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Case Report

A Rare Case of Disseminated Extra Nodal B Cell Marginal Zone Lymphoma of Larynx in an Adolescent Girl

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Abstract

Reporting a case of 14 year old girl with disseminated extra nodal B cell marginal zone lymphoma of larynx. She presented with history of hoarseness of voice and stridor on lying down of 2 months duration. Underwent endoscopic resection. PET CT showed dissemination to lung parenchyma. The clinicopathological characteristics and rational treatment of these lymphomas are still unclear and there is lack of evidence based treatment protocols. Radiotherapy is the first option for limited-stage primary laryngeal MALT lymphoma because of excellent treatment outcome. And chemotherapy is usually reserved for disseminated and recurrent disease. We treated with 6 cycles of RCHOP chemotherapy (Rituximab, Cyclophosphamide, Vincristine, Doxorubicin and prednisolone). Presently child is in complete remission and under regular follow up.

Key words: *Lymphoma, PET CT, chemotherapy, radiotherapy*

Case Report

In Oct 2019, 14 year old girl, presented with history of frequent episodes of Light headedness with husky voice and stridor on lying down of 2 months duration. Seen by ENT specialist, found to have supraglottic laryngeal mass.

MRI neck was suggestive of large fairly well marginated soft tissue mass seen involving left pyriform fossa,ary epiglotic fold with involvement of left free margin of epiglottis. The medial edge of the ipsilateral vocal fold was not well made out. A large Smooth surfaced soft Mass is seen extending into the Hypopharynx with moderate to severe luminal narrowing. Lesion measured 2.7 X 2.8 X 4.5 cm (About 16 ml).Large lymph nodes are seen in bilateral level 1 B, left level 2 and left level 5 groups(MRI neck images Appendix 1).

She underwent endoscopic excision with prior tracheostomy with Coblation Wand (cold radiofrequency ablation) assistance under general anaesthesia. Coblation Facilitates resection with less intra operative Bleed. The patient was decannulated around the 10th post-operative day uneventfully.

Histopatholgy (Images in Appendix 3) showed stratified squamous epithelium and smooth muscle bundles with a diffuse cellular infiltrate in the sub epithelial tissue. The cells were reported as small and dyscohesive. The cytoplasm was scanty and nuclei round with condensed chromatin. Plasmacytoid lymphocytes were seen.

Small calibre blood vessels were present. There is no significant mitosis or necrosis. IHC showed diffuse and strong positivity for CD 20 and bcl2. CD 3 and CD 5 stains in T cell pattern. Negative for CD 10, bcl6 and cyclin D1.KI 67 around 30%. Suggestive of extra nodal marginal zone B cell lymphoma possibly MALT type. CT guided biopsy of the lung nodule was done and histopathology was suggestive of non-Hodgkin's lymphoma. PET CT (Images in Appendix 2) showed heterogeneously enhancing FDG avid lesion in left pyriform fossa measuring 27 X 32 X 37 mm with max SUV 5.6 with local extension. Multiple enlarged cervical lymph nodes seen with max SUV 3. Metabolically active lesions noted in right upper lobe and left lower lobe of lung. Suggestive of stage IV disease. There were no clinical features suggestive of autoimmune disorders or infective etiology.

This is a very rare case of extra nodal marginal zone B cell lymphoma with stage IV disease in an adolescent girl. The case was discussed in the unit. Parents refused for bone marrow aspiration and biopsy. She was given IT Methotrexate along with first lumbar puncture. . CSF was negative for malignant cells. She was started on RCHOP (Rituximab, Cyclophosphamide, Vincristine, Doxorubicin and prednisolone) chemotherapy. PET CT was repeated after 3 cycles of RCHOP and was suggestive of complete metabolic response (Deauville 1). She was given total 6 cycles of RCHOP chemotherapy. PET CT done after completion of chemotherapy on Mar 2020 showed complete response (Deauville 1).

Post-surgery she had good relief from stridor but the voice quality was suboptimal due to a small anterior glottis scar band. Plan to release scar at a later date under the guidance of video laryngoscopy. Presently she is undergoing voice therapy and planned for review once in 3 months.

Discussion

The non-Hodgkin lymphoma subtype of marginal zone lymphoma represents a group of lymphomas that arises from post-germinal center marginal zone B cells and share a similar immunophenotype: positive for B cell markers CD19, CD20, and CD22, and negative for CD5, CD10, and usually CD23.

Extranodal marginal zone lymphoma (EMZL) arises in a number of epithelial tissues, including the stomach, salivary gland, lung, small bowel, thyroid, ocular adnexa, skin, and elsewhere. While it has a tendency to remain localized to the tissue of origin for long periods of time, it is a clonal B cell neoplasm that frequently recurs locally and has potential for systematic spread and transformation to an aggressive B cell lymphoma.

They occur in older patients with a median age at presentation ranging from 55 to 65 years, with females affected more commonly than males 1,2,3 .The incidence of EMZL is more than fourfold greater than primary NMZL1. The stomach is the most frequent site of involvement, followed by salivary glands, orbit, and lung

4, 5. An association with autoimmune disease, including Sjogren's syndrome and more rarely systemic lupus erythematous, has been shown in up to 30% of the patients3 .EMZL tend to present as localized disease and have a good prognosis.

Laryngeal MALT lymphomas are extremely rare in children. Literature search did not reveal many case reports of Laryngeal EMZL with involvement lung parenchyma. Our case was unique in this aspect. Laryngeal lymphoma presents clinically with symptoms such as hoarseness, dyspnoea, a foreign body sensation in the throat, or stridor. Uncommonly, it may present catastrophically with acute airway obstruction requiring immediate surgical intervention. Systemic symptomatology is unusual, since laryngeal lymphomas tend to remain localized for prolonged periods, though more aggressive forms tend to spread earlier6 .Interestingly, these tumors usually spread to other mucosal sites such as bowel, lung, and orbit rather than nodal sites 7, 8. MALT lymphomas present with a series of recurrent genomic lesions, including chromosomal translocations and unbalanced aberrations. In our case we didn't do translocation studies because of financial constraints.

18F-FDGPET/CT has been used for staging laryngeal MALT lymphomas and has provided a basis for both altering the therapeutic strategy and also evaluating the response to radiotherapy 9. However, despite being proven valuable in detecting sites of disease in some types of lymphomas, 18F-FDG PET/CT is not yet the standard method of staging and cannot substitute bone marrow examination. In our case we couldn't perform it because of parent's refusal.

These lymphomas are highly radiosensitive. For localized disease, radiotherapy is the most appropriate treatment for larynx preservation. Chemotherapy is reserved for recurrent or disseminated disease 10.There is no well-defined chemotherapy regimen for disseminated disease. Hence the case was discussed in the unit and RCHOP chemotherapy was chosen. The response was good in our case. Plan to follow up once in 3 months for recurrence and adverse effects of chemotherapy.

Conclusion

In conclusion, laryngeal MALT lymphomas represent rare clinical entities without any clear risk factors. Due to the small number of cases reported, there is no definite consensus regarding best management of laryngeal EMZL. Nevertheless, the follow-up period in our case was relatively short, and long-term follow-up is necessary to determine whether these lymphomas recur later in life.

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