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Case Report

Solitary Extramedullary Plasmacytoma of the Trachea: A Case Report

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Abstract: The extramedullary plasmacytomas is a plasma cell tumour arising outside the bone marrow and constitutes around 4 % of all plasma cell neoplasms. The Extramedullary Plasmacytoma of the Trachea (EPT) is an extremely rare condition and presents as a tracheal expanding mass. The most common symptoms consist of coughing, dyspnoea, voice change, stridor and expiratory wheezing. The diagnosis is done by computed tomography, bronchoscopy and histological examination which demonstrates neoplastic monoclonal plasma cells expressing specific antigens. The treatment of the EPT remains still speculative, current options are radiotherapy or surgery alone and surgery followed by radiotherapy. The adjuvant chemotherapy is considered only in case of relapse or systemic spread. The prognosis is comforting: the complete remission occurs in 60 % of cases, the recurrence in 22 %, the progression to multiple myeloma in 16 %. We report the case of a 51-year-old female affected by EPT and treated with the removal of the first four tracheal rings and subsequent PAD chemotherapy (bortezomib, adriamycin and dexamethasone). Although the stage of the tumour was early, we decided to administer chemotherapy conceiving it as a preventive systemic treatment.

Keywords: Tracheal Tumours, Tracheal Resection, Extramedullary Plasmacytoma, Airway Obstruction

1. Introduction

The extramedullary plasmacytomas are rare plasma cell malignancies arising in soft tissues outside the bone marrow. They constitute around 4 % of all plasma cell neoplasms and in 80 % of cases occur in the upper aerodigestive tract. [1-5]

The trachea is a rare site of primary cancer onset, its tumours constitute around 1-2 % of all respiratory tract neoplasms and 0.1 % of all malignancies. Two-thirds of all primary tracheal neoplasms are squamous cell carcinomas and adenoid cystic carcinomas; the remaining third is a heterogeneous group composed of malignant, intermediate and benign lesions. [1-3]

The coexistence of these two situations, namely an Extramedullary Plasmacytoma of the Trachea (EPT), is an extremely rare condition and less than 20 cases are described in the Medical Literature until 2013. [1]

We report our anecdotal experience.

2. Case Report

A 51-year-old female presented to our Unit complaining of a goiter causing dyspnoea.

The neck ultrasound scan confirmed the goiter but did not show tracheal compression. The chest and neck computed tomography with 3D reconstruction found a subglottic oval lesion of 3 cm in size protruding into the tracheal lumen (Figures 1A and 1B). The bronchoscopy showed an endoluminal oval mass arising from the anterior wall of the trachea and occupying over 60% of the tracheal lumen (Figure 1C). No biopsy was done because of the potential risk of bleeding.

The patient underwent a total thyroidectomy, a removal of the first four tracheal rings and a direct anastomosis of the airway as described in the following paragraph.

The postoperative hospital stay lasted 10 days and was uneventful, at day 4 the patient started a liquid diet with a gradual recovery of a normal diet.

The definitive histological examination found a solitary plasmacytoma of the trachea without tumour presence at the margins of the extended resection and the patient was assigned to the Haematology Unit.

The multiple myeloma diagnosis was excluded by routine blood counts, serum protein electrophoresis, assessment of immunoglobulin, estimation of urine Bence Jones Protein, skeletal investigations and bone marrow biopsy.

The total body computed tomography excluded a systemic spread of disease and the patient underwent 4 cycles of PAD chemotherapy (bortezomib, adriamycin and dexamethasone).

She is recurrence free after 24 months of follow up.

3. Surgical Strategy

The patient signed a detailed informed consent for surgery and was prepared for doing a permanent tracheostomy if the extent of resection had requested. The surgery was divided into two parts.

The first part. It was performed an orotracheal intubation with a thin tube and under bronchoscopic guidance to prevent the traumatism and dissemination of tumour; the extremity of tube was placed beyond the mass. The first surgical time consisted of total thyroidectomy and excision of the tumour with the subglottic anterior wall of the trachea from where it arose (Figures 2A and 2B).

Since the impromptu histological examination found a tracheal plasmacytoma and tumour presence at the surgical margins, to obtain the oncological radicality, it was mandatory a more extended resection and it began the second part of intervention.

The second part. It was made a wide tracheotomy below the fourth tracheal ring (above the fifth), a tube for mechanical ventilation (a tracheostomy tube) was placed through it in the lower stump of trachea and concomitantly the orotracheal tube was retracted (Figure 3A). In this way, the surgeons were able to mobilize the proximal trachea and resect the first four rings.

After tracheal resection, it was taken off the tracheostomy tube and performed a new orotracheal intubation during which the surgeon manually pushed the tube in the distal trachea and subsequently, with the airway protected, an "end-to-end" anastomosis was done between the tracheal stump and the larynx with single stitches by absorbable material (Figure 3B). Both laryngeal nerves were identified and saved.

A paratracheal drain was positioned to monitor a possible bleeding or fistula and it was removed at postoperative day 8 without sequela.

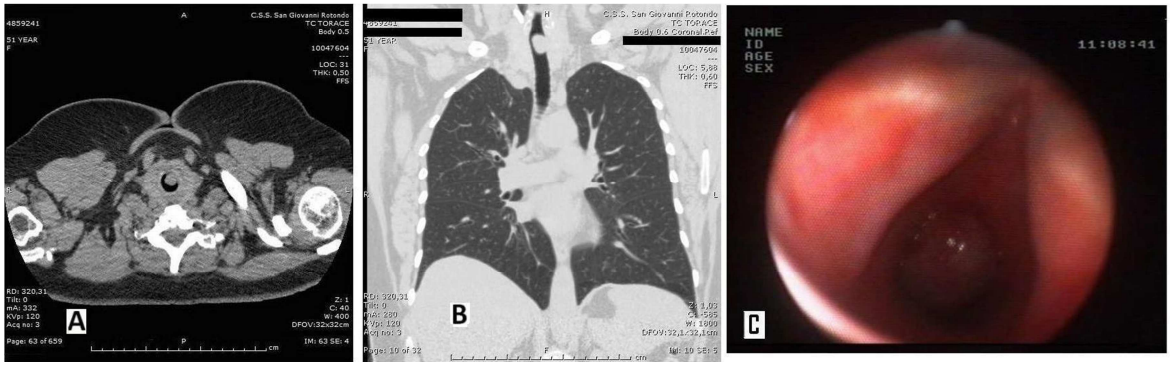

Figure 1. *A. Transversal sections of chest and neck computed tomography. B. Frontal section of chest and neck computed tomography. C. Endoscopic view of ETP.*

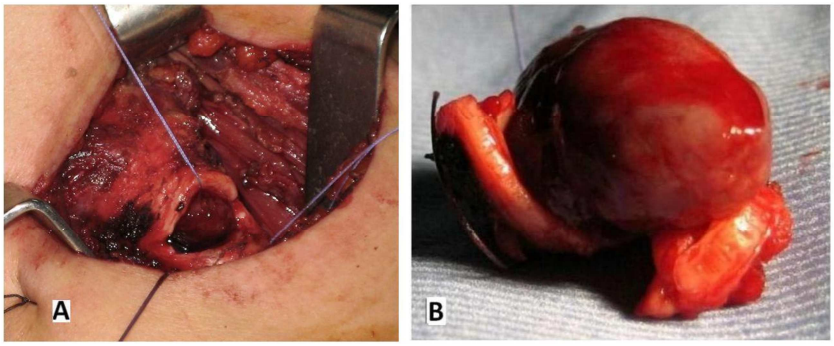

Figure 2. *A. Intraoperative view after tumour removal with the anterior wall of the trachea from where it arose (before extended tracheal resection). B. Macroscopic view of the surgical specimen.*

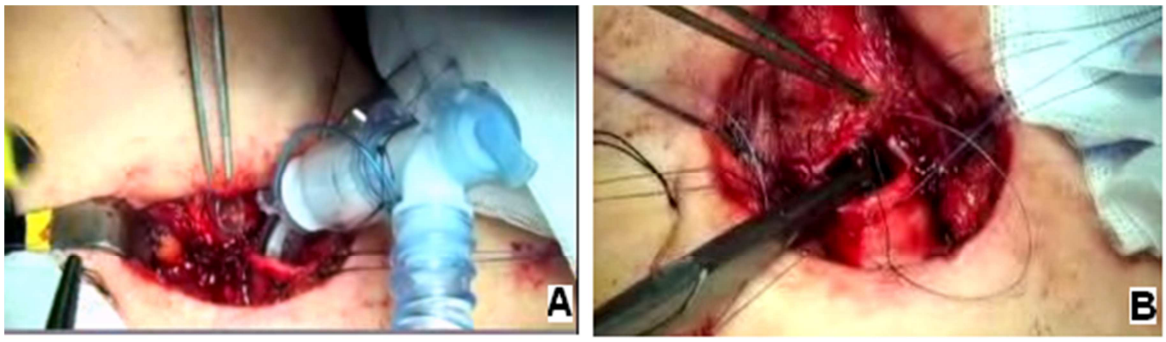

Figure 3. *A. Intraoperative view showing the wide tracheotomy below the fourth tracheal ring, the tube for mechanical ventilation placed in the lower stump of trachea and concomitantly the not working orotracheal tube retracted in the larynx. B. Intraoperative view showing the performing time of the anastomosis.*

4. Discussion

The extramedullary plasmacytoma of the trachea (EPT) presents as a tracheal expanding mass and becomes symptomatic when narrows over 75 % of the tracheal lumen. [2, 3]

The symptoms are owing to chronic airflow obstruction and consist of coughing, dyspnoea, haemoptysis, voice change, stridor and expiratory wheezing. They are frequently misdiagnosed as asthma or sleep apnoea syndrome. [2, 3, 6]

The computed tomography with contrast enhancement assesses the location of the tumour, the distance from the cricoid cartilage and to the carina, the extent of the narrowing within the lumen, the airway wall, the mediastinal structures and the possible systemic spread. The bronchoscopy shows an endoluminal view of the tumour and allows to perform a biopsy. [7]

The final diagnosis is obtained by the histological examination which demonstrates sheets of neoplastic monoclonal plasma cells expressing cytoplasmic immunoglobulin with light chain restriction and specific antigens. [1, 7, 8]

When an EPT is suspected, it is mandatory to exclude a multiple myeloma and some kind of lymphomas.

The multiple myeloma is excluded by routine blood counts, skeletal investigations, immunoglobulin assessment, serum protein electrophoresis, estimation of urine Bence Jones Protein and bone marrow biopsy.

The lymphomas with extensive plasmacytic differentiation that more frequently should be differentiated from a primary plasma cell dyscrasia (as EPT) are lymphoplasmacytic lymphoma, extranodal marginal zone lymphoma of mucosa associated lymphoid tissue (MALT lymphoma), follicular lymphoma, chronic lymphocytic lymphoma and diffuse large B cell lymphoma. In these cases the differentiation is obtained by cell surface antigens, specifically CD45 and CD19 are typical of lymphoma whereas CD56, CD38 and CD138 are typical of a primary plasma cell dyscrasia. [1, 7, 8]

The debate about the treatment of the extramedullary plasmacytomas and EPT remains still open in Medical Literature: radiotherapy alone, surgery alone or surgery followed by radiotherapy are the three current options in case of localised disease, instead, the adjuvant chemotherapy is considered only in case of relapse or systemic spread. [1, 2, 9]

In addition, by a model utilizing bone marrow flow cytometry and light-chain analysis, some authors demonstrated the presence of occult marrow disease in 68 % of their patients with solitary plasmacytoma of bone with no evidence of systemic spread. Since the occult marrow disease resulted to be predictive of progression, they claim that the adjuvant systemic therapy is warranted in these patients even if the tumour is localized. [10]

In our case, given the rarity of the EPT, the progression rate of 40% [1], the absence of guideline about its treatment (only 20 cases reported to date) and the young age of the patient (51 years), we preferred to approach it as relapsed or advanced disease although its stage was early.

In fact, we treated locally with surgery alone, achieved a radical excision by performing an extended tracheal resection, and administered PAD chemotherapy (bortezomib, adriamycin and dexamethasone) as prevention of recurrence or systemic spread.

We avoided radiotherapy because we obtained the oncological radicality, as demonstrated by the definitive histological examination, with extended tracheal resection.

The prognosis is comforting: around 60 % of patients with plasmacytoma of upper aerodigestive tract present complete remission, 22 % have recurrence and 16 % show progression to multiple myeloma. The 5-year survival rate is between 30 and 82%. [1, 2, 5]

5. Conclusion

To sum up, the EPT is a malignant neoplasm and may recur or disseminate as myeloma although its prognosis is better than that of other malignancies, therefore a close and long term follow up is mandatory.

About its treatment,

- given the empirical and pioneering context because of the absence of specific series studies in Medical Literature,
- given the demonstrated presence of occult marrow disease in a large number of patients with solitary plasmacytoma of bone with no evidence of systemic spread and its prediction of progression [10],

in our opinion, it could be wise to give a preventive systemic treatment even in case of localized disease.

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