Xanthogranuloma of the sellar region diagnosed by frozen section

1 Introduction

The incidence of xanthogranuloma (XG) of the sellar region is quite rare. Jung’s report showed that only 2 patients were diagnosed as xanthogranuloma of the sellar region in 159 patients with an intrasellar or parasellar lesion [1]. And Hermann’s research indicated that 5-year overall survival (OS) and 5-year event-free survival (EFS) were 1.00±0.0 in 14 patients who suffered xanthogranulomas of the sellar region [2]. Xanthogranuloma of the sellar region can cause pituitary dysfunction or visual change because of compression of the optic chiasm [3]. Xanthogranuloma (XG) of the sellar region was first described and classified by Paulus as a distinct entity in 1999, which was different from classical adamantinomatous craniopharyngioma [4]. In 2000, the World Health Organization (WHO) accepted this disease as a specific brain tumor classification [4-5]. However, the 2007 WHO Classification of Tumors of the Central Nervous System mentioned xanthogranulomas of the sellar region as a clinicopathologically distinct entity that “is not yet fully defined [6].” Subsequently, sellar xanthogranuloma was mentioned in the 3rd and 4th editions of the WHO classification of central nervous system tumors [5-6]. Xanthogranulomas of the sellar region display obvious histological characteristics including cholesterol clefts, macrophages, chronic-inflammatory infiltrates, necrotic debris, multinucleated giant cells, and hemosiderin deposits [1,4,7-9]. When we searched PubMed with the terms “Xanthogranuloma and sellar region”, only 18 papers were retrieved which covered 82 patients including 54 patients from Germany, 17 from Japan, 4 from China, 3 from USA, 1 from Croatia, 1 from Taiwan, 1 from India, and 1 from Tunisia. Although xanthogranulomas of the sellar region has been mentioned in some reports, however, we reported a case of xanthogranuloma in the sellar region from China, which was diagnosed by intraoperative frozen section and postoperative pathology, after the mass had been removed by surgery.
2 Case report

A 43-year-old Chinese woman presented with a history of headache and extended menstrual cycles over the course of the past half year. On clinical examination, the patient had obvious upper visual-field defects. Initial laboratory examinations were conducted to detect hormone status and the result showed that the patient’s prolactin levels were higher than normal (750 mIU/L, range 109.8 to 562.4 mIU/L). MRI results showed a clearly defined sellar lesion with a diameter of 16 × 14 × 16 mm. A high intensity zone was observed in the grey matter of the lesion (Fig. 1A-E) and non-homogeneous contrast enhancement images were discovered (Fig. 1F-G). Our preoperative diagnosis was a prolactin-producing adenoma associated with hemorrhage.

The lesion was completely removed by surgical exploration. Pathological examination result showed that the solid mass was friable and brown to yellow-green. Intraoperative frozen sections showed that the tumor was composed of small epithelial cells with foamy macrophage and lymphoid infiltration, a portion was undergoing necrosis; hemosiderin deposits and cholesterol clefts were also observed (Fig. 2A&B). Thus, the patient was diagnosed as having a xanthogranuloma by intraoperative pathologic examination. Histologic examination of the mass revealed distinct granulomatous inflammation consisting of macrophages and hemosiderin-laden macrophages with a few lymphocytes and plasma cells admixed with large regions of cholesterol cleft deposition and fibrin (Fig. 2C-G).

Immunohistochemical analysis of the lesion tissue was performed; the results revealed that there were a few small and low cuboidal epithelium, showed positive immunoreactivity on staining for cytokeratin (Fig. 2H) and negative expressions of prolactin (Fig. 2I). After surgery, the patient’s visual field defects and headache were relieved. The patient’s post-operation MRI confirmed that the mass had been completely removed and no evidence of tumor remnants was revealed (Fig. 1H&I). Three months later, the patient’s prolactin levels continued to drop to the normal range and recurrence was not discovered at 36 months follow-up.

Ethical approval: The research related to human use complied with all the relevant national regulations, institutional policies, and in accordance with the tenets of the Helsinki Declaration. The report was approved by the authors’ institutional review board or equivalent committee.
**Informed consent:** Informed consent was obtained from the patient before submitting this report.

### 3 Discussion

Xanthogranuloma (XG) has been reported in both humans and domestic animals [10-12]. The duration of symptoms has been noted in some records which is 18 months on average (range: 1-96 months) in earlier patients of XG [2], which is similar to the duration of symptoms reported in this case. Masses which occur in the sellar region result in various clinical signs. Headache has been the most frequently recorded initial symptom in 63% of XG patients, and approximately 15% of patients had visual impairment due to compression of the visual pathway [2]. In this case, the patient had a history of headache and initial signs of upper visual-field defects. Furthermore, there was a report that the patient of xanthogranuloma presented the disturbance of consciousness which might be associated with hydrocephalus [3,13]. Although a sellar location of this disease is characteristic, suprasellar xanthogranulomas have been noted in a few case reports [14]. For the diagnosis of xanthogranuloma, there has been no typical imaging criteria defined until recently. In this patient's case, the sellar mass had the volume of 3.584 cm³, less than 3.9 cm³, which is the average volume of xanthogranulomas reported in the literature [14]. The typical pathologic features of xanthogranuloma were observed in this case. Surgical resection is the best treatment measure for these lesions. Recurrences after complete resection are very rare [2], but have been known to occur. Because xanthogranulomas of the sellar region are rare, their nature and clinical course remain somewhat unclear. To the best of our knowledge, neither sellar lesions nor suprasellar lesions have been described by frozen section during a surgical operation. This is the first report of a xanthogranuloma diagnosed by frozen section.

### 4 Conclusion

Because xanthogranulomas of the sellar region is uncommon, the nature of the disease and its clinical course remain unclear to some degree. Thus, further reports are essential in order for clinicians to gain greater insight into the clinical course and prognosis regarding xanthogranulomas of the sellar region.

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