

Case Report

# Unique case of trigeminal neuralgia due to Epstein-Barr-virus-associated B-cell lymphomatoid granulomatosis of the Meckel’s cave and cavernous sinus: Important clinical and therapeutic implications

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## Abstract

**Background:** Trigeminal neuralgia (TN) represents one of the most disabling pain syndromes. Several diseases have been described as etiological triggers of TN, vascular compression of the trigeminal nerve being the most frequent cause. Here, we describe for the first time a rare case of TN caused by an infiltration of an isolated Epstein–Barr virus (EBV) B-cell lymphomatoid granulomatosis (LYG) mass into the Meckel’s cave and cavernous sinus.

**Case Description:** A 51-year-old woman undergoing immunosuppressant treatment for Crohn’s disease presented due to right-sided TN. Magnetic resonance imaging (MRI) scans revealed an isolated lesion affecting the right Meckel’s cave and lateral wall of the cavernous sinus. We accomplished tumor resection through a subtemporal extradural approach and the patient recovered successfully from surgery. Histological examination revealed an LYG, and a blood test confirmed low but positive EBV counts. The immunosuppressant therapy was discontinued and we assumed a watchful waiting management. During a 41-months’ follow-up there was neither evidence of LYG recurrence nor an increase of EBV counts.

**Conclusions:** LYG, an angiodestructive disease associated with EBV reactivation in the context of immune dysfunction and often associated with an aggressive behavior or even malignant transformation, should be considered as a rare differential diagnosis of TN associated with skull base lesions. The management of this rare disease is still controversial and varies from limiting the treatment to correcting immune dysfunction up to chemotherapy. In this case of

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an isolated mass, surgical excision and discontinuation of immunosuppressants were effective to prevent the relapse of the disease in a long-term follow-up.

**Key Words:** B-cell lymphomatoid granulomatosis, cavernous sinus, Meckel's cave, trigeminal neuralgia

## INTRODUCTION

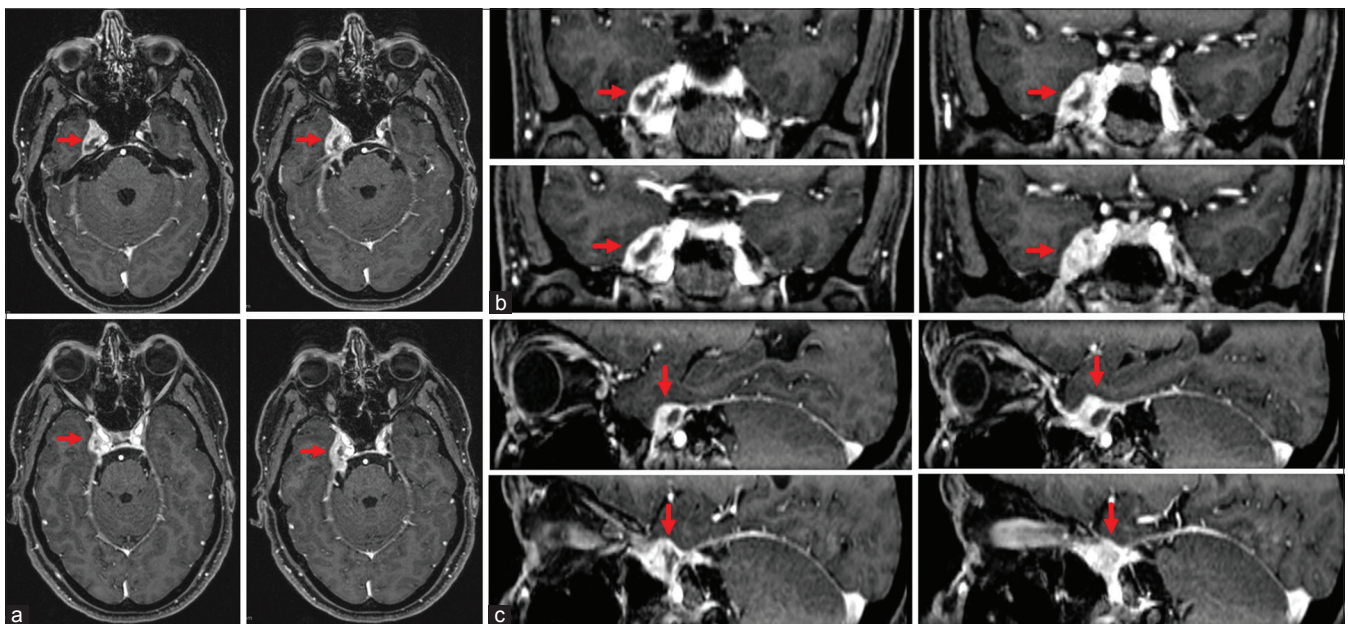
Trigeminal neuralgia (TN) represents one of the most disabling pain syndromes that patients can experience. The identification of its etiology in individual patients is mandatory to rule out diseases that may primarily require special evaluation and treatment besides the pharmacological therapy directed to relief neuropathic pain. TN is associated in a vast majority of cases with a vascular compression of the trigeminal nerve, followed more infrequently by inflammatory and tumor processes affecting the nerve or its central pathways.<sup>[1,2,5-7,9,15,16,18,24,29,33]</sup> Although granulomatous diseases such as sarcoidosis, tuberculosis, and Wegener's granulomatosis have been described to affect the trigeminal nerve and trigger TN due to infiltration of the skull base.<sup>[1,2,5-7,16,24,33]</sup> Till now, there are no reports of B-cell lymphomatoid granulomatosis (LYG) showing the infiltration of structures within the middle fossa or causing TN. We present for the first time the case of a 51-year-old woman receiving immunosuppressant treatment due to Crohn's disease and presenting TN caused by an infiltration of a LYG mass into the Meckel's cave and lateral wall of the cavernous sinus. LYG is an angiodesstructive disease associated with EBV reactivation in the context of immune dysfunction. Its identification

may be critical due to the potential of this disease to show a very aggressive behavior or even malignant transformation.<sup>[10,23,28,35]</sup> In this report, we discuss the important clinical and therapeutic implications of recognizing this entity as a rare but possible differential diagnosis of lesions affecting the middle fossa and causing TN. We also review the literature available on LYN to provide an insight into the management and therapy that might be applicable in these cases.

## CASE REPORT

A 51-year-old woman presented to our clinic because she had been suffering for the last 3 weeks from TN affecting the right V1-V2 branch. The patient reported a strong progression of pain as well as diplopia. Her neurological examination was inconspicuous apart from the presence of a right abducens nerve paresis and sensory loss corresponding to the V1 and V2 dermatome. According to her clinical records, she suffered from Crohn's disease and had received azathioprine as immunosuppressive therapy.

Magnetic resonance imaging (MRI) scans revealed a heterogeneous T1 contrast enhancing lesion in the right Meckel's cave extending to the lateral wall of the cavernous sinus [Figure 1]. The lesion was isointense



**Figure 1:** Preoperative axial (a), coronal (b), and sagittal (c) MRI scans showing a heterogeneous T1 contrast enhancing lesion involving the right Meckel's cave and the lateral wall of the cavernous sinus corresponding in the histopathological analysis to an Epstein-Barr-virus-associated B-cell lymphomatoid granulomatosis

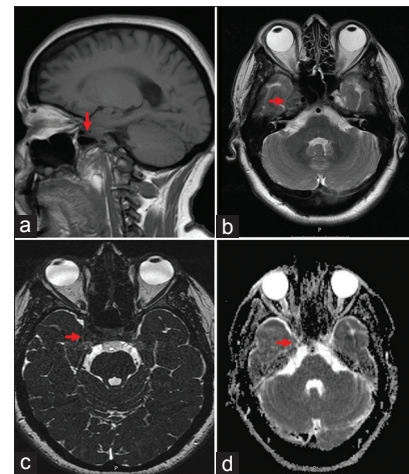
in T2 and constructive interference in steady state (CISS) sequences and did not show diffusion restriction. There was no evidence of vascular compression of the trigeminal nerve in the cerebellopontine angle [Figure 2]. Blood probes did not show any abnormal values, and protein levels in cerebrospinal fluid (CSF) were slightly increased, especially IgA and IgG antibodies. Oligoclonal bands were not detectable.

We suggested surgical exploration and trigeminal nerve decompression in the Meckel's cave using a right subtemporal extradural Dolenc approach. During surgery, the lesion was amenable to be separated from the dural envelope of the cavernous sinus and was completely resected [Figure 3]. After surgery, the patient recovered successfully without any new neurological deficits. Although the facial sensory deficit remained unchanged, facial pain and diplopia from the abducens nerve paresis resolved. No corticosteroids were administered to the patient, neither before nor after the surgery.

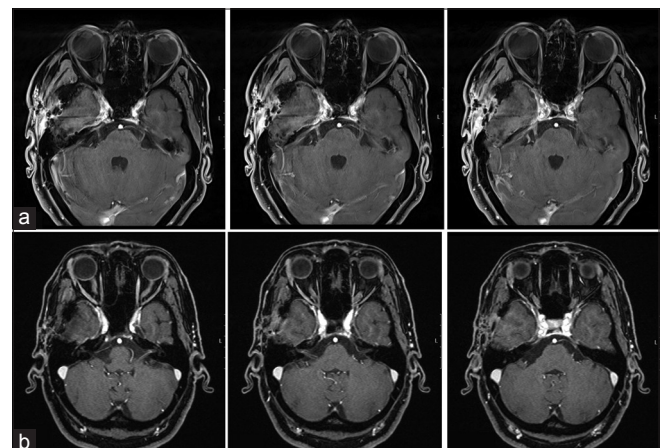
Histopathological examination established the diagnosis of Epstein–Barr-virus-associated B-cell lymphomatoid granulomatosis (LYG) by showing the typical transmural vascular infiltration of lymphocytes, epithelioid cells, and histiocytes with oval nucleus and discrete nucleoles, necrotic epithelioid, and giant cells on the borders and reticulin fiber infiltration [Figure 4]. Mycobacterial infection was excluded by Ziehl–Neelsen staining, and immunohistochemical analysis revealed T-cells as well as clusters of CD20-positive B- lymphocytes (some of them necrotic), small CD1a- positive and S100- negative cells, and CD68+ macrophages.

As expected, analysis of Epstein–Barr virus (EBV) revealed a viral load of 380 copies/mm<sup>3</sup> in blood samples. Abdominal and thoracic computed tomography (CT) scans revealed very small intrapulmonary lesions without affecting the lymph nodes. The patient did not present any systemic or respiratory symptoms. A multidisciplinary conference with the colleagues of the hematology and gastroenterology department decided to discontinue azathioprine and start a treatment with rituximab only if the viral load exceeded 10<sup>3</sup>/mm<sup>3</sup>. The patient was followed up every 3 months in the first year, every 6 months in the second, and then once yearly. In the follow-ups, no corticosteroids were prescribed.

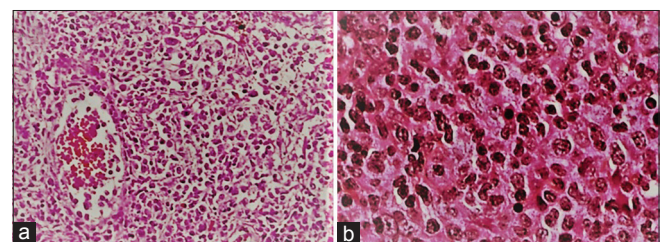
Under surveillance after resection, there was no cranial granuloma recurrence in further neuroimaging studies performed every 6 months during the follow-up [Figures 3 and 5], and EBV load remained lower than 10<sup>3</sup>/mm<sup>3</sup>; hence, there was no need of other specific therapies. Pain recurred after a period of approximately 6 months in which the patient was free of TN. A combined pharmacological therapy was initiated to reduce the pain and showed initial success, but unfortunately failed to achieve a complete symptomatic control in the long term. Because MRI scans consistently ruled out a lesion



**Figure 2:** LYG lesion was isointense in native T1 (a), T2 (b), and CISS (c) sequences and showed no diffusion restriction (d) in preoperative MRI scans



**Figure 3:** Axial contrast enhanced T1-weighted MRI immediately postoperative confirmed a successful removal of the lesion (a) and there was no evidence of LYG relapse (b) 6 months after surgery

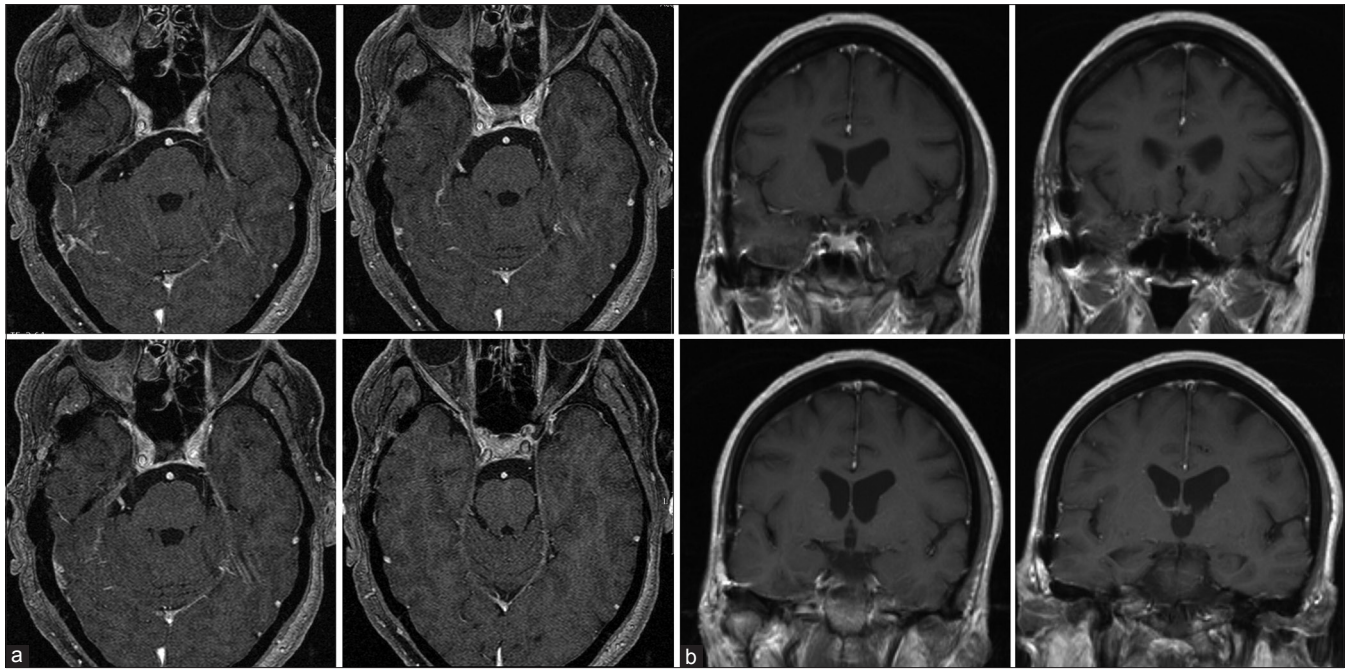


**Figure 4:** Histological tissue sections showing transmural vascular infiltration of lymphocytes, (a) epithelioid cells and histiocytes (b) characteristic of LYG

recurrence and EBV load persisted below 10<sup>3</sup>/mm<sup>3</sup>, we offered a percutaneous thermocoagulation of the Gasserian ganglion, which was refused by the patient.

## DISCUSSION

TN is known as one of the most disabling pain syndromes. Although vascular compression of the trigeminal nerve root



**Figure 5: Axial (a) and coronal (b) contrast weighted T1 MRI images after 41-months' follow-up showing no local recurrence of LYG**

at the Obersteiner Redlich zone in the cerebellopontine angle can be identified as the cause of TN in 80–90% of the cases, several other conditions, such as demyelinated plaques, tumor and pseudotumor lesions, aneurysms, and arteriovenous malformations, have been identified as triggers of TN.<sup>[1,2,5-7,9,15,16,18,24,29,33]</sup> Neuroimaging is mandatory to investigate the etiology of trigeminal pain in each individual patient. It rules out diseases that may primarily require special evaluation and treatment exceeding the standard pharmacological therapy to relieve neuropathic pain and contributes to the assessment of the optimal surgical treatment option if the pain becomes refractory to medication.

Among tumor processes which can affect the trigeminal nerve and trigger TN, neurinoma and meningioma are the most common, although many other entities, included lymphoma, have been already described.<sup>[3,9,12,17,22,25,30,34,36,37,40]</sup> More rarely, granulomatous diseases, such as Wegener's granulomatosis,<sup>[1,7,24]</sup> tuberculosis,<sup>[16]</sup> and sarcoidosis<sup>[2,5,6,33]</sup> involving the Meckel's cave have been described as the etiology of TN. However, to our knowledge, there are no previous reports in the literature of LYG affecting the Meckel's cave or describing this entity as an etiological trigger of TN.

LYG constitutes an extremely rare angiocentric and angiodestructive process described for the first time by Liebow and colleagues in the early 1970s. It is characterized by nodular lesions composed of a mixed population of lymphoreticular cells showing vascular infiltration and necrosis.<sup>[10,28]</sup> Its natural history can vary from a relatively indolent course up to a rapidly fatal evolution resembling high-grade T-cell lymphomas.<sup>[23,35]</sup>

Indeed, cases of true transformation into an aggressive lymphoma have been documented.<sup>[14,23,28]</sup> Therefore, the suspicion and correct diagnosis of this entity is fundamental for the adequate management of each individual patient.

More than 90% of patients suffering from LYG show a symptomatic bilateral involvement of the lungs.<sup>[23,35]</sup> Most patients consult due to systemic complains such as fever, weight loss, and malaise, which are followed by respiratory symptoms such as cough and shortness of breath and present in approximately 50% and 30% of cases, respectively.<sup>[23]</sup> Blood examinations usually show normal leucocyte counts and inflammatory parameters.<sup>[35]</sup> We may remark that no systemic or pulmonary symptomatology was present in our patient at the time of diagnosis or during the follow-up. Other organs such as the kidneys, skin, and central nervous system (CNS) are also frequently affected by LYG. Indeed, an involvement of CNS might occur in 25–50% of patients,<sup>[23,35]</sup> although in such cases affection of the CNS has been always described either as multiple, small, intraparenchymatous brain lesions, as larger heterogeneous ring-enhancing brain masses, or as a leptomeningeal infiltration along the basal cisternal space.<sup>[32]</sup> These lesions are characterized by abnormally increased signal intensity on FLAIR and T2-weighted MR images, as well as by punctate or linear central contrast enhancement in T1 sequences.<sup>[32]</sup> If leptomeninges are affected, MRI scans will show a diffuse contrast enhancement within the basal cisterns. In the series published by Patsalides *et al.*, only one patient developed a lesion in the suprasellar area.<sup>[32]</sup> An isolated affection of

the skull base, as in our patient, has not been reported yet in LYG patients.

Confusion, ataxia, hemiparesis, and seizures are among the most common manifestations of neurological involvement. Others, including cranial nerve dysfunction and peripheral neuropathy affecting usually the lower extremities, have been reported only sporadically. Bell's palsy, diplopia, transient blindness, proptosis, deafness, and vertigo are already described manifestations of cranial nerve dysfunction associated with LYG.<sup>[23,35]</sup> In all patients, leptomeningeal involvement was responsible for the symptoms, not isolated masses within the skull base, as in our patient. No case of TN has been reported earlier in the published series of LYG and the only case existing of trigeminal infiltration was again caused by a leptomeningeal involvement of the basal cisterns.<sup>[23,35]</sup>

In the histological examination, mononuclear cells comprising T-lymphocytes, plasma cells, and histiocytes are accompanied by only a small number of B-lymphocytes showing EBV positivity. However, the key feature comprehends the angiocentric and angiodestructive nature of the lesions with transmural vascular infiltration of T-cells and histiocytes showing various degrees of necrosis.<sup>[10,35]</sup> These characteristics should be meticulously considered to differentiate this entity from other diseases which can affect the cavernous sinus or Meckel's cave, can trigger TN, and may histopathologically resemble some features of LYG, such as sarcoidosis, Wegener's granulomatosis, or lymphoma.

The pathophysiology of this disease has been linked to a lack of control of EBV-infected B-cells. In this context, it is agreed that recruitment of intravascular T-cells induced by EBV infection may mediate the vascular damage seen in histological specimens.<sup>[20]</sup> Apart from EBV load counts, a search for IgG and IgM antibodies directed against EBV should be performed in all patients. Underlying this EBV reactivation, a functional immune dysregulation has been seen in almost all patients suffering from LYG<sup>[20,41]</sup> and, consistent with this observation, the occurrence of LYG has been associated with several autoimmune diseases, congenital immunodeficiencies, and conditions following either leukemia therapy or organ transplantation.<sup>[23,27,31]</sup> In addition, discontinuing immunosuppressants has been observed to induce a remission in patients with LYG, and this option may be considered for each patient, if reasonable.<sup>[35]</sup> In more severe cases with evidence of a progressive disease, patients have been treated with corticosteroids, rituximab, interferon- $\alpha$ -2b, and chemotherapeutic agents such as prednisone, etoposide, vincristine, cyclophosphamide, and doxorubicin.<sup>[4,8,13,14,19,21,23,38]</sup> Although progression and malignant transformation justify the use of these strategies, because an immunodeficiency underlies the pathophysiology of this disease, there are concerns whether

treatment with corticosteroids and chemotherapy could also have the potential of worsening the clinical course. Unfortunately, due to the infrequent presentation of this disease, there is a lack of medical studies systematically proving the effectiveness of these treatments, and a few published series have reported conflicting results. Furthermore, lack of cases reporting an involvement of the skull base makes it difficult to predict the effectiveness of these agents for the treatment of patients like the one we present in this article. Discontinuation of azathioprine, regular control of EBV viral counts, as well as clinical and neuroimaging follow-up seem to have been a reasonable strategy for our patient who did not show evidence of relapse or progressive disease during 41-months of follow-up. We assumed that the recurrence of TN after 6 months must have been associated with postsurgical adhesions/scarring and not to a relapse of LYG or EBV reactivation. It is important to consider that MRI scans performed at the time of pain recurrence and further on repeatedly demonstrated no LYG mass relapse. Furthermore, EBV counts remained  $<10^3/\text{mm}^3$ , and there were no clinical features of active systemic EBV infection along the complete follow-up. Although a local EBV reactivation cannot be completely excluded as a possible cause of TN recurrence (due to the fact that EBV counts had already been  $<10^3/\text{mm}^3$  at the time of diagnosis), in a scenario of local EBV activation we would have expected progressive findings in consecutive MRI scans as well as a further neurological deterioration of the patient, neither had occurred.

The role of surgery may be debated in cases such as the one presented. First, in our patient, surgery was clearly indicated to obtain material for histopathological diagnosis. The atypical localization of a LYG lesion within the Meckel's cave and cavernous sinus had not been described before, thus making the preoperative tentative diagnosis of this disease unlikely in our case. Furthermore, the MRI characteristics shown by this lesion were unspecific. The inhomogeneous contrast enhancement in T1-weighted images resembled features that are commonly described in other diseases which have been reported to affect the cavernous sinus and surrounding structures, such as lymphoma, tuberculoma, sarcoidosis, or Wegener's granulomatosis.<sup>[1,2,5-7,16,24,26,33]</sup> Furthermore, the nodular involvement of the lungs is also a common feature that might occur in all these diseases. Interestingly, we may remark the absence of diffusion restriction in MRI shown by the LYG mass in our patient. Although unspecific, a restriction in diffusion-weighted MRI is usually observed in inflammatory processes such as sarcoidosis or tuberculosis, as well as frequently in dense cellular tumors such as lymphomas.<sup>[26]</sup> Far from being able to generalize our findings, this characteristic, albeit nonspecific feature could eventually help to suspect this disease in future cases. Perhaps MRI spectroscopy

performed on similar lesions could be of great interest in the future to identify possible key features that may be helpful to differentiate and individualize these lesions.

In addition, the determination of EBV antibodies or a positive viral load can be helpful to suspect this disease, however, these parameters do not replace the need of histopathological examination. EBV infection infects the majority of world population and remains asymptomatic in most cases.<sup>[11,39]</sup> Furthermore, other entities which have been described to affect the cavernous sinus, such as Burkitt lymphoma, can be also associated with EBV infection, making the differentiation of these entities possible only by tissue examination.<sup>[12,17,22,40]</sup>

As surgery was indicated in our patient, the next dilemma we faced was whether just a biopsy for diagnostic purposes or an attempt to remove the mass, followed in both cases by immunosuppressant discontinuation and/or chemotherapy, would be the most reasonable approach for cases similar to the one we describe. This question may be difficult to answer given the infrequent presentation of LYG and it is consequentially difficult to predict a response by only adopting a conservative strategy, considering the relatively poor knowledge we have about the natural history of the disease as well as the lack of similar cases affecting the skull base. Although there have been sporadic reports of LYG remission after discontinuing immunosuppressants or chemotherapy,<sup>[35]</sup> none of these cases were dealing with lesions located in the skull base. On one hand, in our experience, the feasibility to remove the mass by an experienced neurosurgeon enabled us not only to get enough material for histopathological analysis but also to resolve the problem of mass effect versus relevant neurovascular structures as well as to avoid leaving behind *in situ* a lesion which is known to have the potential of aggressive malignant transformation.<sup>[14,23,28]</sup> On the other hand, biopsy and immunosuppressant discontinuation added or not to chemotherapy and followed by a narrow wait-and-see strategy could be reasonable in case of lesions difficult to remove. In those cases, a strict follow-up with MRI at short-term intervals must be mandatory.

## CONCLUSIONS

The case we present here provides relevant clinical and therapeutic implications. We describe for the first time LYG as a rare but existing differential diagnosis of Meckel's cave and cavernous sinus masses associated with TN. As the perioperative evaluation and treatment may significantly differ between LYG and other tumor and inflammatory entities, we recommend being aware of this disease, especially in patients with a prior history of EBV infection, autoimmune disease, and/or undergoing immunosuppressive therapy. In such patients,

the presence of systemic symptoms as well as signs of pulmonary, skin, kidney, eye, or even intracerebral and leptomeningeal involvement may be helpful to suspect the diagnosis, but their absence does not rule out LYG, as has been observed in our patient. If LYG is suspected, searching for EBV titers may also be useful to orientate the diagnosis, along with thoracic and abdominal CT scans to detect whether other organs, especially the lungs, are affected. However, in all cases, a histopathological examination is mandatory to confirm the diagnosis.

In our case, we have observed that the removal of LYG localized in the Meckel's cave and cavernous sinus may be feasible and safe. Performed by experienced hands, surgery may correlate with transient pain relief. In our patient, discontinuing immunosuppressive therapy was followed by the reduction and stabilization of EBV counts and a prevention of a relapse or progression of the disease. Whether corticosteroids, rituximab, interferon- $\alpha$ -2b, or chemotherapy should be considered in case of recurrent disease is still an open question. We cannot provide any recommendation for the use of these agents based on our experience.

## Ethical standards and conflict of interest

The manuscript does not contain clinical studies or identity patient data and the authors declare that they have no conflict of interest.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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