Skull base paraganglioma with local spread to the first cervical vertebrae

Dear Editor,

A 36-year-old male patient presented to our department with headache, dysphagia, vertigo, hypoacusis and swelling in the neck. After CT scan of the head & neck and angiography the diagnosis was skull base paraganglioma with local spread to the first cervical vertebrae.

After consultation with surgeons, external beam radiation therapy (EBRT) was delivered (total dose 54 Gy, 1.8 Gy/day). The first evaluation was 2 months after the end of EBRT; semi-annual follow up continued for the first 3 years and then the follow up became annual. The follow up consisted of physical examination and annual head & neck contrast CT scan. Local disease control was effective for 4 years (absence of tumor progression and clinical improvement of the patient). After 4 years the patient complained of headache and vertigo. A head & neck CT scan revealed disease progression.

Three sessions with radioactive indium-111 were administered via catheter with effective local control for 6 years.

Due to new progression of disease, the patient is now scheduled to receive treatment with radioactive yttrium.

Skull base paragangliomas are low grade tumors with their predominant pattern of spread being local [1]. The diagnostic approach is conducted with imaging which consists of head & neck contrast CT scan. The combination of a characteristic angiography and typical CT/MRI findings is considered equivalent to biopsy [2]. It is obvious, that in case of uncertainty, a biopsy can be performed in addition [3]. If there are questionable features of the imaging studies, biopsy may be necessary [4].

In general, one of the treatment options of skull base paragangliomas is EBRT with long-term effectiveness in local control and overall survival [5]. In case of disease progression the standard treatment options are either surgery or stereotactic RT or even radiosurgery. In the reported case, treatment was with radioactive indium-111 administered via catheter, with effective outcomes in local control and patient's neuro-logical symptoms.

Due to their location, many tumors of the skull base are treated with EBRT only, with a 10-year local control of 90% and 25-year local control of 73%.

Disease progression after RT can be managed either with surgery or with stereotactic radiosurgery (SRS), intensity modulated radiation therapy (IMRT), proton beam therapy or systemic treatment (the latter with no significant outcomes though). For the patient reported, the treatment chosen for disease progression after EBRT was radioactive indium-111 administered via catheter with effective local control during a follow-up period of 6 years. Treatment approach of new second disease progression can be with radioactive yttrium.

References

- 1. Hertel F, Bettaq M, Morsdorf M, Feiden W. Paragangliomas of the parasellar region. Neurosurg Rev 2003; 26: 210-214.
- Unal M, Polat A, Pata YS, Vayisoglu Y, Yildz A, Ismi O. Paraganglioma of the skull base: A case report. Auris Nasus Larynx 2007; 34: 427-430.
- Steel TR, Dailey AT, Born D, Berger MS, Mayberg MR. Paragangliomas of the sellar region: report of two cases. Neurosurgery 1993; 32: 844-847.
- Levy RA, Quint DJ, Devaney KO. Unusual tumours of the skull and skull-base: a pictorial essay. Radiol J 1996; 47: 436-443.
- Peltier J, Fichten A, Lefranc M, et al. Paraganglioma of the cavernous sinus. Case Report. Neurochirurgie 2007; 53: 391-394.

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