

## Case Report

# Primary non-hodgkin lymphoma of lateral skull base mimicking a trigeminal schwannoma: case report

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**Abstract:** Primary extranodal lymphoma is a common malignant tumor of head and neck but rarely presents in lateral skull base. We reported such a case of lymphoma categorized as diffuse large B-cell lymphoma (DLBCL) subtype in a 74-year-old Chinese female. She has experienced an acute course of continuously trigeminal neuralgia and Horner's syndrome. The lesion was diagnosed as trigeminal schwannoma based on symptom and medical image before operation then confirmed to be DLBCL pathologically.

**Keywords:** Lateral skull base, primary extranodal lymphoma, b-cell, trigeminal neuralgia

### Introduction

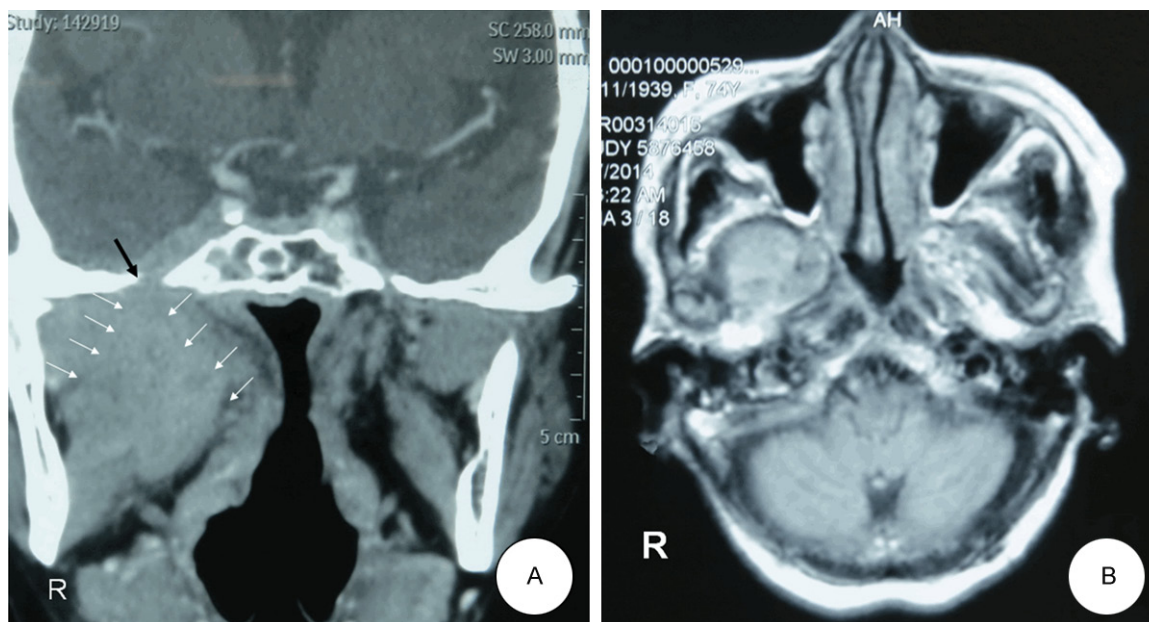
Extranodal non-Hodgkin lymphoma of the head and neck is usually seen in the Waldeyer's ring [1]. Generally, it presents as a slow-growing mass and the patient would be aware of the primary compression symptoms of nearby cranial nerves. Primary non-Hodgkin lymphoma in lateral skull base is extremely rare. It would be misdiagnosed as a neurogenic tumor at first, with whether or not neurological manifestations presented. A few case reports of lymphomas in lateral skull base demonstrated chronic unilateral paralysis of multiple cranial nerves as initial signs [2]. This case offers an acute onset of isolated classic prosopalgia complicated with Horner's syndrome, which implies the entity as nerve sheath tumor [3]. To our best knowledge, the clinical features of this case are extremely rare and there is no similar case description documented in literature.

### Case report

A 74-year-old Chinese woman was admitted to the department of neurology of the West China Hospital in June 2014, complaining of sudden onset of a 20-day history of alternate paroxysmal pain and numbness on the right face. The pain tended to be continuous, knife-like and lightning-touching on the right corner of mouth

and preauricular skin which wouldn't be triggered by any movement of the face. Her appetite exacerbated and her weight lost heavily. Her previous medical history revealed scleroderma for 20 years, hyperglycemia and diabetes for 2 years.

Neurological examination on admission revealed right trigeminal nerve dysfunction. Right facial sensation diminished and the jaw deviated to the right during mouth opening. Right ptosis advanced during the hospitalization period while her eyes movement was not restricted to all directions. No positive signs of other cranial nerves were explored. Magnetic resonance imaging (MRI) and enhanced computed tomography (CT) (**Figure 1A, 1B**) revealed a soft tissue mass with faint boundary in the right lateral skull base with pterygoid muscles, foramen ovale, cavernous sinus and trigeminal nerve involvement. The skull base bone wasn't significantly destroyed. After consultation with specialists in the field of neurosurgery and otolaryngology head and neck surgery, the patient and his family tended to accept a surgical resection through a transcervical approach. Then she received the surgery of resection of the tumor and selective ipsilateral neck dissection. The tumor sized 4.0 × 3.7 cm with naked-eye visualized margin was resected smoothly. The foramen ovale was enlarged and the tri-



**Figure 1.** A. Coronal enhanced CT demonstrating right skull base neoplasm of faint margin (white arrow) connected with thickened trigeminal nerve through enlarged foramen ovale (black arrow). B. Axial T1-weighted MR image demonstrating the right skull base neoplasm pushing the lateral pterygoid muscle inwards.

geminal branche was interrupted. The lateral and medial pterygoid muscle left intact. A few small lymph nodes were collected. Postoperative diagnosis proved to be diffuse large B-cell non-Hodgkin lymphoma for the tumor and reactive hyperplasia of the lymph nodes pathologically (**Figure2A-C**). After surgery, the patient underwent systemic scan for any tumor-like abnormality but got negative results, indicating that the lymphoma was primary in the lateral skull base. The patient was discharged one week after operation with persistent numbness and ptosis in the right face. She gave up standard chemotherapy because she could not tolerate the side effects and was tortured by other systemic chronic diseases. The patient died of multiple organ failure 3 months after operation.

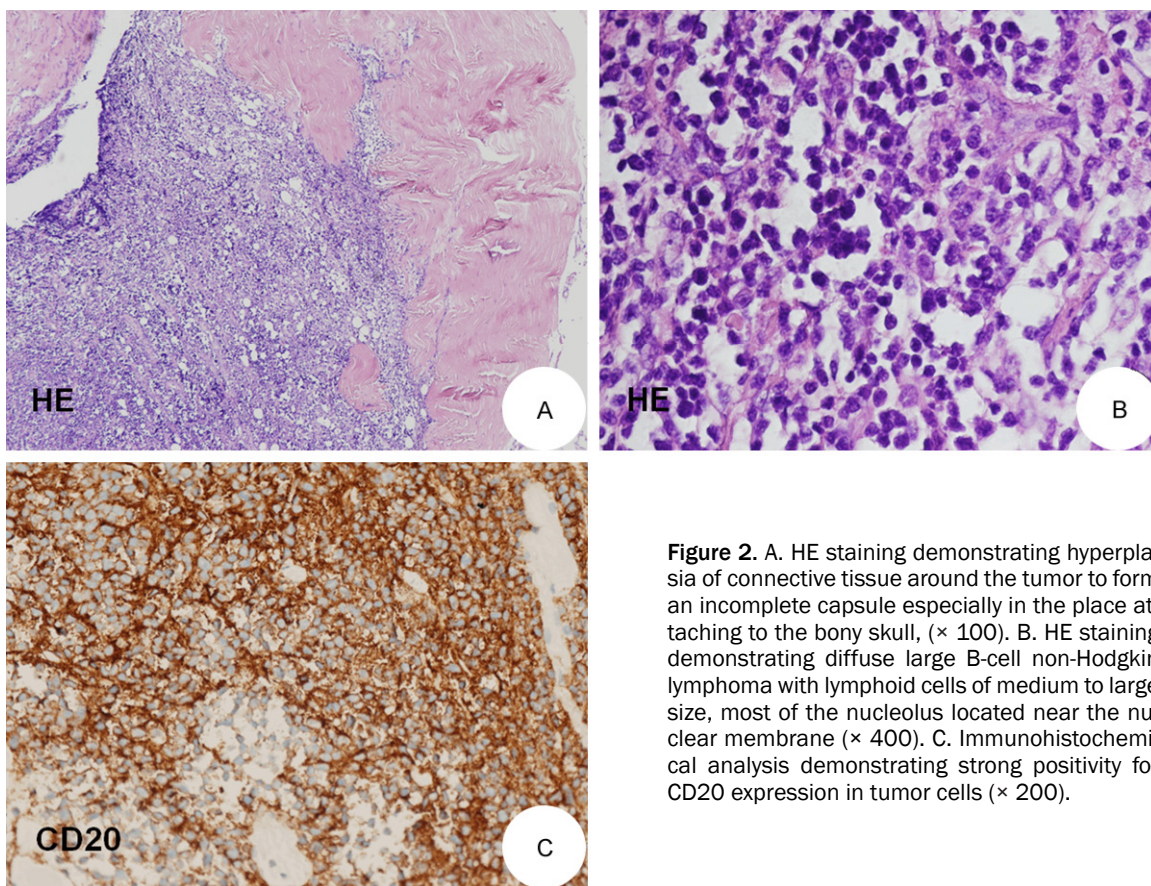
### Discussion

About 20%~25% extranodal lymphomas occur in the head and neck region and most of them are non-Hodgkin lymphomas [1, 4]. Waldeyer's ring is the most common site with more than half of the primary extranodal lymphomas (PEL) of the head and neck take place. A few cases of PEL of skull base were reported sporadically, most of them existed in clivus or sellar region and usual initially presented with diplopia, limited eye movement and hemianopsia due to II,

III, IV and VI cranial nerve involvement [5-8]. There were only three cases PEL of lateral skull base reported in literature, the pathological patterns of them were confirmed as DLBCL without exception [2, 9, 10]. Unlike the PEL in other sites of head and neck, the DLBCL of lateral skull base seems to be more progressive with a poor prognosis though it was not evidently confirmed in this case because the patient died of multiple organ failure [1, 11]. Along with growth of a tumor, all the cranial nerves of the same side would be palsied which is called Garcin syndrome, branches of trigeminal nerve also could be involved.

Apart from cranial nerves paralysis, the imaging of the skull base lymphoma has its some characteristics, it spreads exophytically into the lacuna of skull base rather than infiltrate into the deep tissues and destroy skull base bone, which is the common performance of other types of cancers [12]. In Addition, the MRI of most of lymphomas in head and neck region is similar to that of gray matter of brain on both T1 and T2 with clear boundary which makes it difficultly to differentiate them from neurogenic tumors though exceptions exist [3, 10]. Neurogenic tumors like trigeminal schwannoma, metastatic lesion, meningioma, chordoma, giant cell tumor, myxoma and nasopharyngeal carcinoma of submucosa type should be includ-

## Lymphoma of lateral skull base



**Figure 2.** A. HE staining demonstrating hyperplasia of connective tissue around the tumor to form an incomplete capsule especially in the place attaching to the bony skull, ( $\times 100$ ). B. HE staining demonstrating diffuse large B-cell non-Hodgkin lymphoma with lymphoid cells of medium to large size, most of the nucleolus located near the nuclear membrane ( $\times 400$ ). C. Immunohistochemical analysis demonstrating strong positivity for CD20 expression in tumor cells ( $\times 200$ ).

ed in the differential diagnosis of this kind of skull base tumor [13-15].

In addition to open surgery, fine needle aspiration, ultrasound guided needle biopsy and endoscopic skull base surgery should also be considered for diagnosis in selected cases. As this case indicated, the lesion has a near clear margin for resection, so we recommend a en-block surgery followed with postoperative chemotherapy and radiotherapy [5]. Pathological, immunohistochemical even gene analysis should be guaranteed to meet the need for the treatment measures and prognosis prediction.

### Disclosure of conflict of interest

None.

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