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Unusual Site of Langerhans Cell Histiocytosis Involving Nasopharynx

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Abstract

Langerhans cell histiocytosis (LCH) is a rare proliferative disorder that originates from myeloid-derived precursor dendritic cell, not the Langerhans cell in the skin. The etiology and subsequent development of LCH are idiopathic and not well understood. A 28-day full term baby girl referred her to high tertiary care center due to mass in nasopharynx for further management. Patient was intubated with normal size tube in low sitting mode, no facial dysmorphic feature, fiberoptic nasal scope showed patent nasal cavity bilaterally, mass was encountered at the lower end of nasopharynx with normal covering mucosa. MRI brain and sinuses with contrast demonstrates a single relatively well-defined mass, its epicenter seen at the nasopharynx posterior wall. Its showing extension to the adjacent osseous structures including C1 anterior arch, inferior clivus and bilateral occipital condyle. Transoral biopsy done in the operating room under general anesthesia which confirm the diagnosis of LCH. Case was discussed in the pediatric tumor board, and it was decided to start her on Vinblastine and prednisolone. Patient received Vinblastine 6 mg/m2 IV weekly bolus for 6 weeks, with systemic prednisone 40 mg/m2/day in three divided doses for 4 weeks and then tapered over the following 2 weeks. A follow-up FDG PET was performed for the whole body and MRI of head and neck Interval improvement of the nasopharyngeal and clival mass without significant residual, no enlarged or suspicious cervical lymph nodes. LCH is a rare condition and prevalent among early age group. Occur in any part of body including nasopharynx and oropharynx.

Keywords: Langerhans'cell; Histiocytosis; Nasopharynx

Introduction

Langerhans cell histiocytosis (LCH) is a rare proliferative disorder that originates from myeloid-derived precursor dendritic cell, not the Langerhans cell in the skin [1,2]. The estimated annual incidence of LCH in children are four to eight per million [3-5]. The etiology of LCH is unknown and there has been considerable debate whether LCH represents an inflammatory or a neoplastic disease. The discovery of recurrent mutations in the mitogen activated protein kinase (MAPK) pathway (i.e., BRAF and MAP2K1 mutations) indicates that it is a neoplastic disease [6,7]. The clinical presentation is variable and ranging from localized single-system involvement to disseminated life-threatening multisystem disease [8]. The purpose of reporting this case in a 28-day old baby is that it is rare and unusual site of LCH involving the nasopharynx.

Case Report

Full term baby girl delivered Lower segment caesarean section with good APGAR score. Patient was intubated immediate after birth with normal size tube due to respiratory distress for few days then extubated, on day six reintubated due to respiratory issue. Seen by the ENT team and recommended to do CT scan of head and neck to rule out congenital malformation as baby unable to wean off from ventilation. CT scan showed mass in nasopharynx, so they referred her to high tertiary care center for further management. The case was referred to a tertiary care center, King Fahad Medical City (KFMC) and admitted in NICU. They consulted our Pediatric Otolaryngology service to evaluate the 28 days full term baby girl, with no known medical illnesses, prenatal history was unremarkable. patient was intubated with normal size tube in low sitting mode, no facial dysmorphic feature, examination of oral and oropharynx reveled soft mass in posterior pharyngeal wall causing narrowing of the pharynx. Fiberoptic nasal scope showed patent nasal cavity bilaterally, mass was encountered at the lower end of nasopharynx with normal covering mucosa with no ulceration or discharge. MRI brain and sinuses with contrast demonstrates a single relatively well-defined mass, its epicenter seen at the nasopharynx posterior wall. Its showing extension

to the adjacent osseous structures including C1 anterior arch, inferior clivus and bilateral occipital condyle

The lesion demonstrates homogenous iso T1-WI and intermediate to low T2-WI signal intensity, with enhancement and some restricted diffusion, Figures 1&2. No calcification or hemorrhage, and no cystic or necrotic component. The mass is bulging towards the nasopharynx leading to complete obstruction together with mild obstruction of the oropharynx Figure 1. The parapharyngeal fat is intact. The hypopharynx is clear. No intracranial extension. There is encasement of both internal carotid arteries most significant at the right side, Figure 2. No enlarged cervical lymph nodes. The brain MRI scan was normal, without underlying lesion or abnormal enhancement.

After discussing the case between multiple teams, the plan was to perform transoral biopsy in the operating room under general anesthesia, biopsy taken and send for histopathology which confirms the diagnosis of LCH Figure 3. The patient's case was discussed in the pediatric tumor board, and it was decided to start her on Vinblastine and prednisolone. Patient received Vinblastine 6mg/m2 IV weekly bolus for 6 weeks, with systemic prednisone 40 mg/m2/day in three divided doses for four weeks and then tapered over the following two weeks. A follow-up FDG PET was performed for the whole body and MRI of head and neck Interval improvement of the nasopharyngeal and clival mass without significant residual, No enlarged or suspicious cervical lymph nodes.

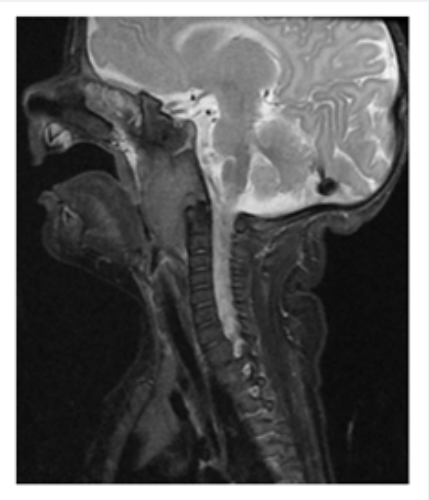

Figure 1: Sagittal T2 STIR (2); An iso- to low T2-WI signal intensity lesion, centered at the nasopharynx bulging towards the naso- and oro-pharynx, with invasion to the adjacent osseous structures including the C1 and inferior clivus, as well bilateral occipital condyle (not included in the images).

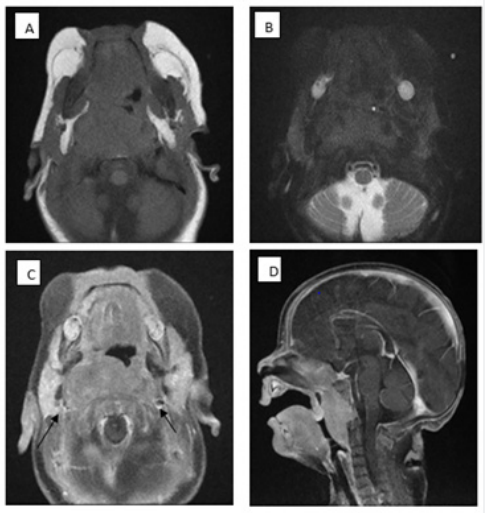

Figure 2: (A) Axial T1; Isointense T1-WI signal intensity lesion, parapharyngeal fat space is preserved. Axial T2 fat sat (B); Isointense T1-WI signal intensity lesion. Axial T1 post contrast (C); Post contrast images shows homogenous enhancement, no cystic or necrotic component. Internal carotid artery (Arrow) encasement worst at the right side with mild narrowing. Sagittal T1 post contrast (D); no intracranial extension.

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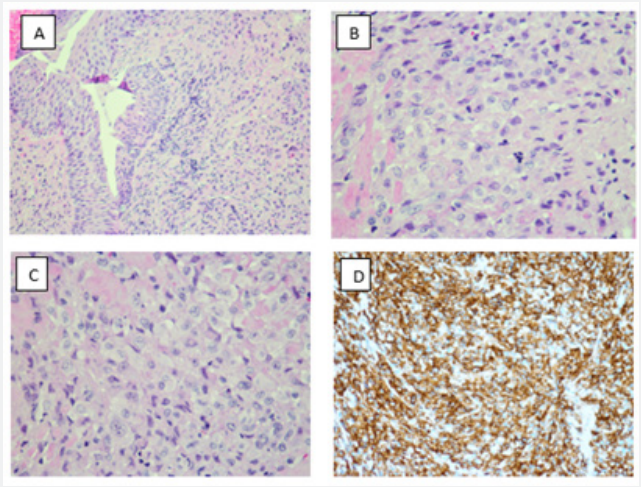

Figure 3: Microscopic examination of hematoxylin-eosin–stained section shows (A) subepithelial proliferation of neoplastic cells (magnification x20). (B, C) These cells have irregular, reniform nuclei with clefts and grooves, fine chromatin, and abundant pale eosinophilic cytoplasm (magnification x40). (D) Immunohistochemical staining demonstrated strong and diffuse immunoreactivity for CD1a (magnification x20).

Discussion

LCH is a rare condition, and it presents clinically in different ways. The etiology and subsequent development of LCH are idiopathic and not well understood. It is suggested that this disease might be seriously affected by predisposing factors that are viral or genetic. However, here's no conclusive prove for it [9]. The clinical presentation of LCH is highly heterogeneous. LCH may involve many organs (skin, bone, central nervous system, spleen, lungs, liver, or gastrointestinal tract) [10]. Definitive diagnosis depends on the identification of characteristic immunohistochemical (IHC) or ultrastructural features of the biopsy specimen [11]. Langerhans cells show strong positivity by IHC studies for S100 protein and CD1a [12]. The optimal management requires interdisciplinary collaboration between specialists (oral surgeon, dentist, ENT, radiologist, pathologist, and oncologist)[13].

In our case report biopsy were necessary to diagnose the patient's condition, which shows a subepithelial proliferation of neoplastic cells with irregular, reniform nuclei with clefts and grooves, fine chromatin, and abundant pale eosinophilic cytoplasm. The cells are admixed with varying proportions of eosinophils and other inflammatory cells. Immunohistochemical staining demonstrated strong and diffuse immunoreactivity for CD1a, S100 and CD68 confirming the diagnosis of LCH Figure 3. Highlighting the challenge of radiology including MRI which can give various differential diagnosis. Manifestations of LCH in nasopharynx and oropharynx regions in Literatures are limited and mostly consists of case reports or retrospective series [14]. In severe cases with systemic LCH presentations, chemotherapy improves the outcome, however due to the rareness of the disease, it is still unclear which ways are best suited for different clinical situations [13]. In our case, the patient received six cycles chemotherapy with no relapsing after three months.

Conclusion

LCH is a rare condition and prevalent among early age group. Occur in any part of body including nasopharynx and oropharynx. Multidisciplinary team need to reach the diagnosis and treatment.

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