Available online at www.sciencerepository.org

Science Repository

Case Report

A Child with Primary Leptomeningeal Melanoma

Álvaro Pimenta Dutra[1*], Marcos Grijo Pitangui[1], Angelo Raphael Tolentino de Rezende[2], Renato Pacheco de Melo[2], Eric Grossi Morato[3] and Luiz Fernando Fonseca[2]

[1]*General Pediatric Service of the João Paulo II Children's Hospital/FHEMIG, (Hospital Infantil João Paulo II/Fundação Hospitalar de Minas Gerais), Brazil*

[2]*Pediatric Neurology Service of the João Paulo II Children's Hospital/FHEMIG, Brazil*

[3]*Neurosurgery Service of the Clinical Hospital in Federal Universitiy of the Minas Gerais (HC-UFMG), Brazil*

ARTICLE INFO

Article history:
Received: 14 June, 2020
Accepted: 29 June, 2020
Published: 3 July, 2020

Keywords:
Leptomeningeal tumor
melanoma
children

ABSTRACT

Primary malignant melanoma of the meninges is an exceedingly rare neoplasm. Usually its symptoms include raised intracraneal pressure resulting from hydrocephalus seconday to tumoral obliteration of cisternal basal cisterns, but the passage of time from initial symptomalogy to diagnosis is frequently delayed. A 12-year-old male with primary letomeningeal melanoma is reported. At the beginning, he presented with vomiting, headache, complex seizures, fever four months before the admission in the hospital where progressive loss of consciouness after admission. Lumbar puncture reveled high protein level, normal glucose level and 50 leucocytes/mm^3 with 86% polymorphic cells. Magnetic ressonance imaging of the brain was referred and in T1-weighted images revealed a diffuse enhancement of the leptomeninges on right frontal cortical. An open brain biopsy trough was performed, after exclusion of the infectious nervous system disease. Histological examination revealed massive infiltration of meninges with brown tumor cells. These cells stained positive for HMB-45, S-100 protein and vimentin. The patient received post operative radiation therapy, but died after three months of the diagnosis with septic shock and epileptical status.

Introduction

Primary leptomenigeal melanoma (PLM) is a rare and agressive tumor in children and accounts for less than 1% of all pediatric malignacies, that originates from pial melanin-bering cells typically along the cerebral convexities or at the base of the brain [1].

Since the first description by Virchow in 1859, approximately 230 cases of the prymary leptomeningeal melanoma have been reported [2, 3]. A population study in Norway covering a 30-year period (1955-1984) identified six definite cases – an incidence of 0.005 new cases per year [4]. In this study, there was only one patient who presented under the age of 20 years, illustrating the rarity of this lesion in children. The gender incidence in these tumors seems equal, and there are no convincing data suggest a racial predominance [4].

Aproximately 75% of patients with primary leptomeningeal melanoma lack any significant concomitant lesion [5]. The typical neurological manifestations of leptomeningeal melanoma include seizures, psychiatric disturbances, and symptoms and signs of raised intracranial pressure secundary to hydrocephalus [3, 6-8].

In this report, we describe a child in whom the diagnosis of primary letomenigeal melanoma was made by an open brain biopsy, after exclusion of the infectious nervous system disease.

Case Report

A 12-year-old boy was admitted previously well, presented with 4-month history of vomiting, intermittent fever and headache. He had been admitted to another hospital six times with same history but in the last admission, he presented with generalized seizures.

*Correspondence to: *Álvaro Pimenta Dutra, Alameda Ezequiel Dias 345, Santa Efigênia - Belo Horizonte/Minas Gerais, Zip Code- 30130110, Third Floor In Medical Residence at João Paulo II Children´s Hospital (Hospital Infantil João Paulo II) - FHEMIG (Fundação Hospitalar do Estado de Minas Gerais), Brazil; Tel: 55011950211559; E-mail: alvaropdutra@yahoo.com.br, aloapd@hotmail.com*

http://dx.doi.org/10.31487/j.COR.2020.07.06

On admission he was drowsy, obeyed commands, left hemiparesis, depressed deep tendon reflexes, no signs of papilledema and cranial nerves were normal. Lumbar puncture revealed high protein level 90 mg/dl, normal glucose level 53 mg/dl and 50 leucocytes/mm^3 with 86% polymorphic cells, 25 erythrocytes/mm^3 and no malignant cells. He was initially treated with intravenous Ceftriaxone and Acyclovir, but six hours after admission he suffered a dramatic decline in consciousness (Glascow coma scale 3), then was admitted to the pediatric intensive care unit. Cranial computed tomography (CT) disclosed diffuse meningeal enhancement consistent with diagnosis of meningoencephalitis without signs of raised intracranial pressure. Further extensive infection screnn, including CSF and blood studies, was negative (including bacteria, viruses, atypical organisms, and fungi).

Cranial magnetic resonance imaging (MRI) (Figure 1) was obtained, which revealed diffuse enhacement of the leptomeninges on T1 images with gadolinium contrast, note the large right frontal cortical enhancing. An open brain biopsy was performed (Figure 2) and revealed a dura brown pigmented appearence. Histological examination demonstrated a typical malignant melanoma with expansion over the subaracnoides spaces by tumoral cells and infiltration of the brain, these cells stained positive for HMB-45, protein S-100 and vimentin. A search for cutaneus nevi and retinal tumor involvement was not revealing. In the absence of an alternative primary site of origin for the tumor, a diagnosis of primary leptomeningeal melanoma was made. Three days after surgery the patient presented a sudden deterioation with signs of raised intracranial pressure and unilateral right blindness, then was followed by a ventriculoperitoneal shunt insertion for development of obstructive hydrocephalus.

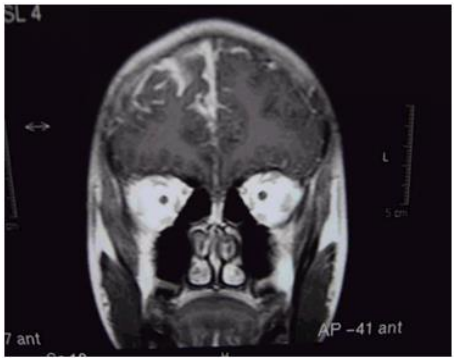

Figure 1: Coronal T1-weighted magnetic resonance imaging scan of the brains showing diffuse postgadolinium enhancement of the meninges upper right frontal lobe.

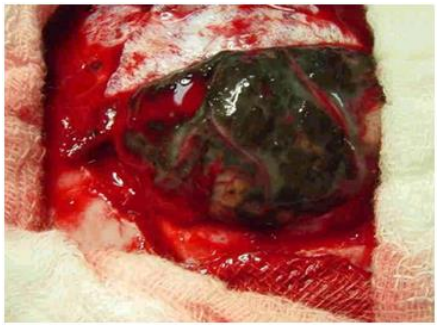

Figure 2: Macroscopic antemortem during surgery demonstrates brown-pigmented appearance of the leptomeninges.

The patient received postoperative radiation therapy with 5400 cGy using fractionated irradiation in high doses, for 30 days. There was no objective response to therapy, and he died fifteen days after concluded the treatment and two months after diagnosis due to septic shock and seizures in an Oncology Center.

Discussion

Primary leptomeningeal melanoma has been described as ocurring in a number of different forms. All forms of leptomeningeal melanoma are uncommon in children [1, 3, 4]. Acute presentation in infancy usually involves hydrocephalus and other symptoms of raised intracranail pressure, such lethargy, iritability, poor feeding and seizures [1, 3, 5-8]. The most commom age at presentation is on 0-24 months [1, 3]. The development of symptons can predate the appearance of malignatn change in the leptomengeal lesions. The clinical course, once symptomatic, even in the absence of malignant transformation, is rapidly progressive in patients with neurocutaneous melanosis [1, 3].

Primary leptomeningeal melanoma of the cetral nervous system hás been classified pathologically into two types: 1) that which diffusely invades the pia mater and spreads into the subarachnoids space; and 2) that which causes nodular tumors [3, 5, 6]. In both forms, primary leptomeningeal melanoma is highly malignant tumor characterized histopathological by marked cellular pleomorphism, mitoses, necrosis and haemorrhage [5, 6].

Diagnosis premortem always has been difficult, but the combination of cytologic examination of cerebral fluid, biopsy and modern imaging techniques has proved helpful [1, 3, 7, 9]. The direct neurosurgical extirpation of tumor has a minimal role to play, as the leptomeninges are usually diffusely involved with melanomatous change. Children, however, frequently require a treatment of their hydrocephalus like in our patient, but blindness has been reported as a late feature [3, 6, 7]. Other central nervous system tumors should be considered in the differential diagnosis, inclunding lymphoma, leukemia, neuroectodermal tumors and pigmented medulloblastoma. Characteristic findings have been described on magnetic resonance imaing (MRI), with hypertense signal on T1-weighted images and hypoinense signal on T2-weighted images, as so in our patient [3, 6, 7, 9].

Generally, solitary leptomenigeal melanomas have a better prognosis than diffuse melanomas, direct surgical removal of tumor is usually possible with slitary leptomenigeal melanomas, which may account for the longer survival periods of the disease [3]. However, therapy combined after craniotomy and chemotherapy or radiotherapy has been used, but the patient response is poor like our patient [1, 3, 7-9].

Although primary leptomeningeal melanoma continues to be a highly agressive, malignant tumor with poor survival, it is hoped that ealier detection by MRI, surgical intervention and newer clinical trials will lead to increased survival for children with this cancer.

REFERENCES

1. G W Makin, O B Eden, L S Lashford, J Moppett, M P Gerrard et al. (1999) Leptomeningeal melanoma in childhood. *Cancer* 86: 878-886. [Crossref]
2. Virchow R (1859) Pigment und diffuse melanose der Archnoides. *Arch Pathol Anat* 16: 180-182.
3. D Allcutt, S Michowiz, S Weitzman, L Becker, S Blaser et al. (1993) Primary leptomeningeal melanoma: an unusually aggressive tumor in childhood. *Neurosurgery* 32: 721-729. [Crossref]
4. A Helseth, E Helseth, G Unsgaard (1989) Primary meningeal melanoma. *Acta Oncol* 28: 103-104. [Crossref]
5. Rubinstein LJ (1972) Neurocutaneous melanosis and primary meningeal melanomas, in AFIP, Rubistein LJ (Eds): Tumors of the central nervous system 309-311.
6. J López Castilla, F Díaz Fernández, J A Soult, M Muñoz, R Barriga et al. (2001) Primary leptomenigeal melanoma in a child. *Pediatr Neurol* 24: 390-392. [Crossref]
7. P Nicolaides, R W Newton, A Kelsey (1995) Primary malignant melanoma of meninges: atypical presentation of subacute meningitis. *Pediatr Neurol* 12: 172-174. [Crossref]
8. D E Crisp, J A Thompson (1981) Primary malignant melanomatosis of the meninges. Clinical course and computed tomographic findings in a young child. *Arch Neurol* 38: 528-529. [Crossref]
9. O Flodmark, C R Fitz, D C Harwood Nash, S H Chuang (1979) Neuroradiological findings in a child with primary leptomeningeal melanoma. *Neuroradiology* 18: 153-156. [Crossref]