

Case Report

A rare case of carotid body tumor associated with near complete cerebral sinus thrombosis and idiopathic intracranial hypertension. Management strategy and review of the literature

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ABSTRACT

Background: Carotid body tumors (CBTs) are rare hypervascular lesions with critical location which makes them very challenging to treat. In rare occasions, compression of the jugular vein from the tumor mass could predispose to progressive thrombosis of intracranial venous sinuses. The latter consequently leads to intracranial hypertension (pseudotumor cerebri) with the accompanying danger to the vision. Herewith, we present our management strategy for this rare presentation of CBTs.

Case Description: A 38-year-old woman, with no medical history, was admitted in the emergency unit with acute onset of headache, dizziness, and vomiting. On the diagnostic imaging studies (CT venography and MRI) a near total occlusion of all cerebral venous sinuses and a large CBT (Shambin Type II) were diagnosed. Initially, the patient was treated with anticoagulants for the thrombosis and with lumbo-peritoneal (LP) shunt for the management of pseudotumor cerebri. At a second stage, after resolution of the cerebral sinus thrombosis, the CBT was completely resected under electrophysiological monitoring, without preoperative embolization. At 1-year follow-up, the patient is neurologically intact with functioning LP shunt, patent cerebral venous sinuses, without tumor recurrence.

Conclusion: We present a rare case of CBT with intracranial complications, which was managed successfully by staged treatment. Careful study of the preoperative radiological and laboratory data, thorough preoperative planning of the tridimensional lesion anatomy, as well as meticulous microsurgical technique under intraoperative electrophysiological monitoring was essential for the successful outcome of the case.

Keywords: Carotid body tumor, Cerebral dural venous thrombosis, Idiopathic intracranial hypertension, Intraoperative electrophysiological monitoring, Pseudo tumor cerebri, Tridimensional planning, Horos software

INTRODUCTION

Carotid body tumors (CBTs) or carotid paragangliomas are complex vascular lesions, which are derived from chromaphin cells within the adventitia of the carotid bifurcation. These tumors account for 0.6% of all head and neck tumors and with incidence of 1:30,000–1:100,000 in the general population.^[15] Although rare, slowly growing and rarely malignant (app. 5% of all CBT)^[10]

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