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Hereditary paraganglioma syndrome associated with *SDHD* gene mutations: a patient with multicentric presentation treated with radiotherapy. Case report

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ABSTRACT

Introduction. Extra-adrenal paragangliomas are rare tumors arising from neuroendocrine cells. Sporadic and hereditary forms have been recognized. Among the latter, the PGL1 and PGL4 syndromes are associated with a high risk of multiple localizations. The treatment of choice for paragangliomas is surgical resection, but in some cases surgery can be difficult due to particular or multiple tumor sites or may result in severe neurological deficits. In such cases radiotherapy can be an effective alternative. In this paper we describe the case of a patient affected by hereditary paraganglioma syndrome with multicentric presentation who was treated at our center by external radiotherapy.

Case report. A 55-year-old man presented in April 2008 with multiple paragangliomas: one in the left pontocerebellar angle, two in the middle neck, one mediastinal paraaortic mass, and an abdominal paraaortic lesion. The left pontocerebellar and mediastinal tumors were treated with three-dimensional conformal radiotherapy (3D-CRT) at total doses of 50.40 Gy and 55.80 Gy, respectively. The neck lesions were treated with intensity-modulated radiotherapy (IMRT) at a total dose of 55.80 Gy. The abdominal paraaortic lesion was surgically resected.

Results. No severe acute or late toxicity as evaluated with the EORTC-RTOG scale was observed. Fourteen months after the end of radiotherapy a whole body CT scan showed that the tumor lesions were stable in size and in their relations to contiguous structures. The arterial pressure was controlled by medical therapy and urine catecholamine levels were within the normal range.

Conclusions. We believe that in patients affected by unresectable paragangliomas radiotherapy is a safe and effective alternative to surgery. The use of high-dose conformity techniques such as 3D-CRT and IMRT will allow higher local control rates with relatively few side effects thanks to the possibility of dose escalation and reduction of the amount of irradiated healthy tissues.

Key words: familial paraganglioma syndrome, multiple paragangliomas, chemodectoma, 3D-conformal radiotherapy, intensity-modulated radiotherapy.

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