OPHTHALMOLOGICAL FINDINGS IN PEDIATRIC BRAIN NEOPLASMS: 58 CASES

MANIFESTACIONES OFTALMOLÓGICAS EN TUMORES CEREBRALES PEDIÁTRICOS: 58 CASOS

SANTAMARÍA A¹, MARTÍNEZ R², ASTIGARRAGA I², ETXEBARRÍA J², SÁNCHEZ M¹

ABSTRACT

Purposes: To describe the visual manifestations of brain neoplasms, and to analyze the effect of tumor control on these.

Methods: This is a descriptive retrospective study, which includes patients under 14 years of age, suffering from different brain neoplasms in our hospital between 1996 and 2005 inclusive.

Results: In the group of patients with low visual acuity, 44% had organic amblyopias. In 28% of cases, the amblyopia was secondary to the strabismus/nystagmus produced by the developing tumor. Corrective treatment was successful in some cases of partial or total organic amblyopia. Ophthalmologic evaluation (including perimetry and fundoscopy) enabled detection of 3 tumor relapses.


Key words: Brain neoplasms, amblyopia, perimetry, visual acuity, astrocytoma.

RESUMEN

Objetivos: Describir las manifestaciones visuales en neoplasias neuropaediatricas, relacionarlas con la localización tumoral y analizar su relevancia en el seguimiento de la enfermedad.

Material y métodos: Estudio descriptivo retrospectivo que incluye pacientes menores de 14 años con neoplasias cerebrales de cualquier estirpe, registradas en nuestro centro desde 1996 hasta el 2005.

Resultados: En un 44% de los pacientes con disenso en la agudeza visual, la ambliopía fue catalogada como orgánica. En el 28% de los casos, la ambliopía era secundaria al estrabismo/nistagmus producido por el tumor. El tratamiento corrector resultó efectivo en varios casos de ambliopía parcial y totalmente orgánica. Tres recidivas tumorales fueron detectadas gracias a la exploración de fondo de ojo y campimetría, antes de que las pruebas de imagen fueran concluyentes.

Conclusión: El examen oftalmológico (agudeza visual, campimetría, funduscopía) es una pieza clave en el seguimiento de la patología tumoral cerebral. Las ambliopías orgánicas parciales/totales son susceptibles de tratamiento corrector.

Palabras clave: Tumores cerebrales, ambliopía, campimetría, agudeza visual, astrocitoma.
INTRODUCTION

Eyes are an open window to the central nervous system. This window may provide us with many different signs, which are the result of alterations at different levels of the optic way and/or of the numerous associated structures which take part in the complex process of vision. A case review is provided below, with a description and an analysis of different intracranial neoplasms amongst a certain paediatric population. The information will be categorized, and interpreted as a component of a severe disfunction, with potential involvement of more than one organ.

SUBJECTS, MATERIAL, AND METHOD

A retrospective descriptive study was performed in order to study the ophthalmological manifestations of brain tumours for a certain subgroup in a paediatric population. The study was configured to cater for all types of brain tumours (including intracranial metastases with a distant primary focus) for children under the age of 14 in our centre. The study included patients controlled in the last ten years, based upon discharge dates registered between 1996 and 2005 (both included). The objectives of the study were:


– Studying ophthalmological signs and symptoms for each patient.

– Studying the relationship between tumour locations and ophthalmological clinical status.

– Interpreting complementary ophthalmological tests.

– Assessing the value of ophthalmological examination for disease diagnosis and follow-up.

– Analyzing the different ophthalmological treatments provided, and the responses for each case.

Data was gathered using the following material: clinical records from the hospital archive; clinical records from the hospital IT systems, scanned due to patient’s death or other non-specified reasons; records submitted to the ophthalmology department or to other hospital areas (paediatric oncology, neuropaediatrics, children’s orthopaedics, endocrinology), active at the time of the study. Data was then collected and categorized by means of a template, where the following sections were included: personal details; brain pathology, with information on tumour type, location, and extension, age of presentation, and number of recurrences if applicable; pathologies secondary to the tumour or the treatment received; visual symptoms (subjective/objective visual acuity, amblyopia and correction for each case, findings from ocular/biomicroscopy/ocular fundus examination, optic nerve disorders, other cranial pair disorders, and abnormalities in eye motility); complementary ophthalmological tests (field of vision, visual evoked potential, colour tests, diplopia test, etc); representative image tests (brain MRI and brain CT scan); ophthalmological/general treatment received during the study period; date of death (when applicable), and patient’s age.

RESULTS

Once data was collected, information was analyzed and categorized for the following criteria: tumour histology; visual acuity/ametropia; cranial pair disfunction; nystagmus; findings from biomicroscopy/ocular fundus test; field of vision; death.

Tumour Histology (table I, fig. 1)

The most frequent tumours of those reviewed belonged to the neuroglial category. This group included 13 astrocytomas, 3 of which were suprasellar, 4 posterior fossa cerebellar neoplasms, 4 anterior fossa hemispheric tumours, one giant-cell astrocytoma in the 3rd ventricle, and one pilocytic cervical medullary neoplasm. Gliomas included 3 encephalic brainstem tumours, 2 optic nerve/chiasm neoplasms, one thalamic tumour, and one diffuse gliomatosis cerebri in a type I neurofibromatosis syndrome. Lastly, 5 ependimus-derived neuroglial tumours were included in this group.

The next most frequent tumours were undifferentiated neuroepithelial, including hemispheric neoplasms derived from primitive neuroectodermal tumours (PNET) (3 cases), and cerebellar vermis medulloblastomas, the largest subgroup, with 13 cases.

Hypophysary tumours found in the paediatric population under study were exclusively craniopharyngiomas (5 cases).
2 meningiomas were found with meningothelial histology; one nervous sheath tumour was detected (schwannoma). Two processes corresponded to patients suffering from type 2 neurofibromatosis. The 3 cases of cerebral hamartomatosis described involved one patient with tuberous sclerosis, and one patient with type 1 neurofibromatosis.

Two of the brain neoplasms were metastases from distant primary focii (Wilms' renal tumour, and suprarenal neuroblastoma). In a similar manner,
10 tumours produced leptomeningeal metastatic spread. Six of them were cerebellar medulloblastomas, with one of the most unfavourable prognosis. Amongst all categorized neoplasms, 42% originated in the posterior cranial fossa.

Visual Acuity/Ametropia

Records of visual acuity were taken for 36 patients only. Subjective and objective loss of vision was determined for 77% of all cases. After a comprehensive analysis of the etiopathogenesis of visual impairments (fig. 2), the conclusion was reached that 44% of all cases showed a visual organic alteration which accounted for the disorder, and which was produced directly by the tumour, or was secondary to its expansion, to the distortion of adjacent structures, or to the increase in intracranial pressure. However, a second group was identified (28%), where the tumour produced an indirect reduction in visual acuity. This consisted of strabismic or nystagmic amblyopia produced by damaged oculomotor nuclei-pairs, as a consequence of the evolution of the intracranial mass. There is no doubt that these patients might have been included under the section entitled “Organic amblyopia or amblyopia secondary to the tumour” since these are indirect tumour sequels. They were however considered to be a group in its own right, due to their specifics and their peculiar physiopathology.

An earlier visual acuity disorder was found for 19% of all subjects. The most prevalent condition was amblyopia derived from accommodating convergent strabismus in hypermetropic patients. This group was called ‘Earlier disfunctions’ or ‘Non-organic amblyopias’. It must be pointed out that visual acuity improved in all cases here after suitable correction was applied (glasses; occlusions; penalties; botulinic toxin injections; strabismus surgery). Moreover, a remarkable improvement in vision was achieved after applying correction to a number of patients with a partial organic amblyopic component (associated to an earlier visual disorder, prior to tumour development) or an all-organic component (fig. 3).

Cranial Pairs

The second cranial pair, or optic nerve, was considered as a separate group in this study, due to the large diversity of signs/symptoms which may appear. The most characteristic alterations included the following: pupillary alterations (18 cases); dyschromatopsia (4 cases); self-limited amaurosis (3 cases); legal blindness (visual acuity under 0.1) in 3 cases, and anophthalmos secondary to exenteration in an optic nerve menigioma. As regards the appearance of the optic disc upon ocular fundus examination, 17 patients showed papilledema at some point in the process, whilst 14 showed residual papillary atrophy. Three cases were recorded where papilledema was the first indication of tumour recurrence at a stage where MRI and the rest of image tests were not clear enough.

On the remaining cranial pairs (see fig. 4 below), the one most frequently involved was abducens,
(VI) because of its great sensitivity to compressions and variations in intracranial pressure (1). The second most frequent pair involved was the facial one (VII). This fact may be explained due to the proximity of both cranial pairs at the exit from the brain stem and throughout their intracranial trajectory (2). These are followed by pair III, with a prevalence of 18%, and pairs IV, V, and VII.

Nystagmus (fig. 5)

The most frequent nystagmus for the population under study was the horizontal component type, exacerbated in lateroverions. Bilateral horizontal nystagmus was found for 61% of cases, and unilateral for 24%. This fact may be explained by the prevalence of involvement of the sixth pair in intracranial neoplasms, where abduction is limited for one or both eyes, and a secondary nystagmus is generated. Vertical nystagmus is much less common.

Biomicroscopy/Ocular Fundus

Findings were rather numerous and varied after ocular examination, biomicroscopy, and ocular fundus tests. Papillary edema and papillary atrophy were common in the study group, as mentioned above. Papilledema is usually presented as one of the early signs of the disease, as a consequence of direct compression of the optic nerve and/or an increase of intracranial pressure, compromising ocular venous return. In general terms, papilledema and the symptoms associated to intracranial hypertension may be resolved after a ventricular-peritoneal derivation valve is implanted. Papillary atrophy, which is characterized by paleness and an increased excavation, is usually consecutive to papillary edema, thereby appearing at later stages (1). The study includes other manifestations, as shown by table I.

Field of Vision (figs. 6-9)

A number of specific patterns for the visual field were found in the study for different tumour locations (3). These patterns are very useful, not only for diagnosis and anatomic location of neoplasms, but also for disease follow-up and recurrence detection. On two concrete cases it was possible to perform an early diagnosis of tumour recurrence due to
the modified visual fields of the patients, and to the recent appearance of binocular diplopia in one of them.

Death

20 patients were deceased (34%); 5 patients of ages between 0 and 4, 5 patients between 5 and 9, and 10 patients between 10 and 15. The average age at the time of death was 6.16. A quarter of all cases were cerebellar vermis medulloblastomas. No indication is provided of the cause of death in most cases (60%), where cardiorespiratory arrest was recorded. 20% of children suffered from an objective respiratory deficiency. The remaining 20% included cardiac deficiency, massive brain hemorrhage, septic shock, and convulsion status.

DISCUSSION

The most common solid neoplasms in children are those involving the central nervous system. The most prevalent tumour types differ from those found in adults. The most common pathologies in children include benign gliomas, undifferentiated neuroepithelial tumours, and craniopharyngiomas (4,5). Cerebellous vermis medulloblastomas account for the largest individual group (6), in agreement with the results of the study. As regards tumour location, the proportion of supratentorial neoplasms in children is similar to infratentorial tumours, as opposed to adults, where the first type is more common (4,7).

As shown above, ophthalmological examination is crucial in order to diagnose recurrence of paediatric brain tumours, where no other neurological signs are present, or no conclusions may be derived from image tests. Our series allowed early detection of several tumour recurrences thanks to the results from routine ocular fundus examination (with 3 cases of papilledema), and from recent visual field variations (2 cases). Tumour recurrence is the most frequent cause of death for paediatric patients. Thus, prognosis improves if recurrence is detected in an asymptomatic phase (4). According to our cri-

Fig. 6: Field of Vision I: left anopsia of a suprasellar pilocytic astrocytoma with obstructive hydrocephalia.

Fig. 7: Field of Vision II: Evolution of the visual field for a chaniopharyngioma. A slight improvement is visible for subsequent studies.

Fig. 8: Field of Vision III: left homonymous hemianopsia in a colloid cyst of the third ventricle, with involvement of the right optic tract.
As regards alterations of visual acuity, checks were performed in order to ensure that correction on non-organic ambliopia (‘earlier disfunctions’) showed similar results to those obtained for healthy individuals. Furthermore, ambliopia by direct organic lesion, or produced by strabismus/nystagmus secondary to tumours may potentially be corrected. For all the reasons mentioned above, we believe that the capacity of oncological children to recover through visual rehabilitation must not be underestimated. In some cases children’s responses to treatment are rather unexpected, and it is sometimes feasible to achieve a degree of visual performance which is acceptable for daily life.

REFERENCES