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Clinical Study

Paraganglioma in sella

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Summary

We report a very rare case of a paraganglioma arising from sellar and suprasellar region which has been treated with radiotherapy following multiple surgeries.

Introduction

Paragangliomas are originally derived from neural crest cells to arise rare tumors of the adrenal medulla and paraganglia [1]. These tumors are usually benign and have tendency to occur at multiple localizations within the body, including the carotid body, paranasal sinuses, and vagus nerve [2]. Although uncommon, they affect the central nervous system; most commonly the glomus jugulare and the filum terminale [3]. Paragangliomas which arise from sellar and parasellar area are very rare and there have been reported only a few cases in the literature up to date [2].

In this context, we report a new case of a paraganglioma arising from sellar and suprasellar region which has been treated with radiotherapy following multiple surgeries.

Case report

A 37-year-old man presented to our clinic on October 2003 with complaints of decreased hearing capacity and visual acuity with diplopia accompanied by a long history of recurrent sellar mass since 1995. Initially, he was diagnosed with a contrast enhancing $50 \times 25 \times 50$ mm^3 mass filling the sellar and parasellar region and suppressing the cavernous sinuses on his cranial magnetic resonance imaging (MRI) in 1995. The mass was initially excised gross totally. Then he underwent gross total and subtotal tumor resections in 1997 and 1999 respectively due to mass recurrence in the same localization. Subsequently on 2002, his follow up MRI demonstrated a recurrent $6 \times 6.5 \times 5$ mm^3 mass infiltrating the cavernous sinuses. He underwent a fourth operation for gross total resection. The histopathological examination revealed a neoplasm of neural crest origin which was immunocytochemically positive with diffuse chromogranin and focal cytokeratin staining. The cells were negative for S-100 protein, glial fibrillary acidic protein, epithelial membrane antigen (EMA), vimentin, and HMB45.

His complaints of diplopia increased 1 year after surgery and follow up MRI showed an enlarged sella and a contrast enhancing parasellar mass filling sellar and suprasellar cistern and infiltrating both cavernous sinuses (Figure 1a–b). The histopathological reevaluation of the specimen strongly supported paraganglioma with positive staining for chromogranin which is characteristic for neoplasm of neural crest origin and cytokeratin.

On admission, physical and neurological examination and laboratory findings were unremarkable except diplopia. We prescribed an external beam radiation dose of 50 Gy (2 Gy/day, 5 days/week) encompassing radiographically visible tumor as CTV plus 5-mm margin for setup uncertainty, making up the planning target volume (PTV). He has completed his treatment without any complications and has been stabile without progression for 1 year now.

As the patient was a referral from an outside community institution, we could not gather detailed information related with his previous endocrinologic status and laboratory tests before and after repetitive surgeries; however the laboratory tests at admission to our clinic revealed adeno-hypophysis hormones within normal range (Prolactin: <15 μg/l, GH: <5 μg/l, FSH: 15 IU/l, LH: 20 IU/l, TSH: 2.5 mU/l) and he has not experienced any endocrinologic problems after radiotherapy in 1 year follow up. The patient had initial reported complaints of decreased hearing capacity and visual acuity with diplopia. The hearing deficit remained stable through the course of radiotherapy. His diplopia was reported to deteriorate progressively before radiotherapy. He had no increase in his diplopia after radiotherapy in 1 year, but improvement in comparison to his admission examination at our radiation oncology clinic.

Discussion

We report a rare location of paraganglioma arising from sellar and suprasellar region. Paragangliomas arise from the paraganglion system of which

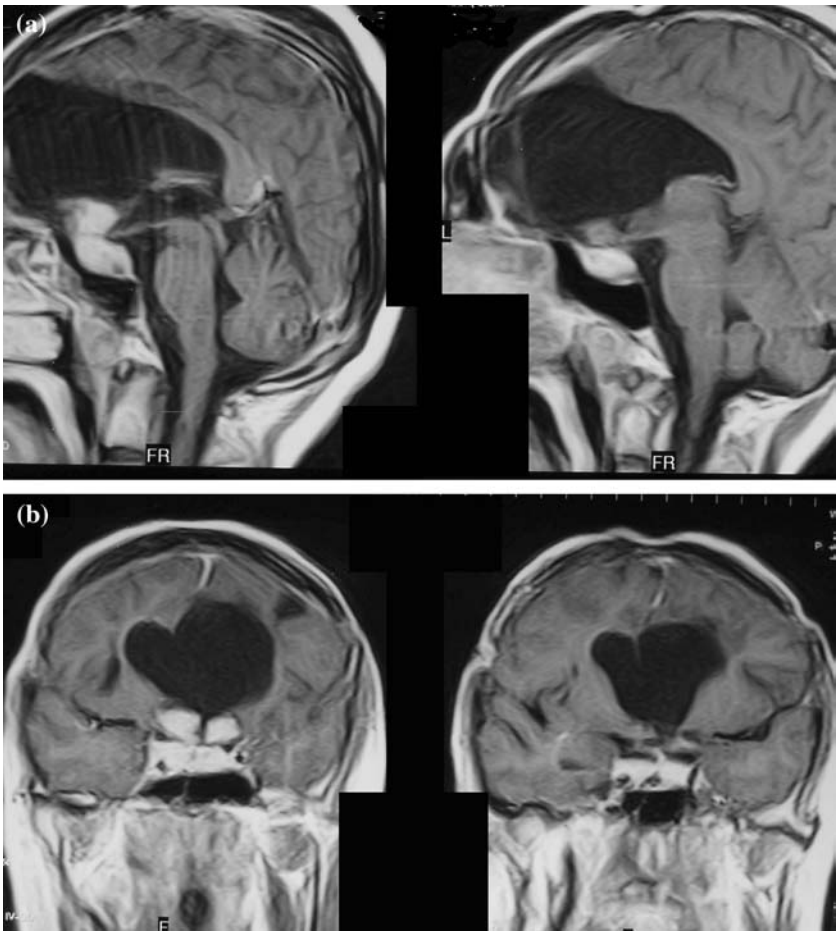

Figure 1. (a) Sagittal MRI showing an enhancing parasellar mass filling sellar and suprasellar cistern, (b) Coronal MRI showing a contrast enhancing parasellar mass filling sellar and suprasellar cistern and infiltrating both cavernous sinuses.

approximately 90% are in the adrenal gland (pheochromacytoma) with the largest collection of chromaffin cells. The remaining 10% arise from extra adrenal sites [4]. Sellar and parasellar paragangliomas are also very rare neoplasms with reported 11 cases in the literature up to date; 2 suprasellar, 2 intrasellar and 7 parasellar localizations [2]. To the best of our knowledge, this case who presented with sellar and suprasellar location seems to be one of the fewest reported cases in the literature.

Paragangliomas might be found in areas that do not normally contain paraganglionic cells in the adult, such as the duodenum, small bowel, kidney, cauda equina, and pineal region owing to considerable extensiveness of the paraganglion system in the fetus and neonate [1]. Bilbao et al. suggested that neural crest or paraganglionic tissue may be included in the developing adenohypophysis, on the basis of avian embryo pituitary studies [5].

It is a neurotologic neoplasm like acoustic neuroma, and named as glomus tumors, chemodectomas, non chromaffin tumors and carotid body tumors. It originates from special neural crest elements, the paraganglion cells, which, with autonomic ganglion cells, form the paraganglia. It is composed of two types of cells: chief cells and sustentacular cells (modified Schwann cells). These two types of cells function as chemoreceptors and their histology consist of characteristic Zellballen pattern [6]. Unlike adrenal paragangliomas, extra adrenal paragangliomas are uncommonly functional. Functional tumors secrete catecholamines, and approximately 1% to 3% of paragangliomas produce catecholamines [7]. These tumors usually are benign and slow growing but destructive. Paragangliomas are usually benign and vascular tumors. In about 3% of patients, the tumor is malignant [8].

The clinical presentation, neuroimaging characteristics, and behavior of sellar region paragangliomas are much the same as those of sellar meningioma and pituitary adenoma [3]. The paragangliomas differ from meningioma and adenomas with their different growth patterns. Meningiomas express EMA, and pituitary adenomas may express some pituitary hormones. Meningiomas are negative for chromogranin and synaptophysin stain, and both meningioma and adenomas do not have sustentacular cells.

The management of paraganglioma historically involved exclusive primary surgery or primary radiation therapy, with a very recent moderating trend toward selecting treatment according to patient profile. The location and extent of the tumor, the age of the patient, the presence of multiple tumors are conditions that affect the choice of the treatment. The goal of surgery is complete tumor removal, whereas the goal of conventionally fractionated, external beam radiation therapy and stereotactic radiosurgery is long-term tumor control

by preventing tumor growth and regional extension that could lead to progressive symptoms and neurologic deficits.

Total surgical removal ensures local tumor control rates varying from 0% to 90% [9]. Surgical removal can be associated with considerable morbidity and mortality [10–14]. In general, gross total surgical resection has been achieved in 40% to 80% of cases in different series [15–17]. Progressive growth of paragangliomas was arrested by conventional external beam radiation therapy at rates of local control ranging from 85% to 100%, with reversal of symptoms in some patients and complication rates of 0% to 10% [18–21]. In general, surgery and radiation therapy appear to have similar rates of long-term local control (~90%); despite the limitations and biases of retrospective, predominantly single-institution reported cases. This is true even though patients treated with radiation therapy generally have larger or more infiltrative tumors that are not readily subject to surgical resection. Because outcome after subtotal resection and radiotherapy is similar to that after radiotherapy alone [22], combined treatment is not common. But postoperative adjuvant therapy might be recommended if the symptoms or signs of cranial nerve compression persist following maximal tumor removal, or if there is evidence subsequent growth of residual tumor [23]. Due to recurrences in his sella with four previous resections, we decided to administer external beam radiotherapy to achieve local control in our case. He has no recent complaints or progression at first year follow up work up.

In the background of scarce literature data related with radiotherapy in this context, we report our case of parasellar paraganglioma to remind that these tumors might present with persistent recurrences following subtotal or even gross total resections and it is important to keep radiotherapy in mind for further management.

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