Management and prognosis of rare tumors in the sellar region

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Abstract: Six rare cases of sellar region treated by microsurgery from Jan 2000 to Jan 2010 were reviewed to study the management and prognosis of rare tumors in the sellar region. Subsequent treatments were according to the instruction of preoperative alpha fetal protein (AFP) and human chorionic gonadotropin (HCG) measurement as well as confirmed histopathological examination in all patients. Variety of histological types were observed in present series, which include leiomyosarcoma, malignant yolk sac tumor, mixed germ cell tumor, embryonal carcinoma, pilocytic astrocytoma and fungal pseudotumor, the contents of AFP and HCG were elevated to some extent in patients with malignant yolk sac tumor or mixed germ cell tumor or embryonal carcinoma, follow-up was conducted in all patients varied from 1 month to 3 years, patients with malignant yolk sac tumor and embryonal carcinoma as well as leiomyosarcoma died in 5 , 6, 10 months after operation. Conclusions: rare nongerminomatous malignant germ cell tumors were predominantly susceptible to the sellar region, Furthermore, High misdiagnose and poor prognosis occurred in present study, dynamic AFP and HCG detection play an important role in nongerminomatous malignant germ cell tumors located in sellar region, awareness of present rare lesions of sellar region is emphasized.

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1. Introduction

The sellar region is one which of spots tumor most often occurs in Neurosurgery. The common pathological change includes: Pituitary tumor, craniopharyngioma, saddle tubercle meningioma, reproduction cytoma, optic nerve glioma, hypothalamic hamartoma and so on. Halbauer DJ (2003) firstly reported 3 examples rare tumor involved in sellar region, that's the ganglion cell differentiation pituitary adenoma, intracranial granular cell tumor and germinoma in hypophysial fossa. Few of reports were seen about this after that. The author collects 6 examples of rare tumor involved in sellar region after the pathology confirmation from Jan 2000 to Jan 2010. All of these were misdiagnosed completely before operation. We review and analyze these examples in this study in order to improve recognition to the complication.

2. Material and Methods

A total of six cases with rare tumors involved in sellar region were identified. This group included four males and two females between the ages of 13 and 23 years (mean 23 years). The duration of initial manifestations until diagnosis ranges from 15 days and 2 years. The symptoms were the binocular vision carries on the drop, drink, the polyuria (1/6); headache, simple eye vision descender (2/6); headache, disgusting, vomiting, menstruation anomalous (1/6); left eyelid sagging, ocular movement barrier after pituitary tumor technique latter 14 years(1/6); headache, low heat , say a word unclearly (1/6). The fundus examination demonstration regards the nipple edge clearly to wither palely were seen in four cases; binocular field of vision nie side damage in two cases; oculomotor nerve paralysis in one cases. The diameter of tumors were 3~6.5cm(mean 4.5cm.).

CT scan showed: the saddle area equilibrium density, promiscuous density's perch venereal diseases change. The signal intensity of tumors in suprasellar region was iso- or hyperintense in T1-weighted imaging, whereas it was usually hyperintensity in T2 weighting imaging. On the enhanced images, there was contrast enhancement. Pouch changes, the necrosis signal were in three cases. The tumors downward cerebral ganglion, the fossa interpeduncularis, three ventricles of the brain direction grows, varying degree hydrocephalus were in two cases. And the mixing property reproduction cytoma presents the honeycomb-shaped enhancement; The sellar region smooth myosarcoma enhanced scanning demonstrated enhanced evenly. They were infiltrative growth obviously to cabernous sinus, and circumvolutioed the internal carotid artery. For lack of specificity of preoperative imaging , it is easily to misdiagnosed as Invasive pituitary adenoma. Inflammatory pseudotumor of fungal was homogeneous enhanced after enhancement, and extensive destruction of tumor invasion to the

sphenoid sinus, ethmoid sinus and the cavernous sinus and skull base and other parts of the former in the extensive growth. Preoperative tumor markers in 3 cases: embryonal carcinoma, yolk sac tumor and mixed germ cell tumor patients were AFP: 71.4ng/ml (0-20) and HCG: 189.70mIU/ml (0-5), AFP 105 ng / ml and HCG: 12.1 mIU / ml, AFP: 4.45ng/ml and HCG: 53.8mIU/ml.

Table 1： Rare sellar lesions detailed clinical information

	Age	Gender	Complaints	Preoperative diagnosis	Pathology	Follow-up
1	20y	male	Headache Decreased vision	goniolma	malignant yolk sac tumor	Died after operation 5 months later
2	13y	male	Decreased vision Diabetes insipidus	craniopharyngioma	mixed germ cell tumor	recurrence has not yet
3	54y	female	Decreased vision Right ptosis	hypophysoma	leiomyosarcoma	Died after operation 10 months later
4	16y	male	Decreased vision Headache	craniopharyngioma	pilocytic astrocytoma	good
5	15y	female	Headache Irregular menstruation	Invasive pituitary adenoma	Embryonal carcinoma	Died after operation 6 months later
6	21y	male	Low heat Headache Slurred speech	Meningioma	Inflammatory pseudotumor of fungal	SAH after operation 2 months later

All patients were treated by microsurgery. Pterion approach was utilized in three cases. Incision starting from the right side of the zygomatic arch before the tragus 1cm, The midline 1~2cm, the dura surrounding the sphenoid ridge arc of cut, coagulation butterfly cut to the top of sinus drainage draining veins, and then open the side of the cistern arachnoid slow release of CSF, and then application of section II, III gap block removal of lesions under the microscope. Subfrontal approach was utilized in two cases. The arc incision is in the right amount of hair over the center line. Then turn the pedicled bone flap about 6x7cm to the temporal bone of skull base side. When the tension of the dural is not high, the CSF from the lateral fissure is released. The frontal lobes are gently elevated. Focus is then removed from I gap. Lamina terminalis approach was utilized in one case. The incision is same to the subfrontal approach. For the I gap is small and the prefixed optic chiasm, after reveal the bilateral optic tract and lamina terminalis, then cut the lamina terminalis longitudinally shows soft taupe tumor in the third ventricle which blood supply amply. And the tumor was removed block under a microscope.

3. Results

6 patients in this group were all misdiagnosed before operation, 2 cases of imaging and clinical diagnosis of craniopharyngioma, the pathological were mixed germ cell tumor and pilocytic astrocytoma; two cases of imaging diagnosis pituitary tumor, 1 case of previous surgery and pathology confirmed the pathological smooth muscle sarcoma, and embryonal carcinoma; 1 patients with a preoperative diagnosis of germ cell tumors and experimental radiotherapy given week, review head CT and MRI showed hydrocephalus increased with increasing tumor After microsurgery, pathology of malignant yolk sac tumor. 1 case of consideration for the huge space-occupying meningioma saddle, the pathological diagnosis of fungal inflammatory pseudotumor (Table 1).

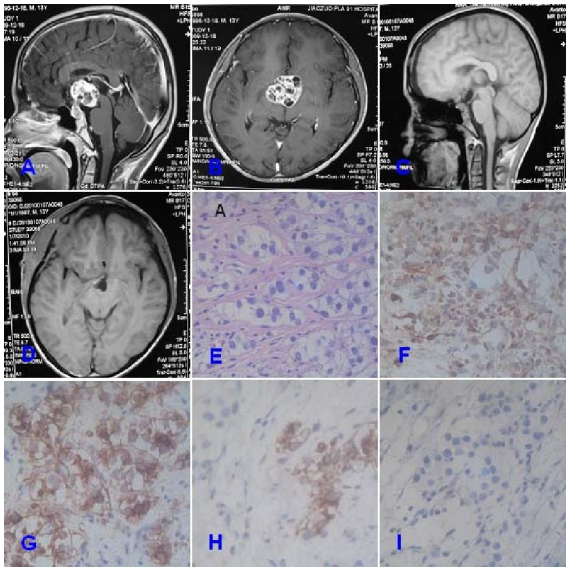

Fig 1: mixed germ cell tumor of the skull sagittal and axial MRI scan shows sellar lesions enhanced irregular enhancement, cyst containing necrotic tissue (FigA, B), review of surgery MRI showed total removal of the tumor (FigC , D), a large number of conventional HE staining of tumor cells (FigE), strongly positive immunohistochemical expression of HCG (Fig F), PLAP strong expression (Fig G), AFP weak expression (Fig H), GFAP-negative expression (Fig I) (Fig E-I x 400)

Total resection was performed in two patients under endoscopic, subtotal resection in four patients. The preoperative detection of AFP and HCG were in three patients. Embryonal carcinoma was significantly higher AFP and HCG, AFP only yolk sac tumor and mixed germ cell tumor patients only HCG significantly increased tumor markers, after reviewing the corresponding decline in varying degrees; immunohistochemistry showed embryonal carcinoma GFAP (-), EMA (-), S-100 (-), PLAP (+), AFP weak (+) , HCG (+); yolk sac tumor of AFP

(++), PLAP (++), Ki 67 (+), P53 (+) GFAP (-), EMA (-); mixed germ cell tumor of HCG (++), PLAP (++), AFP weak (++), GFAP (-) [Fig 1]. Leiomyosarcoma of the actin SMA (+), MSA (+), Vimention (+) [Fig 2]; fungal inflammatory pseudotumor of the acid-fast staining and silver staining were six ammonium (-), PAS staining showed a small number of fungal spores [Fig 3].

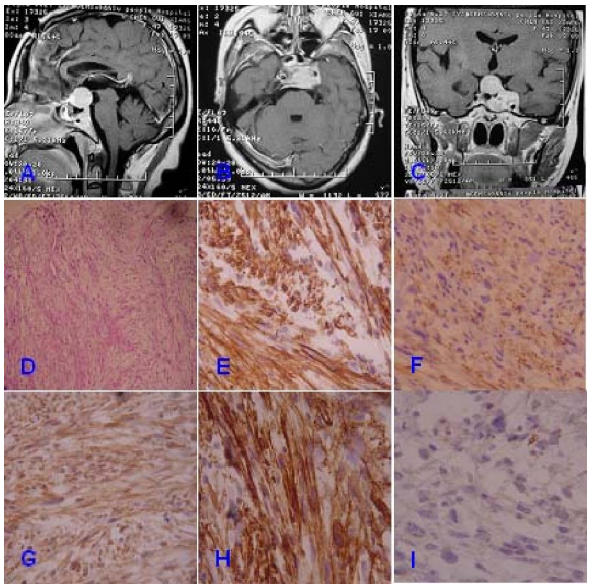

Fig 2: leiomyosarcoma in sellar region head MRI, axial and coronal significantly enhanced scan shows homogeneous enhancement sellar lesion, the direction of invasion to the cavernous sinus growth, particularly in the left cavernous sinus and carotid artery were surrounded tumor (Fig AC). A large number of conventional HE staining of tumor composed of spindle smooth muscle cells (Fig D), immunohistochemistry smooth muscle actin antibody SMA (Fig E), MSA strong expression (Fig F), Vimention was strong expression (Fig G), cerebral aneurysms SMA positive control of vascular smooth muscle (Fig H), SMA negative control (Fig I)

4 cases of postoperative diabetes insipidus, electrolyte disturbance in 3 cases, transient oculomotor nerve palsy in 1 case, malignant pathology after 2 weeks for comprehensive chemotherapy with Nemo Sting, DDP, etc. The course was from 4 to 6 months. And they were given radiotherapy after 1 month. The duration of follow-up ranged from 1 month to 3 years. The patients with malignant yolk sac tumor, embryonal carcinoma, and leiomyosarcoma respectively dead after May, June and October. The patients with inflammatory pseudotumor appeared SAH after 2 months. Then secondary MRA showed left side of the vertebral artery fusiform aneurysm, the current ventilator to maintain breathing in a critical condition. The patient with mixed germ cell tumor is still under close follow-up. No recurrence of this tumor was found. The patients with pilocytic astrocytoma were followed up for good now.

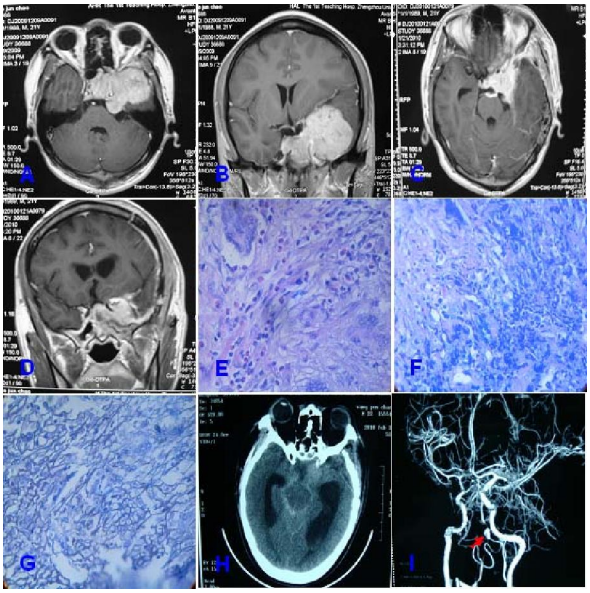

Fig 3: Inflammatory pseudotumor in sellar region Head axial and coronal MRI scan shows sellar enhanced lesions were irregular enhancement huge, invasion, sphenoid, ethmoid and move forward in the cavernous sinus and skull base growth (FigA, B), review of surgery MRI showed the tumor remnant (FigC, D), a large number of conventional HE staining lymphocytes, plasma cells and eosinophils (FigE), acid-fast staining negative (FigF), PAS staining showed a small number of fungal hyphae and spores (FigG), 2 months after the head CT showed SAH (FigH), MRA showed a left vertebral artery fusiform aneurysm (Fig I, arrow)

4. Discussions

Currently the most common pathological includes: sellar pituitary tumors and craniopharyngioma (Heaney AP. 2011), but other less rare disease reported in the literature (Arai A and Nishihara M,. 2010) especially the rare case of sellar region due to improper treatment can lead to catastrophic consequences, such as early hospital depending on the saddle area bit due to inadequate understanding of germ cell tumor surgery taken, after the hypothalamus due to severe reactions in children have died .in recent years by giving radiotherapy to be confirmed after the experimental radiotherapy can be given only to achieve the desired clinical effect. Instead, the group 1 patients for the imaging diagnosis of germ cell tumors, and to give experimental aggravated sexual symptoms after

radiotherapy to take surgery, postoperative pathology confirmed a rare malignant yolk sac tumor, preoperative AFP was significantly higher after 1 Review the week decreased tumor markers, AFP immunohistochemical staining further confirmed strong positive after 1 month after the head MRI showed tumor recurrence and reoperation, occurred after five months extensive metastasis within the spinal canal and upper cervical cord compression produces a large occipital herniation and death; followed by the group 1 patients with a preoperative diagnosis of recurrent hypophysoma, leiomyosarcoma confirmed pathologically

Leiomyosarcoma in sellar region is very rare foreign J. Niwa (1996) first reported by the after surgery and hypophysoma radiotherapy can be transformed into leiomyosarcoma, the disease so far reported a total lack of foreign literature in 10 cases, the current domestic not been reported. Leiomyosarcoma of the cases were due to a history of postoperative radiotherapy for hypophysoma, suggesting that the leiomyosarcoma may be caused by secondary cancer radiotherapy, the clinical manifestations of leiomyosarcoma saddle area, the lack of specificity of imaging features, preoperative misdiagnosis rate is extremely high by immunohistochemistry after SMA, MSA, Vimention were positive to confirm that most of the patients by surgical resection and postoperative radiotherapy supplemented by close follow-up of patients after 10 months was also died.

Malignant goniolmas such as yolk sac tumor, embryonal carcinoma and mixed germ cell tumors occur most often the site is the ovary, and the prognosis is good, 5-year survival rate of 96.7% (Lee KH and Lee IH,. 2009). Intracranial malignant goniolmas mainly occur in the pineal region can also occur in the sellar region, but the incidence rate is low. Wu Maochun (2006) , Beijing Tiantan Hospital were collected between 10 to 19 cases of intracranial high grade non-germ cell tumor goniolmas, only 3 cases of sellar regions, the site of yolk sac tumor is the occurrence of pineal region and the cerebellar hemispheres and lateral ventricles, the 1-year survival rate was 25.0%. While the yolk sac tumors was occurred in the sellar region in this case, although given standard postoperative radiotherapy and chemotherapy, the patient still died 5 months after operation. Huang Xiang(2008) reported that the Shanghai Huashan Hospital, 12 to 39 years were collected non-reproductive cell of cases of intracranial malignant goniolmas, 14 cases in pineal region, only 2 cases of yolk sac tumor.

Only 1 case of embryonal carcinoma was occurred in sellar region. Due to the incidence rate of the high grade non-germ cell tumor goniolmas is low in sellar region. And lack of information on preoperative imaging specificity, combined with lack of understanding of the disease, 3 cases of goniolmas in this group misdiagnosed before surgery. All through this group Preoperative tumor markers with immunohistochemistry after sellar region can be clearly malignant germ cell tumor subtypes, the AFP was significantly elevated, suggesting that the endodermal sinus (yolk sac tumor) or sinus within the embryo based mixed germ cell tumor; HCG was significantly higher for those who should be considered as choriocarcinoma or choriocarcinoma components with mixed germ cell tumor, the AFP and HCG were significantly increased in the same time, the first consider the embryonal carcinoma as embryonal carcinoma with syncytiotrophoblast and endodermal sinus component which allows for both AFP and HCG.

3 patients in this group 1 week after surgical resection of tumor markers review the corresponding decline in varying degrees, but the yolk sac tumor and embryonal carcinoma patients were reviewed after 1 month at the head MRI showed tumor recurrence, review the re-rise of tumor markers high, the full description of preoperative tumor markers AFP and HCG on the diagnosis, prognosis and recurrence of some significance.

Presently, because of yolk sac tumor and embryonal carcinoma high degree of malignancy, postoperative easy to relapse, although 2 patients in this group given standard radiotherapy and timely chemotherapy, patients were respectively died after 5 months and 6 months.1 case of mixed germ cell tumor is a good survival, suggesting that the tumor may be mainly composed of germ cell tumors. Recent studies have shown radiotherapy combined with cisplatin and bleomycin-based combination chemotherapy can effectively delay the highly malignant germ cell tumor survival time of patients (Cheon HC and Jung S,. 2006).

In addition to the height of this group of rare malignant goniolmas (yolk sac tumor, embryonal carcinoma and mixed germ cell tumor in 1 case), and pituitary adenoma into leiomyosarcoma after radiation therapy, the less can be malignant tumors, such as pilocytic astrocytoma, the pilocytic astrocytoma is most common in the cerebellum (Desai KI and Nadkami TD,. 2001) very little occurred in the saddle. This group after the cases were pathologically confirmed as received no follow-up radiation therapy, clinical results were satisfactory, follow-up no recurrence. In addition, 1 patient in this group there are the most specific granulomatous fungal disease that saddle area, preoperative imaging before the highly suspected clinoid meningiomas, pathological diagnosis of

fungal inflammatory pseudotumor, but tuberculosis or a fungal difficult to distinguish inflammatory pseudotumor, after confirmed by PAS staining fungal mold inflammatory pseudotumor.

Currently inflammatory lesions occurred only 0.5% of pituitary lesions in sellar region(Rao S and Rajkumar A, 2008) clinical very rare, as many as case reports of pituitary abscess, clinical is not very common, and the mycosis occurs in sellar region is rare, due to fungal Inflammatory pseudotumor occurs mostly in immunocompromised patients, this group of young patients without immune system dysfunction, or underlying diseases such as diabetes, so little consideration for the preoperative inflammatory pseudotumor, high rate of misdiagnosis. SAH occurred 2 months later, secondary search MRA shows left vertebral artery aneurysm, now in critical condition, breathing machine needed to maintain breathing. Patients with a history of fungal inflammatory pseudotumor, the author believe that the Department of vertebral artery aneurysms and more spread fungal diseases due to the great vessels of skull base Willis ring, therefore, in this case gives the biggest lesson is that once clear of intracranial fungal lesions exists, early MRA or DSA, and early treatment may prevent catastrophic SAH.

In short, this group of 6 cases of rare lesions of the sellar region of clinical data showed that the extremely complex pathology, preoperative misdiagnosis rate is high, especially for young people sellar lesions, a rare highly malignant germ cell tumor should be attention to the preoperative imaging studies should be strengthened, while tumor markers AFP and HCG test in order to determine the nature of the tumor and prognosis for the first time under the microscope during surgery for total resection of tumor and given standard radiotherapy and chemotherapy to improve clinical efficacy.

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