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RESEARCH ARTICLE

LARYNGEAL PLASMACYTOMA IN KAHLER'S DISEASE: A CASE REPORT

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Abstract

Multiple myeloma or Kahler's disease is a malignant hematological disorder due to a monoclonal plasma cell proliferation primarily invading the hematopoietic bone marrow. Extra-medullar involvement occurs in 18% of patients with multiple myeloma. Laryngeal Localization remains rare and has been described in a few cases after a literature review.We report the case of a 44-year-old woman who presented an inspiratory dyspnea caused by a laryngeal tumor. The pathological study and bone marrow examination confirmed the laryngeal localization of multiple myeloma. Treatment consisted of chemotherapy with good clinical outcomes over a 2-year follow-up period.

Introduction:-

Plasmacytoma is a solitary malignant tumor of monoclonal plasma cells, developing either within the bone (solitary bone plasmacytoma) or in soft tissues (extramedullary plasmacytoma or EMP). EMP occurs in 18% of patients with multiple myeloma (1). The most common sites of EMP are the head and neck region, upper respiratory tract, gastrointestinal tract, and central nervous system. Extramedullary plasmacytomas account for less than 1% of all malignant tumors of the head and neck (1). Multiple myeloma (MM) typically presents with anemia, bone pain, and renal insufficiency (9) (10). Laryngeal Involvement in multiple myeloma is rare, with only a few cases reported. We present the case of a woman with extramedullary laryngeal plasmacytoma leading to discover a multiple myeloma after further investigations.

Case Report:-

A 59-year-old female patient, followed for high blood pleasure under treatment, has presented a progressive inspiratory dyspnea and dysphonia over the past 9 months. The course of her symptoms was marked by a subacute deterioration, requiring an urgent tracheostomy.After the patient's condition was stabilized, a nasofibroscopy was done, which identified notable oedema of the ventricular bands, obstructing the view of glottic area. As a result, we initiated adrenalin nebulization and systemic corticosteroid therapy.

Further evaluation included a cervical and thoracic enhanced CT scan, with coronal and sagittal reconstructions. It reveals a tumor at the laryngotracheal junction extending 18mm in height and 23mm in width. This process was found to be in contact with the arytenoid cartilages and the cricoid arch, showing signs of lysis, although it remained

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distant from the glottic area. Furthermore, a small tumor extension was observed in the right lateral wall of thoracic trachea, measuring approximately 10mm. (Figure 1).

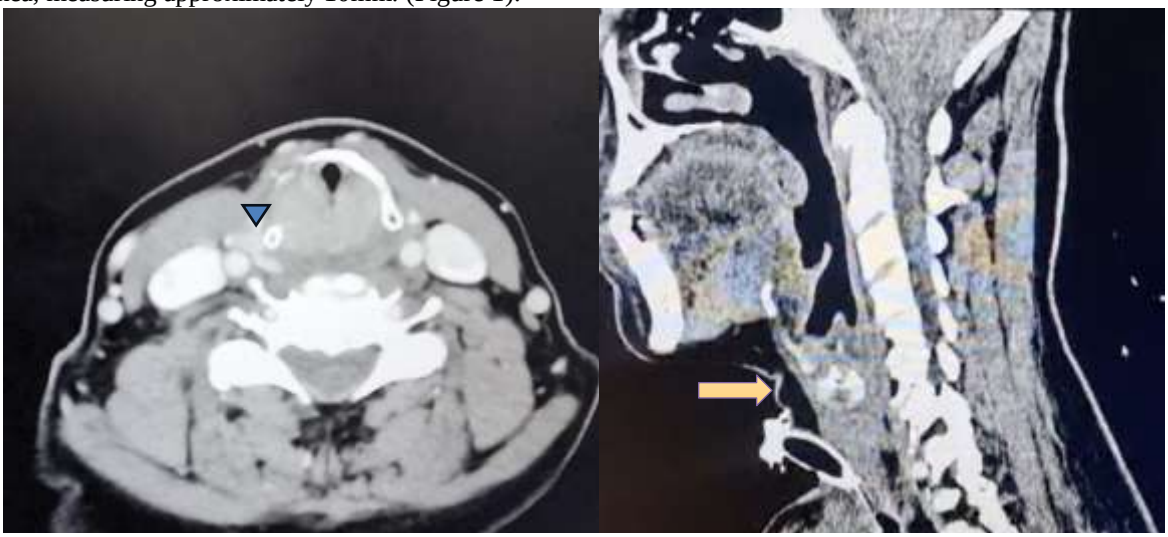

Figure 1:- Sagittal and axial CT scan showing a tumor centered on the subglottic region (arrow) with lysis of the cricoid cartilage.

During direct laryngoscopy, no lesions were detected at the glottic level.However, the subglottic lumen was narrowed by a tissue process occupying nearly 80% of the airway. Multiple biopsies were taken.

Histologically, the laryngeal mucosa showed diffuse proliferative tumor growth (Figure 2), comprising small-sized cells with round nuclei, dense chromatin, and scant basophilic cytoplasm (Figure 3). Immunohistochemical analysis was performed, showing intense and diffuse expression of CD138 and kappa light chains by the tumor cells (Figure 4 and 5). Additionally, there was negativity for lambda light chains (Figure 6). The histological and immunohistochemical findings were consistent with a plasmacytoma.

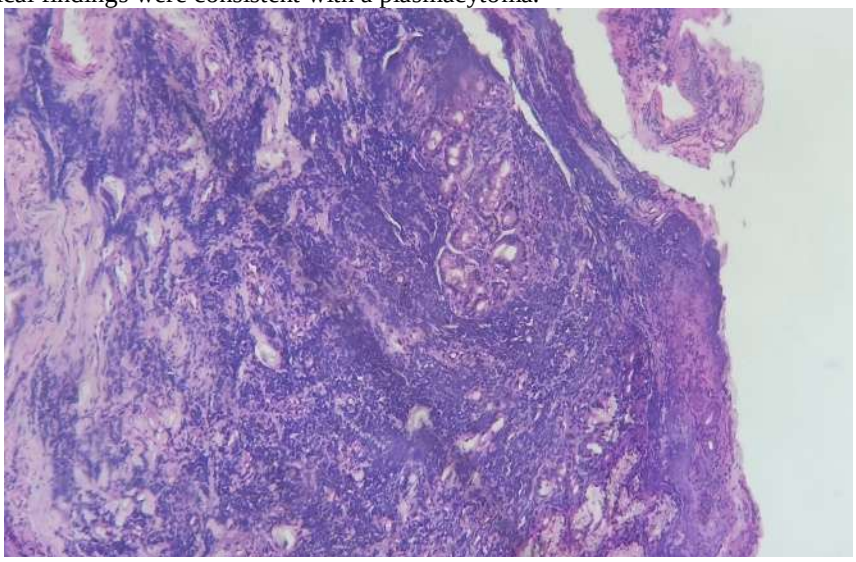

Figure2:- Tumoral proliferation arranged in sheets (HESx100).

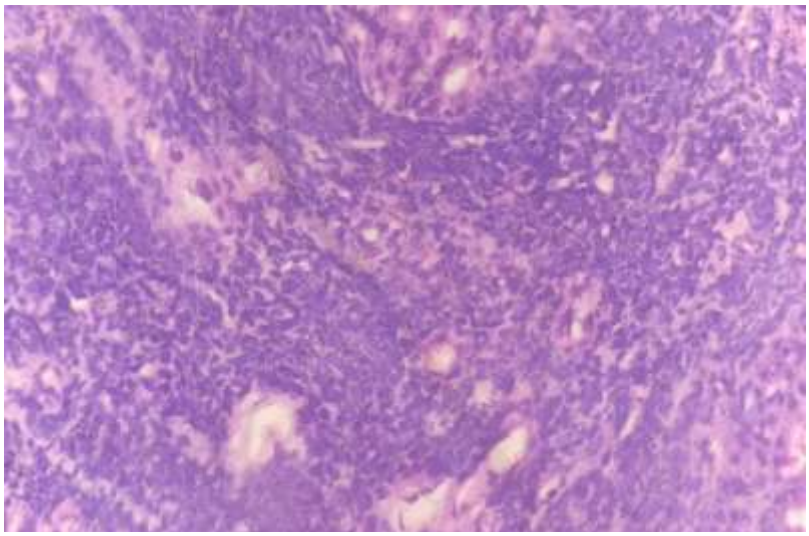

Figure 3:- The tumor cells are small to medium-sized andshow crush artifacts (HESx200).

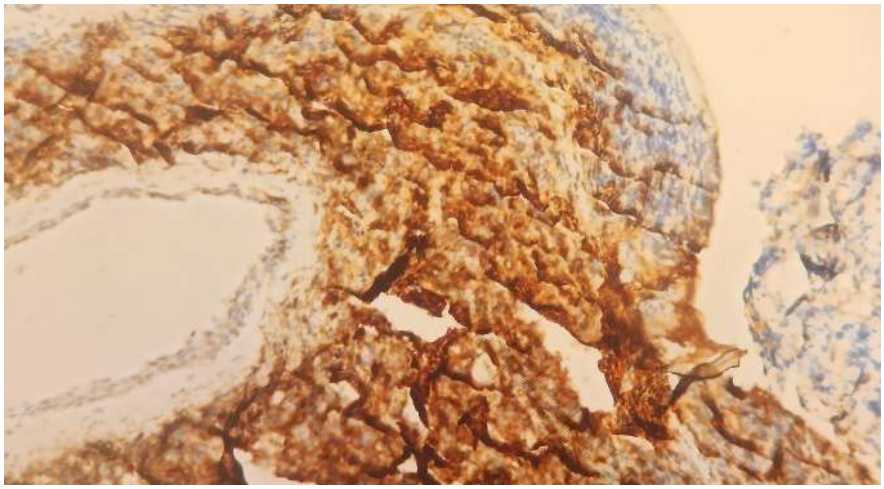

Figure 4:- Immunohistochemical study showing expression of Kappa light chains.

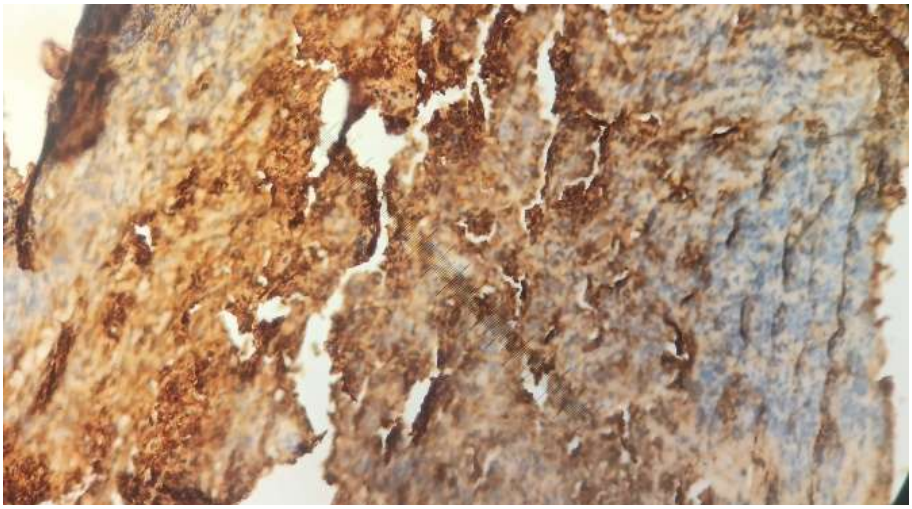

Figure 5:- Immunohistochemical study showing diffuse expression of CD138.

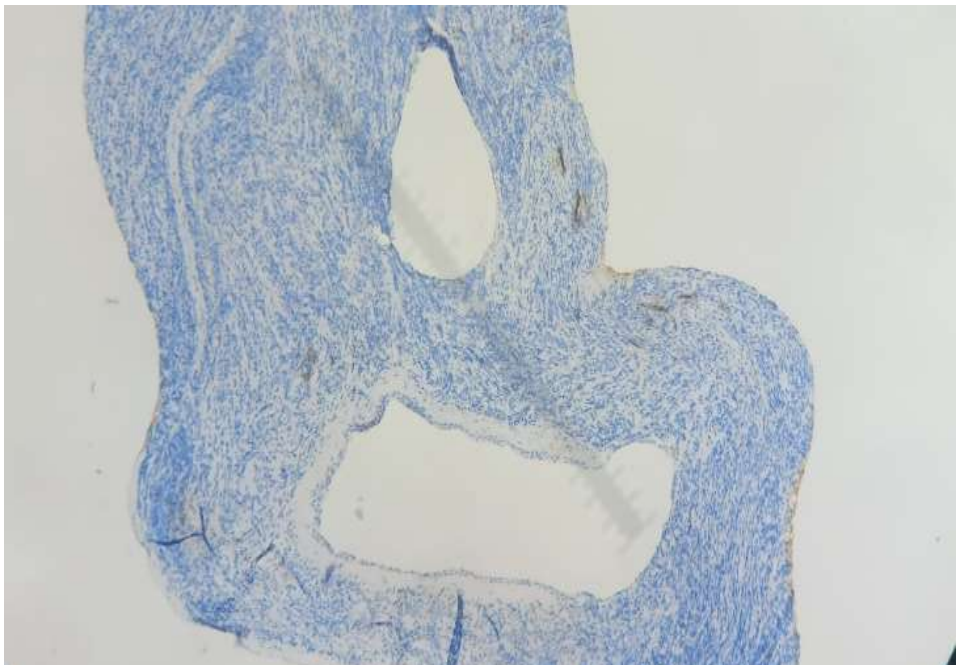

Figure 6:- Immunohistochemical study showing negativity for lambda light chains.

The evaluation was concluded in collaboration with the hematology-oncology department. It involved a bone marrow biopsy, a PET scan, and a phosphocalcic assessment with serum protein immunoelectrophoresis. This was done to distinguish between a solitary plasmacytoma and an extramedullary laryngeal manifestation of multiple myeloma.

The complete blood count (CBC) was normal. A blood smear revealed anisopoikilocytosis, but no circulating blasts were noted.The bone marrow biopsy showed a heterogeneous normocellular marrow infiltrated by a plasma cell population estimated at 14%, with a blast count estimated at 3%.Serum protein immunoelectrophoresis (SPEP) revealed monoclonal spike of immunoglobulin G with kappa light chain.A PET scan with 18FDG showed pathological hypermetabolism at thelaryngeal tumor, without any other suspicious hypermetabolic fixation elsewhere.

Based on cytological and histological data, the diagnosis of laryngeal plasmacytoma inmultiplemyelomadisease was established.

The decision was made to initiate chemotherapy consisting of CDT: Cyclophosphamide + Dexamethasone + Thalidomide. The patient showed a good therapeutic response with regression in size of the laryngeal mass on follow-up scans. The patient's general condition was good, and the tracheostomy was removed after 12 months.

Discussion:-

Plasmacytoma is a single, distinct tumor composed of monoclonal cells(1). These neoplastic plasma cells are either found within the bone as isolated cells (bone plasmacytoma) or in soft tissues outside the bone marrow (extramedullary plasmacytoma or EMP) (1).

Plasmacytomas are uncommon in the head and neck region, with nearly 80% of extramedullary plasmacytomas affecting the paranasal sinuses, pharynx, nasal cavity, and oral mucosa. Only about 10% occurs in the larynx(2). Extramedullary laryngeal plasmacytomarepresents a small proportion, ranging from 0.04% to 0.45% of laryngeal tumors (2).

EMP has a higher prevalence in men, with a male-to-female ratio of 3:1, and typically occurs in individuals aged 50 to 70 years(3).

Symptoms depend on the tumor's location and its impact on laryngeal structures, including dysphonia, dyspnea, dysphagia, and hemoptysis(4). The most frequently affected sites in the larynx, in order of frequency, are the epiglottis, ventricles, vocal cords, aryepiglottic folds, arytenoids, and subglottis(5). These lesions can present with various endoscopic appearances, ranging from small polyps with narrow bases to larger sessile tumor lesions (5).

EMP develops in approximately 18% of individuals with multiple myeloma(1). The diagnostic distinction between EMP and multiple myeloma relies on the absence of systemic symptoms in EMP cases, despite histological similarity between these two entities(6). It's important to note that 20% to 30% of extramedullary plasmacytomas progress to multiple myeloma(2) (7).

Differential histological diagnosis can pose challenges, as plasma cell infiltrates can also be observed in various benign conditions such as chronic inflammatory diseases, inflammatory polyps, and amyloid deposits(8).

Establishing a diagnosis of multiple myeloma requires at least 10% clonal plasma cell infiltration in the bone marrow, along with the presence of M protein in serum or urine (except in cases of non-secretory myeloma), and symptoms resulting from lesions in other target organs of myeloma (e.g., hypercalcemia, renal insufficiency, anemia, or bone lesions) (9) (10).

Immunohistochemistry plays a crucial role in evaluating the presence of the CD138 marker in B lymphocytes and plasma cells, whether in the bone marrow or extramedullary sites(8).

Computed tomography (CT) with or without positron emission tomography (PET) and magnetic resonance imaging can be used to further evaluate bone and soft tissue lesions, revealing additional clinically undetectable lesions or involvement of cervical lymph nodes(11).

Management of plasmacytomas can involve radiotherapy or chemotherapy. Radiotherapy is the preferred treatment modality for EMP, as plasma cell neoplasms are highly radiosensitive(7). In contrast, multiple myeloma is considered a systemic disease, requiring chemotherapy and possibly bone marrow transplantation(7) (9).

Conclusion:-

Multiple myeloma of the larynx is extremely rare. However, when patient presents with submucosal disease in the larynx, it is important to consider the possibility of rare histological diagnoses, including plasmacytoma. Otherwise, a delay in diagnosis can be fatal for the patient.

References:-

1. Narayanan G, Rajan V, TR P, Soman L V. Multiple Myeloma Presenting as Respiratory Stridor. Iranian Journal of Blood and Cancer 2017; 9 (2) :64-66
2. Liebross RH, Ha CS, Cox JD, Weber D, Delasalle K, Alexanian R. Clinical course of solitary extramedullary plasmacytoma. Radiother Oncol. 1999 Sep; 52(3):245-9. Doi: 10.1016/s0167-8140(99)00114-0. PMID: 10580871.
3. Gromer RC, Duvall AJ 3rd. Plasmacytoma of the head and neck. J Laryngol Otol. 1973 Sep; 87(9):861-72. Doi: 10.1017/s0022215100077732. PMID: 4728300.
4. Peng J, Zeng Q. [Extramedullary plasmacytoma of the larynx：a case report]. Lin Chuang Er Bi Yan Hou Tou Jing Wai Ke Za Zhi. 2022 Jul; 36(7):553-556. Chinese. Doi: 10.13201/j.issn.2096-7993.2022.07.015. PMID: 35822386; PMCID: PMC10128390.
5. Pratibha CB, Sreenivas V, Babu MK, Rout P, Nayar RC. Plasmacytoma of larynx--a case report. J Voice. 2009 Nov; 23(6):735-8. Doi: 10.1016/j.jvoice.2008.03.009. Epub 2008 Jul 10. PMID: 18619786.
6. Kumar SK, Callander NS, Alsina M, Atanackovic D, Biermann JS, Chandler JC, Costello C, Faiman M, Fung HC, Gasparetto C, Godby K, Hofmeister C, Holmberg L, Holstein S, Huff CA, Kassim A, Liedtke M, Martin T, Omel J, Raje N, Reu FJ, Singhal S, Somlo G, Stockerl-Goldstein K, Treon SP, Weber D, Yahalom J, Shead DA, Kumar R. Multiple Myeloma, Version 3.2017, NCCN Clinical Practice Guidelines in Oncology. J Natl ComprCancNetw. 2017 Feb; 15(2):230-269. Doi: 10.6004/jnccn.2017.0023. PMID: 28188192.
7. Nofsinger YC, Mirza N, Rowan PT, Lanza D, Weinstein G. Head and neck manifestations of plasma cell neoplasms. Laryngoscope. 1997 Jun; 107(6):741-6. Doi: 10.1097/00005537-199706000-00007. PMID: 9185729.

8. Allegra E, Marino N, Modica D, Emmanuele C, Saita V. Primary laryngeal localization of multiple myeloma: A case report. Mol Clin Oncol. 2017 Feb; 6(2):154-156. Doi: 10.3892/mco.2017.1122. Epub 2017 Jan 2. PMID: 28357083; PMCID: PMC5351711
9. McKenna RW, Kyle RA, Kuehl WM, Grogan TM, Harris NL, Coupland RW. Plasma cell neoplasms. In: Swerdlow SH, Campo E, Harris NL, et al., editors. WHO classification of tumours of haematopoietic and lymphoid tissues. 4th ed. Lyon, France: IARC; 2008. pp. 200–213
10. International Myeloma Working Group. Criteria for the classification of monoclonal gammopathies, multiple myeloma and related disorders: a report of the International Myeloma Working Group. Br J Haematol. 2003 Jun; 121(5):749-57. PMID: 12780789.
11. Grobman AB, Vivero RJ, Campuzano-Zuluaga G, Ganjei-Azar P, Rosow DE. Laryngeal involvement of multiple myeloma. Case Rep Oncol Med. 2012; 2012:257814. Doi: 10.1155/2012/257814. Epub 2012 Nov 18. PMID: 23213583; PMCID: PMC3505634.