American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2024; 25: e942681
DOI: 10.12659/AJCR.942681

Received: 2023.09.25
Accepted: 2024.01.25
Available online: 2024.04.02
Published: 2024.XX.XX

Langerhans Cell Histiocytosis in Sphenoid Sinus: Uncommon Bone Involvement

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABEF 1,2 Taynara Luisa de Mello Heliodoro
AB 3 Paulo Ronaldo Jubé Ribeiro
BC 3 Luciana Ximenes Salustiano
AEF 3 Leandro Azevedo de Camargo
EF 1,2 Fayez Bahmad Jr.

1 Faculty of Health Sciences, University of Brasília, Brasília, DF, Brazil
2 Department of Otolaryngology, Brasiliense Institute of Otorhinolaryngology, Brasília, DF, Brazil
3 Faculty of Medicine, Federal University of Goias, Goiânia, GO, Brazil

Corresponding Author: Fayez Bahmad Jr., e-mail: fayezbjr@gmail.com
Financial support: None declared
Conflict of interest: None declared

Patient: Male, 6-year-old
Final Diagnosis: Langerhans cell histiocytosis
Symptoms: Diplopia • headache • ptosis • strabismus
Clinical Procedure: Adjuvant chemotherapy • surgical resection
Specialty: Neurosurgery • Ophthalmology • Otolaryngology • Pediatrics and Neonatology

Objective: Rare disease

Background: Langerhans cell histiocytosis (LCH) is a rare and uncontrolled proliferation of dendritic cells of myeloid origin. The incidence of LHC was estimated at 5 cases per million children ages 0-15 years old. The most common places for this tumor are the jaw, vertebra, pelvis, and the extremities. The disease with multisystem involvement can present a mortality rate of 20% and one-third of children have multisystem involvement. We present a case with unusual bone involvement of the anterior cranial base with a challenging diagnosis and a complex surgical approach.

Case Report: We report the case of a 6-year-old boy who manifested the disease with daily holocranial headache, worse in the frontal region and refractory to analgesia for 10 days, strabismus homonymous, diplopia, and right palpebral ptosis. The tumor affected the sphenoid sinus, internal carotid artery, and sella turcica, and made contact with the pituitary gland. A joint surgery with Otorhinolaryngology and Neurosurgery was performed by nasal endoscopic access to the skull base by means of the right medial turbinectomy (for the access) and right sphenoid opening, septectomy and opening of the left sphenoid to work with 4 hands and, after resection of lesion, inside the sphenoid.

Conclusions: This patient had rare bone involvement from LCH and atypical clinical presentation next to the important and delicate structures of the anterior skull base, but had a satisfactory outcome.

Keywords: Dendritic Cells • Headache • Myeloid Cells • Skull Base Neoplasms • Strabismus

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/942681

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

Introduction

Langerhans cell histiocytosis (LHC) is a rare and uncontrolled proliferation of dendritic cells of myeloid origin [1,2]. The incidence of LHC was estimated at 5 cases per million children. This neoplasm presents a very rare involvement of the skull base [3,4]. This tumor is most common in bones (77%), mainly the jaw, vertebra, pelvis, and extremities [5]. Two-thirds of affected children have involvement of a single system, most commonly in the bones, but also lymph nodes or the skin. In one-third of the remaining cases, the disease with multisystem involvement can present a mortality rate of 20% [6]. We present a case with an unusual bone involvement of cranial base and challenging diagnosis. The patient was a boy with rare bone involvement from LCH and atypical clinical presentation, but who had a satisfactory outcome. Earlier diagnosis of this rare tumor in a child was essential to establish an effective treatment without sequelae.

Case Report

A 6-year-old boy was admitted to for intense daily holocranial headache, worse in the frontal region and refractory to analgesia for 10 days, evolving strabismus, homonymous diplopia, and right palpebral ptosis. There were no reports of rhinorrhea,

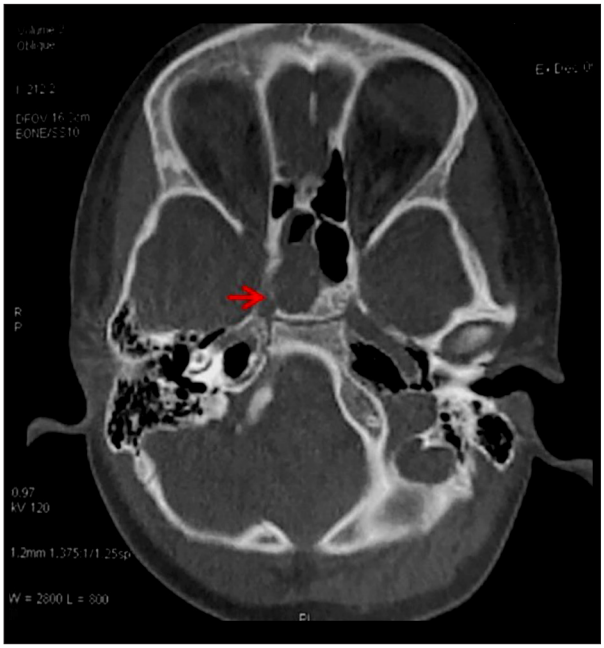


Figure 1. Axial computed tomography scan. The picture shows an expansive lesion in the right sphenoid sinus, causing cortical rupture in its posterior and lateral wall and extending to the intracranial compartment next to the right cavernous sinus. The size of the lesion was 1.5×1.2 cm.

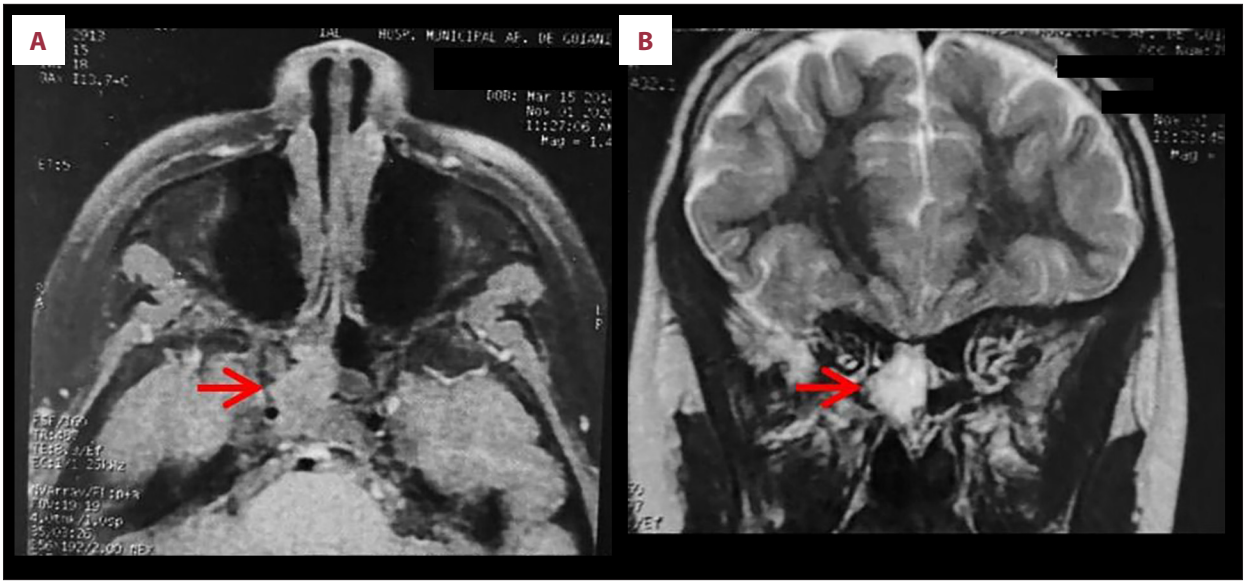


Figure 2. Magnetic resonance imaging. (**A**) Magnetic resonance imaging in axial T1 section. The red arrow points to a solid extensive/infiltrative lesion in the sphenoid body, on the right side of the midline, occupying a large part of the sphenoid sinus and causing bone erosions on the posterior, lateral, and superior walls. There was also extension to the right cavernous sinus, where it involved about 180° of the cavernous segment of the right internal carotid. (**B**) Magnetic resonance imaging coronal T2 section. The red arrow indicates a solid extensive/infiltrative lesion in the sphenoid body, on the right side of the midline, occupying a large part of the sphenoid sinus, causing bone erosions on the posterior, lateral, and superior walls. There was also extension to the right cavernous sinus, where it involved about 180° of the cavernous segment of the right internal carotid artery.

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

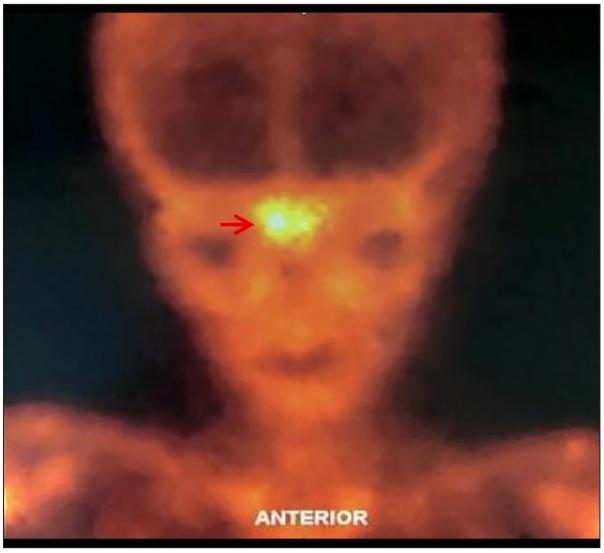


Figure 3. SPECT/CT bone scintigraphy with MDP-99mTc. The red arrow shows increased osteogenic activity in the sphenoid bone to the right of the midline.

epistaxis, nasal itching, fever, or sneezing. His parents reported that he had not had any previous symptoms, comorbidities, surgeries, or long-term use of medication. Anterior rhinoscopy and endoscopy did not show any alteration. Computed tomography revealed an expansive lesion in the right sphenoid sinus, which caused cortical rupture in its posterior wall and extended to the intracranial compartment next to the right cavernous sinus. The size of the lesion was 1.5×1.2 cm (**Figure 1**). Nuclear magnetic resonance imaging (MRI) showed a solid expansive and infiltrative lesion in the body of the sphenoid on the right side of the midline, occupying a large part of the sphenoid sinus, and bone erosions on the posterior, lateral and superior walls. There was also extension to the right cavernous sinus involving about 180° of the cavernous segments of the right internal carotid artery, and this artery was narrowed. There was erosion of the sella turcica floor and contact with the hypophysis, and foramen rotundum involvement in the V2 pathway (**Figure 2**). Technetium-99m bone scintillography showed increased osteogenic activity in the sphenoid bone on the right side of the midline, corresponding to bone lesions disseminated by contiguity, and there were no secondary implants (**Figure 3**). A joint surgical approach by Otorhinolaryngology and Neurosurgery was performed by nasal endoscopic access to the skull base. The steps of surgery were right medial turbinectomy (for the access) and right sphenoid opening, septectomy and opening of the left sphenoid to be able to work with 4 hands and, after the resection of lesion, to work inside the sphenoid. There was no neuronavigation system available. All the macroscopically visible lesions appeared to have been removed. The anatomo-pathology exam indicated a poorly differentiated neoplasia of indeterminate histogenesis analysis on stored preparations stained with hematoxylin-eosin (**Figure 4**). Immunohistochemistry (IHC) was compatible with Langerhans cell histiocytosis (positive markers were anti-macrophages CD68, CD163, Ki67, CD1A, and S-100 protein) (**Figure 5**). Subsequently, the patient underwent 12 sessions of Vinblastine adjuvant chemotherapy. There was complete resolution of headache and diplopia on the thirtieth postoperative day. Follow-up was carried out for 2 years following the surgery, with biannual bone scintigraphy, videonasoscopy,

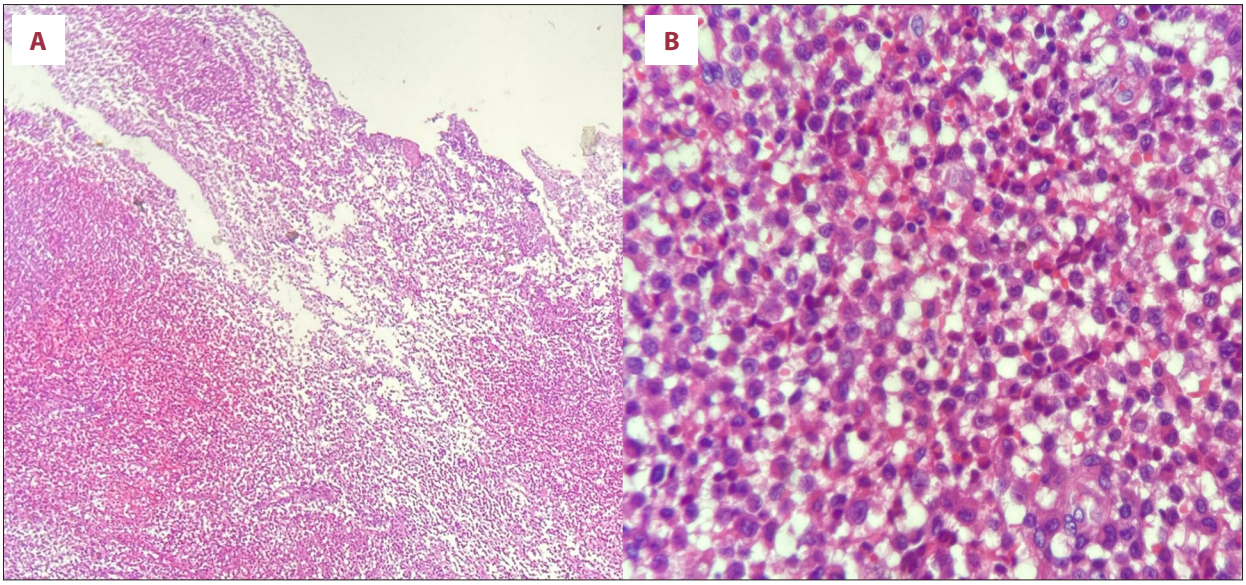
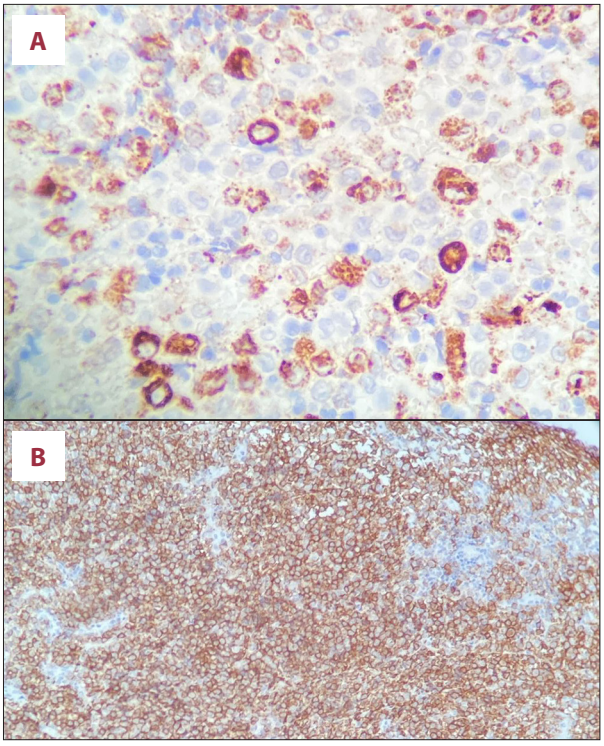


Figure 4. (**A**) A photomicrograph of the resected surgical lesion, stained by hematoxylin and eosin (H&E), magnification 10×. The anatomo-pathology indicated a poorly differentiated neoplasia of indeterminate histogenesis analysis. (**B**). Photomicrograph of the resected surgical lesion, stained by hematoxylin and eosin (H&E). In a bigger magnification of 40×. The anatomo-pathology indicated a poorly differentiated neoplasia of indeterminate histogenesis.

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]



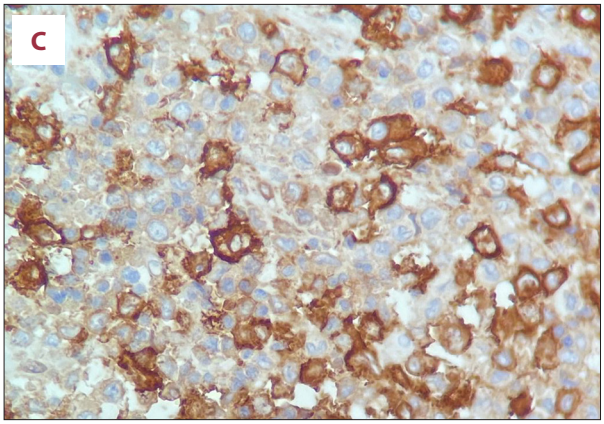


Figure 5. Immunohistochemistry (IHC) compatible with LCH. (**A**) Immunohistochemistry (IHC) with the antibody maker S100, compatible with LCH. (**B**) Immunohistochemistry (IHC) with the antibody maker CD1A, compatible with LCH. (**C**) Immunohistochemistry (IHC) with the antibody maker CD163, compatible with LCH.

and MRI, without evidence of any recurrence or sequelae. The patient is still being followed up.

Discussion

Histiocytosis is identified by abnormal proliferation of cells of the dendritic lineage monocytes and lymphocytes, differentiated from myeloid progenitor cells [7]. These neoplasms are classified as Langerhans cell-related, non-Langerhans cell-related, or malignant. LHC is derived from an immature myeloid progenitor and can occur in all age groups, but is most frequent in children aged 1-3 years [8]. This differs from our patient, who presented clinical symptoms at age 6 years. It is more common in White and male children. The male-to-female ratio is about 2.5: 1 [9]. It affects about 3-5 per million children aged 0-15 years [10]. Although there are silent cases, signs and symptoms vary according to the location and extent of the disease. The most frequent symptom is localized pain. It is limited to a single organ system – eg, skin, lymph nodes, thymus, lungs, spleen, liver, bone marrow, or central nervous system (CNS) – in about half of patients, and in children, in up to two-thirds of cases [11]. Bone involvement occurs in most cases, and it can involve any bone tissue in the body. Cutaneous involvement occurs in up to 40% of cases, and was not observed in our patient. Significant risk factors for LCH can include maternal urinary tract infection during pregnancy, feeding problems or blood transfusions during infancy, Hispanic ethnicity, crowding, low education level, neonatal infections, solvent exposure, family history of thyroid disease, and in vitro fertilization [12], but our patient did not have any of these. Protective factors appear to be Black race, childhood vaccinations, and supplemental vitamins [13]. Our patient had all vaccines on the Brazilian public health system calendar up to date and he was supplemented with vitamins A and D until he was 2 years old. Imaging exams demonstrate lytic areas and erosions, as well as a possible adjacent soft-tissue mass. The most sensitive diagnostic test is high-resolution CT, which can demonstrate cysts and nodules characteristic of LCH, and it was the initial exam performed in this case. MRI was important in the anatomical study and delimitation of the lesion for surgical programming [14]. The risk of affecting the CNS varies according to bone involvement. Injuries to facial bones and the anterior and middle cranial fossa present a higher risk, affecting almost 25%. The most common symptoms of CNS involvement are diabetes insipidus, ataxia, and cognitive dysfunction. LCH is diagnosed from the pathological evaluation, interpreted in association with the clinical context [15].

The possible differential diagnosis is benign tumors of the anterior skull base, which can originate from intracranial, cranial, or extracranial sites, as well as fibro-osseous lesions like osteoma, ossifying fibroma, and fibrous dysplasia. The most common extracranial neoplasms that can extend to the cranial base include inverted papilloma and angiofibroma. The diagnosis is often delayed because symptoms are nonspecific. Small asymptomatic tumors (eg, meningioma, osteoma) may be only observed, while others must be treated due to potential for malignancy (eg, inverted papilloma) or continued destructive growth (eg, angiofibromas) [16]. More rare differential diagnoses include epithelial adenomatoid hamartomas and schwannoma of the anterior skull base [17,18].

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

Biopsy of the osteolytic bone or cutaneous lesion is preferred when possible. Langerhans cell histiocytes may be suspected based on morphologic criteria. Their identity should be confirmed by positive immunohistochemical staining for CD1a and CD207 or identification of Biberck granules by electron microscopy [19]. LHC can be challenging to diagnose due to its rarity and the possibility of affecting many systems, with a wide range of symptoms. Histological and immunophenotypic differentiation from other histiocytic diseases, such as dendritic cell diseases, lymphohistiocytic and macrophage hemophagocytic activation syndromes, and solid metastatic or hematopoietic neoplasms is required [18,19]. The prognosis of the single-system form is better than the form affecting multiple systems, with a 5-year survival of close to 100%. The 5-year recurrence rate is under 20%. Bone involvement usually occurs in the same location as the primary involvement [19]. In this case, there was no recurrence in 2 years of follow-up. Despite the possibility of spontaneous remission in the bone lesion within 3 months, the recommended treatment was surgical resection due to its location next to important and delicate structures. Although the surgery was risky, our patient had a very satisfactory outcome, without evidence of any recurrence or sequelae.

Conclusions

We report a case of LCH involving the sphenoid sinus, internal carotid artery, and sella turcica, making contact with the pituitary gland. CT and MR images, particularly with contrast, were helpful in defining extent before treatment. LCH should be considered in the differential diagnosis of lesions in this location in the pediatric age group. Despite the possibility of spontaneous remission, in bone lesions within 3 months, the recommended treatment was surgical resection due to its location next to important and delicate structures. Although the surgery was risky, our patient had a very satisfactory outcome, without evidence of any recurrence or sequelae.

Department and Institution Where Work Was Done

Department of Otolaryngology, Brasiliense Institute of Otorhinolaryngology, Brasília, DF, Brazil.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

References:

1. Leung AKC, Lam JM, Leong KF. Childhood Langerhans cell histiocytosis: A disease with many faces. World J Pediatr. 2019;15(6):536-45
2. Krooks J, Minkov M, Weatherall AG. Langerhans cell histiocytosis in children: History, classification, pathobiology, clinical manifestations, and prognosis. J Am Acad Dermatol. 2018;78(6):1035-44
3. Kobayashi M, Tojo A. Langerhans cell histiocytosis in adults: Advances in pathophysiology and treatment. Cancer Sci. 2018;109(12):3707-13
4. Liu ZF, Dai Q, Yu HM. [A case report of Langerhans cell histiocytosis in sphenoid sinus.] Zhonghua Er Bi Yan Hou Tou Jing Wai Ke Za Zhi. 2018;53(1):63-65 [in Chinese]
5. Yu G, Huang F, Kong L, et al. Langerhans cell histiocytosis of the sphenoid sinus: A case report. Turk J Pediatr. 2010;52(5):548-51
6. Kim BE, Koh KN, Suh JK, et al. Clinical features and treatment outcomes of Langerhans cell histiocytosis: A nationwide survey from Korea histiocytosis working party. J Pediatr Hematol Oncol. 2014;36:125-33
7. Baumgartner I, von Hochstetter A, Baumert B, et al. Langerhans'-cell histiocytosis in adults. Med Pediatr Oncol. 1997;28(1):9
8. Emile JF, Abla O, Fraitag S, et al. Revised classification of histiocytoses and neoplasms of the macrophage-dendritic cell lineages. Blood. 2016;127:2672
9. Stromberg JS, Wang AM, Huang TE, et al. Langerhans cell histiocytosis involving the sphenoid sinus and superior orbital fissure. Am J Neuroradiol. 1995;16(4 Suppl.):964-67
10. Krishna H, Behari S, Pal L, Chhabra AK, et al. Solitary Langerhans-cell histiocytosis of the clivus and sphenoid sinus with parasellar and petrous extensions: Case report and a review of literature. Surg Neurol. 2004;62(5):447-54
11. Bagnasco F, Zimmermann SY, Egeler RM, et al. Langerhans cell histiocytosis and associated malignancies: A retrospective analysis of 270 patients. Eur J Cancer. 2022;172:138-45
12. DiCaprio MR, Roberts TT. Diagnosis and management of Langerhans cell histiocytosis. J Am Acad Orthop Surg. 2014;22:643-52
13. Liu H, Stiller CA, Crooks CJ, et al. Incidence, prevalence and survival in patients with Langerhans cell histiocytosis: A national registry study from England, 2013-2019. Br J Haematol. 2022;199:728-38
14. Singh J, Rajakulasingam R, Saifuddin A. Langerhans cell histiocytosis of the shoulder girdle, pelvis and extremities: A review of radiographic and MRI features in 85 cases. Skeletal Radiol. 2020;49:1925-37
15. Rodriguez-Galindo C. Clinical features and treatment of Langerhans cell histiocytosis. Acta Paediatr. 2021;110:2892-902
16. Snyderman CH, Lavigne P. Benign tumors of the anterior cranial base. Adv Otorhinolaryngol. 2020;84:106-13
17. Cascio F, Basile GC, Felippu AWD, et al. Diagnosis and treatment of bilateral respiratory epithelial adenomatoid hamartomas with and without sinonasal polyposis. Ear Nose Throat J. 2021;100(5 Suppl.):495S-97S
18. Poma S, Modica DM, Cascio F, et al. Endoscopic endonasal resection of a schwannoma of the anterior cranial fossa. Ear Nose Throat J. 2022;101(2):NP41-NP44
19. Hu X, Buhtoiarov IN, Wang C, et al. Langerhans cell histiocytosis: A population-based study of anatomical distribution and treatment patterns. J Bone Oncol. 2022;36:100454

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]