CASE STUDY

Extramedullary Plasmacytoma of the Larynx. A Case Report

Isabel González Guijarro, Laura Díez González, Nieves Rodríguez Acevedo, Estrella Pallas Pallas

Service de Otorrinolaringología, Hospital Xeral-Cíes de Vigo, Vigo, Spain
Servicio de Otorrinolaringología, Hospital del Bierzo, Ponferrada, Spain

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Abstract Extramedullary plasmacytoma of the larynx and localised laryngeal amyloidosis are two entities that are extremely rare in children. We report the case of an 11-year-old child presenting with progressive dysphonia, with a diagnosis of extramedullary plasmacytoma and localised laryngeal amyloidosis. The treatment he received and subsequent follow-up were compared with the few cases found in the literature.

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PALABRAS CLAVE
Plasmocitoma extramedular; Amiloidosis; Plasmocitoma laríngeo; Población pediátrica

Plasmocitoma extramedular de laringe. A propósito de un caso

Resumen El plasmocitoma extramedular de laringe y la amiloidosis localizada laringea son dos entidades infrecuentes y extremadamente raras en niños. Presentamos el caso de un niño de 11 años con disfonía progresiva, diagnosticado de plasmocitoma extramedular y amiloidosis localizada laringea. Comparamos su tratamiento y posterior evolución con los escasos casos encontrados en la literatura.

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Clinical Case

We present the case of an 11-year-old boy sent to the reference hospital due to dysphonia of 2 months’ evolution, without respiratory and deglutitive disruptions.

Physical examination showed a right hemilaryngeal tumour that bulged towards the glottal lumen, reducing ipsilateral mobility. We were unable to define its extension to the right vocal fold (RVF) and the remainder of the ENT exploration was normal (Fig. 1A). The MRI revealed a submucosal glottic tumour centred on the RVF, which deformed the right aryepiglottic fold and obliterated the ipsilateral pyriform sinus, discarding vascular aetiology (Fig. 2A).

We performed laryngeal microsurgery, with intraoperative amyloidosis biopsy and reduction of the tumour mass with CO₂ laser.
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Figure 1 (A) Preoperative video stroboscopy: right hemi-laryngeal submucosal tumour bulging into the glottal lumen associated to RVF paresis. (B) Postoperative video stroboscopy: disappearance of tumour with preserved RVF mobility.

Figure 2 (A) Before surgery: cervical MRI: right submucosal hemi-laryngeal tumour. 1) H- - -E: plasma cells along with extracellular, hyaline and eosinophilic material. 2) Congo red: apple-green birefringence with polarised light. 3) Immunohistochemistry: expression of lambda chains and negative for kappa.
(B) After reoperation: cervical MRI: decrease in mass effect with persistence of the lesion. 1) H- - -E: large masses of amyloid material. 2) Congo red: positive without accumulation of plasma cells. 3) Immunohistochemistry: absence of plasma cells.

The final anatomopathological (AP) results were plasmacytoma, B lymphoid monoclonality and localised amyloidosis (Fig. 2A).

The patient was referred to the haematology service for an extension study, which was negative and rejected the existence of multiple myeloma and plasmacytoma at other levels.

In the postoperative control, there was a significant reduction in tumour mass, with good glottic passage and recovery of right vocal fold mobility (Fig. 1B). The MRI showed persistence of the right hemi-laryngeal mass, although smaller in volume compared to the previous study.

After assessing treatment options in conjunction with the haematology and radiotherapy services, radiotherapy was applied with curative intent, with a total dose of 45 Gy. The patient showed good tolerance and clinical improvement.

Laryngeal microsurgery was performed 3 months after radiotherapy to reduce the tumour mass and to confirm eradication of plasmacytoma, with an AP result of localised amyloidosis and no evidence of plasmacytoma (Fig. 2B).

Regular follow-up controls during the past 3 years have evidenced persistence of the right hemi-laryngeal amyloid lesion (Fig. 2B), although with clinical improvement in both dysphonia and in tolerance to physical exercise.

Discussion

Extramedullary plasmacytoma is an uncommon entity, which is extremely rare in children as its mean age of presentation is 56–59 years, with a predominance in males. We have found only one case of coexistence of amyloidosis and laryngeal plasmacytoma in children described in the literature: a 12-year-old girl, published by Nagasaka et al.

The symptoms are caused by local growth, with hoarseness, dysphagia, cough and dyspnoea being the main manifestations. Pribitkin et al., in a study in patients with laryngeal amyloidosis, pointed to hoarseness as the main manifestation, while Riazza et al. pointed to dyspnoea.

The diagnosis of extramedullary plasmacytoma is fundamentally histological, based on the presence of plasma cells showing apple-green birefringence with Congo red staining, presenting cytologically benign traits; however, immunohistochemical study shows monoclonality, pointing to their neoplastic nature. In our case, immunohistochemical techniques expressed lambda chains, with a negative expression of kappa chains, and the molecular study showed B lymphoid monoclonality.

An extension study is required to establish a differential diagnosis with multiple myeloma and solitary bone plasmacytoma, considering that extramedullary plasmacytoma is generally a localised entity associated with long survival.

In the treatment of laryngeal amyloidosis, excision of the lesion with CO2 laser is effective because, despite having invasive capacity, altering the functionality of the vocal fold, the lesion behaves like a benign tumour and does not usually present recurrence in localised forms. Nevertheless, some authors advocate an expectant attitude, reserving surgery for specific cases.

Extramedullary plasmacytoma is highly radiosensitive, with local control being achieved in 95% of cases with total doses of 40–60 Gy. In the patient described, we excised the lesion with CO2 laser in the first surgical step, obtaining significant clinical improvement and reduction of the tumour mass. After the definitive AP report of extramedullary plasmacytoma, we decided to apply radiotherapy, achieving the eradication of the plasmacytoma, although the amyloid mass persisted. Given the patient’s limited symptoms, we chose to conduct periodic reviews, observing good clinical evolution.
Conclusion

- The presence of localised laryngeal amyloidosis and laryngeal plasmacytoma could be the two infrequent entities in children.
- The diagnoses are mainly histological and immunohistochemical.
- Treatment should be aimed at eradicating the plasmacytoma and reducing the mass effect. However, in some cases with few symptoms, it may be limited to regular controls.

References