

▪ **Case Report**

Pineal germinoma presenting with Parinaud's syndrome

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Abstract

Intracranial germinomas typically occur in the suprasellar, pineal, and other locations. Pineal germinoma presenting with Parinaud's Syndrome is a group of abnormalities of eye movement and pupil dysfunction. It is a rare tumor and its clinical and imaging features can help clinicians in the management of these lesions.

Keywords: Pineal germinoma, Parinaud's syndrome, Computed tomography, magnetic resonance imaging.

Introduction

Germinomas are rare neoplasms of the central nervous system and account for 0.2 to 1.7% of all primary intracranial tumors.¹ Most of these tumors occur in children and young adults; the mean age of onset is about 17 years.^{2,3} Intracranial germinomas typically occur in the suprasellar region (49%), followed by the pineal region (38%), both pineal and suprasellar regions (8%), and other locations (5%).⁴ Parinaud's syndrome, also known as dorsal midbrain syndrome is a group of abnormalities of eye movement and pupil dysfunction. It is caused by lesions of the upper brain stem and Henri Parinaud was first to describe the disease.^{5,6}

Case presentation

A 30 years old male patient presented with complaints of intermittent headache since 3 months. There is no history of blurring of vision, trauma or fever. No history of hypertension, diabetes mellitus. On examination, he was conscious, well oriented to time, place and person. Blood pressure, pulse rate and respiratory rate were within normal limits. Ophthalmological examination revealed restriction in the upward gaze, however there was preservation of the downward gaze with bilateral papilloedema. Laboratory investigations were within normal limit.

Contrast-enhanced computed tomography (CT) scan of head showed a well defined, slightly hyperdense mass in the pineal region, surrounding and engulfing the calcified pineal gland (Figure 1a and 1b). The mass

was abutting and compressing the posterior third ventricle and the cerebral aqueduct resulting in mild hydrocephalus. There was mild homogeneous enhancement of the mass on contrast-enhanced CT scan. On post-contrast T1-weighted magnetic resonance imaging (MRI) in coronal and sagittal planes, the mass was homogeneously enhancing with mild hydrocephalus (Figure 2a and 2b).

Coronal and (B) sagittal postcontrast T1-weighted images reveal a somewhat ill-defined, homogeneously enhancing tumor in the suprasellar region. The diagnosis of pineal germinoma presenting as Parinaud's syndrome was been made. Patient was referred to oncology centre for radiotherapy.

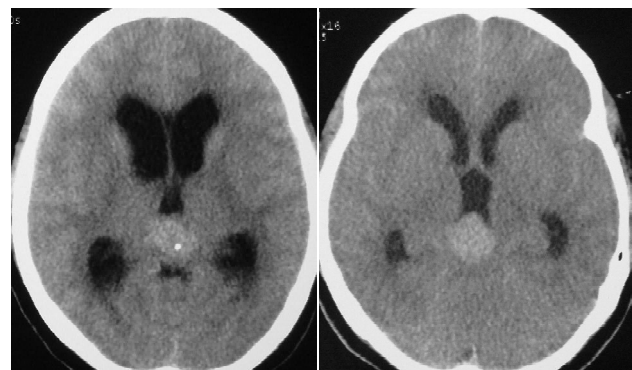


Fig 1a

Fig 1b

Figures 1a and 1b: Axial section CECT of head showing a well defined, mildly enhancing mass in the pineal region, surrounding and engulfing the calcified pineal gland along with mild hydrocephalus.

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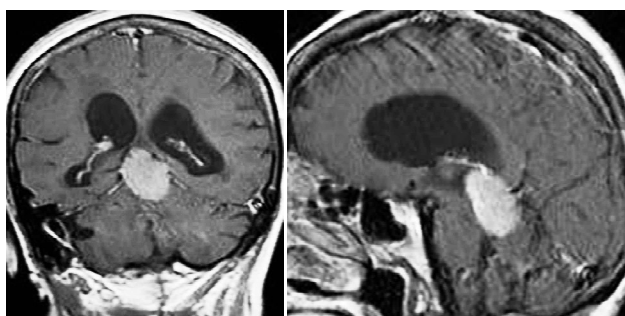


Fig 2a

Fig 2b

Figures 2a and 2b: Post-contrast T1-weighted MR images in coronal (Figure 2a) and sagittal (Figure 2b) planes showing homogeneously enhancing mass in pineal region with mild hydrocephalus.

Discussion

Pineal germinoma with classical Parinaud's syndrome presents with various abnormalities of eye movement and pupil dysfunction. It is characterized by paralysis of up-gaze, Pseudo-Argyll Robertson pupils, convergence-retraction nystagmus, eyelid retraction (Collier's sign) and conjugate down gaze in the primary position i.e. "setting-sun sign".⁷ Neurosurgeons will often see this sign most commonly in patients with failed ventriculoperitoneal shunts. Pineal germinoma is also associated with bilateral papilledema, spasm of accommodation on attempted upward gaze, pseudoabducens palsy (also known as thalamic esotropia) or slower movements of the abducting eye than the adducting eye during horizontal saccades, see-saw nystagmus and associated ocular motility deficits including skew deviation, oculomotor nerve palsy, trochlear nerve palsy and internuclear ophthalmoplegia.⁶

Pineal region neoplasms can be classified into 3 major groups according to their cellular origin: tumors of germ cell origin, tumors of pineal cell origin and tumors of other cell origin. Tumors of germ cell origin include germinoma, mature teratoma, malignant teratoma, embryonal cell carcinoma, endodermal sinus tumor, choriocarcinoma, and mixed germ cell tumors.^{5,6}

Computed tomography (CT) scanning may be needed to evaluate a calcified pineal gland that is associated with a pineal germinoma or tumor calcification associated with other neoplasms in the pineal region.^{5,7} CT reveals a large (typically $e \approx 3$ cm), lobulated, typically hyperdense mass. The pineal calcifications may appear displaced at the periphery of the lesion. Nearly 100% of patients have obstructive hydrocephalus.⁷ MRI is the preferred imaging modality and it enables the

accurate delineation of pineal masses before surgery. MRI allows true pineal masses to be distinguished from parapineal masses that impinge on the pineal gland. MRI is not perfect in the detection of calcifications. On MR evaluation, signal intensity is homogeneous and similar to that of gray matter on both T1- and T2-weighted sequences. The mass frequently engulfs a densely calcified pineal gland.

Germinomas are well-circumscribed lesions that demonstrate a two-cell pattern of lymphocytes and large polygonal primitive germ cells. The abundance of lymphocytes contributes to the hyperdensity seen at CT and the reduced diffusion at diffusion-weighted MR imaging. Germinomas can be divided into two subtypes: pure germinoma and germinoma with syncytio-trophoblastic cells. Those containing syncytio-trophoblastic giant cells have a higher recurrence rate and decreased long-term survival and demonstrate elevated cerebrospinal fluid levels of hCG.⁷

The pineal germinomas are exquisitely radiosensitive, and 90% of patients with pure intracranial germinomas can be treated successfully by radiation therapy. Radiosurgery is now considered to be another treatment option, along with chemotherapy and radiation, for germinomas. Hydrocephalus is a serious complication of pineal region tumors and has to be managed appropriately. Ventriculostomy is considered a good alternative to ventriculoperitoneal shunt to relieve the hydrocephalus, and it does not carry the risk of peritoneal metastasis in the future.⁸

Summary

In conclusion, pineal germinomas are rare tumors and its clinical and imaging features can help clinicians in the management of these lesions. Once the initial histopathologic diagnosis is established as germinoma, debulking should proceed only as needed to avoid undue risk to the patient. Long-term control of the lesion is ultimately achieved via radiation therapy.

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