Parinaud's Syndrome

Also known as dorsal midbrain syndrome, pretectal syndrome.

Parinaud's syndrome (Paralysie des mouvements associe´s des yeux named for Henri Parinaud (1844–1905). [1] is a supranuclear paralysis of vertical gaze [2] resulting from damage to the mesencephalon [3].

Associations

Progressive supranuclear palsy, with preservation of vestibuloocular reflex

Lid retraction:(Collier's sign)

Convergence palsy

Accommodation palsy

Pseudoabducens palsy.

See saw nystagmus.

Fixed pupils.

Variants

Skew deviation may be a unilateral variant of Parinaud's Syndrome.

Sylvian aqueduct syndrome.

Etiology

Parinaud's Syndrome results from injury, which compresses the the quadrigeminal plate at the rostral interstitial nucleus of medial longitudinal fasciculus (riMLF), specifically, compression or ischemic damage of the mesencephalic tectum, including the superior colliculus adjacent oculomotor (origin of cranial nerve III) and Edinger-Westphal nuclei, causing dysfunction to the motor function of the eye.

Pineal region tumors.

A dilated suprapineal recess can compress the mesencephalic tectum, due to hydrocephalus.

Stroke or brainstem hemorrhage in the upper part.

Guillain-Barré syndrome

Myasthenia gravis

Botulism

Hypothyroidism

Gradual benign loss of upgaze in senescence.

Sylvian aqueduct syndrome

Aberrant regeneration of the third nerve

Limited upgaze in elderly patients

Progressive supranuclear palsy

Niemann-pick disease

Whipple’s disease.

Classically, it has been associated with three major groups:

Pineal region tumor: Pinealoma (intracranial germinomas) are the most common lesion producing this syndrome).

Women in their 20s-30s with multiple sclerosis.

A 26 year old Pakistani lady with first presentation of a demyelinating event, presenting as Parinaud's syndrome. A video demonstrates a convergence-retraction nystagmus on upgaze and failure of accommodation, and her brain imaging confirms a corresponding pre-tectal contrast enhancing T2 hyperintense lesion suggestive of demyelination [4].

Older patients following stroke of the upper brainstem However, any other compression, ischemia or damage to this region can produce these phenomena: obstructive hydrocephalus, midbrain hemorrhage, cerebral arteriovenous malformation, trauma and brainstem toxoplasmosis infection. Neoplasms and giant aneurysms of the posterior fossa have also been associated with the midbrain syndrome.

Vertical supranuclear ophthalmoplegia has also been associated with metabolic disorders, such as Niemann-Pick disease, Wilson's disease, kernicterus, and barbiturate overdose.

Isolated Oculomotor Nerve Nucleus Infarct [5].

Unilateral vascular ischemic lesion [6].

A case of longstanding, undiagnosed spontaneous intracranial hypotension (SIH) with an acute presentation of Parinaud's syndrome, in whom serial imaging demonstrated development of a midbrain mass. The patient was ultimately diagnosed with tumefactive venous infarction secondary to SIH. However, this patient underwent a brainstem biopsy, which in retrospect may have been avoidable. This case demonstrates the imaging features of tumefactive venous infarction in SIH and highlights the risk of misinterpretation as a neoplasm with potentially catastrophic consequences [7].

Migraine [8].

Miller Fisher syndrome [9].

Clinical features

Parinaud's syndrome, includes multiple clinical signs, with the most prominent being paralysis of upward gaze.

Parinaud's Syndrome is a cluster of abnormalities of eye movement and pupil dysfunction, characterized by:

Paralysis of upgaze: Downward gaze is usually preserved. This vertical palsy is supranuclear, so doll's head maneuver should elevate the eyes, but eventually all upward gaze mechanisms fail.

Pseudo-Argyll Robertson pupils: Accommodative paresis ensues, and pupils become mid-dilated and show light-near dissociation.

Convergence-Retraction nystagmus: Attempts at upward gaze often produce this phenomenon. On fast up-gaze, the eyes pull in and the globes retract. The easiest way to bring out this reaction is to ask the patient to follow down-going stripes on an optokinetic drum.

Eyelid retraction (Collier's sign) Conjugate down gaze in the primary position: “setting-sun sign”. Neurosurgeons will often see this sign most commonly in patients with failed ventriculoperitoneal shunts. It is also commonly associated with bilateral papilledema. It has less commonly been associated with 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.

Diagnosis

MRI is very helpful in determining the cause and thus guiding appropriate treatment

Differential Diagnosis

Etiologies

1. masses pressing directly on quadrigeminal plate (e.g. pineal region tumors)

2. elevated ICP: secondary to compression of mesencephalic tectum by dilated suprapineal recess, e.g. in hydrocephalus

3. stroke or hemorrhage in upper brainstem

4. multiple sclerosis (MS)

5. occasionally seen with toxoplasmosis Conditions affecting ocular motility that could mimic the upgaze palsy of PS:

1. Guillain-Barré syndrome

2. myasthenia gravis

3. botulism

4. hypothyroidism

5. there may be a gradual benign loss of upgaze with senescence

Case reports

A 3-year-old boy presented with headache, disturbance of consciousness, and Parinaud's syndrome. Magnetic resonance (MR) imaging revealed a pineal mass lesion, and total resection of the tumor was achieved. The histological diagnosis was mature teratoma. He did not receive further treatment, and did well without recurrence for 20 years. However, he suffered headache 21 years after resection, and MR imaging revealed a homogeneously enhanced pineal mass with low minimum apparent diffusion coefficient value and proton MR spectroscopy showed a huge lipid peak. The levels of tumor markers were not elevated. Cerebrospinal fluid (CSF) cytology found atypical cells with large nuclei and irregularly shaped nucleoli. To elucidate the relationship between the primary and recurrent tumors, we reviewed the histological specimens and CSF cytology at the initial treatment and found a subset of incompletely differentiated components resembling fetal tissues in the histological specimen and atypical large cells in the CSF. Based on these radiological and histological findings, Mano et al., presume that the recurrent disease was disseminated germinoma after the resection of disseminated IMT. He received chemotherapy and craniospinal radiation therapy, and the enhanced lesion and atypical cells in the CSF disappeared. This case demonstrates that disseminated IMT can be controlled for the long term without adjuvant therapy, but may recur as germinoma. Tumor dormancy may

account for this unusual course [10].

1)
Parinaud H. Paralysie des movements associe´s des yeux. Archives de Neurologie 1883;5:145–72.
2)
Pierrot-Deseilligny C, Chain F, Gray M, et al. Parinaud’s syndrome. Brain 1982;105:667–96.
3)
Pearce JM. Parinaud's syndrome. J Neurol Neurosurg Psychiatry. 2005 Jan;76(1):99. PubMed PMID: 15608003; PubMed Central PMCID: PMC1739319.
4)
Gnanapavan S, Sillery E, Acheson JF, Toosy AT. Parinaud's syndrome - A rare presentation of clinically isolated syndrome. Mult Scler Relat Disord. 2014 May;3(3):398-401. doi: 10.1016/j.msard.2013.09.003. PubMed PMID: 25876480.
5)
Kumar Y, Hooda K, Sapire J. A Case Report of Isolated Oculomotor Nerve Nucleus Infarct: A Rare Cause of Parinaud's Syndrome. Conn Med. 2016 Mar;80(3):167-8. PubMed PMID: 27169301.
6)
Serino J, Martins J, Páris L, Duarte A, Ribeiro I. Parinaud's syndrome due to an unilateral vascular ischemic lesion. Int Ophthalmol. 2015 Apr;35(2):275-9. doi: 10.1007/s10792-015-0045-y. PubMed PMID: 25649259.
7)
Bray TJ, Chandrashekar H, Rees J, Burke A, Merve A, Thust S. Venous infarction mimicking a neoplasm in spontaneous intracranial hypotension: an unusual cause of Parinaud's syndrome. J Surg Case Rep. 2016 Mar 17;2016(3). pii: rjw037. doi: 10.1093/jscr/rjw037. PubMed PMID: 26987945; PubMed Central PMCID: PMC4794942.
8)
Jumma OK, Hamdalla HM. Parinaud's syndrome due to migraine. Can J Neurol Sci. 2013 Jan;40(1):125. PubMed PMID: 23427358.
9)
Mallia M, Chircop C, Aquilina J. An unusual case of Parinaud's syndrome. BMJ Case Rep. 2012 Nov 21;2012. pii: bcr2012006722. doi: 10.1136/bcr-2012-006722. PubMed PMID: 23175005; PubMed Central PMCID: PMC4544412.
10)
Mano Y, Kanamori M, Kumabe T, Saito R, Watanabe M, Sonoda Y, Tominaga T. Extremely Late Recurrence 21 Years after Total Removal of Immature Teratoma: A Case Report and Literature Review. Neurol Med Chir (Tokyo). 2017 Jan 15;57(1):51-56. doi: 10.2176/nmc.cr.2016-0241. PubMed PMID: 27928096; PubMed Central PMCID: PMC5243165.