumor capsule, which was firmer and less bloody than the usual for germinomas, was dissected, and subtotal resection was achieved.

Histopathological examination using hematoxylin and eosin staining showed large, epithelioid cells with abundant cytoplasm and large, round nuclei, irregular and pleomorphic nuclei, and prominent nests of lymphocytes. Immunohistochemical studies using OCT4 demonstrated strong and diffuse nuclear stain, and CD117/C-KIT exhibited positive diffuse cytoplasmic stain. The specimen was negative for chromogranin A, CD56, NSE, and neurofilament protein. The conclusion was a germ cell tumor with features and an immunoprofile consistent with germinoma (Fig. 3a–d).

The next day, the patient developed anisocoria with unreactive pupils bilaterally. A postoperative brain CT showed a subtotal resection along with postoperative changes with no complications (Fig. 4a–b). The patient was managed conservatively with help of steroids, with which he eventually made a complete recovery with no residual hearing deficits or pupillary dysfunction. The patient was referred to a tertiary care center to proceed with radiotherapy.

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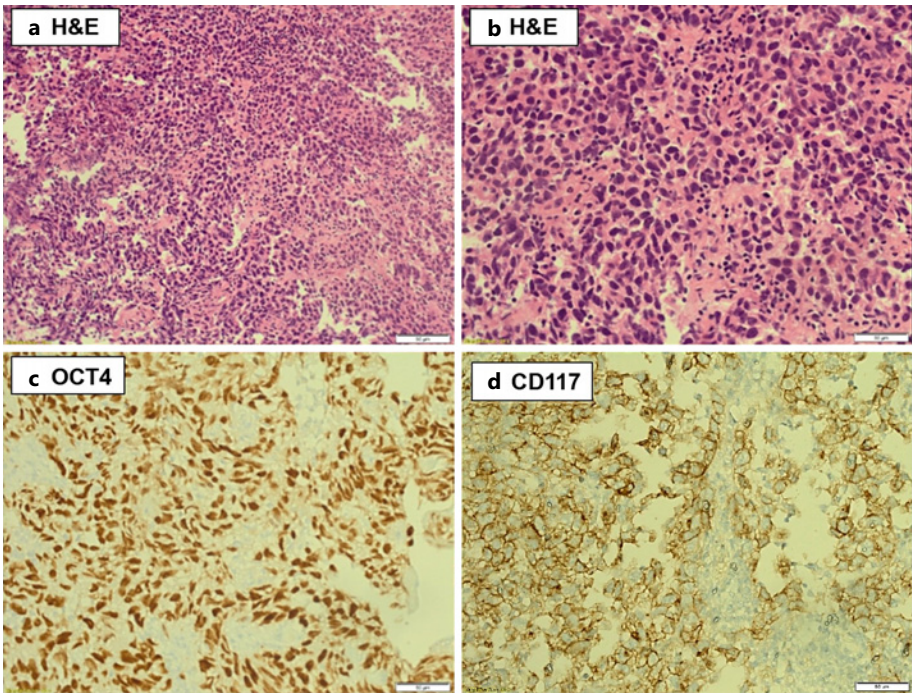

Fig. 3. The figure shows H&E staining at low (**a**) and intermediate (**b**) power, revealing large epithelioid cells with abundant cytoplasm, round nuclei, irregular and pleomorphic nuclei, and prominent nests of lymphocytes. Immunohistochemical staining demonstrates strong and diffuse nuclear staining for OCT4 (**c**) and positive diffuse cytoplasmic staining for CD117/C-KIT (**d**). H&E, hematoxylin and eosin.

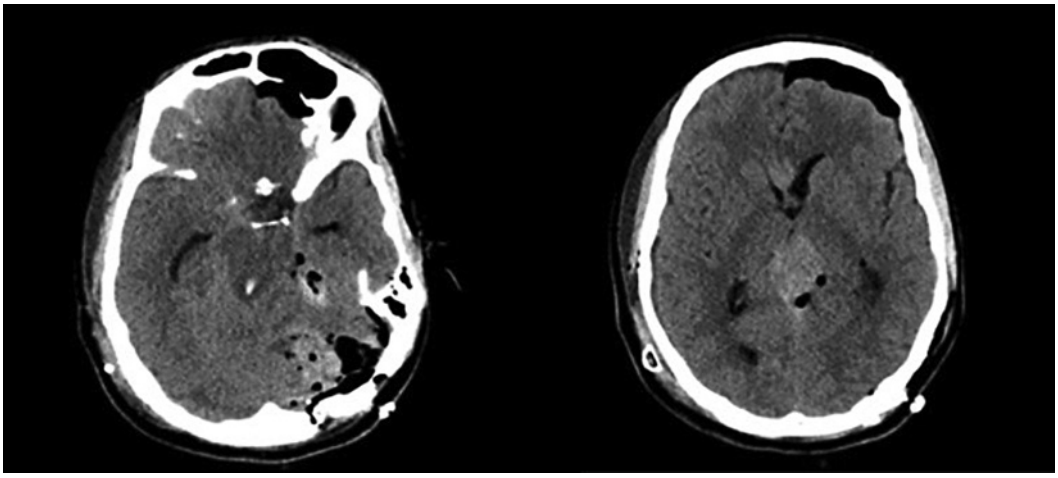

Fig. 4. Postoperative non-contrasted brain CT, showing expected postoperative changes.

Discussion

Primary central nervous system (CNS) germ cell tumors (GCTs) are a complex group of cancers whose etiology and pathophysiology remain inadequately understood. They affect males significantly more than females. While the pineal region represents a major site of CNS GCTs, they can also occur in other locations of the CNS, such as the sellar region [2].

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Table 1. Reported cases of pineal region tumors presenting with hearing impairment

Country	Author, year	Journal	Age, years/ sex	Signs and symptoms	Pathology	Laterality	Management	Outcome
USA	Hersh et al. [14] (2021)	*Neurosurgical Focus: Video*	6/male	Decorticate, hearing impairment, inability to open eyes	Mature teratoma	Bilateral	Tumor resection via posterior interhemispheric transcallosal approach	Complete recovery
USA	Aaroe et al. [15] (2021)	*Neurology*	62/male	Hearing impairment, visual disturbance, Parinaud's syndrome	Melanoma	Bilateral	Right frontal endoscopic biopsy, chemotherapy, radiotherapy	Incomplete recovery
Japan	Mizobuchi et al. [18] (2021)	*NMC Case Report Journal*	13/male	Hearing impairment	Glial cyst	Bilateral	Cyst fenestration via right occipital transtentorial approach	Incomplete recovery
Turkey	Damgacı et al. [17] (2020)	*Current Medical Imaging*	17/ female	Nausea, vomiting, hearing impairment, Parinaud's syndrome	Papillary tumor	Bilateral	Tumor resection	Not reported
Switzerland	Joswig et al. [10] (2015)	*Clinical Neurology and Neurosurgery*	19/male	Headache, Parinaud's sign, hearing impairment, tinnitus	Germinoma	Bilateral	Endoscopic transventricular biopsy and third ventriculostomy, radiotherapy	Complete recovery
Japan	Shinsato et al. [12] (2012)	*Surgical Neurology International*	49/ female	Hearing impairment, tinnitus	Melanoma	Bilateral	Tumor resection via left occipital transtentorial approach, radiotherapy	Complete recovery[1]
France	Gaspar et al. [1] (2003)	*Journal of Neurosurgery*	12/male	Headache, hearing impairment, photophobia	Malignant germ cell tumor	Bilateral	Chemotherapy, tumor resection, radiotherapy	Incomplete recovery[2]
Japan	Haque et al. [6] (2002)	*Acta Neurochirurgica*	34/male	Headache, hearing Impairment, Parinaud's sign, abnormal gait	Meningioma	Bilateral	Tumor resection via left occipital transtentorial approach	Complete recovery
Japan	Islam et al. [3] (2002)	*Neurologia medico-chirurgica*	18/male	Headache, hearing impairment, tinnitus, diplopia	Germinoma	Bilateral	Tumor resection via occipital interhemispheric subtentorial approach, chemotherapy, radiotherapy	Incomplete recovery
India	Mukherjee et al. [13] (1999)	*British Journal of Neurosurgery*	70/male	Headache, forgetfulness, abnormal gait	Pineal cyst	Bilateral	Cyst excision via occipital transtentorial approach	Incomplete recovery[3]

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Table 1 (continued)

Country	Author, year	Journal	Age, years/ sex	Signs and symptoms	Pathology	Laterality	Management	Outcome
Italy	Missori et al. [11] (1995)	*Acta Neurochirurgica*	47/ female	Headache, visual disturbance, tinnitus, papilledema, upward gaze, +ve audiometry	Meningioma	Bilateral*	Tumor resection via occipital transtentorial approach	Incomplete recovery[4]
Italy	Missori et al. [11] (1995)	*Acta Neurochirurgica*	48/ female	Truncal ataxia, Parinaud's sign, nystagmus, third nerve palsy, hearing impairment	Melanoma	Bilateral	Tumor resection via infratentorial supracerebellar approach, radiotherapy	Incomplete recovery
Italy	Missori et al. [11] (1995)	*Acta Neurochirurgica*	40/ female	Blurry vision, papilledema, Parinaud's sign.+ve audiometry	Pineocytoma	Bilateral*	Tumor resection via infratentorial supracerebellar approach	Complete recovery
USA	DeMonte et al. [7] (1993)	*Neurosurgery*	63/male	Headache, hearing impairment	Meningioma	Bilateral	Tumor resection via suboccipital transtentorial approach	Complete recovery[5]
USA	Sekhar and Goel, [16] (1992)	*Surgical Neurology*	45/male	Hearing impairment, loss of balance	Meningioma	Bilateral	Tumor resection via two-stage combined supratentorial and infratentorial approach 15 days apart	Incomplete recovery[6]
USA	Reger [19] (1987)	*Annals of Otology, Rhinology and Laryngology*	23/male	Headache, hearing impairment, unsteady gait	Not reported	Bilateral	Tumor resection via suboccipital craniectomy, radiotherapy	Incomplete recovery
USA	Toshniwal et al. [9] (1986)	*Journal of Neuro-Ophthalmology*	8/ female	Headache, hearing impairment, tinnitus, abnormal gait	Pineoblastoma	Bilateral	Tumor resection, radiotherapy	Incomplete recovery

*The patients did not report hearing problems, but audiometry showed impaired acoustic sensations.
[1]The patient received no chemotherapy due to renal failure, yet MRI 56 weeks after treatment showed no of recurrence or metastasis.
[2]All auditory functions returned to normal, but the patient continued to have mild left-eye diplopia.
[3]The patient made an initial recovery but 2 months later had chronic subdural hematoma drainage.
[4]Audiometry showed severe bilateral neurosensory hypoacusia and on follow-up, audiometric examination was unchanged.
[5]Hearing improved clinically but brain stem evoked responses showed increased latencies pre- and post-op.
[6]Right abducens nerve paresis.

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Anatomically, the central auditory pathways run from the medial geniculate body and the inferior colliculus to Heschel's gyrus. These fibers are in close proximity to the pineal body, forming extensive interconnections and bilateral innervations of the brain stem structures responsible for hearing. These bilateral interconnections make hearing impairment an atypical symptom of the disease [3].

Our case is a germinoma in an adult patient presenting with hearing impairment as the chief complaint, which is rarely seen in pineal region tumors. The typical presentation of pineal region tumors is related to the mass effect on adjacent structures. This includes symptoms and signs of increased intracranial pressure secondary to hydrocephalus. In 80% of cases, hydrocephalus occurs due to direct compression on the cerebral aqueduct. In neurological examination, findings usually include Parinaud's syndrome, in which the involvement of the superior colliculi of the midbrain tectum can explain [1].

The diagnosis of CNS GCTs is based on radiological findings from brain CT and magnetic resonance imaging scans, as well as cerebrospinal fluid analysis for tumor markers such as human chorionic gonadotrophic hormone and alpha-fetoproteins. Germinomas are specifically associated with increases in levels of human chorionic gonadotrophic hormone and placental alkaline phosphatase [4].

Although intracranial germinomas are highly radiosensitive and can be cured with radiotherapy alone, the current standard of care involves neoadjuvant platinum-based chemotherapy along with lower doses of radiation [4]. In a literature review, surgical resection as a management strategy in a germinoma was reported by Islam et al. [3], who published a case of an 18-year-old male who had incomplete recovery following gross total tumor resection.

Literature Review

Including our one institutional case, a literature review was conducted, and cases with a similar presentation were included going back to 1986. Cases of hearing impairment associated with pineal region tumors have been reported since 1879 when Dr. Gowers first reported this phenomenon [5]. In 1989, Luo et al. [6] reported 64 cases of pineal region tumors, 9 of which presented with hearing impairment. De Monte et al. reviewed the literature from 1879 to 1992 and found only fourteen cases [5, 7]. Konovalov et al. reported 10 cases of pineal region meningioma in 1996, with hearing impairment noted in two of them [8].

Individual cases of pineal region tumors presenting with hearing impairment are summarized in Table 1 [1, 3, 5, 7, 9–19]. Seventeen similar cases were collected, all of which had bilateral hearing impairment. The included cases consisted of 11 males and 6 females, with ages ranging from 6 to 70 years old. Of the 17 cases, 4 were meningiomas, 3 were melanomas, and 2 were germinomas. The cases were reported from various countries, including 6 from the USA, 4 from Japan, and 3 from Italy (with all 3 cases reported by one author). One case was reported from each of Turkey, France, Switzerland, and India. The oldest reported case dates back to 1986 by Toshniwal et al., while the most recent was reported in 2021 by Aaroe et al. The majority of patients, 10 out of 17, had incomplete recovery, while 6 had complete recovery. One case reported by Damgacı et al. had no clearly stated outcome. *Acta Neurochirurgica* was the journal with the highest number of publications for the included cases, with 4 cases in 2 articles.

Conclusion

The presented case emphasizes the significance of considering CNS GCTs as a plausible differential diagnosis even in adult patients presenting with sudden hearing loss with no accompanying neurological manifestations, particularly in the absence of an identifiable

etiology. Prompt recognition of these tumors is crucial for the timely initiation of treatment, which can profoundly influence patient prognosis. The outcomes of the review of relevant literature provide a valuable point of reference for clinicians and future researchers who might explore possible associations between disease or patient variables and presentation and prognosis.

Statement of Ethics

This research was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. Written informed consent was obtained from the patient for publication of this case report and any accompanying images. Ethical approval is not required for this case report in accordance with local guidelines.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

The authors made substantial contributions to the conception of the idea, design, data collection, analysis, and drafting the manuscript. A.H.: literature review, data acquisition, writing, and revision. H.M.: literature review, methodology, editing, and revision. A.A. and T.A.A.: conceptualization and revision. A.A.A.: literature review and writing. A.S.: data acquisition and revision. F.R.A.: data acquisition, analysis, and revision. All authors read the manuscript before submission, provided critical feedback, and approved the final version. All authors agreed to be accountable for all aspects of the work.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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