Relato de Caso

Opalski Syndrome: A Case Study

Síndrome de Opalski: Um Relato de Caso

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ABSTRACT

Opalski Syndrome (OS) is an unusual presentation of Lateral Medullary Syndrome (LMS) with ipsilateral hemiparesis. We report the case of a 65-year-old woman with a background of unprovoked deep venous thrombosis and unintentional weight loss that presented after a one-week-long episode of sudden onset vertigo, frontotemporal headache, blurry vision, and weakness. A head magnetic resonance imaging (MRI) revealed subacute ischemic event, with flow-void loss in the right vertebral artery and absence of blood flow, suggesting OS secondary to likely right posterior inferior cerebellar artery (PICA) occlusion. An abdominal and thoracic computed tomography (CT) scan showed suggestive findings of stage IV pancreatic tail cancer. Because of its non-classical presentation, OS might be a diagnostic challenge to most physicians. As in this patient, the existence of concomitant cancer probably led to a hypercoagulable state, which could explain the ischemic stroke while on anticoagulation therapy.

Keywords: Hemiparesis; Lateral Medullary Syndrome; Ischemic Stroke; Pancreatic Neoplasms.

ABSTRACT

A Síndrome de Opalski (OS) é uma apresentação incomum da Síndrome Medular Lateral (LMS) com hemiparesia ipsilateral. Relatamos o caso de uma mulher de 65 anos com história de trombose venosa profunda não provocada e perda de peso não intencional que se apresentou após um episódio de uma semana de vertigem de início súbito, cefaleia frontotemporal, visão turva e fraqueza. Uma ressonância magnética (RM) de crânio revelou evento isquêmico subagudo, com perda de fluxo vazio na artéria vertebral direita e ausência de fluxo sanguíneo, sugerindo OS secundária à provável oclusão da artéria cerebelar inferior posterior direita (PICA). Uma tomografia computadorizada (TC) de abdome e tórax evidenciou achados sugestivos de câncer de cauda pancreática em estágio IV. Devido à sua apresentação não clássica, a OS pode ser um desafio diagnóstico para a maioria dos médicos. No caso dessa paciente, a existência de neoplasia concomitante provavelmente induziu um estado de hipercoagulabilidade, o que poderia explicar o acidente vascular cerebral isquêmico em regime de anticoagulação.

Palavras-chave: Hemiparesia; Síndrome Medular Lateral; AVC Isquêmico; Neoplasias Pancreáticas.

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INTRODUCTION

Wallenberg syndrome (WS) is a neurological disorder caused by injury to the lateral and posterior segments of the medulla. Also known as Lateral Medullary Syndrome (LMS) or Posterior Inferior Cerebellar Artery (PICA) Syndrome, this corresponds to the most common syndrome of posterior circulation infarction[1], leading to symptoms associated with compromise of the lateral medulla functions. Ischemic stroke affecting this area may present with vertigo, central nystagmus, dysphagia, dysphonia and dysarthria, facial numbness, ipsilateral cerebellar ataxia and, in some cases, Horner syndrome. On the contralateral side, impaired temperature and pain sensation are described[2].

Opalski Syndrome (OS), a rare variant of LMS, is characterized by similar findings, but ipsilateral hemiplegia is seen as well[3]. This report aims to describe a case of an OS presenting to our hospital.

Case report

This case report was submitted and approved by the Research Ethics Committee of the University Hospital of the Federal University of Juiz de Fora under the number 6.880.377.

Herein present the case of a 65-year-old woman with a history of hypertension, osteoporosis and poor controlled type 2 diabetes mellitus. She had a previous history of unprovoked deep vein thrombosis (DVT) in her left leg, for which she was taking rivaroxaban 20 mg daily, and had lost 15 kilograms of weight within the past six months.

She presented to the hospital after a one-week-long episode of sudden onset vertigo, frontotemporal headache, blurry vision, and weakness. She described that the symptoms started abruptly as she woke in the morning. After that, she developed dysarthria, right-sided hemiparesis, facial weakness, and vomiting.

Motor system examination showed full-proportioned right-side hemiparesis, right side dysmetria and cerebellar ataxia, with tendency to fall to the right side. Cranial nerve examination was remarkable for a right eye multidirectional nystagmus and ptosis, alongside a hemianesthesia for pain and temperature contralaterally. History and neurological exam were compatible with LMS, besides the fact that it also involved ipsilateral hemiplegia considering the initial topographic diagnosis made.

The initial hypothesis was an ischemic stroke affecting the posterior circulation, more specifically, the PICA territory. A head magnetic resonance imaging (MRI) revealed subacute ischemic event, with flow-void loss in the right vertebral artery and absence of blood flow, suggesting arterial occlusion (Figure 1). Additionally, it showed several supratentorial lesions on both anterior and posterior circulation, suggesting cardioembolic etiology at first.

A previous electrocardiogram was negative for atrial fibrillation, and further investigation for identifying cardioembolic sites was initiated. Transthoracic echocardiogram revealed no alterations significant for cardioembolic events, and a cervical computed tomography (CT) revealed absence of contrast opacification of the distal V3 segment and a large part of the intradural segment of the right vertebral artery, suggesting occlusion.

During the etiologic investigation, corresponding to OS secondary to right-PICA occlusion (atherothrombotic ischemic stroke in posterior circulation), an abdominal and thoracic CT scan was performed, showing a primary neoplastic lesion in the tail of the pancreas, with lymph node implants in the upper abdomen and mediastinum, liver metastases, small peritoneal and bone implants; focal areas of infarction in the parenchyma of the kidneys and spleen; chronic thrombosis of the splenic vein, with varicose veins near the gastric fundus (Figure 2); suggestive findings of stage IV pancreatic tail cancer.

Given the limited data in literature suggesting possible benefits beyond 24 hours of last known well, our approach was to decide against reperfusion therapy[4]. The patient was offered a follow-up consultation in our Neurology service for stroke sequelae and secondary prevention, and transferred to a specialized oncology center, in order to continue disease staging and treatment, but passed away two weeks later.

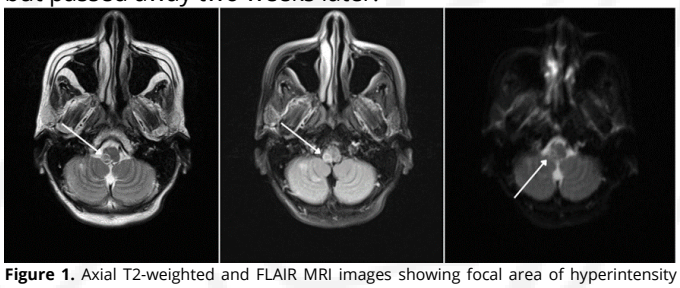

Figure 1. Axial T2-weighted and FLAIR MRI images showing focal area of hyperintensity involving dorsolateral medulla on the right side, suggestive of infarct. Axial diffusion weighted MRI image showing focal area of diffusion restriction in dorsolateral medulla on the right side, suggestive of acute infarct.
MRI - magnetic resonance imaging

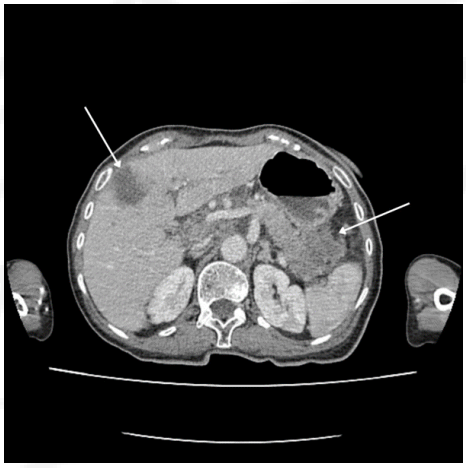

Figure 2. Abdominal CT image showing large expansive and infiltrative lesion in the tail of the pancreas, suggestive of primary neoplastic lesion, and heterogeneous nodular lesion in the liver, compatible with secondary implant.
CT - computed tomography

DISCUSSION

WS is a neurological disorder that results from malperfusion of the vertebral artery (67% of lateral medullary strokes) or its extension, PICA (10%)[6], as seen in the present case, which involved right-PICA occlusion[6]. Around 50% of WS cases stem from large-vessel infarction, with cardioembolism responsible in 5% of instances, while arterial dissections account for 15%, and small vessel infarcts contribute to 13% of WS cases[5].

OS, a rare variant of WS, presents with ipsilateral hemiparesis. It was first described in 1946 by Opalski, who reported two cases of lateral medullary stroke in which patients exhibited ipsilateral hemiplegia, ataxia, Horner syndrome, facial hypoesthesia and diminished superficial sensation of the contralateral side.

The cause of this manifestation remains controversial[3]. Initially, hemiparesis was attributed to the caudal extension of the infarct, involving corticospinal fibers after decussation. However, recent studies propose that involvement of medullary penetrating arteries which supply the pyramidal fibers post decussation may be responsible. These arteries are a branch of the vertebral artery, the most common artery implicated in WS[3].

Pancreatic cancer (PaCa) has a peculiar ability to induce a hypercoagulable state, representing one of the most prothrombotic neoplasms, with an incidence of thrombotic complications of up to 36%[6]. It is associated with high tumoural expression of tissue factor, activation of leukocytes with the release of neutrophil extracellular traps and the dissemination of tumor-derived microvesicles that promote hypercoagulability and increased platelet activation[6].

Previous studies have reported venous thromboembolism (VTE) prevalence rates of 12–36% in patients with PaCa. Arterial thromboembolic events are less common, with an estimated incidence of 2–5%, usually involving myocardial infarction and cerebrovascular events[6].

In this context, metastatic disease appears to be associated with a higher risk of thromboembolism than localized disease[6]. Concerning stroke, an observational retrospective multicenter study observed that, among patients with stroke and PaCa, in 93% of cases, the cancer was metastatic[6]. Regarding the type of PaCa, those involving the body and tail seem to be associated with a higher risk of VTE when compared with tumors of the pancreatic head[6].

Our patient had a primary neoplastic lesion in the tail of the pancreas with metastasis involving multiple organs. In this context, it is possible that the hypercoagulable state of the condition contributed to the development of VTE and subsequent ischemic stroke during anticoagulation therapy.

CONCLUSION

Because of its non-classical presentation, OS might be a diagnostic challenge to most physicians. This case study highlights the importance of identifying atypical variants of WS, such as OS, to ensure accurate diagnosis and effective treatment.

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