Case Report

Extramedullary Plasmacytoma of the Larynx Treated by a Surgical Endoscopic Approach and Radiotherapy

Massimiliano Pino, Filippo Farri, Pietro Garofalo, Fausto Taranto, Andrea Toso, and Paolo Aluffi

1E.N.T. Department, University "Amedeo Avogadro" of Piemonte Orientale, 28100 Novara, Italy
2E.N.T. Department, A.O.U "Città della Salute-Regina Margherita", 10126 Torino, Italy

Correspondence should be addressed to Pietro Garofalo; garofalo.ptr@gmail.com

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Extramedullary plasmacytoma (EMP) is a rare variant of plasma cell myeloma that affects soft tissues. The head and neck region are the most affected sites, although others have also been described. Herein we report an uncommon case of EMP of the larynx in a 65-year-old male who presented with a history of progressive dysphonia and hoarseness. Laryngeal fiberscopy evidenced a reddish pedunculated voluminous mass in the left false cords and ventricle. Microscopic suspension laryngoscopy was performed under general anaesthesia and a 4 W Acublade CO2 Laser was used for transoral resection of the lesion. This was followed by adjuvant radiotherapy, with the widely recommended doses on the supraglottic region, to achieve better local control. Diagnosis of EMP is based on immunohistochemistry and the exclusion of systemic plasma cell proliferative disorders. Diagnosis of solitary EMP can be made only if studies for disseminated disease and X-ray and/or magnetic resonance imaging of the spine, pelvis, femurs, and humerus and bone marrow biopsy are negative. As there are no internationally established guidelines, treatment of EMP is mainly based on consensus of expert opinion.

1. Introduction

Plasma cell neoplasms represent a spectrum of diseases ranging from benign conditions, such as monoclonal gammapathy, to malignant entities, such as plasma cell myeloma and plasma cell leukemia [1]. Plasma cell myeloma (multiple myeloma (MM)), a neoplastic proliferation of plasma cells [2], is the most common plasma neoplasm and is characterized by the involvement of the bone marrow at multiple sites [3]. A solitary plasmacytoma is a single localized mass of neoplastic plasma cells occurring in either bone (medullary) or soft tissue (extramedullary). The localized variant may be either the first evidence of generalized myeloma or an independent solitary lesion, characterized by the production of monoclonal immunoglobulins detectable in the serum and/or urine [2].

Most cases of extramedullary plasmacytoma (EMP) are seen in older men (male:female ratio 3:1, with a peak incidence in the 50–70-year-old group [4]). Extramedullary plasmacytomas have been reported in various sites in the body, such as the airway passages, gastrointestinal tract, and soft tissues [5]. However, about 80% of extramedullary plasmacytomas occur in the head and neck region [6], mainly in the nasal cavity, paranasal sinuses, or nasopharynx [4], whilst EMP of the larynx is quite rare.

2. Case Report

P. L., a 65-year-old male, presented to the Ear, Nose and Throat (E.N.T.) Department of the Azienda Ospedaliero-Universitaria “Maggiore della Carità,” Novara, Italy. He was complaining of dysphonia that had worsened continuously over a 6-month period, accompanied with a slight dysphagia of a 3-month onset. Laryngeal fiberscopy evidenced a reddish pedunculated voluminous mass in the left false cords and ventricle (Figure 1), without impairment of the airway
Patency. No lymph node enlargement was detected at neck palpation.

Direct suspension laryngoscopy was performed under general anaesthesia with a binocular microscope having a 400 mm objective lens coupled with a CO2 laser (Acublade SuperPulse 4 W). The lesion was radically resected with CO2 laser, removing the left false cord whilst preserving the left vocal cord. Systemic antibiotics (Cephalosporin) and PPIs were administered to prevent the formation of excessive fibrin and granulation tissue, which might lead to stenosis.

The patient then had adjuvant radiotherapy at a dose of 46 Gy in 24 fractions on the supraglottic region, due to closed tumour-free margins.

Histopathology revealed a tumour with a monomorphic population of atypical plasma cells, which had totally or partially substituted the laryngeal architecture (Figure 2). Higher magnifications showed that plasma cells had eccentric nuclei and atypical cytology (prominent nucleoli, dispersed nuclear chromatin, and a high nuclear-cytoplasmic ratio) (Figure 3). Neoplastic cells showed diffuse membrane positivity for CD138 at immunohistochemical staining (Figure 4).

Both the histopathological and immunohistochemical features were consistent with a diagnosis of extramedullary plasmacytoma. Laboratory examinations (full blood counts, serum chemistry, and serum protein immunoelectrophoresis- (IEP-) serum test) were within the normal range. Further systemic investigations, that is, X-ray of the spine, pelvis, femurs, humerus, and bone marrow biopsy, were performed after surgical excision so as to stage the disease. A diagnosis of solitary extramedullary plasmacytoma was made on the basis of the findings.

3. Discussion

Although EMP of the larynx is rare, it is on the increase, with 4.5% to 18% of EMP of the head and neck occurring in the larynx [7]. Although the clinical presentation varies depending on the organ involved [8], EMP of the larynx often presents with hoarseness and/or dysphagia [7]. Cervical node metastasis and acute laryngeal obstruction have also been reported [7]. Common sites of laryngeal involvement in order of frequency are the epiglottis, ventricles, the vocal cords, false cords, the aryepiglottic folds, arytenoids, and subglottis [7]. Plasmacytomas in the larynx vary from smooth polypoidal tumours with narrow bases to sessile tumours with wide attachments, which may be red or pale pink.

Diagnosis of extramedullary plasmacytoma is based on the exclusion of a systemic plasma cell proliferative disorders and immunohistochemistry results. On light microscopy, extramedullary plasmacytoma must be differentiated from a reactive plasmacytosis, plasma cell granuloma, poorly differentiated neoplasms, immunoblastic lymphoma, or extranodal marginal zone B-cell lymphoma with plasmacytic
differentiation. Plasmacytoid lymphoma has a mixture of lymphocytes and plasma cells. Immunoblastic lymphomas show a cytoplasmic IgM heavy chain and express pan B-cell surface antigen, such as CD19 and CD20 [2].

The diagnosis of plasmacytoma necessitates a detailed evaluation so as to exclude multiple myeloma [4]. Diagnosis of solitary EMP should be made only if studies for disseminated disease are negative. X-ray and/or magnetic resonance imaging of the spine, pelvis, femurs, and humerus, and bone marrow biopsy should also be within the normal range. There should be no signs of serum urine monoclonal protein, anaemia, hypercalcemia, or renal impairment [8]. A prospective study by Schirrmeister et al. has also assessed the accuracy of positron emission tomography (PET) scanning in staging patients with presumed solitary plasmacytoma [9].

There are no generally accepted guidelines for the treatment of patients with EMP and most recommendations remain as consensus of expert opinion [10]. Indeed, due to the rarity of this neoplasm, most studies are retrospective and no randomized trials are available.

There are several strategies which may be adopted, including surgery, radiotherapy, chemotherapy, and combinations of these, but, to date, radiotherapy is the standard treatment for EMP, as it is highly radiosensitive.

Alexiou et al. [11] reviewed 714 cases in literature published between 1905 and 1997: radiation therapy alone was used in 44.3% of cases and surgery alone in 21.9%, with combined therapy (radiation after surgery) in 26.9% of the cases. Liebross et al. [12] reported 19 cases, where all but one had been treated with radiotherapy alone.

The diagnostic criteria for EMP are based on the exclusion of a systemic plasma cell neoplasm (MM) and a thorough radiological investigation as aforementioned, with negative results and no signs of renal impairment and even if it has better prognosis than multiple myeloma, it should always be considered malignant or potentially malignant and treated as such. As plasmacytoma of the head and neck region has better prognosis than other sites, the treatment strategy is to be chosen with care, aiming at the preservation of laryngeal function whenever possible. As it is on the increase, we emphasise the need for EMP to be taken into consideration in the differential diagnosis of laryngeal neoplasms.

Moreover, at this point we may also ask ourselves if this increase is also due to dissemination of knowledge on this rare finding. Therefore, further prospective and randomized trials would help, not only to inform but also to clarify the question.

Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

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References

[1] N. C. Munshi and K. C. Anderson, “Plasma cell neoplasm,” in Cancer Principles and Practice of Oncology, V. T. DeVita Jr., S. Hellman, and S. A. Rosenberg, Eds., pp. 2155–2188, Williams & Wilkins, Philadelphia, Pa, USA, 7th edition, 2005.

[2] R. D. Bruning, “Bone marrow,” in Surgical Pathology, J. Rosai, Ed., pp. 2099–2105, Mosby-Elsevier, St. Louis, Mo, USA, 9th edition, 2004.

[3] J. C. Aster, “Diseases of white blood cells, lymph nodes, spleen and thymus,” in Robbins and Cotran Pathologic Basis of Disease, V. Kumar, A. K. Abbas, and N. Fausto, Eds., pp. 678–681, WB Saunders, Philadelphia, Pa, USA, 7th edition, 2004.

[4] R. C. Gromer and A. J. Duvall III, “Plasmacytoma of the head and neck,” Journal of Laryngology and Otology, vol. 87, no. 9, pp. 861–872, 1973.

[5] A. Gorenstein, H. B. Neel III, K. D. Devine, and L. H. Weiland, “Solitary extramedullary plasmacytoma of the larynx,” Archives of Otolaryngology, vol. 103, no. 3, pp. 159–161, 1977.

[6] Y. Rakover, M. Bennet, R. David et al., “Isolated extramedullary plasmacytoma of the true vocal fold,” The Journal of Laryngology & Otology, vol. 114, pp. 540–542, 2000.

[7] K. A. Maclellan and J. B. Schofield, “Haemopoietic neoplasms,” in Neoplasms of the Larynx, A. Ferlito, Ed., pp. 331–333, Churchill Livingstone, Edinburgh, Scotland, 1993.

[8] A. J. Maniglia and J. W. Xue, “Plasmacytoma of the larynx,” Laryngoscope, vol. 93, no. 6, pp. 741–744, 1983.

[9] H. Schirrmeister, A. K. Buck, L. Bergmann, S. N. Reske, and M. Bommer, “Positron emission tomography (PET) for staging of solitary plasmacytoma,” Cancer Biotherapy and Radiopharmaceuticals, vol. 18, no. 5, pp. 841–845, 2003.

[10] M. Hughes, R. Soutar, H. Luraft, R. Owen, and J. Bird, Guidelines on the Diagnosis and Management of Solitary Plasmacytoma of Bone, Extramedullary Plasmacytoma and Multiple Solitary Plasmacytomas: 2009 Update, UKMF Guidelines Working Group, 2009.

[11] C. Alexiou, R. J. Kau, H. Dietzfelbinger et al., “Extramedullary plasmacytoma: tumor occurrence and therapeutic concepts,” Cancer, vol. 85, no. 11, pp. 2305–2314, 1999.

[12] R. H. Liebross, C. S. Ha, J. D. Cox, D. Weber, K. Delasalle, and R. Alexanian, “Clinical course of solitary extramedullary plasmacytoma,” Radiotherapy & Oncology, vol. 52, no. 3, pp. 245–249, 1999.

[13] E. Medini, Y. Rao, and S. H. Levitt, “Solitary extramedullary plasmacytoma of the upper respiratory and digestive tracts,” Cancer, vol. 45, no. 11, pp. 2893–2896, 1980.

[14] J. Corwin and R. D. Lindberg, “Solitary plasmacytoma of bone vs. extramedullary plasmacytoma and their relationship to multiple myeloma,” Cancer, vol. 43, no. 3, pp. 1007–1013, 1979.

[15] R. Jyothirmayi, V. P. Gangadharam, M. K. Nair, and B. Rajan, “Radiotherapy in the treatment of solitary plasmacytoma,” British Journal of Radiology, vol. 70, pp. 511–516, 1997.

[16] M. Hughes, R. Soutar, H. Lucraft, R. Owen, and J. Bird, “Guidelines on the diagnosis and management of solitary plasmacytoma of bone, extra-medullary plasmacytoma and multiple solitary plasmacytomas: 2009 update,” http://www.bloodmed.com/contentimage/guidelines/3454.pdf.

[17] A. Gorenstein, H. B. Neel III, K. D. Devine, and L. H. Weiland, “Solitary extramedullary plasmacytoma of the larynx,” Archives of Otolaryngology, vol. 103, no. 3, pp. 159–161, 1977.