CASE REPORT

COMPLETE RESPONSE OF PRIMARY LYMPHOMA OF LARYNX, A RARE MALIGNANCY TO CHEMOTHERAPY AND RADIOTHERAPY WITH LITERATURE REVIEW

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PJR October - December 2015; 25(4): 168-171

ABSTRACT

OBJECTIVE: Primary (Non Hodgkin lymphoma) NHL of larynx is extremely rare. Only a few cases have been reported in literature so far. Here such case is presented to share our experience with review of literature. **METHODS:** It was a male patient presented in emergency with gradually progressing shortness of breath (SOB) and had direct laryngoscopy revealing mass in larynx and underwent tracheostomy. He was diagnosed as diffuse large B cell (DLBC) NHL on laryngeal biopsy and immunohistochemistry and referred to us in MINAR cancer hospital, Multan for further management. **RESULTS:** The patient was treated with standard multidrug regime chemotherapy followed by external radiotherapy. Complete response was found on investigations at three weeks after completion treatment and was disease free at one year of follow up. **CONCLUSION:** Although Primary carcinoma of larynx is extremely rare but once diagnosis is made can be treated effectively.

Key words: Chemotherapy, Larynx, Non Hodgkin lymphoma, Radiotherapy.

Introduction

Non-Hodgkin lymphoma (NHL) is a predominantly lymph node tumor. It presents in extra nodal sites in about one third of cases.[1] Although the lymph nodes are the main targets, any organ may be involved. 60% of extra nodal NHL occurs in the head and neck.[2]

Primary involvement of the larynx is extremely rare in this lymph node malignancy, with only a few cases reported in the medical literature so far.[1-4] Laryngeal NHL contributes less than 1% of all laryngeal malignancies[3] and raises diagnostic challenges.

Case History

A male patient presented in emergency with gradually progressing shortness of breath (SOB), had tracheostomy and direct laryngoscopy revealing mass in larynx. On clinical examination no lymph node or abdominal viscera was palpable. Other routine investigations including complete blood examination (CBC), routine urine examination (RUE), fasting blood sugar (FBS), renal parameters (RPMs) and liver function tests (LFTs) were with in normal limits. There was no mediastinal widening on chest x-ray (CXR). No visceromegaly or lymph adenopathy was found on abdominal ultrasonography (USG). CT scan of neck revealed growth in larynx (Fig. 1,2). Biopsy of mass was taken which was reported as non-Hodgkin lymphoma on histopathology. CD 20 (a tumor marker) was positive on imunohistochemistry (IHC). His lactate dehydrogenase (LDH) was raised that was 1088 U/dl (N is >480 U/dl). No other significant present or past medical problem related to this disea-se was found on history and clinical examination. Disease was isolated laryngeal lymphoma so staged as I according to An Arbor classification.

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Submitted 16 June 2015, Accepted 22 June 2015

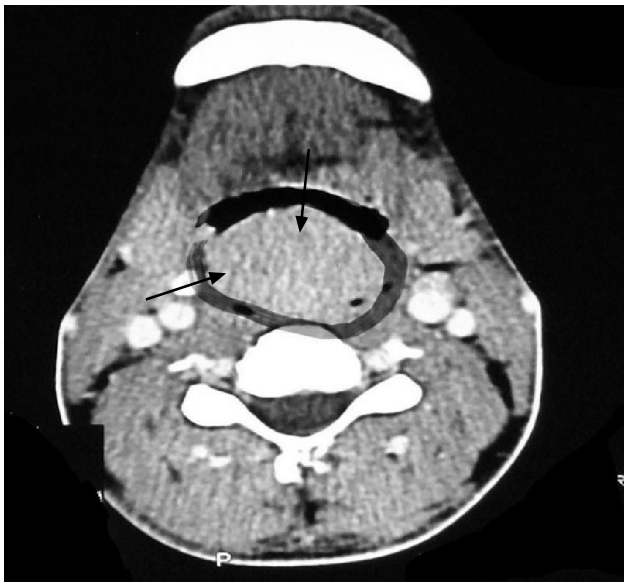

Figure 1: Axial image of CT neck (Black arrows showing large laryngeal mass)

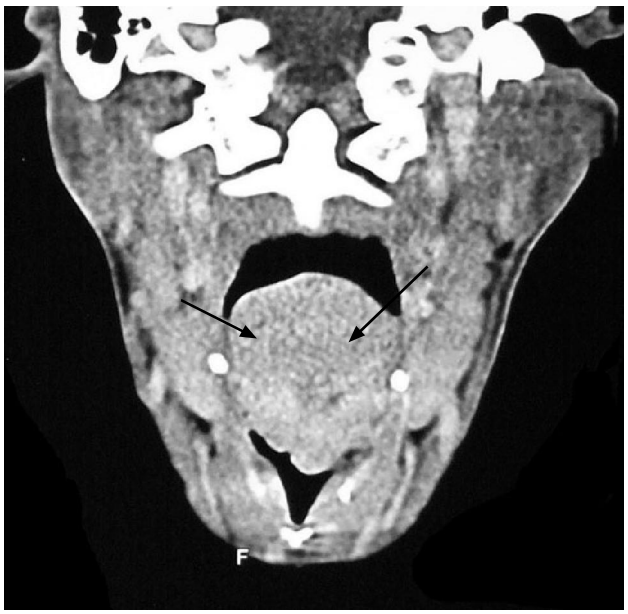

Figure 2: Coronal section of CT neck (Black arrows showing large laryngeal mass)

Patient was treated with multidrug regime chemotherapy including cyclophosphamide, doxorubicin, vincristine and prednisolone (CHOP). Doses were calculated according to body surface area. Six cycles were given at three weeks interval. Chemotherapy was followed by external radiotherapy to neck with standard fractationation that is 200 cGy/Fx x 22 Fxs(total dose of 4400 cGy). Complete response was found on investigations at three weeks after completion of treatment. The patient is now being regularly followed at 3 to 4 months interval with complete clinical examination, routine investigations, LDH levels and abdominal USG. The patient was disease free on CT scan after completion of one year of his follow up (Fig. 3).

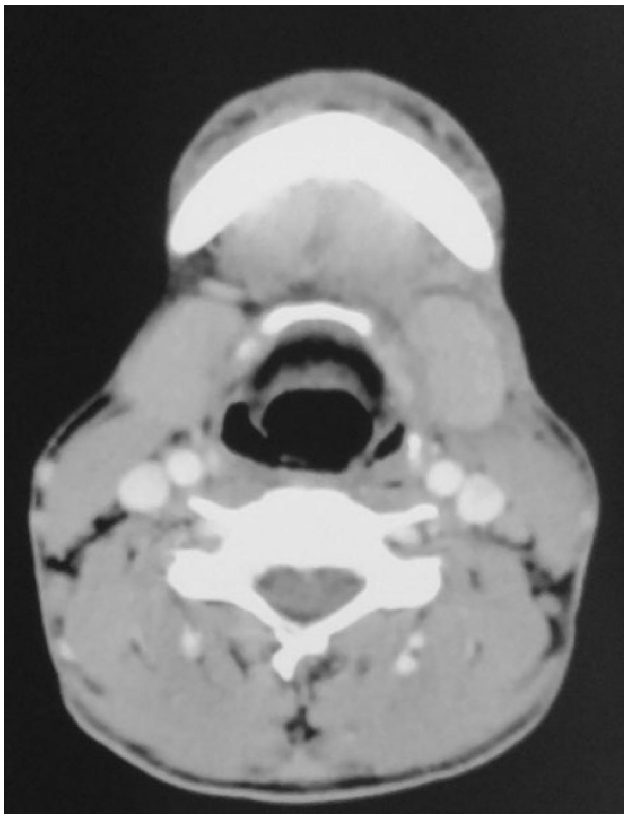

Figure 3: Post Treatment follow up CT scan showing complete resolution of laryngeal mass.

Discussion

Although the lymph nodes are the main targets of NHL but any organ may be involved. The incidence of non-Hodgkin's lymphoma originating in extra nodal sites ranges from 10-35% overall.[4] Extra nodal lymphomas originate not only at sites that normally contain lymphoid tissues such as small intestine and parotid gland but also in non-lymphoid tissue such as thyroid and stomach. Non-Hodgkin lymphoma (NHL) of the head and neck contributes 60% of total extra nodal NHLs[2] and only 5% of all head-and neck malignancies[5] but NHL of larynx in head and neck region is very rare entity. Fewer than 100 cases have been reported in the literature.[6] Laryngeal NHLs con-

tribute less than 1% of all laryngeal malignancies.[5] and raise diagnostic challenges. Prognosis and pattern of progression of extra nodal NHL of head and neck are similar to those of NHL at other sites (spread to the bone marrow and to nodes on both sides of the diaphragm).[3]

The symptoms of extra nodal NHLs are site specific, as opposed to tumor type-specific. The symptoms of laryngeal NHL are related to obstruction of the larynx and consist chiefly of gradually worsening dyspnea and moderate dysphagia. Dysphonia is very rare.

FNAC can give a clear diagnosis of lymphoma but an adequate biopsy should be performed to ensure the accurate histological grading of the lymphoma.[7] Immunohistochemistry determines whether the tumor is composed of B cells or T cells.

The accurate staging of extra nodal NHL is important for effective planning of the treatment. Staging is determined by a detailed history, clinical examination and radiological investigations. It is further recommended that all cases of NHL should undergo a complete evaluation,[8,9] including a complete hemogram, liver and renal functions, serum 2-microglobulions, chest X-ray, bone marrow biopsy, gallium scan and cerebrospinal fluid analysis as well as CT scans of the abdomen, pelvis and bones. As a majority of our patients are low-income, initially we undertake routine investigations, CXR and ultrasonography of the abdomen. Then, if required, other expensive investigations are performed in suspicious cases.

The treatment of NHL is a controversial issue.[8,10] It con-sists of chemotherapy, radiotherapy, or surgery. Sometimes, in cases of limited extra nodal NHL only surgery without chemotherapy or radiotherapy is used.[8] However, the main role of surgery is diagnostic only and it is rarely used for a cure. The treatment of low-grade, non metastasized, symptomatic NHL rests on radiation therapy (30- 45 Gys with a classic schedule, according to the site of the tumor).[3,11,12] A lasting complete remission is achieved in 50 to 90% of patients with localized stage I or II tumors. Survival after 10-15 years is 50 to 60%.[11] Chemotherapy is the treatment of choice for intermediate- and high-grade NHLs.[8,11] The addition of monoclonal antibody (rituximab) in the CHOP regimen (R-CHOP) has demonstrated promising results and is now frequently used for treatment of lymphomas. A total of 3-4 cycles of this regimen followed by radiotherapy are quite effective in the treatment of stage I and non-bulky stage II NHL; however, bulky stage II-IV tumors need 6-8 cycles of this chemotherapy. In our set up belonging to developing country rituximab being expensive drug is not used in routine. So we give standard regime of CHOP as in this case followed by external radiotherapy.

Prognosis is calculated based on age, LDH levels, performance status, and the number of extra nodal sites involved. The prognosis declines with an increase in the patient's age, LDH levels, histological grade, or stage of the NHL.[8] The case we presented here was diagnosed in stage I with isolated laryngeal lymphoma and had complete response with combination of chemotherapy followed by radiotherapy.

Concisely Laryngeal NHL is an exceedingly rare tumor but has considerable good prognosis after simple and well tolerated course of chemotherapy and radiotherapy. Presenting symptoms are very non specific. Therefore it should be considered routinely in differential diagnosis of laryngeal mass and adequate deep biopsy must be collected.

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