



Occipital Condyle Syndrome Revealing Non-Small Cell Lung Cancer in a Myasthenia Gravis Patient

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Commentary

A 44-year-old woman presented with a new-onset tension-like headache, pleuritic chest pain and a gradually deteriorating dysphagia (without dysarthria). The patient, with a history of heavy smoking (30 pack-year), had an one-year diagnosis of anti Muscle-Specific receptor tyrosine Kinase (MuSK) positive Myasthenia Gravis (MG), with excellent response on treatment with oral steroids.

On admission, she was lying in bed with the head rotated to the side of the pain, which was exacerbated by neck flexion and rotation of the head to the left. A right hypoglossal palsy was identified. No other symptoms or signs specifically related to a possibly worsening MG were elicited from history or neurological examination.

An MRI of the skull base revealed a hyperintense, enhancing mass infiltrating the right occipital condyle and extending to the hypoglossal canal and adjacent part of the inferior clivus (Figures 1A-1E). The thoracic CT findings were consistent with a possible lung tumor (Figures 2A-2C). The bronchoscopic biopsy confirmed a poorly differentiated non-small cell lung adenocarcinoma.

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Received Date: 06 Dec 2021

Accepted Date: 24 Dec 2021

Published Date: 05 Jan 2022

Citation:

Mastorodemos V, Papadaki E, Plaitakis A. Occipital Condyle Syndrome Revealing Non-Small Cell Lung Cancer in a Myasthenia Gravis Patient. Clin Oncol. 2022; 7: 1889.

ISSN: 2474-1663

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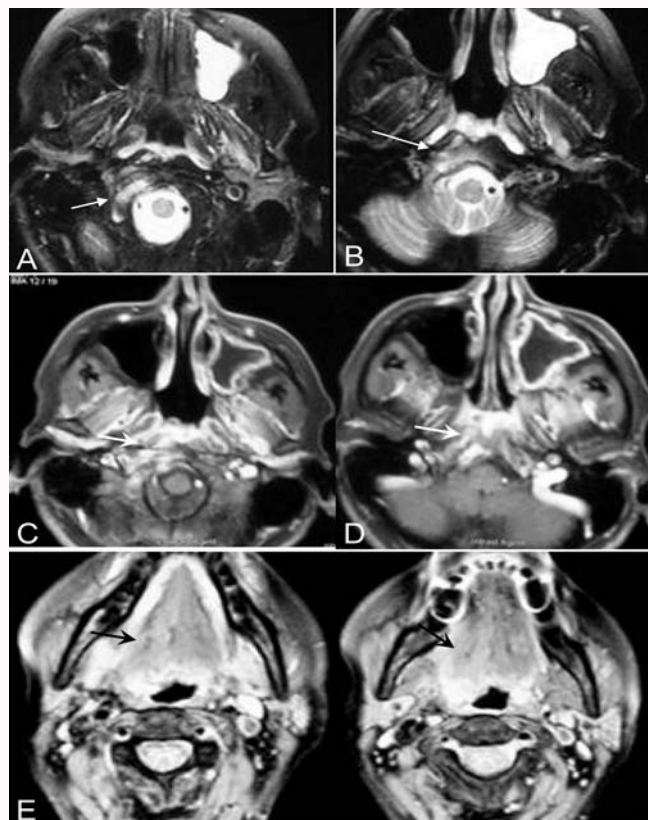


Figure 1: Fat-saturated T2 (A, B), post-Gadolinium T1 (C, D) and proton density (E) sequences of the skull base showed an hyperintense, enhancing infiltrating mass at the right condyle, that extended to the ipsilateral hypoglossal canal and the right inferior part of the clivus (A-D, arrows). In addition, a diffuse mild increase of signal intensity at the right part of the tongue, with effacement of the ipsilateral vallecula, indicative of right hypoglossal nerve involvement was demonstrated (E, black arrows).

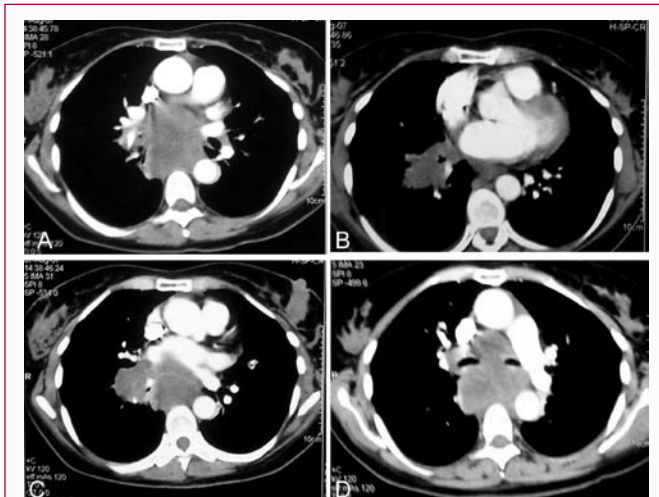


Figure 2: A contrast-enhanced CT of the thorax, 11/2 months after initial symptoms, revealed a solid, hypodense, 35 mm x 27 mm lung mass at the apical segment of the right lower lobe, with irregular margins and mild enhancement, that extended to the right hilum and infiltrated the right inferior pulmonary veins (A-C). An extensive mediastinal lymphadenopathy producing luminal narrowing of the trachea and the central bronchi was also observed (D).

Despite intense chemotherapy, she deteriorated rapidly and died of acute pulmonary failure, approximately four months after initial symptoms, due to mass enlargement and total obstruction of the right main bronchus (Figure 2D).

Comment: Occipital Condyle Syndrome (OCS) represents a stereotypic syndrome (unilateral occipital pain and ipsilateral hypoglossal paralysis), associated with skull base metastases in patients with a known history of cancer, mainly breast cancer in females and prostate adenocarcinoma in males [1,2]. However, only rarely has been reported as the initial manifestation of an underlying neoplasia [3,4].

The co-existence of cancer and myasthenia gravis raises the possibility of an etiopathogenetic correlation of these two entities. A recent study showed an increased association of MG with extrathymic malignancies, which may be explained by a primary immune dysregulation [5]. As our patient was anti-MuSK positive, which is thought to have a reduced response to conventional immunosuppressive therapies and has never been linked to cancer coincidence, it is tempting to relate the recent evolvement of myasthenia gravis with the aggressive course of the tumor.

In conclusion, the constellation of the characteristic symptoms in occipital condyle syndrome, though infrequently seen today compared to the older literature [1], should lead to early recognition and prompt a thorough search for possible primary cancer.

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