Upper Aero-Digestive Tract Lymphomas Presenting as Polypoidal/Pedunculated Lesions: Case Report and Review of Literature

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Abstract

Aim To describe a case of diffuse large B cell lymphoma (DLBCL) presenting as a pedunculated/polypoidal mass in upper aero-digestive tract and to review pertinent literature.

Methods Using Pubmed advanced search, case reports and literature were reviewed for this condition.

Results A 16 year old female presented with history of shortness of breath when lying down, voice change, progressing odynophagia and feeling of throat lump. Direct visualization showed flower like pedunculated mass, attached to the left lateral oropharyngeal wall. Excision of

this mass showed DLBCL and no disease anywhere. She received four cycles of Rituximab + CHOP and is disease-free for 4 years. Only 21 cases of pedunculated upper aero-digestive tract lymphomas have been reported in literature. Median age 53 years (16–90 years), males 80 %, females 20 %, localized disease (65 %). Common sites were endobronchial polypoid lesion (7 cases), nasal polyps (7), nasopharyngeal polyps (2), nose and paranasal sinus (1), laryngeal polyp (2) and one each as gum, esophagus and oropharyngeal (current case) polyp. Pathology showed aggressive lymphoma in 94 %.

Conclusion DLBCL as an extranodal pedunculated lesion in the oropharynx is very rare. Careful pathological evaluation is essential for these lesions for proper diagnosis.

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Introduction

Lymphoid malignancies represent approximately 5 % of all malignant neoplasms of the head and neck. It may involve both nodal and extranodal sites. The head and neck region is the second most frequent extranodal lymphomas site after the gastrointestinal tract [1].

Diffuse large B cell lymphoma (DLBCL), a Non-Hodgkin lymphoma (NHL), presenting as an extranodal pedunculated/polypoidal mass in the oropharyngeal/nasopharyngeal or trachea is very rare. There are limited case reports in adults and only two in children/adolescents. We are reporting a case of oropharyngeal pedunculated DLBCL in an adolescent.

Fig. 1 Axial (a), Sagittal Reconstruction (b) and Coronal Reconstruction (c, d), showing the isodense minimally enhancing pedunculated mass in the region of left hypopharynx attached to the left

lateral wall partially encroaching on the lumen. Corresponding post operative axial (e) and Coronal reconstruction (f) of the same area showing complete excision with no residual mass lesion

Material and Method

Case Report

Patient is a 16 years old single Saudi female with no past medical history. She had bilateral tonsillectomy at the age of 4 years. She presented with history of voice change, progressing odynophagia, feeling of throat lump and foreign body sensation in the throat that is moving with the respiration. She also complained shortness of breath when lying down. He denied fever, night sweat or weight loss.

Her brother was diagnosed at the age of 15 with classical Hodgkin lymphoma and was treated here (alive with no disease). Two of her father's sisters had breast cancer, diagnosed at the ages of 63 and 50, both were treated at our institution and died of metastatic disease. Her first cousin (from father side) had composite lymphoma (features of both NHL and Hodgkin lymphoma) diagnosed at the age of 15, treated at an outside institution and then had recurrence disease for which he underwent high dose chemotherapy and transplant (Hodgkin lymphoma at relapse) at our institution. He is alive and disease free. Her grandfather also had history of lymphoma diagnosed at an old age (pathological type is not know, diagnosed and treated outside) and died of disease.

Her history indicates a possible familial predisposition to lymphoid malignancies. There is no history of consanguinity in her father and mother.

She was seen in a local hospital with tentative diagnosis of lingual cyst. She was sent to our institution for further evaluation. Her performance status was 1. Her physical exam was unremarkable. Direct visualization showed flower like pedunculated mass, hanging with a 2 mm pedicle attached to the side of the lower pole of the tonsillar fossa/left lateral oropharyngeal wall, which was the site of previous tonsillectomy. Computed tomographic examination (Fig. 1) of head and neck showed small, oval-shaped, globular lesion noted within the oropharyngeal space between the uvula and terminal part of the soft palate superiorly, the epiglottis and the left aryepiglottic fold inferiorly, the posterior part of the left side of the tongue anteriorly and the left posterior oropharyngeal soft tissues posterolaterally. It elicits isodense texture to the soft tissues. It was approximately $17 \times 19 \times 27$ mm along its maximum anteroposterior, transverse and craniocaudal dimensions respectively.

She had complete excision of this mass under general anesthesia. Intra operative laryngoscopy showed normal laryngeal and the hypopharyngeal appearances. A small

Fig. 2 (a) Sheets of neoplastic cells in the subepithelial tissue. H & E stain; original magnification: $\times 100$. (b) High power magnification showing the lymphoid tumor cells. H & E stain; original

magnification: $\times 400$. (c) The lymphoid tumor cells staining positively with CD20 immunohistochemical marker. (d) Positivity of the vast majority of tumor cells for the Ki-67

sub-centimeter neck lymph node was identified and fine needle aspiration was negative for malignancy.

Pathologic examination of the excised left pharyngeal mass showed a mucosa-lined tissue, measuring $25 \times 20 \times 15$ mm. Microscopic examination demonstrated sheets of large neoplastic cells in the subepithelial tissue (Fig. 2a, b). The surgical margin (base of resection) was positive (in case of lymphoma diagnosis, we generally do not comment on that). The specimen was received in fragments so we cannot comment on the status of stalk of the tumor was positive or negative.

Immunohistochemical study revealed that these cells are positive for CD20, CD10, and MUM 1; BCL-6 showed patchy positivity, and MIB-1 (Ki-67) proliferative index was high (90 %) (Fig. 2c, d). Most of the cells were negative for BCL-2. CD3 showed reactive T-cells. CD30 and TdT were negative and testing for Epstein-Barr virus by in situ hybridization method was negative. Cytogenetic testing for Myc translocation was attempted on the paraffin embedded slides but was technically unsuccessful. Flow cytometry analysis revealed monoclonal B-cells, expressing CD10, and with Kappa light chain restriction. The final diagnosis was DLBCL.

Her final stage was stage IAE (Ann Arbor/Cotswolds modification staging system). She was started on Rituximab + cyclophosphamide, adriamycin, vincristine and prednisone

(R-CHOP) every 3 weeks. After developing severe ileus due to vincristine that required hospitalization, vincristine was omitted from the remaining three cycles. She is disease-free for 4 years.

Discussion

Non-Hodgkin lymphoma represents for approximately 7 % of cancers in children younger than 20 years [2]. DLBCL is a mature B-cell neoplasm that represents 10–20 % of pediatric NHL [2, 3].

Diffuse large B cell lymphoma presenting as an extranodal polypoidal/pedunculated/polyps in nasopharynx/oropharynx or tracheo-bronchial area is very rare. There are limited case reports in the literature [4–23].

Using PubMed advanced search, we used search term lymphoma (in humans/all languages) and combined it with polypoidal (8 reports), pedunculated (9 reports) and polyp (484 reports). All relevant abstracts and full text if available were reviewed to identify the clinical presentation, management and outcome of lymphoid malignancies presented as polypoidal/pedunculated/polyps in nasopharynx/oropharynx or tracheo-bronchial area. Cross references were also reviewed if needed. We identified 21 cases as shown in Table 1. Median age at diagnosis was 53 years

Table 1 Cases reported in the literature

No.	Author	Year/ Reference	Gender/age in yrs	Location/presentation	Extent of disease	Treatment/comments	Pathology
1.	Chorianopoulos D	2010 / [4]	Male/51	Nasal polyps	Systemic	CHOP → progressed and died	NK/T
2.	Yoruk O	2009 / [5]	Female/32	Nasal polyps	Local	CHOP → alive Pregnant at diagnosis	Burkitts lymphoma
3.	Kaira K	2008 / [6]	Male/63	Endobronchial polyp	Local	R-CHOP x3 → XRT. Alive	NHL – no details
4.	Chen SC	2008 / [7]	Male/17	Endobronchial polypoid mass	Systemic	CHOP → alive	ALCL
5.	Tzankov A	2005 / [8]	Male/63	Gingival polyp	Local	NA	Plasmablastic NHL
6.	Kandoğan T	2004 / [9]	Male/36	Nose and paranasal sinuses polypoid	Local	NA	NK/T
7.	Kiani B	2003 / [10]	Male/44	Endobronchial polypoid mass	Regional	ABVD → XRT → alive	Hodgkins lymphoma
8.	Gordin A	2003 / [11]	Male/61	Nasopharyngeal and occluding left external ear polyp	Local	DVIP chemo → sepsis and died	DLBCL + CLL
9.	Yaqoob N	2002 / [12]	Male/50	Nasal polyp	Local	NA	DLBCL + schwannoma
10.	Loo CK	2002 / [13]	Male/55	Nasal polypoid lesion	Local	Chemo → XRT Relapsed → Chemo → XRT → alive	NK/T
11.	Kim KI	1999 / [14]	Male/23	Endobronchial polypoid mass	Regional	ABVD → alive	Hodgkins lymphoma
12.	Kim DH	1998 / [15]	Male/28	Endobronchial polypoid mass	Regional	XRT → CHOP → alive	ALCL
13.	Miyata Y	1997 / [16]	Female/58	Endobronchial polypoid mass	Local	Chemo → alive	DLBCL
14.	Dasgupta A	1997 / [17]	2 cases, no other information	Nasal polyps	NA	NA	Pathological review of nasal polyps
15.	Kawaida M	1996 / [18]	Male/86	Laryngeal pedunculated mass	Local	Chemo → alive	DLBCL
16.	Melato M	1994 / [19]	Male/18	Esophageal polyp	Local	NA	Hodgkins lymphoma
17.	Zrunek M	1993 / [20]	Male/66	Nasopharyngeal + gastrointestinal polypoid	Systemic	CHOP → alive	NHL
18.	Bickerton RC	1988 / [21]	Male/79	Soft palate + Nasal polyps.	Local	XRT → alive	Lymphoplasmacytic lymphoma
19.	Ho AD	1985 / [22]	Female/74	Endotracheal polypoid mass	Relapsed	Chemo → alive	High grade lymphoma
20.	Noon M	1979 / [23]	Male/90	Laryngeal polyp	Local	XRT → alive	LBCL
21.	Current case	2012	Female/16	Oropharyngeal pedunculated mass	Local	Resection → R-CHOPx4, alive, 4 years	DLBCL

CHOP cytotaxan, vincristine, adriamycin, prednisone, *NK/T* natural killer/T cell lymphoma, *R-CHOP* Rituximab + CHOP, *ABVD* adriamycin, bleomycin, vinblastin, dacarbazine, *XRT* radiation therapy, *DVIP* dexamethasone, etoposide, ifosfamide, cisplatin, *CLL* chronic lymphocytic leukemia, *ALCL* anaplastic large cell lymphoma, *DLBCL* diffuse large B cell lymphoma, *NA* not available

(16–90 years), males were 16 (80 %) and females were 4 (20 %). Most commonly reported location and presentation was endobronchial polypoid lesion in 7 cases, nasal polyps in 7, nasopharyngeal polyps in 2 (one with gastrointestinal polyposis and one with ear involvement), nose and paranasal sinus in 1, laryngeal polyp in 2 and one each as gum, esophagus and oropharyngeal (current case) polyp. Most patients had localized disease 13/20 (65 %), 3/20 had regional involvement and 4 had systemic involvement by lymphoma. Pathological details are available for 18 patients and 17 (94 %) of them had aggressive lymphoma (6 with DLBCL, 2 with anaplastic large cell lymphoma, 2 NHL with no details, 3 with NK/T cell lymphoma, 2 high grade, 1 low grade lymphoma, 1 plasmablastic and 3 Hodgkin lymphoma). Fourteen of these patients received chemotherapy and 4 patients also had radiation therapy. Two patients received radiation therapy as the sole management. Details of treatment are not available for six patients. Sixteen patients had survival status available; 14/16 were reported alive in the case report.

Despite the fact that these tumors, in most cases were limited, almost all of them were aggressive lymphoid malignancies except one lymphoplasmacytic lymphoma (a low grade lymphoma) presenting as pedunculated mass in upper aerodigestive track. Because of the location of these tumors, they were causing symptoms in most cases that most likely required proper evaluation and early diagnosis in most cases. Most cases were not suspected to be malignant and clinical impression was benign polyp. Some cases required multiple biopsies/resections as the symptoms were not relieved/recurred after initial resection.

Lymphoid malignancies presenting as an extranodal pedunculated/polypoidal mass in the oropharyngeal/nasopharyngeal or trachea is very rare. Clinical presentation varied but usual clinical impression was benign etiology. Except one, all cases of lymphomas in this region were aggressive lymphomas as reported. Careful pathological evaluation is essential for these pedunculated lesions for proper diagnosis. Despite short follow-up of most cases, majority is alive and indicating longer survival in these patients.

Conflict of interest None.

Ethical standards Approved by Institutional Research Advisory Counsel.

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