Subependymal giant cell astrocytoma treatment

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Surgical resection has been the standard treatment for SEGA, however, medical management through mTOR inhibitors has now predominantly replaced surgery as the primary treatment modality. Additionally, newer treatment modalities have emerged with the hopes of providing safer methods for treating the tumour such as laser interstitial thermal therapy (LITT). However, very few reports have addressed these newer methods and analysed the results [1)]

Technological advances in neuroendoscopy and the more recent use of Laser interstitial thermal therapy have significantly enlarged the range of available management opportunities.

In 2020 A thorough review of the literature has been performed. Accordingly, current views in open surgical treatment, medical therapy, endoscopic tumor removal, and new trends (such as laser interstitial thermal therapy) are discussed.

The risk of significant neurological morbidity (5-50%) complicating open surgery has been for a long time representing a main drawback in the management of SEGAs. More recent series report a significant reduction of morbidity and mortality. The mTOR inhibitors have demonstrated efficacy in both warranting a tumor reduction by up to 60% of the tumor size and helping the control of seizures. However, the reported rate of side effects is as high as 30% and tumor recurrence is a documented occurrence at the time of mTOR inhibitor discontinuation. Endoscopic tumor removal has been more extensively considered an option due to the acquisition of new tools. Limits are still represented by tumor size (< 3 cm) and broad attachment of the tumor to the basal ganglia. Laser interstitial thermal therapy (LITT) is the more recently considered option. Though promising, only short follow-up is available so far, while data on medium- and long-term results of this treatment are completely lacking to date.

Surgical treatment remains a mainstay of the management of SEGAs. The indication for an open craniotomic approach should be balanced with an endoscopic tumor removal or LITT according to patient conditions, the presence or not of active hydrocephalus, and extension of the attachment of the tumor to the basal ganglia. The mTOR inhibitors do have a definite role both as primary and as

adjuvant treatment, but consistent limitations are represented up to now by a not negligible rate of complications and the uncertainties related to the possibility of tumor recurrence once the medical treatment is discontinued [2].

Laviv et al.reported two cases of recurrent shunt malfunctions in adult TSC patients with protein-secreting SGCTs and describe the complexity of treating such patients with an emphasis on the role mTOR inhibitors may have in their management [3].

SEGAs have been reported to regrow if mTOR inhibitor therapy is stopped, raising the possibility that long-term medication may be required to prevent tumor growth and hydrocephalus. The question of regrowth following medication withdrawal will need to be addressed in more patients to help establish the optimal duration of therapy. The risks of surgery include acute morbidity and the permanent need for ventriculoperitoneal shunting, which must be balanced against the adverse effects of mTOR inhibitors, including immunosuppression (infections, mouth sores), hypercholesterolemia, and the need for chronic drug monitoring. Some additional benefits of mTOR inhibition in patients with tuberous sclerosis complex, however, may include shrinkage of angiofibromas and angiomyolipomas as well as a possible decrease in seizure burden. Recent reports of successful nonsurgical treatment of SEGAs are promising, and it is hoped that further specifics on dosing, duration, and long-term outcome will help patients and physicians to make informed therapeutic choices.Present treatment recommendations for SEGAs include routine surveillance neuroimaging and close clinical follow-up, paying particular attention to signs and symptoms of acute hydrocephalus. If symptoms arise, or if serial neuroimaging demonstrates tumor growth, neurosurgical intervention is recommended. When gross total resection is impossible, rapamycin and everolimus should be considered, but may not offer a durable response.

In a phase 1–2, open-label study in 28 patients with evidence of serial subependymal giant cell astrocytoma growth, the mTOR inhibitor everolimus (Afinitor, Novartis, East Hanover, NJ) was associated with a reduction in SEGA volume and improved quality of life [4].

Arroyo et al. present a seven-year-old boy with a large, symptomatic SEGA which was treated acutely with everolimus.

Everolimus treatment resulted in rapid reduction in tumor size, symptomatic improvement, and decrease in cerebrospinal fluid protein.

Everolimus can effectively reduce tumor size, decrease cerebrospinal fluid protein, and allow successful ventriculoperitoneal shunt placement without the need for surgical resection of a symptomatic SEGA [5].

References

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