Reminder of important clinical lesson

Eye twist and tongue twist: a rare neurological syndrome

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Summary

A 27-year-old man presented with diplopia without features of raised intracranial pressure. He had left sixth cranial nerve (CN) palsy. Initial investigations in the form of blood tests, cerebrospinal fluid (CSF) opening pressure including CSF analysis and CT head were normal. He represented with paraparesis after 3 weeks. Examination revealed sixth CN palsy (eye twist) and new left-sided twelfth CN palsy (tongue twist), and hence Godtfredsen syndrome was diagnosed. MRI showed vertebral and clivus metastases. He showed partial response to high dose steroid. The statement 'sixth CN palsy may be a false localising sign' in neurology, should be followed by '...it can also be a true initial sign of clivus mass.'

BACKGROUND

This case highlights the importance of considering sixth cranial nerve (CN) palsy as an initial dangerous sign of clivus metastasis which might be missed with CT head given, the early diagnosis and management of this condition decreases the morbidity. It also clarifies the confusing rare neurological Godtfredsen syndrome.

CASE PRESENTATION

A 27-year-old man presented to us with acute onset history of diplopia on horizontal gaze looking towards left side. He denied any history suggestive of raised intracranial pressure. He had undergone operation for carcinoma rectum (adenocarcinoma) 6 months back and received chemotherapy and radiotherapy. He looked healthy and pleasant. Detailed neurological examination revealed left-sided abducens nerve palsy. Other neurological examination was normal. He had abdominal scar with colostomy bag in situ. His CT scan of head, cerebrospinal fluid (CSF) opening pressure and CSF analysis turned out to be normal. He was discharged on steroid from outpatient department. Four days later at home, he developed paraparesis with sensory loss below T10 thoracic level and bladder retention, for which he was treated by traditional healer (common in our country). He revisited us after 3 weeks of paraparesis. He had additional left-sided twelfth CN palsy (figure 1). His MRI revealed clival metastasis (figure 2). He also had peritoneal and pleural metastasis. He was finally diagnosed to have adenocarcinoma rectum with widespread metastasis.

OUTCOME AND FOLLOW-UP

After treatment with steroid, he showed mild improvement. The patient has not developed other CN palsy. He was discharged on steroid and referred to nearby oncology center for radiotherapy.

DISCUSSION

Multiple CN palsies is one of the ominous condition seen in neurological clinics and ward. Sixth CN palsy has been

emphasised from early medical school days as a false localising sign and is considered in many instances before the complete work up of a case. Among numerous non-false localising causes of sixth CN palsy, clivus metastasis from adenocarcinoma of prostate has been reported as an important cause.¹ Sixth combined with twelfth CN palsy is rarely seen. It was Godtfredsen who first described this combination in cases of nasopharyngeal carcinoma, where he emphasised that, the findings of sixth CN paresis with twelfth CN paresis are not by a chance. There was also occurrence of trigeminal neuralgia in majority of such cases.² This combination, when caused by nasopharyngeal carcinoma, is described as Godtfredsen syndrome.³ Our patient did not have any evidence nasopharyngeal carcinoma and trigeminal neuralgia. Out of 21 patients, Kean and James reported five patients to have combination of sixth and twelfth CN palsy without other CN involvement. However, it has to be noted that, his series constituted 1262 patients with sixth nerve and 116 patients with twelfth nerve palsy. Chordoma of clivus, metastases (prostate, pancreas, ?ovary) and self-limited cranial polyneuropathy were the causes found in above five patients.⁴

Clivus metastases are extremely rare and represent 0.18% of all intracranial tumours and 0.42% of skull base tumours (overestimation is likely in this report).⁵ In a study by Kean and James, 41% had clival marrow metastases.⁴ Metastases is reported to have originated from prostate, thyroid, kidney, lungs, skin, bone, liver and gastric tissue.⁵ Our patient had rectal adenocarcinoma with clivus metastases and this association with sixth and twelfth CN palsy is reported for the first time to our knowledge.

In an attempt to describe the involvement of relatively separated sixth and twelfth CNs without other distal CNs involvement, Godtfredsen attributed sixth CN palsy to direct invasion of the tumour around the cavernous sinus and twelfth CN to the retropharyngeal lymphnode metastasis causing nerve dysfunction at hypoglossal canal, as all of the patients had cervical lymphadenopathy.² The anterior exiting nature of these two nerves is attributed to the involvement by clivus tumour which can describe our patient's condition, sixth nerve being compressed at

Figure 1 Left abducens and left hypoglossal nerve palsy.

Figure 2 Sagittal T1-weighted MRI showing clivus metastasis (arrow) with mild flattening of the pons (arrowhead). Normal MRI for comparison.

Dorello's canal by the clival mass. Noteworthy in our patient is that the first involvement of sixth CN followed by twelfth CN, which can be explained by the metastases involving relatively higher part of clivus first and

progressing to involve the lower part. Is this the predilection of tumour to involve the upper part first or occurrence by chance is yet to be determined. As sixth CN paresis may be the first sign of a clivus mass, the conventional

teaching of sixth CN palsy as a false localising sign should be followed by a statement that, it may be also a sign of serious underlying condition as this may be the first clinical sign of clivus mass and moreover we know that clivus metastases carries a very poor prognosis.

Learning points

- ▶ Evaluate for clival metastasis in unexplained sixth CN palsy
- ▶ 6 + 12 CN dysfunction is an important sign of clivus involvement
- ▶ Clival metastasis is a rare clinical entity
- ▶ Term 'Godfredsen syndrome' should be limited to CNs lesion due to nasopharyngeal carcinoma.

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Competing interests None.

Patient consent Obtained.

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