CASE REPORT

Trilateral Retinoblastoma

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SUMMARY

Trilateral retinoblastoma is the association of hereditary bilateral or unilateral retinoblastoma with a pineal neuroblastic tumour. We describe two cases of trilateral retinoblastoma from a total of 141 cases of retinoblastoma seen over an 8.5 year period. Both had a fatal outcome, with survival times of only 4 and 8 months respectively.

KEY WORDS:
Trilateral, Retinoblastoma, Pineal gland tumour, CT scan

INTRODUCTION

Trilateral retinoblastoma (TR) is characterised by an intracranial neuroblastic tumour arising in the pineal region, associated with hereditary retinoblastomas which are usually bilateral but may also be unilateral. TR is rare, occurring in 3% of cases and is usually fatal [1]. From August 2001 till December 2009, a total of 141 cases of retinoblastoma were seen at our institution. During this time, two children progressed to trilateral disease (1.4%).

CASE REPORT

Case 1

A 6-week old baby girl was diagnosed to have bilateral intraocular retinoblastoma. The right eye was enucleated. The left eye was Reese-Ellsworth (R-E) group I and was treated with laser photocoagulation only, no chemotherapy was given. At the age of 22 months, she presented with vomiting and generalised tonic-clonic seizures. A computed axial tomographic (CT) scan of the brain showed a tumour in the pineal region causing hydrocephalus (Figure 1). A ventriculo-peritoneal shunt was inserted and the child was treated with nine courses of vincristine, carboplatin and etoposide (JOE). She was also given intrathecal chemotherapy once the intracranial mass resolved. However, at 30 months of age, she developed status epilepticus due to progressive neurological disease and died.

Case 2

A 2-year old boy was diagnosed to have bilateral intraocular retinoblastoma. The left eye was enucleated, the right eye was R-E group II. The child was given nine courses of JOE for chemoreduction. At 40 months of age, the child presented with vomiting. A CT scan showed a pineal gland tumour causing hydrocephalus. A biopsy and and ventriculostomy was done. The histopathology confirmed retinoblastoma. The child was given palliative chemotherapy in the form of vincristine, actinomycin and cyclophosphamide. He died of progressive neurological disease at the age of 44 months. In both cases, there was no active ocular disease at the time of presentation of TR; fundal examination showed areas of scarring only, due to the previous photocoagulation.

DISCUSSION

Both our cases of TR occurred in children with bilateral disease but neither had a positive family history. Both children did not have genetic analysis performed due to lack of facilities in our institution. It has been postulated that the use of chemotherapy as chemoreduction in bilateral retinoblastoma may prevent the development of trilateral disease [2]. We did not find this to be true as our second case developed TR despite nine courses of chemoreduction. In a large series of TR, it was found that the median time of progression from retinoblastoma to TR was 21 months [3]. The time of progression in our two cases was 21 months and 16 months respectively. TR can also precede the development of retinoblastoma. TR is usually fatal, leptomeningeal disease is the main cause. Strategies include the use of intrathecal chemotherapy (as in our first case) as well as craniospinal radiotherapy. However, even with these modalities, the outcome in the past has been dismal. Recently, more aggressive chemotherapy including autologous stem cell transplant has resulted in some survivors [4,5]. It has been

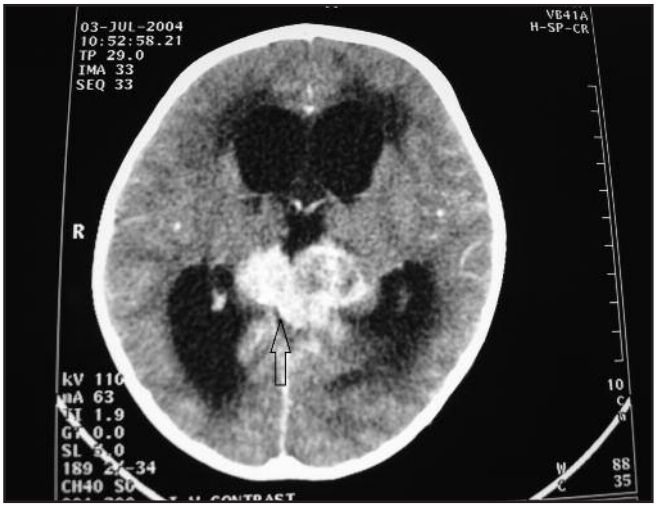

Fig. 1: Computed axial tomographic (CT) scan of the brain showing a calcified tumour (arrow) in the pineal region with hydrocephalus.

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suggested that screening children with bilateral retinoblastoma for TR may improve survival by detecting smaller tumours which are easier to cure. The current recommendation is routine neuroimaging every six months for the first five years of life. In conclusion, trilateral retinoblastoma is a rare and often fatal development in a child with hereditary retinoblastoma. The ideal treatment approach is yet to be elucidated.

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