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Rare Case of Giant Cervical Paraganglioma

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Abstract **Clinical Image**

Paragangliomas are rare, mostly benign, adult neuroendocrine tumors that develop at the expense of the autonomic nervous system along major vascular axes. They are most frequently located in the cervico-facial region, and the diagnosis is usually made by the suggestive clinic and imaging workup. We present the case of Mr C.A., aged 55, without a notable history, who consults for a left laterocervical swelling that has been evolving for four years and gradually enlarging. The examination found a deterioration of the general condition and an evening heat without excessive sweating. Tinnitus such as ringing in the ears and pulsatile earache associated with hearing loss has been reported. The physical examination found ipsilateral facial paralysis, as well as a large swelling of the left parotid region measuring 7cm in long axis, firm, painless, mobile in the horizontal plane and fistulated in the skin, without palpable cervical lymph nodes. Imaging examinations including cervical ultrasound and a cervico-facial tomodensitometry found a hypervascularized deep tissue cervical mass arising at the left carotid bifurcation suggesting a tumor of the left carotid glomus. Initial workup for metastases did not find a second tumor location. The laboratory test for excretion of fractionated urinary metanephrines and normetanephrines in a 24-hour urine sample was negative, ruling out the diagnosis of secreting paraganglioma. Due to the size of the tumor lesion and the potential morbidity of a surgical procedure, surgery was not a therapeutic option. The patient was treated with arterial embolization followed by local radiotherapy. At two years follow up, reduction in tumor volume and regression of neurosensory symptoms.

Keywords: Giant cervical paraganglioma, Late diagnosis, Arterial embolisation, Radiation therapy.

Figure 1: Photographs of the patient: Front view [A] and Side view [B]

COVER LETTER

We present a photograph of a rare case of giant paraganglioma of the cervico-facial region. It is a benign tumor for which the curative treatment of choice is surgery. This treatment option could not be considered due to the tumor´s large size. The cause was most certainly the delay between symptomatology and treatment (4 years). An early diagnosis could have allowed a total resection of the tumor and thus avoid the installation of permanent complications such as facial paralysis. This clinical case illustrates the lack of concern, both of the population and of potential caregivers, for a condition that can initially be seen as simple cosmetic damage before the occurence of morbidity. This photograph would permit to sensitize the reader about investigating a cervico-facial mass, and raise the diagnosis of paraganglioma when this swelling is accompanied by an evocative symptomatology.

Conflict of interest: The authors declare no conflict of interest.