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Multidisciplinary approach in the treatment of malignant paraganglioma of the glomus vagale: a case report

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ABSTRACT

Malignant paraganglioma of the glomus vagale is a rare tumor entity originating from paraganglia or glomus cells. It typically affects middle age. It differs from benign paraganglioma because of its rapid growth and more aggressive clinical behavior. We report the case of a 40-year-old man presenting with a 5 cm lesion in the upper right cervical region detected by computed tomography (CT) and magnetic resonance imaging (MRI), which also showed enlargement of ipsilateral spinal and jugulodigastric lymph nodes with contrast enhancement. Clinical manifestations at diagnosis included a partial neurological deficit involving the right cranial nerves X, XI and XII. Tumor vascularization was assessed by digital angiography. The tumor mass was entirely removed by a right cervical approach with *en-bloc* resection with the regional lymph nodes. Histopathological examination showed a paraganglioma with cellular pleomorphism, necrotic microfoci, perineural infiltration and angiogenesis. Massive metastases in two of three jugulodigastric and one spinal lymph nodes on the right side were also detected. Postoperative workup included MRI, positron emission tomography (PET)/CT, meta-iodine-benzyl-guanidine (MIBG) scan, liver ultrasound and chest radiography. Subsequently, the patient underwent conformal radiotherapy with concomitant cisplatin administration. At the last clinical and radiological follow-up examination 5 years after completion of treatment, the patient was free of tumor recurrence.

The integrated treatment by surgery and chemoradiation was feasible and effective in the management of this rare case of malignant paraganglioma of the glomus vagale. Multicenter studies should be done to increase the knowledge of tumor presentation and natural history and to analyze the possible treatment options.

Key words: paraganglioma, glomus vagale, radiotherapy.

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