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CASE REPORT Open Access

Plasmacytoma of the larynx: case report and literature review of laryngeal extra-medullary plasmacytoma

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Abstract

Background: Monoclonal plasma cell neoplasms are rare; laryngeal solitary plasmacytoma is very rare.

Case presentation: This is the case of a 46-year-old man who has been treated for chronic laryngitis for 8 months; he presented first with dysphonia, dyspnea, and dysphagia. The initial tests showed normal erythrocyte counts, elevated erythrocyte sedimentation rates, elevated CRP, and a thrombocytopenia at 40.000. The rest of the test panel was without incidence. Whole body MRI was performed which showed no additional lesion besides the laryngeal location. Bone marrow biopsy was normal. The patient underwent pan-endoscopy under general anesthesia. The biopsy result showed plasmacytoma. The patient had radiotherapy, after which the patient needed tracheotomy with a good outcome. This work presents a very rare case of laryngeal solitary plasmacytoma, with a literature review of differential diagnosis, as well as current trends in diagnosis, and therapy.

Conclusion: The essential take-home message is that however, this is a rare entity, it should be considered and treated as a malignant disease that could progress to diseases with worse diagnoses. Timely diagnosis and treatment can allow good outcomes and prevention of progression.

Keywords: Myeloid neoplasm, Larynx, Extramedullary plasmacytoma, Acute myeloid leukemia

Background

Monoclonal plasma cell neoplasm represents 1% of head and neck neoplasms; extra-medullary plasmacytoma are very rarely localized in the larynx, and only 164 cases have been reported in English, Chinese, and Indian literature [1, 2] (Fig. 1).

Case presentation

A 46-year-old male came to emergency consultation for dyspnea, dysphonia, and dysphagia. He has been having a similar episode of mild dyspnea and dysphonia in the last 8 months, for which chronic laryngitis was being diagnosed.

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Laryngeal fibroscopy showed a thickening of the mucosa in the ventricular bands of the vocal cords, which extended to the sub-glottic space. The patient was admitted for a tracheotomy to control airways as his respiratory distress worsened. The patient is in direct suspension laryngoscopy under general anesthesia. A biopsy was conducted in several localizations.

The patient underwent a cervicothoracic CT scan, which showed a thickening in laryngeal mucosa in epilaryngeal, glottic, and subglottic regions (Figs. 2 and 3). A comprehensive ear, nose, and throat exam, a blood test panel, and radiological assessment were performed.

The clinical exam did not find any lymph node involvement, and no other abnormalities were found.

The blood test panel showed an elevated erythrocyte sedimentation rate, an elevated C reactive protein, and a thrombocytopenia at 40.000. The rest of it was without abnormalities.

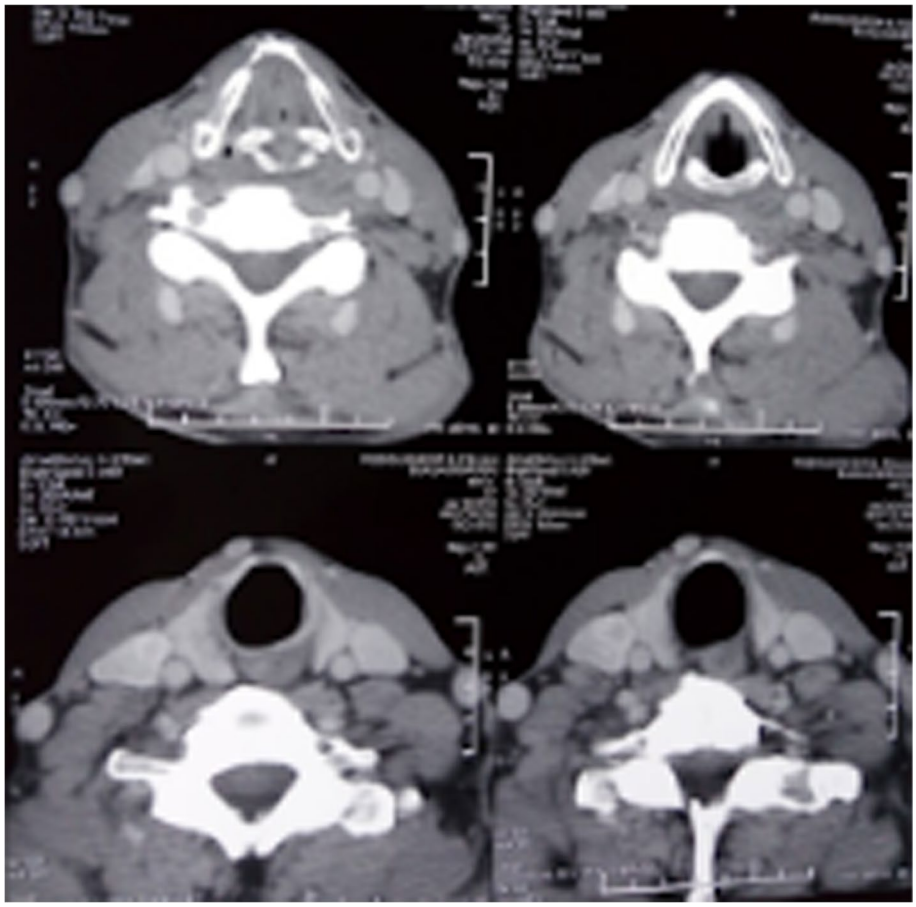

Fig. 1 Relationships between myelomatoses (Mochimatsu et al.)

Immuno-electrophoresis serum test was within normal range.

The radiological assessment was made of cervical, thoracic, and pelvic MRI, which showed no other lesions other than the laryngeal ones.

The patient was referred to radiotherapy, to a dose of 50 Gy.

Histopathology revealed plasma cells had eccentric nuclei and atypical cytology (prominent nucleoli, dispersed nuclear chromatin, and a high nuclear-cytoplasmic ratio). Immuno-histo-chemical exam of the specimen was positive for CD 138 and CD 38.

The diagnosis of solitary laryngeal extra-medullary plasmacytoma was established after the elimination of most plamacytoses.

The outcome was satisfying with a stabilization of lesions in the endoscopic follow-up.

The 3 years follow-up was satisfying.

Discussion

Relationships between myelomatoses regarding clinical evolution overlap [3] (Fig. 3). These tumors are histologically indistinguishable from plasma cell tumors arising from the bone marrow; they can be found anywhere in the body. The first case was discovered in 1905 by Schridle [4, 5].

Solitary plasmacytoma is an immuno-proliferative mono-clonal disease of B cells. They are composed of clones of a single cell [2].

While plasma cell myeloma (multiple myeloma) is the most common plasma cell neoplasm that includes the involvement of multiple localizations and bone marrow [2], solitary plasmacytoma is a singly localized mass. It could be in the bone marrow or in extra-medullary localizations. However, it could be the first manifestation of plasma cell plasmacytoma [2]. Plasmacytoma has a predilection to head and neck, and 80% are located herein

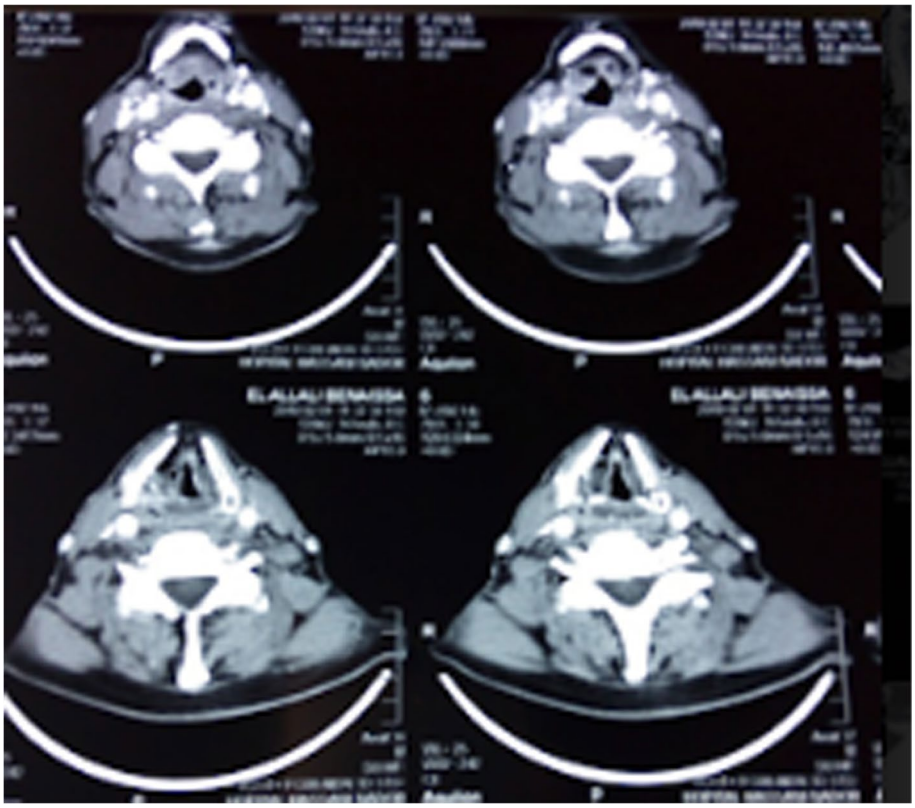

Fig. 2 Axial CT scan sections: thickening of laryngeal mucosa at the level of the thyroid cartilage and cricoid cartilage, with complete obstruction

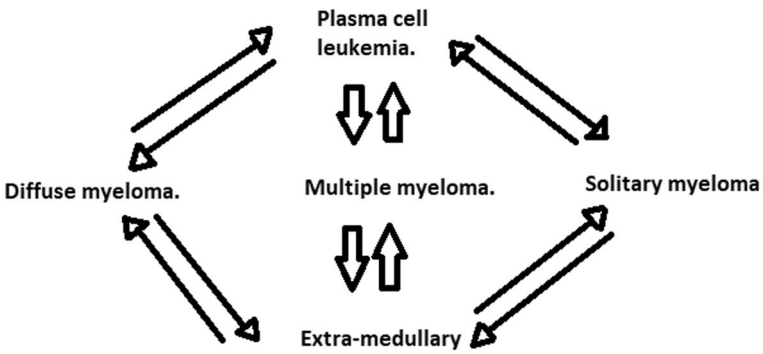

Fig. 3 Axial CT scan sections: thickening of laryngeal mucosa at the level of the epilaryngeal space with restriction of the laryngeal lumen

[6]. The oto-rhino-laryngological exam should be as complete as possible [7, 8].

The most common age is between 50 and 70 years old, although it had been reported to appear since the second decade of life, with a predilection to males [1, 2, 5, 6].

Since diagnosis is established after the elimination of other systemic myelomatoses. A detailed evaluation is necessary. Imaging studies and pan-endoscopy is important for the elimination of these diagnoses [6]. X-ray and MRI of the spine, pelvis, humerus, and femur should eliminate the existence of other lesions in order to eliminate systemic conditions [5, 6]. A whole body scintigraphy can bear the same results as a bone marrow biopsy [5].

Diagnosis is only established after the absence of other lesions is confirmed [6] (Table 1).

Extramedullary plasmacytoma is generally submucosal. The macroscopic aspect is polypoid sessile masse

Table 1 Most important tests in eliminating a differential diagnosis of extramedullary plasmacytoma

Biological tests:	Radiological tests: (Pinto et al.)
- Blood count. - Urinary tests for Bence Jones proteins. - Protein electrophoresis. - Bone marrow biopsy.	- X-ray spine, pelvis, humerus, and femur. - MRI. - And/or total body scintigraphy. - And/or PET scan.

with an epithelial surface and diffuse thickening of yellow grayish color to dark brown [4].

Symptoms of the extramedullary plasmacytoma are mainly dysphonia, dysphagia, cough, dyspnea, stridor, cough, sore throat, hemoptysis, and laryngeal foreign body sensation [7, 8].

This tumor could be unilateral or bilateral; the sites of predilection are epiglottis, supra-glottis, hemi-larynx, false vocal cords, and arythenoid folds, and multiple sites could be found [8].

Cervical node involvement has been cited in the literature [5, 7, 8].

Granuloma is generally a polymorphous infiltrate of plasma cells with lymphocytes and macrophages [2].

A microscopic aspect shows diffuse monomorphic infiltrate in a fine reticular stroma with plasma cells with abundant cytoplasm [5]. Distinguishing medullary plastocytoma from undifferentiated epithelial tumors relies on immuno-histo-chemical profiling [5]. The first step is a difference between monoclonal and polyclonal tumors [5].

Immuno-histo-chemical profile showed CD138, CD 38, and CD 56 [1, 8, 9]. Sometimes, immunoglobulin gene rearrangement analysis could contribute to the diagnosis; it is not yet the gold standard [8].

Different malignant and benign tumors are to be eliminated before establishing the diagnosis (Table 2).

The most important objective of treatment is the preservation of laryngeal function as long as possible [6].

Radiotherapy is the treatment of choice (50 Gy). Surgical modalities have been recommended in the most recent years [5, 8]. Adjuvant chemotherapy may be used to prevent the progression of the disease [5, 8].

Table 2 Differential diagnosis of extramedullary

	Diseases:
Benign differential diagnosis (C.B Pratibha) (Pino et al.).	- Granuloma - Monoclonal gammopathy. - Waldenstrom's macroglobulinemia
Malignant differential diagnosis (C.B Pratibha) (Pino et al.).	- Plasma cell myeloma. - Plasma cell leukemia. - Plasmacytoid lymphoma. - Large cell lymphoma of immunoblastic type (C.B Pratibha) (Pino et al.).

Radiotherapy; surgery; surgery and radiotherapy; radiotherapy and chemotherapy; surgery and chemotherapy; and surgery, radiotherapy, and chemotherapy are the possibilities of treatment [8].

Risk factors for progression to multiple myeloma are large size, M-protein and high chain, amyloid deposit, and high nuclear grade [1]. There is a higher risk of progression to multiple myeloma in the first 2 years [8].

Prognosis of extra-medullary plasmacytoma progressing to multiple myeloma is better than cases of primary multiple myeloma [10]. However, solitary extra-medullary should be considered malignant or potentially malignant and treated as such [6]. Single localization is associated with a better prognosis [8].

Follow-up is recommended for long periods of time, cases of progression to multiple myeloma after 15 years [1, 5, 8].

Five years survival is of 18% in multiple myeloma and of 68% in solitary myelocytoma [5].

Conclusion

Solitary plasmacytoma of the larynx is rare, and accurate diagnosis is established after elimination of hematological neoplasms and neoplasm of the larynx [6].

This is the case of a patient with extramedullary laryngeal solitary plasmacytoma, with timely diagnosis and management which allowed good outcomes.

The essential take-home message is that however, this is a rare entity, it should be considered and treated as a malignant disease that could progress to diseases with worse diagnoses. Timely diagnosis and treatment can allow good outcomes and prevention of progression.

Abbreviations

CT scan: Computed tomography; MRI: Magnetic resonance imaging; PET scan: Positron emission tomography scan; CRP: C-reactive protein.

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Authors' contributions

KC: conceived and designed the analysis, collected the data, performed the analysis and wrote the paper. NB: contributed data or analysis tools. MNE: contributed data or analysis tools. All authors have read and agreed to its content.

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Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication
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Competing interests
The authors declare they have no competing interests, no financial or non-financial conflict of interest.

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