INTRINSIC TECTAL LOW GRADE ASTROCYTOMAS

IS SURGICAL REMOVAL AN ALTERNATIVE TREATMENT?

Long-term outcome of eight cases

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ABSTRACT - Low-grade gliomas arising in dorsal midbrain in children and young patients usually present few neurological symptoms and findings, and patients' management is controversial. Some authors propose only clinical observation until the patient present signs of increased intracranial pressure when a shunt with or without biopsy, is inserted; others recommend radiotherapy after stereotactic or open biopsy. Microsurgical total removal of tumor may be curative. We present a retrospective analysis of eight patients (mean age 16.6 ±11.5 years-old) with low-grade astrocytoma of the tectal region operated on using an infratentorial/supracerebellar approach between 1981 and 2002. All patients presented hydrocephalus and had a shunt insertion before surgical resection of the lesion. The tumour could be totally resected in seven patients. In one case radical removal was not possible due to infiltrative pattern of the lesion. Postoperative radiotherapy was performed in two cases, one patient at the beginning of this series and in the case with infiltrative tumor. This patient presented progressive tumor growth and died five years after surgery. No recurrence occurred after total removal. Post-surgical follow-up time ranged from 2 1/2 to 22 1/2 years (mean 9.9 ± 5.9 years). Radical microsurgical removal of non invasive tumors is possible without mortality or significant morbidity. It may be curative and should remain as an alternative to be discussed with the patient.

KEY WORDS: astrocytoma, tectal tumours, glioma, hydrocephalus.

Astrocitomas tectais de baixo grau: o tratamento cirúrgico é uma alternativa? análise de oito casos com longa evolução


PALAVRAS-CHAVE: astrocitoma, tumores tectais, glioma, hidrocefalia.

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Surgical resection of brain stem tumours is currently limited to dorsal exophytic tumours arising from the floor of the fourth ventricle and some intrinsic lesions of the cervicomедullary junction and mid-brain. Radiotherapy and chemotherapy remain alternative therapies for other infiltrative and biologically aggressive brainstem tumours. Long-term prognosis is usually very poor. Low-grade astrocytomas of the tectal region present characteristic neuro-radiological findings (especially in the MRI examination) and are often slow growing lesions causing late-onset aqueductal stenosis and hydrocephalus.

Due to this indolent growth and few clinical symptoms the management of these lesions remains controversial. Some authors recommend only a shunt procedure and clinical observation, others indicate biopsy and radiotherapy, and in some centers radical surgery has been performed.

We reviewed our experience with eight patients harbouring intrinsic tectal low-grade astrocytomas.

**METHOD**

Between 1981 and 2002, eight patients (six males and two females) with low-grade astrocytomas of the tectal plate were operated on in the Instituto de Neurologia de Curitiba (Table 1). Seven patients were younger than 20 years and one was older than 40 years (mean age 16.6±11.5 years-old). During this period of time, 43 pineal region tumors were surgically removed. All patients complained of headache, nausea and vomiting for several months. Three patients had papilledema. Two patients presented Parinaud’s syndrome and one was referred with the diagnosis of communicating hydrocephalus with no tumor visible in a high definition contrast enhanced computerized tomography (CT) examination. One patient had a ventricle-peritoneal shunt surgery 33 years before admission due to communicating hydrocephalus. She presented headache and Parinaud’s syndrome and magnetic resonance image (MRI) examination revealed a lesion in the tectal region. All patients presented hydrocephalus, focal calcification in the pineal region, and in two cases and contrast enhancement in the pineal and tectal region was observed in two patients. The first patient of this series had only CT scan which showed a enhanc-
ing tectal tumor. The other patients underwent (MRI) examination with and without gadolinium administration. The patients studied with MRI presented, in the tectal region, an iso or hypointense mass on T1-weighted studies and hyperintense on T2-weighted images. No enhancement or edema around the lesion were observed.

Hydrocephalus was treated with shunt procedure (seven patients) and endoscopic third ventriculostomy (one case). Tumor removal was performed through an infratentorial/supracerebellar approach in a modified "concorde position". The first patient in this series was irradiated (6 Mv linear accelerator, 5500 cGy) and presented memory impairment probably related to radiotherapy. One patient with recurrent infiltrative astrocytoma received postoperative radiotherapy. Control neurological examination and MRI-examinations were performed (annually) in all cases between 2 1/2 and 22 1/2 years (mean 9.9 ± 5.9 years) after operation.

RESULTS

There were no complications related to the shunt placement or third ventriculostomy in this series. Hydrocephalus resolved in all patients. In the postoperative period, three patients presented incomplete Parinaud's syndrome, which disappeared completely three to six months after surgery. The two patients with preoperative Parinaud's syndrome showed marked improvement of the upward gaze after the operation. Radical tumor removal was possible in seven patients. Total tumor removal of the lesion was confirmed by MRI in seven cases. Sub-total tumor removal, due to infiltration of the brainstem (a small portion in the cerebral peduncle remained), was performed in one patient. Growth of this residual tumour was observed in MRI 3 years after operation. The tumor progressed and the patient died five years after surgery in spite of radiotherapy. There was no major complications in this series, three patients presented postoperative transient abducens palsy, which completely resolved within 3 and 6 months after surgery. No sign of tumor recurrence was observed in the follow-up examination of the remained seven patients. Histological diagnosis of all patients was low grade astrocytoma. A summary of treatment and outcome is presented in Table 2.

ILUSTRATIVE CASES

Case 1 – This 13-year-old boy presented to our clinic with a 6 months history of headache and vomiting. Neurological examination showed no abnormalities. CT scan examination showed hydrocephalus but no tumor even after contrast injection. A shunt was inserted and the symptoms disappeared. MRI performed 6 months after the shunt demonstrated a no enhancing tumor in the tectal region (Fig 1). The lesion was totally removed through a supracerebellar/infratentorial approach. Postoperative the patient developed Parinaud’s syndrome that completely resolved in four months after operation. The histological diagnosis was low-grade astrocytoma. The patient remained asymptomatic and no tumor recurrence was observed in the control MRI 10 years after surgery (Fig 1).

Case 2 – This 17-year-old boy started to complain of headache and diplopia, 8 months before admission to our clinic. The neurological examination revealed Parinaud’s syndrome. MRI revealed hydrocephalus due to aqueduct occlusion by a tectal tumor. The lesion showed no contrast enhancement (Fig 2). A shunt was inserted and the tumor totally removed through a supracerebellar/infratentorial. The postoperative course was uneventful. The histopathological examination revealed a low-grade astrocytoma. A control MRI examination 7 years after operation showed no tumor recurrence (Fig 2).

Case 3 – This 44-year-old woman had a history of hydrocephalus treated by shunt placement 33 years before admission to our clinic. Several CT-scan examinations and two MRI showed normal ventricles and no tumor. Two months before admission to our clinic she presented symptoms of headache and diplopia and Parinaud’s syndrome. MRI revealed a non-enhancing tumor in the tectal region (Fig 3). The lesion could be radically removed through a supracerebellar/infratentorial approach. There was no postoperative complication and the Parinaud’s syndrome disappear completely 6 months after operation. The histological diagnosis was low-grade astrocytoma. The patient remained asymptomatic and a MRI, 2 years after surgery, showed total removal of the lesion (Fig 3).

DISCUSSION

Gliomas of the tectal plate are rare lesions. In a review of 486 brain tumors in children, only 6 cases were observed. They may also occur in adults but are rare and corresponded to 8% in a series of 48 brain stem tumors. These tumors are silent presenting very often only late-onset aqueduct stenosis and hydrocephalus. The diagnosis of low-grade astrocytomas of the tectal region was difficult until the advent of MRI. CT-scan may fail to reveal these tumors, for they may be isodense and non-enhancing. Therefore all patients with aqueduct stenosis should be investigated with MRI. MRI examination of these low-grade astrocytomas led to earlier diagnosis and they can be followed without invasive techniques. Low-grade astrocytomas of this region typically present as isointense or hypointense lesions on T1-weighted images compa-
red to the surrounding brain and hyperintense in T2-weighted images. Tectum enlargement is characteristic and gadolinium enhancement is not always observed. The natural history of these lesions is not well known and the choice of the best therapeutic approach is still controversial. In a series of 16 patients, 4 cases demonstrated clinical signs of tumor progression (Parinaud’s syndrome) with a median interval of 11.5 years from onset of initial symptoms and hydrocephalus. Two cases of spontaneous tumor involution after third ventriculostomy were reported. Daglioglu et al. conclude that intrinsic tectal gliomas of childhood which are less than 2 cm in diameter, without any tumor extension or contrast enhancement, should be managed conservatively.

The treatment of these patients varies from clinical observation with control MRI examinations to radical surgical removal. Increase of intracranial pressure due to hydrocephalus is the most common symptom of these patients. Shunt-procedures are indicated for hydrocephalus due to aqueductal stenosis. Radical removal and/or radiotherapy for these benign lesions are usually indicated when radiological or clinical signs of tumor progression is observed. Total tumor resection is possible with low morbidity. Recently, Pollack et al. recommended biopsy and radiotherapy for patients with progressing symptoms. Biopsy may be performed with neuroendoscopic techniques. A biopsy may, however, produce sampling error and has some risks. The proliferate index studies with Ki-67 labelling is proving to be a valuable tool to confirm the aggressive behaviour of some astrocytic lesions but their routine use as a prognostic index still need to be established.

Radical surgical removal is possible in non infiltrative lesions with no mortality and low morbidity.
ity, it has two main advantages: confirm the diagnosis and may cure the patient. The approach used in this series, was the infratentorial/supracerebellar in a modified “concorde position”31. There is no clear evidence of beneficial effect of radiotherapy ( radiosurgery) for low-grade astrocytomas. Routine use of radiotherapy or chemotherapy is questionable21. In a series of seven patients harbouring low-grade tectal gliomas treated with stereotactic radiosurgery, two developed severe radio-induced edema, one with permanent deficits and one did not respond to that treatment34. Radiation therapy should be avoided in children because it may cause anaplastic transformation of low-grade astrocytomas35,36. Tarlov37 first suggested this complication in 1937 and development of radiation-induced tumours in other areas has been also reported38-40. Other deleterious effects of radiotherapy as psychomotor impairment, cranial neuropathy, leukoencephalopathy, endocrinopathy and vasculopathy have been also described24, 41-47. When an infiltrative pattern is present, a subtotal resection might be done to reduce morbidity. The efficacy of radiotherapy for incompletely resected low-grade astrocytomas remains to be proven48,49. The optimal management of high-grade astrocytomas of this region is unclear too. Attempt of radical surgery in patients with high-grade infiltrative lesions may produce severe postoperative neurological deficits. If gross total removal is, however, possible and followed by radio and chemotherapy, the prognosis of these patients is better50.

In conclusion, low-grade astrocytomas of the tectal region have an indolent clinical course. Most of these lesions are truly focal neoplasms with little tendency to infiltrate surrounding structures. Headache, hydrocephalus and Parinaud’s syndrome are the most common symptoms. Some tumors may suffer transformation to higher grades. Hydrocephalus should be treated by shunt insertion or endoscopic third ventriculostomy. Radical removal of the lesion is possible in the majority of patients, may be curative and present very few complications. Surgical removal should be presented to the patient as a possibility of treatment, specially in growing low-grade tumors.

REFERENCES