Successful Use of Radiotherapy for Pterygopalatine Fossa Amyloidosis

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Amyloidosis is a group of heterogeneous diseases resulting from the accumulation of misfolded proteins, which can be deposited into any organ or tissue. Although amyloidosis is rare, with an incidence of 1 to 5 per 100,000 people, 20% of cases involve structures of the head and neck, therefore making it relevant to otolaryngologists. Current treatment of local amyloidosis focuses around surgical resection; however, optimum treatment is unknown for cases that are not amenable to surgery. We present the first documented case of pterygopalatine fossa amyloidosis successfully treated with radiotherapy. Exemption was granted from the NHS Research Ethics Committee.

Case Report
A 40-year-old woman presented with a 2-year history of gradual left-sided proptosis, maxillary pain, and nasal obstruction. Magnetic resonance imaging showed a lesion in the left pterygopalatine fossa, extending into the orbital apex and through the inferior orbital fissure into the left cavernous sinus (Figure 1). Endoscopic transnasal biopsy led to the diagnosis of localized left pterygopalatine AL amyloidosis.

The patient’s symptoms continued to deteriorate with reduction in her visual acuity. It was therefore decided that a conservative approach would not be appropriate. Discussion at the regional multidisciplinary team considered surgery as an option. However, it was thought that this would be associated with significant morbidity. Instead, a trial of radiotherapy was started. The optimum dose of radiotherapy was unknown in this case. A report by Khaira et al used 3400 and 3000 cGy to treat orbital amyloidosis, without complication.3 Given the near orbital apparatus involvement in this case and visual compromise, it was decided that the patient should undergo a similar but slightly higher dose of radiotherapy. As such, the patient underwent radiotherapy with a total dose 3600 cGy over 20 fractions. This resulted in marked improvement of symptoms, including visual acuity, with no reported complications. Posttreatment and annual surveillance imaging demonstrated a response to treatment with a reduction of abnormal infiltrative soft tissue in the left pterygopalatine fossa. This suggests involution in response to the radiotherapy and demonstrates maintenance of stable appearances with no evidence of progressive disease at 5 years (Figure 2).

Discussion
Amyloidosis is a group of disorders characterized by the accumulation of beta-pleated sheets and deposition of fibrils formed by defective proteins. Presentation varies according to the organs and tissues affected and the amyloid type. Classification of amyloid is described by the prefix “A” followed by the letters that represent the associated subunit protein. Although 25 separate proteins have been discovered that can cause amyloid, only a small number of these are associated with clinically significant disease. The 2 most common types of amyloid are AL and AA. Amyloidosis may also be classified as local, whereby only 1 organ or tissue type is affected, or systemic, where >1 organ/system is affected, and it may be acquired or familial. For all, pathologic specimens will demonstrate apple-green birefringence after staining with Congo red dye, when viewed under polarized light.4

Amyloidosis often affects structures in the head and neck and is therefore of importance to otolaryngologists. Treatment for head and neck AL amyloid would usually involve either a conservative approach or a local surgical excision.1 However, we present a challenging case where the location of disease would make surgical access difficult with close proximity to the orbit, globe, and extension to the cavernous sinus.

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Currently, optimum treatment for such cases is unknown. Given the progressive eye symptoms in this case, conservative management was not an option, and surgical excision would have resulted in significant morbidity for benign disease. There have been reports of successful treatment with radiotherapy alone and in combination with surgery in some cases of head and neck amyloidosis.\(^5\) Radiotherapy therefore provides an alternative treatment option, although currently there is little evidence base to inform the dose that should be used.\(^2\)

**Conclusion**

We present the first documented case of pterygopalatine fossa amyloidosis and demonstrate successful use of
primary radiotherapy as a treatment modality for local AL amyloid. In keeping with recent literature, we have demonstrated a positive response to radiation therapy being used therapeutically to treat head and neck amyloidosis. We recommend, following multidisciplinary discussion, that radiotherapy be considered an alternative treatment option for localized AL amyloidosis.

Author Contributions
Kristina Lee, contributed to the conception and design of the work and interpretation of data; drafting and revising the manuscript for intellectual content; final approval of the version to be published and agreement to be accountable for all aspects of the work; Navin Mani, contributed to interpretation of data; drafting and revising the manuscript for intellectual content; final approval of the version to be published and agreement to be accountable for all aspects of the work; Zhi En Tan, contributed to interpretation of data; drafting and revising the manuscript for intellectual content; final approval of the version to be published and agreement to be accountable for all aspects of the work; Nick Slevin, contributed to interpretation of data; drafting and revising the manuscript for intellectual content; final approval of the version to be published and agreement to be accountable for all aspects of the work

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