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CASE REPORT

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Preservation of vision through an interdisciplinary approach: a case report of intraorbital malignant solitary fibrous tumor

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Abstract

A 70-year-old female with a history of subtotal surgical resection of malignant solitary fibrous tumor (SFT) located in the lateral intraconal compartment of the right orbit was referred to the Oncology Team. The patient underwent surgical treatment with adjuvant radiotherapy of the right orbit. No major complications and no new neurological deficits related to radiotherapy were observed during the 4.5 years of follow-up. The discussed case is an essential source of knowledge for the medical community, demonstrating successful interdisciplinary collaboration involving surgery and high-dose radiotherapy of intraorbital malignant solitary fibrous tumors, particularly for optic nerve protection.

Keywords: solitary fibrous tumour, interdisciplinary approach, case report

Case report

The aim of our report is to present the results of a multidisciplinary approach in the treatment of intraorbital solitary fibrous tumor (SFT) after non-radical resection.

A 70-year-old female underwent surgery due to an intraorbital, intraconal tumor of the right orbit. During the first surgical treatment in 1996, the tumor was diagnosed as neurofibroma. In 2018, the patient appeared with a large tumor (37 × 21 × 25 mm) within the lateral intraconal compartment of the right orbit (Fig. 1). Right-sided exophthalmos and visual field deficits were detected before the second surgery. Using minimally invasive lateral orbitotomy, the tumor was subtotally resected. Only a small capsule fragment attached to the optic nerve was left. Immediately after surgery, no new visual deficit was observed. Histopathological examination did not confirm the initial diagnosis of neurofibroma, and a malignant solitary fibrous tumor was diagnosed [CD34(+), STAT6(+), CD(–), ERG(–)]. A few months later, a significant vision improvement was observed with normalization of the visual field.

Based on the result of histopathological examination, the patient was referred to the Oncology Team and qualified for adjuvant radiotherapy (after surgery). The plan of the treatment was based on preoperative magnetic resonance imagining (MRI) (Fig. 1) and postoperative computed tomography (CT) scan.

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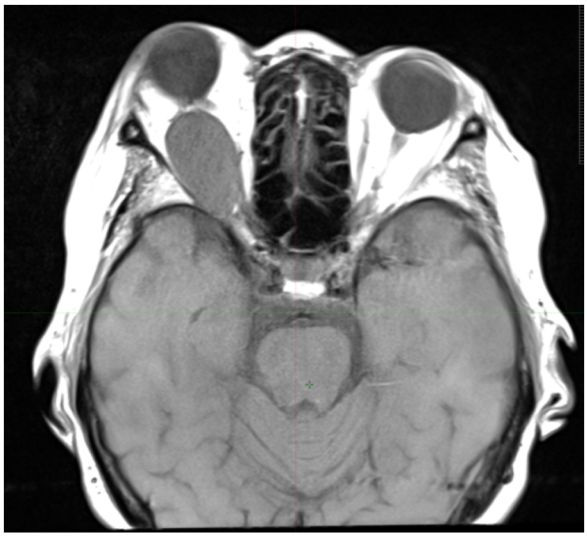

Figure 1. Magnetic resonance imaging before surgery

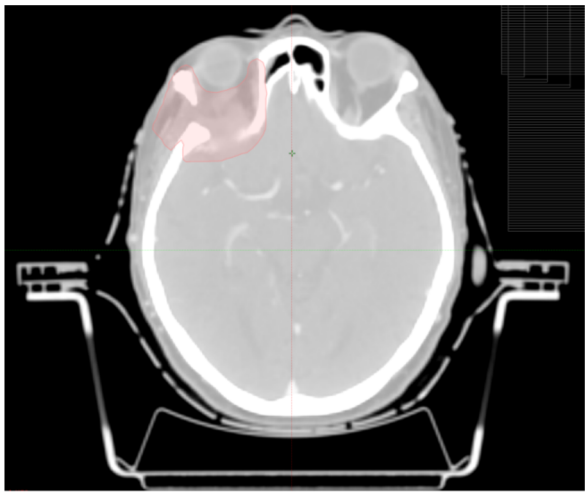

Figure 2. Contouring of clinical target volume (CTV) 60 Gy

Information that it was possible that a small remnant of the tumor was left attached to the optic nerve sheath and contrast enhancement within the orbital cone on postoperative MR resulted in qualifying the patient for adjuvant high-dose radiation therapy. Intensity modulated radiation therapy (IMRT) (Varian Medical Systems) was applied onto the tumor bed (space after the surgical intervention), with the treatment energy of 6 MeV photons. A total dose of 60 Gy was delivered with 30 fractions within 6 weeks. The contoured area of the tumor bed, our clinical target volume (CTV) for a 60 Gy dose, is shown in Figure 2. The distribution of treatment plan dose of 57 Gy (95% total dose) is shown in Figure 3. The maximum dose on the optic nerve was 59.4 Gy, and the mean dose was 56.99 Gy. The maximum dose on the right eye was 59.37 Gy; the mean was 38.28 Gy. The maximum dose on the right lens was 17.9 Gy; the mean dose was 14.05 Gy. The maximum dose on the left lens was 2.28 Gy; the mean dose was 1.75 Gy. The patient accepted the risk of vision deterioration after the high dose of irradiation. Ophthalmological follow-up examination revealed worse visual acuity due to a growing cataract, but there was no regrowth of the tumor within the orbit.

Discussion

Solitary fibrous tumors are rare spindle-cell neoplasms initially documented in pleural locations by Klemperer and Rabin [1]. Westra et al. [2] were the first ones to describe STF in the orbit. The most common symptoms related to SFT are arthralgia, hypoglycemia, effusion, and exophthalmos [3].

Contribution of radiotherapy within a multidisciplinary treatment context has not been directly investigated in the literature [4–9]. In a study conducted by Krengli et al. [10], comparable overall survival (OS) rates between patients who had undergone surgery (Sx) alone and those who had received both surgery and radiotherapy (RT) were observed. The actual 5-year local control (LC) rates were 50.4% after Sx and 91.6% after Sx plus RT ($p < 0.0001$) for LC, and 50.4% after Sx and 83.1% after Sx plus RT ($p < 0.008$) for disease-free survival (DFS). However, radiotherapy demonstrated enhancements in both LC and DFS [10]. Importantly, late local recurrences were identified, even beyond ten years [7].

Complete excision of the tumor remains the preferred treatment for SFT. In our case, it was not possible without damaging the optic nerve and impacting on vision. Radical surgery may be a major prognostic factor for LC and DFS [11, 12]. With R0 and R1 resections, the status of the margin does not affect the result, but in research conducted by Krengi et al. [10], R2 operations without adjuvant treatment resulted in very high baseline local recurrences reaching up to 75%. The role of radiotherapy in SFT treatment is still disputable. According to Salas et al. [7], adjuvant radiotherapy improves LC without affecting OS. Nevertheless, factors such as tumor location, size, adhesion, potential for bleeding, and postoperative complications might occasionally preclude this approach or can be indicators for adjuvant radiotherapy [10, 13].

Different doses of adjuvant radiotherapy ranging from 45 to 68.4 Gy (mean 60 Gy) using different radiotherapy techniques are encountered in the literature [10]. The radiotherapy dose level can be conditioned by the tumor location and the margin status of the postoperative histopathological examination. Based on our review of the literature, we decided to apply the mean irradiation dose. In our case, it was difficult to assess the margin status. Information gathered

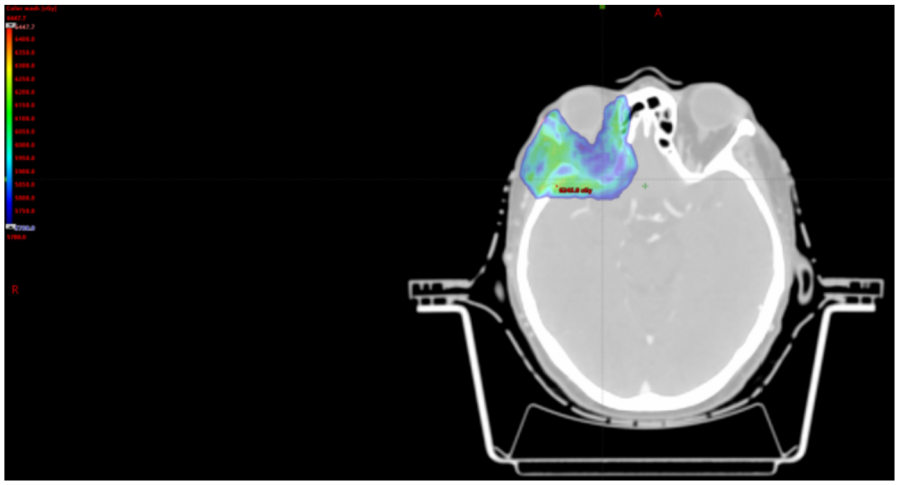

Figure 3. Distribution of a dose of 57 Gy

from the operative theatre and the postoperative MRI findings may have suggested residual remnants of the tumor. Therefore, the patient was qualified for adjuvant high-dose radiotherapy. This appeared to be successful and for 4.5 years of follow-up, no recurrence was diagnosed. Certainly, one can expect late radiation side effects with the risk, according to the literature, at a level above grade 2.

Conclusions

An interdisciplinary approach with minimally invasive surgery of intraorbital malignant SFT preserving eye function combined with high-dose radiotherapy could be effective and safe for patients with nonradical surgical treatment.

Article Information and Declarations

Ethics statement

The patient consented to being part of this study.

Author contributions

All authors made substantial contributions to the study design and data analysis and interpretation. All authors drafted the manuscript, revised it critically for important intellectual content, and read and approved the final version of the manuscript to be published.

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Conflict of interest

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Supplementary material

None.

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