EVEROLIMUS IN MONOTHERAPY AS A THERAPEUTIC OPTION IN PARAGANGLIOMAS/PHEOCHROMOCYTOMAS.

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INTRODUCTION AND OBJECTIVES

Everolimus is a drug selective inhibitor of mTOR. In 2012 a Phase 2 study* of everolimus monotherapy demonstrated modest efficacy in patients with pheochromocytomas/paragangliomas. Our objective was to evaluate the efficacy and safety of everolimus in a patient with paraganglioma. (*Phase 2 study of everolimus monotherapy in patient with nonfunctioning neuroendocrine tumor or pheochromocytomas/paraglangliomas. Do-Young Oh et al.Cancer 2012;15:6162-70).

CLINICAL CASE

We present a 29 years old man with recent arterial hypertension diagnosis (triple therapy), vomiting, and weight loss. A scan study shows left para-aortic retroperitoneal mass of 7.4 x 8.5 x 8 cm involving the celiac trunk, superior mesenteric artery and left renal hilum. The cytology result by endoscopic ultrasound was spindle cell tumor, suspected of GIST (gastrointestinal stroma tumor). The pathological diagnosis after biopsy exploratory laparotomy was infiltration by paraganglioma. A metaiodobenzylguanidine scintigraphy show patholocical uptake in retroperitoneal mass, 9th right rib and left iliac bone, with great elevation of urinary catecholamines (normetanephrine: 12832 µg/ 24h; dopamine: 1345 µU/24 h). The patient was treated with 131I-MIBG, 200 mCi by intravenous infusion. A CT control showing progression of disease with high catecholamines (normetanephrine: 18140 µg/ 24h; Dopamine: 1944 µU/24 h). We tried treatment with cyclophosphamide, vincristine and dacarbazine (CVD) in cycles every 21 days with a partial response by CT and biochemical monitoring (normetanephrine: 11325 µg/ 24h; Dopamine: 830 µU/24 h). However the treatment was suspended after the second cycle for severe toxicity (ventricular systolic dysfunction severe, hepatolisis, neutropenia, anemia, thrombocytopenia and refractory hypertension). In this moment we started treatment with everolimus (10 mg/24 h) . After 10 treatment cycles the disease has remained stable (normetanephrine: 8706 µg/ 24h; Dopamine: 737 µU/24 h) with good blood pressure control and improvement of pain. No important side effects were observed.

FIG 1: METAIODOBENZYLGUANIDINE SCINTIGRAPHY

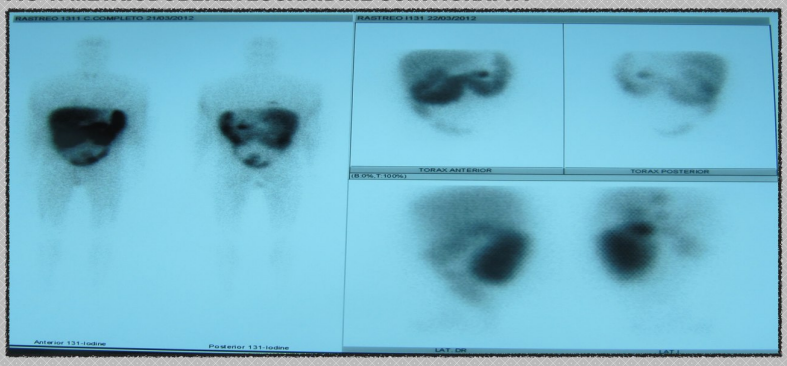


FIG 2: RETROPERITONEAL MASS EVOLUTION BY CT

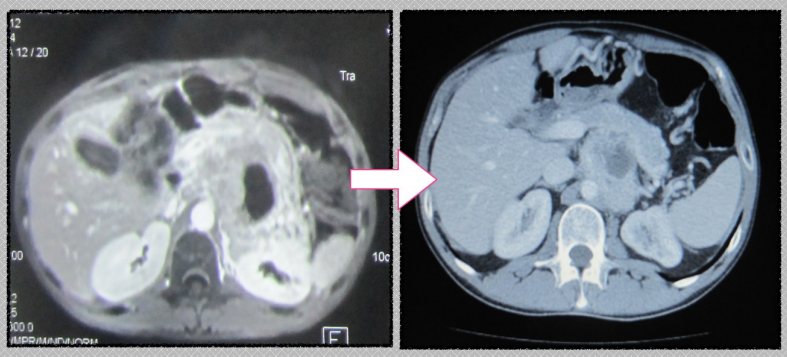
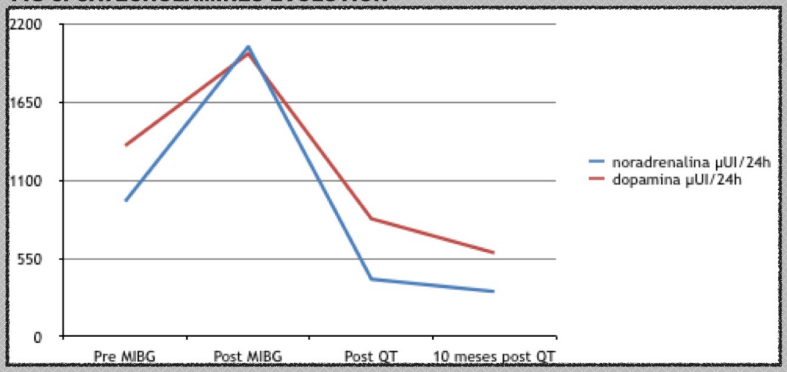


FIG 3: CATECHOLAMINES EVOLUTION



CONCLUSIONS

In cases of pheochromocytomas/paragangliomas with severe toxicity after treatment by chemotherapy CVD , everolimus in monotherapy keep the response with good tolerance , so it may be a good treatment option in this cases.

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