A Case Report of Granular Cell Tumor of the Left Mandibular Canal

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Key Clinical Message

Granular cell tumor (GCT) is a rare benign, soft tissue neoplasm. GCT rarely occurs in the jaw. Here we report the case of a GCT of the left mandibular canal. The patient exhibited a favorable postoperative recovery and remained free of recurrence throughout the 3-year follow-up period.

Key words : Case report, Granular cell tumor, Abrikossoff's tumor, Mandibular canal

Introduction

Granular cell tumor (GCT) is a rare tumor observed in the clinic. GCT was first described in 1926 by Abrikossoff and therefore, also called Abrikossoff's tumor [1]. GCT can involve any part of the body, but in most cases, it localizes in the head-and-neck region, especially the tongue [2]. No previous study of GCT occurring in the mandibular canal has been reported. Thus far, the histogenesis and pathological nature of GCT are controversial. Based on immunohistochemical labeling and ultrastructural studies in recent years, most scholars believe that this tumor originates from Schwann cells of the nerve sheath. Herein, we report the case of a patient with GCT of the left mandibular canal.

Case History

A 62-year-old female presented at our department with swelling, pain, and numbness in the left mandible for more than 4 years. The patient had a history of hypertension and diabetes for many years. On clinical examination, the patient had hard, ill-defined, and painful distension at the left mouth angle. The skin in the region was pigmented, the left lower lip and chin skin were mildly numb, and the buccal cavity and shape were not obviously abnormal (Figure 1). An intra-oral examination revealed a nodular, poorly defined, hard, tender, sessile lesion, with texture similar to that of the adjacent mucosa. The nodule (1.5 cm in diameter) was located on the buccal region of the first and second left mandibular premolars (Figure 2). The crowns of the left mandibular first and second premolars were intact, painless, and showed no loosening. No significant enlarged lymph nodes were found in the neck.

Cone-beam computed tomography (CBCT) scans showed an enlarged left mandibular canal and a markedly distended mental foramen (Figure 3). A contrast-enhanced CT scan showed a locally widened left mandibular

tube, with locally convex and a slightly enhanced soft tissue density shadow in the corresponding area (Figure 4).

Differential Diagnosis, Investigations and Treatment

In this case, diagnosis is difficult. Squamous cell carcinoma of the jaw was considered at first impression because the swelling, pain and numbness in the left mandible for more than 4 years. However, according to the imaging results, the lesion developed along the mandibular canal with clear boundaries, so we believed that the lesion was a neurogenic tumor with high probability of benign. In maxillofacial region, the most common neurogenic tumors are schwannoma and neurofibroma. But the clinical and imaging findings of this case did not support these two diagnoses. To determine the nature of the lesion, we performed a fine-needle aspiration biopsy, but the puncture was not successful, so we decided to proceed directly to exploratory excision.

The tumor was totally excised under general anesthesia. During the operation, the tumor was noted along the long axis of the inferior alveolar nerve; appeared white; and had an intact capsule, cystic cavity, and milky cloudy fluid; the inferior alveolar nerve had denatured and could not be distinguished from the mass (Figure 5). The tumor and the adjacent nerve were completely excised during the operation.

Intraoperative frozen sections were obtained for pathological examination. The pathological examination indicated a cystic wall composed of fibrous connective tissue aggregated with a large number of histiocytic and cytoplasm-rich cells (Figure 6). Postoperative immunohistochemical pathology results showed cytokeratin (CK) (-), CD68 (focal +), CD163 (focal +), Ki-67 (+ <2%), S-100 (+), CD56 (+), synaptophysin (Syn) (-/+), SRY-related HMG-box (SOX)-10 (+), neuron-specific enolase (NSE) (-/+), glial fibrillary acidic protein (GFAP) (-), neurofilament (NF) (partial +), and ETS-related gene (ERG) (-) (Figure 7). Immunohistochemical analysis showed the GCT with slightly irregular nuclei surrounded by a few nerve fibers.

Outcome and follow-up

The patient was followed up 2 weeks after the surgery, followed by telephonic follow-ups at 6 months, 1 year, and every year postoperatively. Thus far, it has been more than 3 years and no sign of recurrence has been noted, but the skin numbness of the left lower lip and chin persisted.

Discussion

GCT is rare in the clinic and can occur in various parts of the human body. More than 50% of the reported cases have occurred in the head and neck, and the tongue is the most frequently involved anatomic site, followed by skin and subcutaneous tissue, breast, respiratory tract, and digestive tract. GCT can occur in patients at any age, but is more common during the fourth to the sixth decades of life and is rare in children [3]. Very few cases of earlier presentation (younger than 20 years old) have been described [4]. GCTs are twice as common in women as in men, and its incidence is more common in the African-American ethnic groups than Caucasians [2]. Familial cases have rarely been reported [3]. Most GCTs are well-circumscribed solitary, asymptomatic nodules that grow gradually [5], but multiple GCTs have also been reported, with an incidence ranging from 3.4% to 20% [1].

GCTs are not easy to detect because they are asymptomatic without swelling and pain or neurological symptoms, and the diagnosis cannot be made only by clinical symptoms and therefore, biopsy or fine-needle aspiration or other pathological examinations may be required [5]. Macroscopic findings showed that the mass is round or oval, generally no more than 2 cm. The capsule is incomplete without a normal film. The texture is hard, and the mass is generally gray yellow; its section may be pale yellow, milky white, or gray white. Under light microscopy, round cells are observed with rich cytoplasm that may have abundant eosinophilic granules. The nucleus is small and round, uniform, centered, and occasionally deviated. The tumor cells are closely arranged in nests or cords, and occasionally striated muscle fibers are closely associated with granulosa cells. Under electron microscopy, the cytoplasm was filled with compound lysosome membrane package with different sizes and shapes. And more than 30% of the cases of tumor surface epithelium are pseudoepitheliomatous hyperplasia [6]. Sometimes, the mass is associated with keratosis because its

morphology is extremely similar with the nest of early invasive squamous cell carcinoma; if neoplastic granular cells beneath the epithelial are not noticed, it may be easy to misdiagnose a GCT as squamous cell carcinoma [7]. Immunohistochemical staining showed strong positive expression of S-100 protein, NSE, and vimentin in tumor cells, and negative expression of NF and GFAP [8]. However, the diagnosis of GCT is mainly based on the typical manifestations of tumor cells under light microscope. Immunohistochemical pathology is often used to distinguish from other tumors.

Most GCTs are benign in clinical behavior and histomorphology, but malignant cases have also been reported. The incidence of malignant GCT is 2%-3%, and such malignant GCTs are most commonly seen in the chest wall, followed by the thigh [5]. Benign GCTs tend to invade surrounding tissues and nerves and therefore, local invasiveness is not the key point in the differentiation between benign and malignant GCTs. In 1998, Fanburg-Smith et al. proposed for the first time that histological features of soft tissue GCT could be benign, malignant, and cell atypia, according to the following six factors: necrosis, spindle tumor cells, vesicular nuclei with distinct nucleoli, the nucleoplasmic ratio was greater than 2:1, mitotic figures > 2/10HP, and cell pleomorphism. GCTs with three or more items are considered as malignant, those with two items are considered as cell atypia, and the rest are considered as benign [1,9].

Benign GCTs are usually locally resected along with the surrounding 1 cm of normal tissue. Tumor recurrence usually occurs within 5 years, and a few recurrences are due to multicentric growth rather than incomplete resection of the primary tumor [6]. The prognosis is generally good, and the malignant transformation rate is low [6]. Malignant GCTs are prone to local recurrence and metastasis, and the most common metastatic sites are regional lymph nodes, lung, and bone. Clinically, large tumor size, rapid growth, and rapid recurrence also suggest the possibility of malignancy [10]. Some studies have shown that chemotherapy and radiotherapy cannot significantly improve the clinical course of malignant GCTs. Extensive local resection and regional lymph node dissection have been performed when necessary [8]. Close follow-up should be performed after the operation.

In this case, clinical and imaging diagnosis were difficult, and diagnosis was confirmed on postoperative pathological results. However, the lesion in this case had a complete capsule, which was contrary to the incomplete capsule without normal film reported in previous cases [7]. In our case, the tumor and the peripheral denatured nerve were completely removed during the operation, and no signs of recurrence were observed up to the 3-year follow-up.

Conclusion

GCT is rare in the clinic. The clinical manifestations and imaging examination for GCT are not specific, and thus, it is easy to misdiagnose. Generally, GCTs are benign, and the prognosis of surgical resection is good, with less recurrence, but a small number of GCTs can be malignant. Close follow-up should be performed after the operation. Similar cases are rare, and few related literature have been reported. The diagnosis, treatment, and prognosis evaluation of GCT should be further studied.

Author contributions

Xing-Hong Zhou: Conceptualization; data curation; formal analysis; writing-original draft. **Zheng-Da Zhu:** Data curation; investigation. **Yue Yan:** Data curation; investigation.**Rui-Juan Zhang:** Supervision. **Shi-Min Chang:** Conceptualization; supervision; writing-review and editing.

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Conflict of interest statement

The authors declare that they have no competing interests.

Data availability statement

The data are not publicly available due to privacy of the patient.

Ethics statement

The study was approved by the ethics review board of Beijing Friendship Hospital, Capital Medical University (BFHHZS20230050). We have obtained written informed consent from patient before beginning of the study.

Consent

The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient.

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Images

Figure 1: A photograph of the left part of the chin showing pigmentation on the surface skin at the left mouth angle.

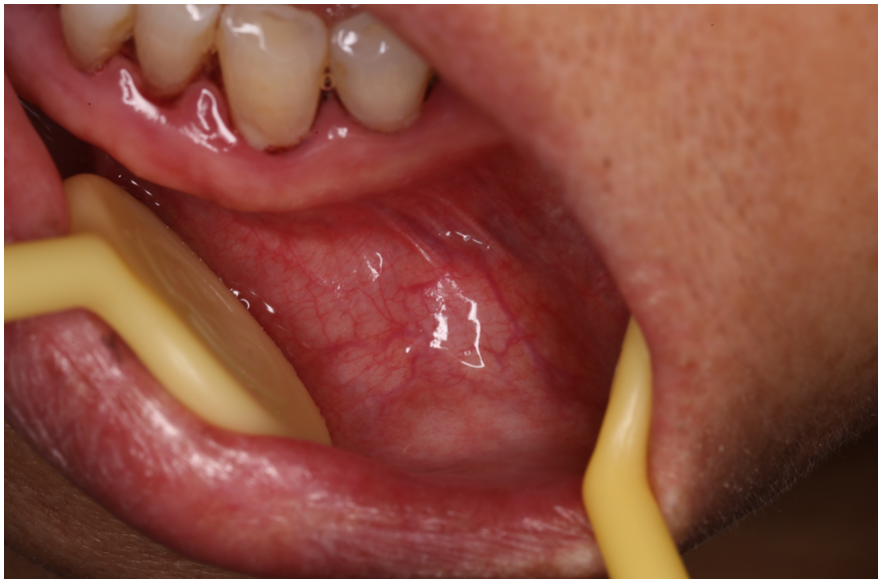
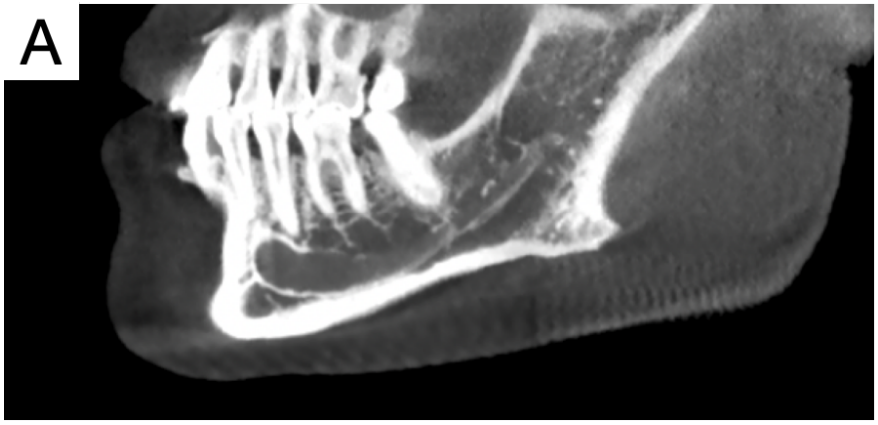


Figure 2: A photograph of inside of the buccal cavity showing a nodule of the first and second left mandibular premolars.



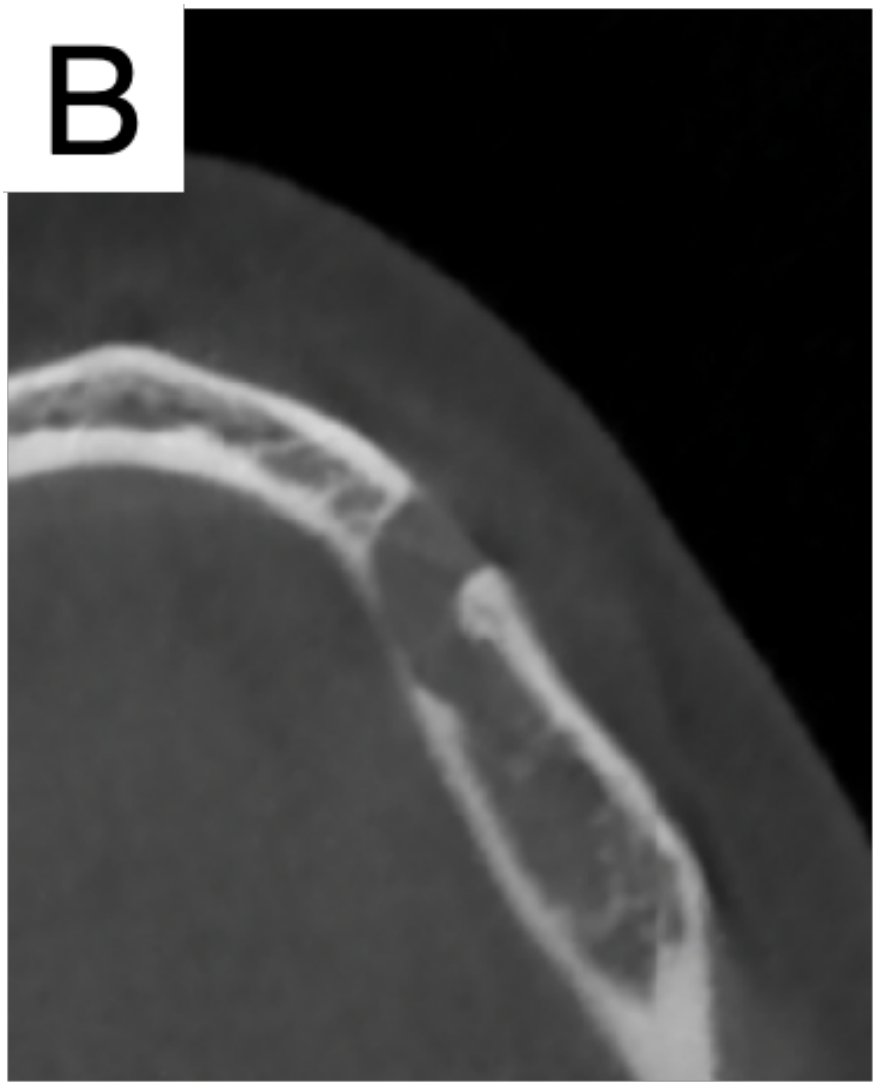


Figure 3: A cone beam computed tomography scan showing (A) an enlarged left mandibular tube and (B) a markedly distended mental foramen.

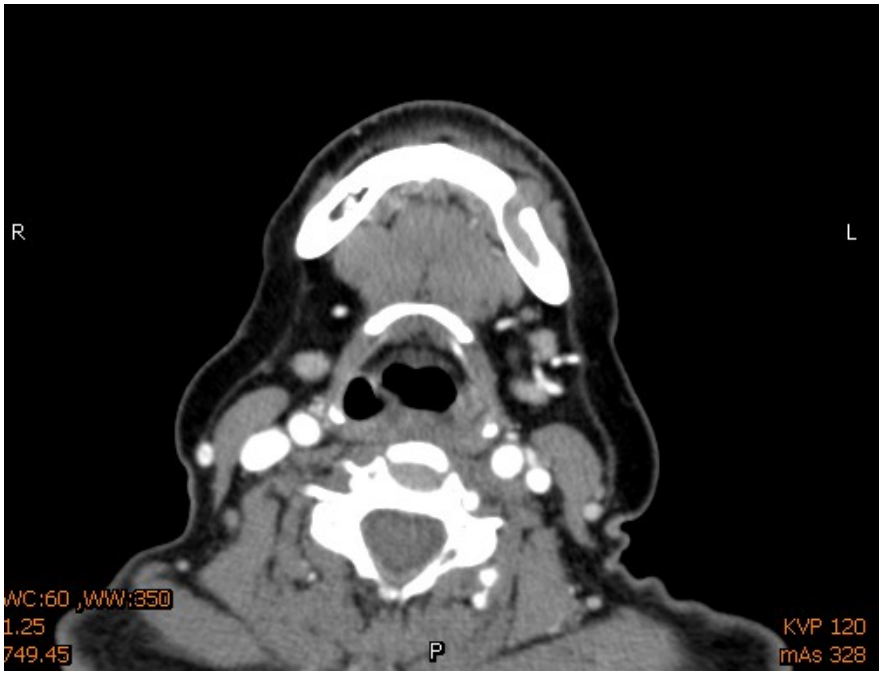
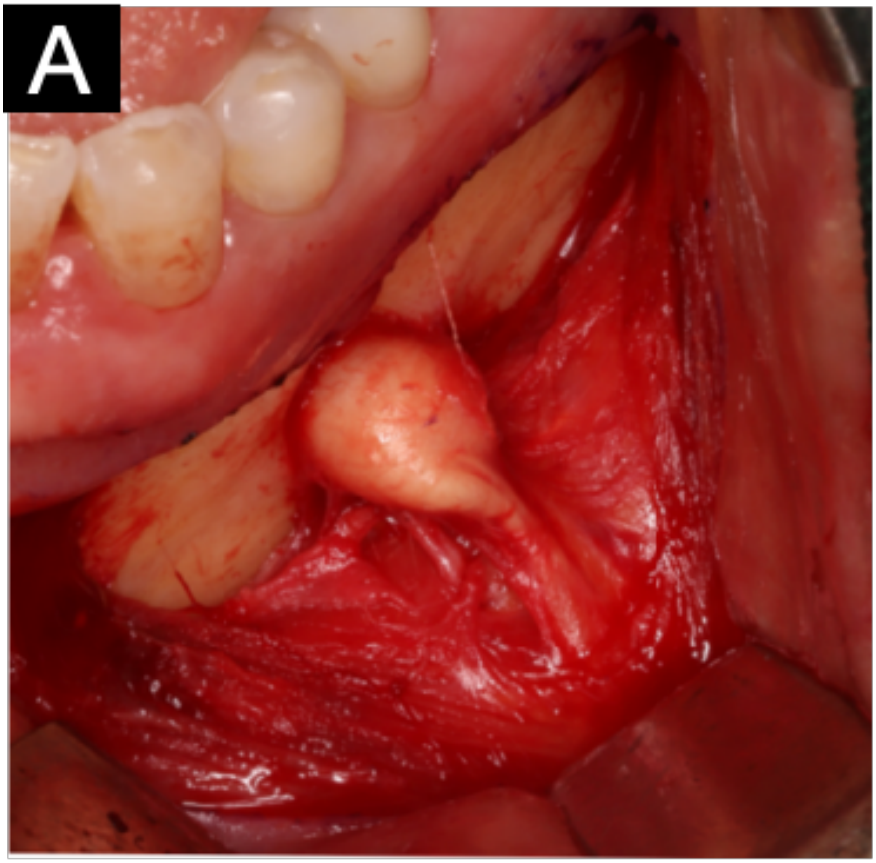
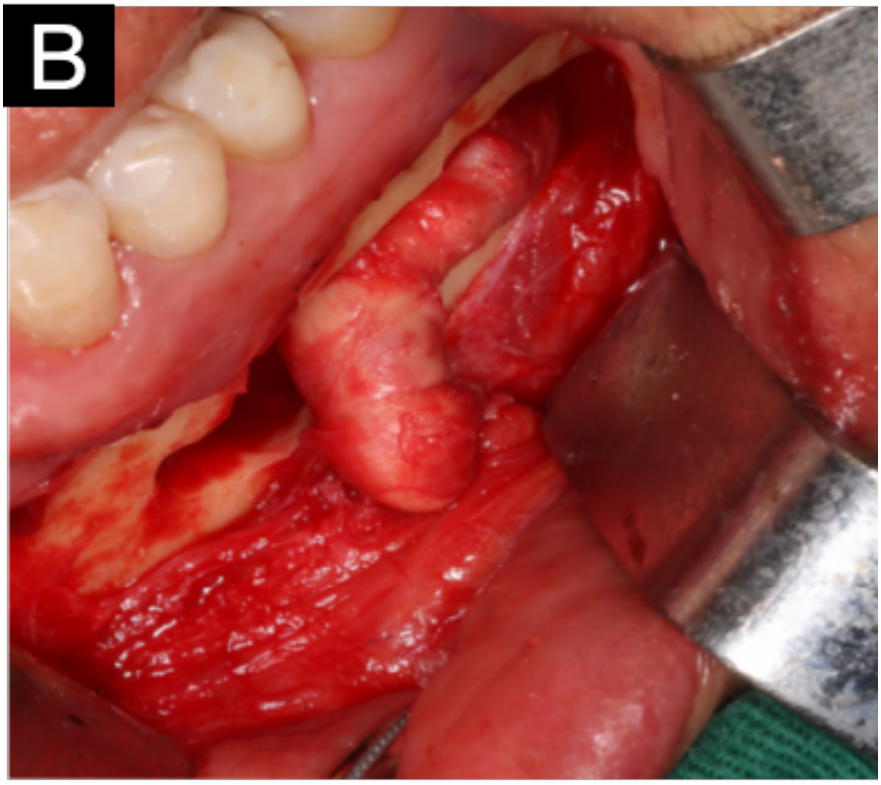


Figure 4: A contrast-enhanced computed tomography scan showing a locally widened left mandibular tube, with locally convex and a slightly enhanced soft tissue density shadow in the corresponding area.





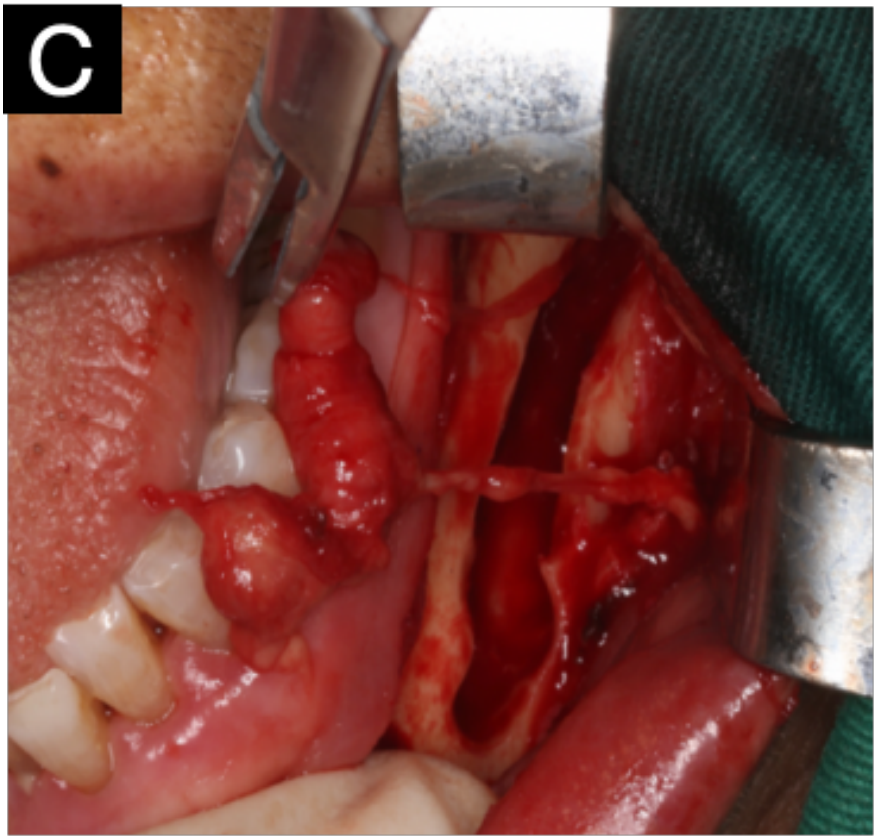


Figure 5: Intraoperative photographs showing (A) swelling in the mental foramen and (B) and (C) showing the mass along with nerve.

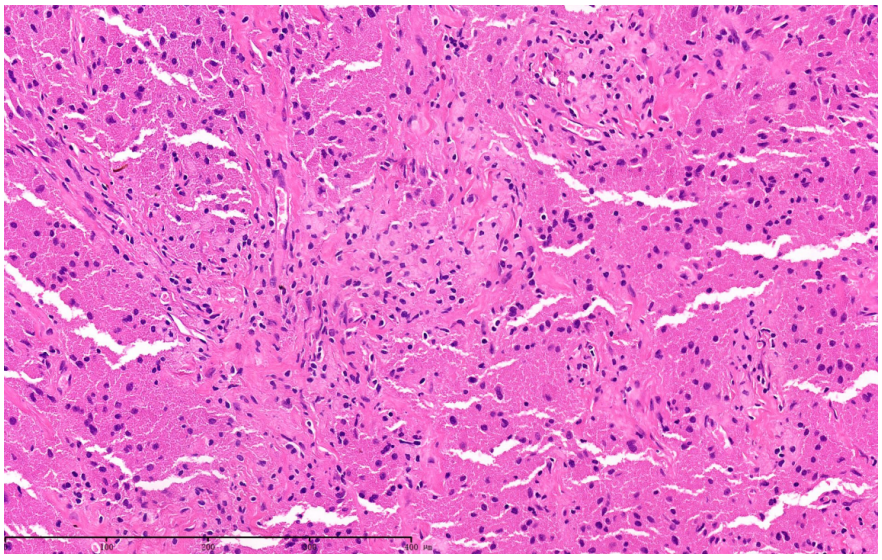


Figure 6: Photomicrograph showing the tumor cells were polygonal or round, with abundant cytoplasm and abundant eosinophilic particles (hematoxylin and eosin staining, magnification, ×20).

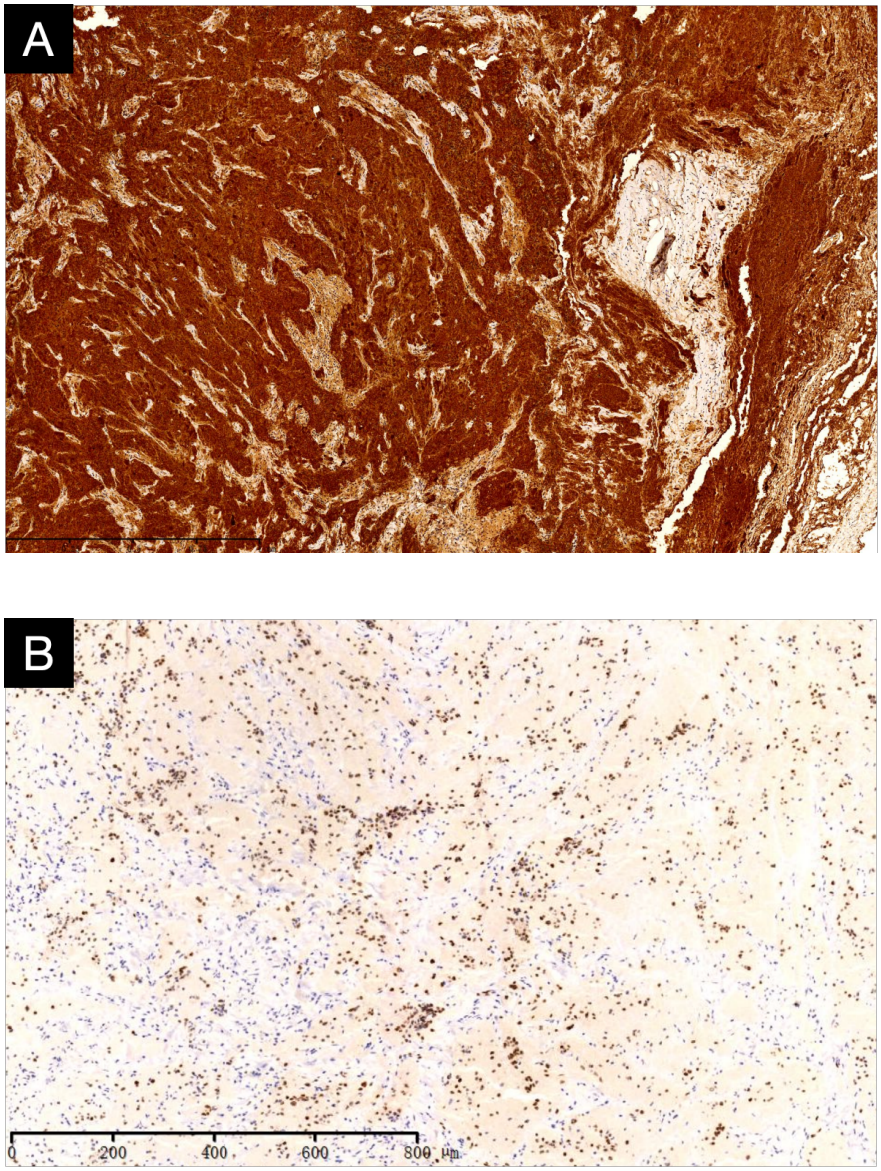


Figure 7: Immunohistochemical staining showing positive expression of S-100 (A) and SOX-10 (B) (magnification, ×5).