Metastatic renal cell carcinoma presenting as one-and-a-half syndrome

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We report a case of 43-year-old male, presented with sudden onset binocular diplopia on lateral gazes. Ocular examination showed features of ipsilateral one-and-a-half syndrome. Comprehensive systemic work in conjunction with magnetic resonance imaging of the brain illustrated irregular mixed solid and cystic lesions in the brainstem, possibly indicative of brain metastases. Further imaging revealed hidden renal cell carcinoma as a primary neoplasm, which led to secondary manifestations.

Key words: Diplopia, metastases, one-and-a-half

“One-and-a-half syndrome” manifests as conjugate horizontal gaze palsy to one side, along with internuclear ophthalmoplegia. We report a unique case of ipsilateral one-and-a-half syndrome; in which serious morbidity such as renal cell carcinoma (RCC) and brain metastasis was initially hidden and could only be diagnosed subsequent to the observation of motility defects. To the best of our knowledge, detection of metastatic RCC subsequent to the diagnosis of isolated one-and-a-half syndrome, without systemic complaints, has not been reported. This case highlights the importance of heightened awareness for the thorough systemic workup and imaging in patients with ocular motility defects.

Case Report

A 43-year-old male patient presented to our centre, with chief complaint of sudden onset, progressively worsening binocular diplopia in the right eye for the preceding 1 month. No history of any trauma or similar complaint was elicited in the past. The patient was found to be adequately built, with vital signs being normal and had no evidence of systemic abnormality. On ocular examination, best-corrected distance visual acuity and near vision in both the eyes were 6/6 and N6, respectively. Anterior and posterior ocular segment evaluation was unremarkable. Although the eyes were aligned in primary position, horizontal gaze palsy was noted with loss of saccades and pursuit movements in levovertical. On attempted dextroversion, the right eye was abducting with associated abduction nystagmus, in the absence of left eye adduction [Fig. 1]. Both vertical gaze and convergence were normal. Based on ocular findings, the diagnosis of ipsilateral one-and-a-half syndrome was made.

Magnetic resonance imaging (MRI) was advised following clinical evaluation, which revealed an irregular complex cystic enhancing space-occupying lesion measuring 42 mm × 39 mm × 35 mm, in the region of fourth ventricle closely abutting the brain stem posteriorly [Fig. 2a and b]. Multiple small ring and disc enhancing lesions were also seen in the bilateral cerebellar hemispheres and right occipital lobe. Radiological findings suggested brain metastasis.

Further evaluation was supplemented with 18F-fludeoxyglucose positron emission tomography/computed tomography (CT) imaging to locate the primary pathology. Abdominal CT scans showed primary neoplastic lesion, arising from the upper pole of the right kidney and another exophytic mass in the lower pole of the left kidney. In addition, metastatic lesions in the right adrenal gland, pancreas, and brain were seen. Based on examination and investigations, diagnosis of Stage 4 RCC (inoperable) with brain metastasis with associated ipsilateral one-and-a-half syndrome was made. The patient was shifted to oncology department and started on palliative radiotherapy after sorting consultation for the same.

Discussion

One-and-a-half syndrome is described as combination of unilateral horizontal gaze palsy along with internuclear ophthalmoplegia. Premotor neurons of two gaze centres are located in the reticular formation and help in controlling the direction of saccades. The paramedian pontine reticular formation (PPRF) is a small ill-defined area in pons, which lies ventral to the abducens nucleus and forms the horizontal gaze center.[1] Frontal eye field in the cerebral cortex initiates voluntary signals for contralateral gaze, which further passes to contralateral PPRF through superior colliculus. To produce a horizontal saccade, contraction of lateral rectus of one eye and medial rectus of contralateral eye occur simultaneously, with the help of innervations from ipsilateral abducens nucleus and contralateral oculomotor nucleus, respectively. Activated neurons from PPRF pass to the abducens nucleus of the same side. From the abducens nucleus, one type of neurons innervates lateral rectus muscle of same side while others cross the midline, innervate the medial longitudinal fasciculus and then a portion of the oculomotor nucleus, innervating medial rectus to produce saccade.[2]
The one-and-a-half syndrome is caused either due to ipsilateral lesion of the PPRF or due to the abducens nucleus or both on one side, which causes conjugate gaze palsy. In addition, due to interruption of internuclear fibers of the ipsilateral medial, after crossing the midline, longitudinal fasciculus causes failure of adduction in same eye.

Brain MRI was suggestive of metastases at dorsal and dorsolateral part toward the left side of the pons and medulla in our patient which explains the left gaze palsy with internuclear ophthalmoplegia. It is pertinent to note that in the present case, convergence was intact, in spite of inability to adduct voluntarily, which pointed toward sparing of the medial recti subnuclei location in the midbrain.

Although our patient was orthophoric on presentation, as per previous reports on the acute stage of internuclear ophthalmoplegia (INO), exotropia is not uncommon. However, it can be frequently overlooked due to short duration. As previously documented, structures vital to vertical and torsional eye movements are rostral interstitial nucleus of the medial longitudinal fasciculus, interstitial nucleus of Cajal, and posterior commissure. In the present case, these structures should have been spared from metastasis, which resulted in normal vertical, torsional movements, and absence of skewing as well as optokinetic nystagmus.

Reported etiological factors for one-and-a-half syndrome include pontine hemorrhage, pontine infarction secondary to thrombosis or spasm of basilar artery, tumors like glioma, metastases, demyelinating conditions like multiple sclerosis, and infective conditions like tuberculosis. In our patient, brain metastases were attributed to one-and-a-half syndrome.

Cases of one-and-a-half syndrome, secondary to metastatic breast carcinoma, acute myeloid leukemia, and multiple infarcts have already been reported in the literature. To the best of our knowledge, detection of metastatic RCC subsequent to the diagnosis of one-and-a-half syndrome has never been reported. The most commonly documented histological type of renal cancer is RCC, and almost 30% of cases are metastatic at the time of diagnosis. Moreover, the 5-year survival rate of metastatic RCC is <10%, and the average median survival rate decreases to less than a year after brain metastasis in RCC.

Conclusion

We report a unique case of one-and-half-syndrome secondary to metastatic RCC.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Bogousslavsky J, Miklossy J, Regli F, Deruaz JP, Despland PA. One-and-a-half syndrome in ischaemic locked-in state: A clinico-pathological study. J Neurol Neurosurg Psychiatry 1984;47:927-35.
2. Menon V, Gogoi M, Saxena R, Singh S, Kumar A. Isolated “one and a half syndrome” with brainstem tuberculosis. Indian J Pediatr 2004;71:469-71.
3. Johkura K, Kudo Y, Amano Y, Kikyo H, Imazeki R, Amari K, et al. Gaze palsy and exotropia in internuclear ophthalmoplegia. J Neurol Sci 2015;353:158-60.
4. Henn V, Hepp K, Vilis T. Rapid eye movement generation in the primate. Physiology, pathophysiology, and clinical implications. Rev Neurol (Paris) 1989;145:540-5.
5. Newton HB, Miner ME. “One-and-a-half” syndrome after a resection of a midline cerebellar astrocytoma: Case report and discussion of the literature. Neurosurgery 1991;29:768-72.
6. Wall M, Wray SH. The one-and-a-half syndrome – A unilateral disorder of the pontine tegmentum: A study of 20 cases and review of the literature. Neurology 1983;33:971-80.
7. Crisostomo EA. One-and-a-half syndrome in a patient with metastatic breast disease. J Clin Neuroophthalmol 1985;5:270-2.
8. Hsu WH, Chu SJ, Tsai WC, Tsao YT. Acute myeloid leukemia presenting as one-and-a-half syndrome. Am J Emerg Med 2008;26:513.e1-2.

9. Verma J, Jonasch E, Allen PK, Weinberg JS, Tannir N, Chang EL, et al. The impact of tyrosine kinase inhibitors on the multimodality treatment of brain metastases from renal cell carcinoma. Am J Clin Oncol 2013;36:620-4.

10. Shuch B, La Rochelle JC, Klatte T, Riggs SB, Liu W, Kabbinavar FF, et al. Brain metastasis from renal cell carcinoma: Presentation, recurrence, and survival. Cancer 2008;113:1641-8.