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Case Report

Burkitt lymphoma masquerading as nasopharyngeal angiofibroma

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ABSTRACT

Primary nasopharyngeal (NP) lymphoma is a very rare tumor. Burkitt lymphoma is a subtype of non-Hodgkin's B cell lymphoma. It is rare in the Waldeyer's ring (3-5%). It is a highly aggressive, highly chemo sensitive lymphoma seen predominantly in childhood. Presenting a unique case of an 11 year old, male child presented to us with the chief complaints of bilateral nasal obstruction since 1 month, 6-7 episodes of nasal bleed since 1 week. Radiologically, a large soft tissue density mass lesion was seen in the region of nasopharynx, extending into the para/retro pharyngeal space. Excisional biopsy of the mass lesion histopathologic evaluation was suggestive of small round blue cell tumor, later confirmed to be Burkitt lymphoma after a bone marrow biopsy. Nasopharyngeal NHL should be kept in mind as a differential diagnosis, apart from inflammatory adenoid hypertrophy and benign tumors such as JNA while evaluating a nasopharyngeal mass in children. Nasopharyngeal NHL are highly chemo sensitive, therefore, an early diagnosis can aid timely commencement of chemotherapy and prevent progression and complications of the disease.

Keywords: Burkitt, Lymphoma, Child, Angiofibroma, Adenoid

INTRODUCTION

Lymphoma is the most common non-epithelial malignancy of the head and neck region. Lymphoma is classified into two types: Hodgkins and non-Hodgkins.[1] Of these, NP lymphoma is a very rare tumor. It has been reported that less than 10% of NHL cases in Western countries involved the WR, designated as WR-NHL while 10-18% of NHL cases were WR-NHL in Asian countries.[2,3] Among WR-NHL, about 35-37% of cases were at the nasopharyngeal subsite, and there was no difference of incidence between Western and Asian countries.

Burkitt lymphoma is a subtype of non-Hodgkin's B cell lymphoma seen most commonly in childhood, and is two times more common in males than females. It is rare in the Waldeyers ring (3-5%). However, it is important to recognize it as an oncologic emergency, as it is a highly aggressive lymphoma characterised primarily by activation of c-myc oncogene, which also renders it highly chemosensitive.[4]

Burkitt lymphoma has three clinical variants: Endemic, sporadic and immunodeficiency related. These types are similar in morphology, immunophenotype and genetic features. The annual incidence has been estimated at 40-50 per million children younger than 18 years, in endemic areas. Mass per abdomen is the most common presentation, followed by the head and neck region.[5]

Here, we present a case of an 11-year-old immunocompetent male child with nasopharyngeal mass, later diagnosed as NP Burkitt lymphoma.

CASE REPORT

A 11-year-old, male child presented to us with the chief complaints of bilateral nasal obstruction since 1 month, 6-7 episodes of nasal bleed since 1 week and 8-10 episodes of blood stained vomitus since 4 days. On

general examination the child was vitally stable, on examination of the oral cavity, a mass was seen posterior to the soft palate, pushing the soft palate anteriorly, mass was seen to be bleeding spontaneously. The child was admitted and started on IV antibiotics and IV fluids while the mass was worked up.

Imaging showed a large soft tissue density mass lesion in the superior and inferior wall of the nasopharynx, extending into the para/retro pharyngeal space (Figure 1). And was reported to be suggestive of nasopharyngeal angiofibroma or adenoid hypertrophy.

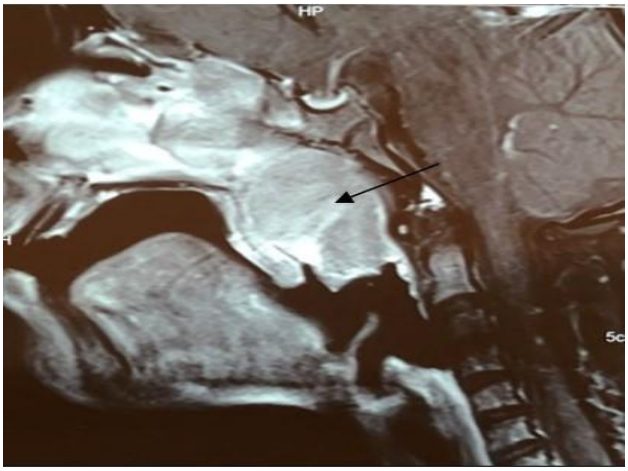

Figure 1: MRI-nasopharynx of a heterogeneously enhancing mass lesion (black arrow) in the posterosuperior wall of the nasopharynx.

Following which, a cerebral angiogram (ICA and ECA) (Figure 2) was done in order to rule out/ preoperatively embolise a nasopharyngeal angiofibroma. However, the angiogram showed no significant contrast blush in the region of the nasopharynx and super-selective catherization of internal maxillary artery also showed no arterial feeders supplying the mass lesion and hence, nasopharyngeal angiofibroma was ruled out.

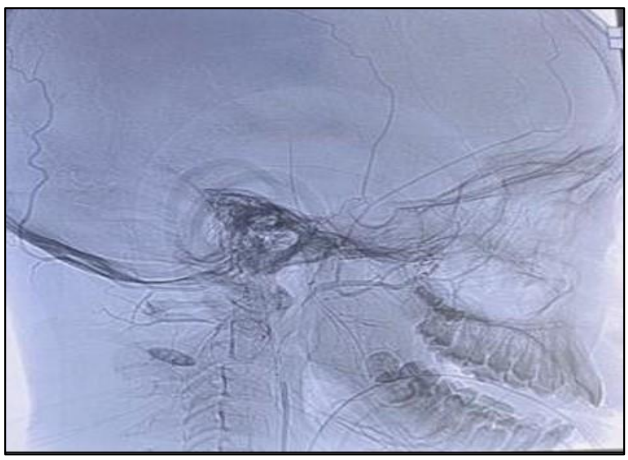

Figure 2: Cerebral angiogram of absence of contrast blush in the region of nasopharyngeal mass.

Since the child had continued episodes of haematemesis and difficulty breathing when lying down, we proceeded with a microdebrider assisted excisional biopsy of the mass lesion. Intraoperatively, the nasopharyngeal mass was seen to be obstructing both choanae and a soft tissue mass was also seen in the left nasal cavity (Figure 3 and 4), below the posterior end of middle turbinate which was also sent for HPR.

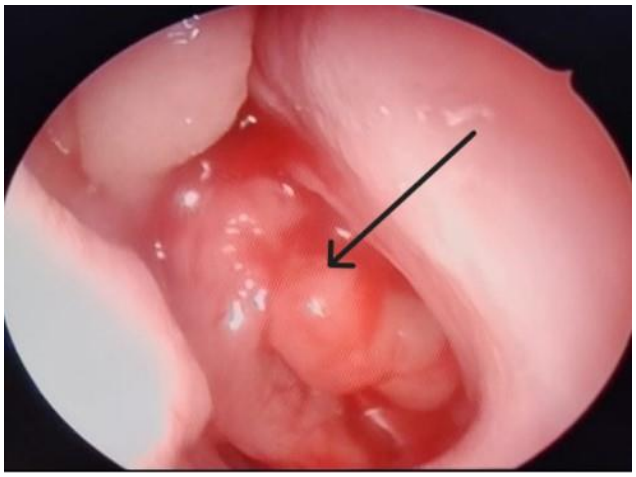

Figure 3: Intra op 0-degree endoscopic view of mass in the nasopharynx fully obstruction the right choana.

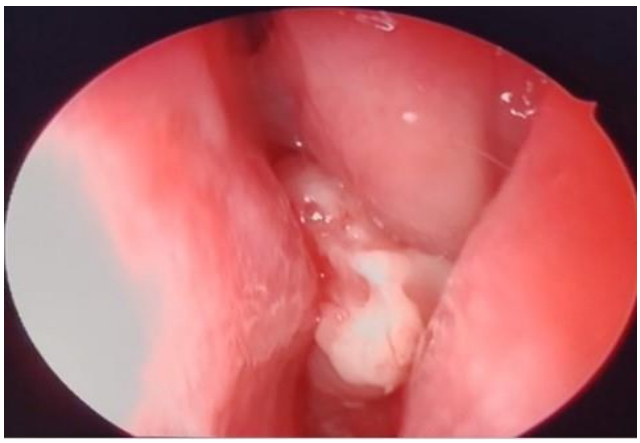

Figure 4: Intra-op 0-degree endoscopic view of mass in the nasopharynx extending into the left cavity.

Histopathologically, both the samples showed subepithelial neoplastic cells in sheets, with focal necrosis. And was suggestive of small round blue cell tumor. Patient was then shifted under the care of a pediatric hemato-oncologist for further management and started on chemotherapy while the sample was sent for immunohistochemistry, the neoplastic cells were shown to express CD45, CD20 and CD79a, and a Ki67 proliferative index of 80%, which favoured a diagnosis of high grade B cell lymphoma. PET-CT was done (Figure 5-7) and a provisional diagnosis of Burkitt lymphoma was made based on these findings. Bone marrow aspirate was negative for neoplastic cells, thus ruling out leukemia. We would have preferred to confirm this with

C-MYC immunohistochemistry but could not, due to poor financial status of the patient. The child was then started on definitive chemotherapy, a regimen that included cyclophosphamide, vincristine, prednisolone, adriamycin, methotrexate- COPAD-M regimen. Patient responded well to the treatment and was clinically better after 5 cycles of chemotherapy. A repeat PET-CT done after the regimen showed complete remission.

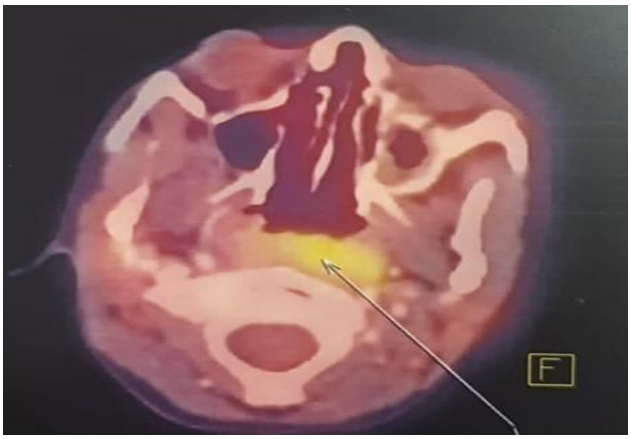

Figure 5: PET-CT of hypermetabolic, ill defined soft tissue density along the left posterior nasopharyngeal wall.

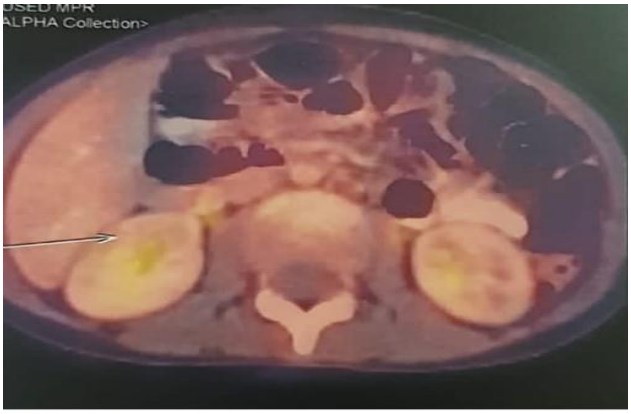

Figure 6: PET-CT of non-FDG avid hypodense area in the inferolateral cortex of right kidney.

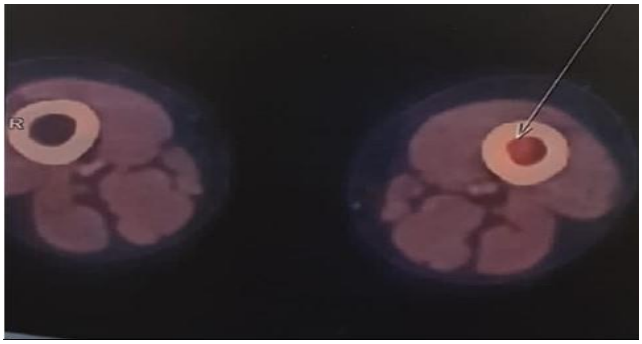

Figure 7: PET-CT hypermetabolic region with soft tissue density in marrow of left distal shaft of femur-likely to be lymphomatous involvement.

DISCUSSION

Adenoids form an important component of the Waldeyer's ring with the basic function of antibody formation, especially in childhood. Due to the location of this lymphoid tissue in nasopharynx, where it frequently interacts with the antigens entering the body, adenoid hypertrophy is a commonly encountered disorder in pediatric population and can present as a spectrum of symptoms, mainly due to physical obstruction to the structures communicating with the nasopharynx, including nasal cavity and eustachian tube. Common presentations include snoring, nasal obstruction, hyponasal speech, obstructive sleep apnoea, chronic rhinosinusitis and recurrent otitis media.[6]

However, in some cases, epistaxis and hematemesis may be the main symptoms, which can lead to a missed diagnosis of adenoid hypertrophy. Adenoidectomy is seen to be curative even in the such peculiar presentations.[7]

Other differentials of a nasopharyngeal mass in similar age group, include benign tumors like juvenile nasopharyngeal angiofibroma (JNA) and nasopharyngeal malignancies, former being a more important differential.[8] JNA is an uncommon fibrovascular tumor, with a usual presentation of nasal obstruction, epistaxis or rhinorrhoea exclusively in adolescent males. Apart from clinical features, CT and MRI scans aid the diagnosis of JNA. Angiography provides additional evidence for diagnosis for vascular tumors such as JNA.[9]

Of malignancies, squamous cell carcinoma is the most commonly found in head and neck region, followed by lymphomas (2.5%), and non-Hodgkin's lymphomas (NHL) represent the majority (65-70%), most frequently involved site being the Waldeyer's ring. The presentation includes nasal obstruction (88.6%), followed by hypoacousia, epistaxis, hearing impairment and rhinorrhea. Only 27% patients of head and neck NHL are reported to have systemic symptoms like fever, sweat, and significant weight loss.[10] Large B-cell lymphomas are seen to form majority of Waldeyer's ring NHL, Burkitt's Lymphoma forming only the majority of 3-5%.[5]

Burkitt's lymphoma is a highly aggressive form of B-cell lymphoma and is the fastest growing human tumor, showing to have a high association with EBV and chromosomal translocation that activates an oncogene (c-myc). It occurs in three major clinical forms: endemic, sporadic (mainly found in regions not affected by malaria) and immunodeficiency associated.[4] Although rare, it is important to recognise Burkitt lymphoma as an oncologic emergency in view of its highly aggressive nature. Patients usually present with a painless, enlarging submucous mass with a healthy-looking hyperemic mucosa, which only become symptomatic on growing large enough to cause obstructive symptoms.

Diagnosis of Burkitt's lymphoma is confirmed by microscopy and immunocytological analysis. On microscopy, it is characterised by monomorphic medium-sized cells with very high proliferation rate (almost 100%).[5] C-MYC rearrangement with one of the immunoglobulin gene loci is the genetic hallmark of BL.[11] Several other investigations in suspected cases like CBC, ESR, renal profile, liver function tests, and serum lactate dehydrogenase, EBV status, and chest radiography are performed further to assess the severity and prognosis of disease. Radiography (CT chest and abdomen) provides additional information about extent of disease lymph nodes involvement (especially mediastinal). PET scanning is also recommended but not essential.[4] Once the diagnosis is confirmed, CSF should be examined to rule out intracranial involvement, as it may change the intensity of treatment.

Treatment of a Burkitt lymphoma is mainly including an induction course including low-dose cyclophosphamide and high-dose methotrexate. Currently used is COPADM regimen (cyclophosphamide, vincristine, prednisolone, Adriamycin, methotrexate). Studies have also shown to improve the outcome with addition of rituximab to the regimens, especially in patients who present with central nervous system involvement.[12] The outcome for sporadic Burkitt's in children is excellent with an overall cure rate of approximately 90%.[4]

CONCLUSION

Although rare, nasopharyngeal NHL should be kept in mind as a differential diagnosis, apart from inflammatory adenoid hypertrophy and benign tumors such as JNA while evaluating a nasopharyngeal mass in children, especially with an unusual presentation. Since they present as submucosal growths, these malignant masses may easily be mistaken as adenoid hypertrophy which are similar in gross appearance. Nasopharyngeal NHL are highly chemo sensitive, therefore, a high suspicion, and hence an early diagnosis can aid timely commencement of chemotherapy and prevent progression and complications of the disease.

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