Unusual presentation of a clinically isolated temporal region mass: Non-Hodgkin lymphoma

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1 | INTRODUCTION

Temporal fossa tumors are a rare entity among head and neck neoplasms. These tumors arise from its various components including muscle, fascia, vessels, bones, nerves, and skin. Temporal bone tumors represent only 0.2% of head and neck cancers, and temporal fossa tumors are not widely found in the English literature.

Eight percent of fine needle aspirates done in the head & neck region are found to be lymphoma. Lymphoma is a malignant proliferation of the immune system cells that is categorized into Hodgkin (HL) and non-Hodgkin lymphoma (NHL). Its clinical presentation may present as nodal, extra-nodal or a combination of both.

Only four cases of temporal fossa NHL have been reported in the English literature. We present a case of NHL presenting as an isolated unilateral temporal soft tissue swelling.

2 | CASE PRESENTATION

Sixty-two-year-old female patient presented to our otolaryngology clinic on November, 2018 with a swelling in the right temporal fossa. The swelling was initially painless but rapidly and progressively increased in size over 2 months, causing pain over the temporal area that radiated to the eye.

Local examination showed a firm, non-mobile, erythematous mass about 4 cm in the right temporal region (Figure 1). The rest of the head and neck examination was unremarkable with no palpable cervical lymphadenopathy.

A non-contrast CT scan of the facial bones showed a subcutaneous mass of intermediate density in the right lateral orbital region measuring 3.2 cm antero-posteriorly, 2 cm in height and 1 cm transversally, surrounded with moderate swelling in the adjacent fat planes. Due to lack of specificity of findings, a brain MRI with gadolinium
followed showing the same mass with intermediate T1/T2 signal with peripheral enhancement and few septations within it (Figure 2).

Complete surgical excision of the 4 × 3 × 1 cm mass and histology findings were consistent with diffuse large B-cell NHL.

A whole body fluorodeoxyglucose (FDG) positron emission tomography-computed tomography study (PET-CT) showed avid bulky adenopathies above and below the diaphragm as well as avid systemic lymphomatous involvement of the palatine tonsils, nasopharynx, lungs, kidney, and adrenals. This revealed an extra-nodal and a nodal involvement of disease.

R-CHOP therapy (Rituximab 650 mg, Cyclophosphamide 1,250 mg, Doxorubicin 80 mg, Vincristine 2 mg, and Prednisolone 60 mg) protocol was initiated for a total of six cycles. An end of therapy FDG-PET CT scan was done revealing no evidence of residual disease. The patient has been placed on protocol surveillance with the oncology team and is disease free up to date.

FIGURE 1 Preoperative imaging of the patient

3 | DISCUSSION

Lymphoma of the head and neck region is the second most common malignancy after squamous cell carcinoma. NHL is more common than HL. The most common histologic type of NHL is diffuse large B cell Lymphoma. NHL of the temporal fossa has rarely been reported in the literature. Other cases of NHL of the temporal bone have been reported, but these were mostly in the middle ear and not in the temporal fossa. The clinical presentation of the differential diagnosis can be used to narrow down possible etiologies before further investigation.

Among the differential diagnosis of temporal fossa tumors are chondrosarcoma, epidermoid cysts, dermoid cyst, temporalis muscle sarcomas, temporalis muscle herniation, myositis ossificans, temporal arteritis, arteriovenous malformations, and metastatic disease.

Dermoid cysts and epidermoid cyst appear in the third and fourth decade and have a similar clinical course. Both are slow growing tumors, round, firm, and mobile.

Dermoid cysts can additionally appear in young children.

Unlike dermoid cyst, epidermoid cyst do not have skin appendages. More than 50% of epidermoid cysts in the head and neck region appear in the upper orbital quadrant. These become symptomatic if inflamed or infected, and can be found in the superficial or deep planes. When they are found in the deep planes, they may be confused with other masses.

Nodular fasciitis is a benign neoplasm of the temporal fascia that presents as a slow growing tumor over several weeks. Seldom, it presents as a rapidly growing tumor that enlarges over hours. Frequent in children secondary to minor trauma or tick bite with Borrelia afzelii. The lesion is embedded in the muscle as opposed to the subcutaneous tissue making the preoperative diagnosis vague.

Temporal bone chondrosarcomas and sarcomas are extremely rare but when they appear, they present as slow growing tumors. Sarcomas may be rapidly progressive depending on the subtype. Children and young adults with Ewing sarcoma of the temporal fossa present with a painful swelling, headaches, and possible facial palsy.

Metastasis to the temporal bone is less common than primary tumors of the temporal bone. Otologic symptoms were the most prominent with hearing loss first in the list. However, symptoms appear when the disease has dispersed widely. According to Gloria-Cruz et al., primary tumors with predilection for the temporal bone included the breast (63%) then the lungs and prostate with the most common type of histopathology being adenocarcinoma.
In our case, it was a lymphoma, which manifested solely as a fast-growing temporal fossa mass without any other systemic signs.

Temporal fossa masses need to undergo extensive assessment, as many may present as the tip of a more serious pathology. Both high resolution CT scan (HRCT) and MRI are needed as HRCT will provide information about bone involvement and MRI will provide detail about soft tissue involvement including meninges, brain parenchyma, and perineural spread. PET CT is required for staging purposes in order to direct therapy, and for assessment post therapy.

Biopsy is essential as histologic type is a major factor determining medical vs. surgical treatment and prognosis.

4 | CONCLUSION

The aim of this case report is to highlight that advanced systemic diseases such NHL can present as an isolated temporal fossa swelling without any other clinical manifestations that may indicate a more severe entity. Therefore, a detailed assessment with a comprehensive clinical evaluation keeping in mind all the possible differential diagnosis, imaging studies, and biopsy of the lesion is essential for proper evaluation, treatment, and patient safety.

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CONFLICTS OF INTEREST
Dr. Mariam Mella, Dr. Fouad Fata & Dr. Jihad Khoury declare no conflict of interest.

AUTHOR CONTRIBUTIONS
Mariam Mella, M.D: Wrote the manuscript, edited and prepared the manuscript, and collected the data. Jihad Khoury, M.D: Edited the manuscript. Fouad Georges Fata, M.D: Was the senior author, wrote and edited the manuscript. Made in Beirut, Lebanon.

ETHICAL APPROVAL
This article was accepted by the International Review Board (IRB) committee of Saint George Hospital University Medical Center.

CONSENT
The patient gave a written consent to the publication of this article.

FIGURE 2 Multiplanar multi-sequential MRI of the brain before and after intravenous gadolinium administration. Subcutaneous right temporal encapsulated lesion, with fairly homogeneous contents showing isointense signal on T1, hypointense signal on T2, and hyperintense signal on diffusion with rim enhancement after intravenous contrast administration.
DATA AVAILABILITY STATEMENT
Data available on request from the authors.

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