To the Editor: Four patients with diagnosis of malignant peripheral nerve sheath tumor (MPNST) was treated at the Department of Oral and Maxillofacial Surgery (Nanjing Stomatological Hospital, Medical School of Nanjing University) from 2005 to 2020. We reviewed the clinical records and evaluated the patient profile data that consisted of age, sex, clinical symptoms, site of the lesion, surgical approach, rate of recurrence, and survival. In all 4 cases, the diagnosis of MPNST could be confirmed. The most common symptom was rapidly enlarging mass with no nerve palsy associated. One of the patients was a 25-year-old male patient was admitted to the hospital in October 2017 due to “swelling and pain in the left submaxillary region with dysphagia for 2 weeks.” The patient was operated in the local hospital in February 2017 due to neurofibroma of the left neck. In August 2017, the patient was operated in the local hospital again due to the tumor in the submaxillary area of the left mouth. The postoperative pathological diagnosis was MPNST, but no further treatment was taken. The male had a family history of neurofibroma. The patient’s maxillofacial appearance was found to be asymmetric on admission examination, and the left submaxillary and neck could be touched. The swelling is hard, the boundary is not clear and fixed, the size is about 10 × 8 cm. The results of examination showed that the patient’s tongue movement was limited, the tongue was tilted, and there was no numbness in the tongue; the patient’s milk and coffee spots were seen in the general examination, and the imaging results were shown in (Fig. 1); after admission, the patient reviewed the pathological sections of the hospital and combined with the opinion of the pathology department of Fudan University Cancer Hospital, the patient was diagnosed as malignant peripheral neurilemmoma. After examinations, the obvious surgical contraindications were eliminated, and the close relationship between the tumor and the important vessels in the neck was eliminated by enhanced computed tomography (CT). Under general anesthesia, the patients underwent extended resection of the lesions under the left tongue and mouth and submaxillary, transfer and repair of the right anterolateral femoral flap, and tracheotomy (Fig. 2). Postoperative pathological results: confirmed malignant peripheral neurilemmoma. One month after operation, he received radiotherapy and chemotherapy in Jiangsu cancer hospital. At present, the prognosis of follow-up showed no recurrence till now.

MPNST is a kind of rare soft tissue malignant tumor. It has been reported in the literature that there is no significant gender difference. Some patients have a family genetic history of neurofibroma. In our 4 patients, 2 were male and 2 was female. One of the male patients has a family history of neurofibromatosis. Malignant peripheral nerve sheath tumor has a high degree of malignancy and active growth of tumor tissue. Patients often show tissue swelling and pain caused by compression of surrounding tissue. At the same time, the tumor originated from the peripheral nerve, which can be converted into local numbness or dyskinesia. Because of the active growth of the tumor, when the patient has symptoms, the tumor is often large and the boundary is unclear. At the same time, the oral and maxillofacial region is adjacent to the brain, and the neck also has important blood vessels and nerves, which undoubtedly makes it more difficult to control the safety margin of MPNST operation. Because of the active growth of the mass, some necrotic areas showed persistent low-density shadow in enhanced CT. Compared with CT, Magnetic resonance imaging (MRI) showed a clearer boundary of MPNST, and the mass showed high density and mixed components in MRI. MRI examination before operation is of great significance to clarify the tumor invasion boundary.
At present, the treatment of MPNST is still mainly to resect tumor tissue by operation. Because the tumor is closely related to the nerve, special attention should be paid to the resection of the nerve involved in the focus during the operation. It is of great significance for the prognosis and survival of the patients to make sure that there is no residual tumor tissue on the nerve and tumor margin through the frozen pathological examination during the operation. Because this kind of tumor is easy to have distant metastasis and the rate of lymph node metastasis is relatively low, if there is no obvious lymph node enlargement in clinic, neck lymph node dissection is generally not allowed. Radiotherapy and chemotherapy after MPNST surgery can improve the survival rate of patients. However, due to the high invasive and malignant nature of the tumor itself, the recurrence rate is still as high as 50% to 80%. The incidence of lung, bone and liver metastasis was 33%.

The factors influencing the prognosis of MPNST were reported in the literature, including tumor size, local recurrence, distant metastasis, history of neurofibroma and family history. It has been reported that the 5-year survival rate of MPNST patients is 53% without neurofibromatosis, whereas the 5-year survival rate of patients with neurofibromatosis is only 16%. Due to the lack of statistical results of large samples, the prognosis and survival of domestic patients with oral and maxillofacial MPNST remains to be verified. In addition to local tissue swelling, pain and neurological symptoms, MPNST in oral and maxillofacial region can also show swelling and language disorders. Because of its high malignancy, local recurrence and distant metastasis of lung and bone tissue often occur, whereas local lymph node metastasis rate is low, so the cervical lymph node dissection is not usually used in maxillofacial MPNST. At the same time, it is of great significance for the prognosis of patients to resect the tumor completely and ensure enough safe margin. It is of great significance for the prognosis and survival of patients to further radiotherapy and chemotherapy or adopt radiotherapy and chemotherapy when the operation cannot be performed. However, due to the low incidence of the disease itself and the small number of oral and maxillofacial cases reported, the survival rates of patients in different literatures are different. The long-term survival still needs close follow-up in order to further deepen the understanding of the diagnosis and treatment of the disease.

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To the Editor: Navigation is an image-guided technologies used by surgeons to integrate previously acquired magnetic resonance imaging or computed tomography (CT) images with the operative field.1 In the article “Extensive polyostotic craniofacial fibrous dysplasia with optic nerve impingement,” Hu et al2 reported the case for surgical treatment of extensive fibrous dysplasia with the navigator. We report here a similar case and share our experience with the domestic navigator.

The 16-year-old girl had twice operative histories of optic nerve decompression via craniotomy. Physical examination showed normal except for mild left forehead protrusion. Neurological examination showed left blurred vision without extraocular motion limitation. The visual acuity of the left eye revealed 0.2. The visual field test showed unilateral superior quadrantanopia of the left eye. Facial bone CT showed progression of local thickening with the ground-glass appearance of the left orbitofrontal bone and narrowing of the left optic canal due to the surrounding bone expansion.

The patient’s position was identified with a digital reference frame that was fixed rigidly with the intermaxillary fixation method via the dental wire without fastening on the Mayfield clamp (Fig. 1). The new method can avoid complications of cranial fixation devices, including skull fractures, needle infections, venous air embolism, and epidural hematoma.3

The patient was approached via the endoscopic transnasal route for left optic nerve decompression. The Retina navigation system (EPED Corp., Kaohsiung, Taiwan)

FIGURE 1. The digital reference frame was fixed rigidly with intermaxillary fixation method.

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