Available Online at http://www.bjbmr.org

BRITISH JOURNAL OF BIO-MEDICAL RESEARCH

Cross Ref DOI: https://doi.org/10.24942/bjbmr.2018.214 Volume 02, Issue 02, March-April 2018

ISSN:2456-9739

Case Study

Suprasellar Pilomyxoid Astrocytoma With Leptomeningeal Dissemination In An Infant: A Case Report And Review Of The Literature

Chiara Carducci[1], Giovanna Stefania Colafati[2], Mariangela Marras[3], Matteo Di Nardo[4], Antonella Cacchione[5], Ioan Paul Voicu[6], Andrea Carai[7], Angela Mastronuzzi[8]

[1-2]Bambino Gesù Children's Hospital IRCCS, Rome, Italy, Neuroradiology Unit, Department of Imaging.
[3]Department of Radiology, Azienda Ospedaliero Brotzu (A.O.B.), Cagliari, Italy.
[4]Pediatric Intensive Care Unit, Children's Hospital Bambino Gesù, IRCCS, Rome, Italy.
[5-6]Department of Hematology/Oncology and Stem Cell Transplantation, Bambino Gesù Children's Hospital, IRCCS, 00165, Rome, Italy
[7]Department of Neuroscience and Neurorehabilitation, Neurosurgery Unit, Bambino Gesù Children's Hospital, IRCCS, 00165, Rome, Italy.
[8]Department of Hematology/Oncology and Stem Cell Transplantation, Bambino Gesù Children's Hospital, IRCCS, 00165, Rome, Italy.

ARTICLE INFO

Article History:
Received on 09th March 2018
Peer Reviewed on 29th March 2018
Revised on 18th April 2018
Published on 30th April 2018

Keywords:
Pilomyxoid astrocytoma, Pilocytic astrocytoma, Children, MRI, CNS tumors

ABSTRACT

Pilomyxoid astrocytoma (PMA) is a rare entity added to the 2007 WHO classification of tumors of the central nervous system for the first time. Recently in the literature, PMA have been further studied with regard to their occurence, clinical and radiological features and treatment strategy. We present a case of a 8-month-old boy presented at the Emergency Department for suddenly lose consciousness, macrocephaly, anisocoria and no previous health problems, CT and MRI showed an obstructive hydrocephalus due to a multilobulated solid lesion centrally located (epicentre) in the suprasellar region with diffuse leptomeninges-based enhancing lesions in the brain, especially in the tentorial region, and spine. A biopsy revealed a glial neoplasm consistent with an aggressive PMA. To the best of our knowledge, there are no other reports of PMA with similar radiological findings in infants younger than 1 year.

INTRODUCTION:

Pilomyxoid astrocytoma (PMA), introduced as a distinctive entity in 1999 by Tihan et al. [1] and firstly described by Janisch et al [2] in 1985, is a rare entity, recently recognized as a variant of pilocytic astrocytoma (PA). It has been added to the 2007 WHO classification of tumors of the central nervous system [3]. The grading of PMA has been changed in 2016 WHO Classification of tumors of the central nervous system [4].

While previously designated as WHO grade II, recent studies have shown extensive histological and genetic overlap between pilomyxoid and PA, with some of the former maturing into the latter over time and less certainty that the pilomyxoid variant always follows a more aggressive course than a more classic appearing suprasellar pilocytic astrocytoma. It is not clear that PMA should be assigned to WHO grade II and the suggestion was made to suppress grading of pilomyxoid astrocytomas until further studies clarify their behavior [4].

PMA predominantly affects the hypothalamic/chiasmatic region and leptomeningeal dissemination occurs more frequently than PA [5].We report the clinical course, comorbidity, radiological, and histological features of a leptomeningeal disseminated pilomyxoid astrocytoma in an 8-month old infant with radiological findings consistent with slow growth. A review of the relevant literature is also presented.

CASE REPORT

Clinical history and radiological findings

An 8-month-old boy presented at the Emergency Department for suddenly lose consciousness. He had macrocephaly, anisocoria with pupils reagents little to light. Hypertonia was documented.

No response to stimuli and language absence were showed. No previous health problems, even during the peri- or neonatal periods. The patient was undergone brain Computed Tomography (CT) that showed an obstructive hydrocephalus due to a cerebral lesion centrally located in the suprasellar region, with no evidence of calcifications. A ventriculoperitoneal shunt was positioned.

Magnetic Resonance Imaging (MRI) of the brain and spine with gadolinium revealed a multilobulated solid lesion centrally located (epicentre) in the suprasellar region associated with perilesional (adjecent) enlargement of cerebrospinal fluid spaces and hypoplasia of the frontal lobe .

The fronto-parietal region of right hemisphere showed subacute hyschemia. Diffuse leptomeninges-based enhancing lesions in the brain, especially in the tentorial region, and spine were detected (Figures.1,2,3).

A biopsy of the lesion were performed and the istological examination of the specimen revealed a glial neoplasm consistent with an aggressive pylomyxoid astrocitomaI with high malignancy features as large vessels proliferation with focal necrosis.

Postoperatively, was performed a CT that disclosed an intralesional hemorrhage with an associated blood component in the lateral ventricles and right hemisphere ischemia.

No therapy was performed in the meantime. A palliative treatment and his transfer to a regional hospital was planned.

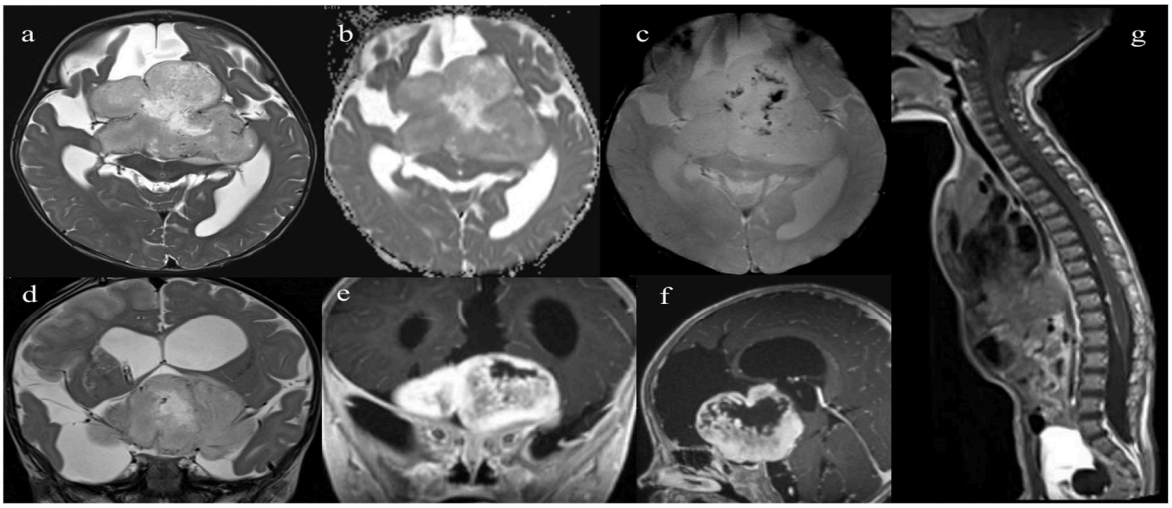

Fig.1: MR Axial and coronal T2-weighted (a,d), ADC map (b), axial GRE (c), coronal and sagittal T1-weighted with gadolinium images of brain show a multilobulated solid lesion, predominantly well-defined, centrally located in the suprasellar region associated with diffuse leptomeninges-based enhancing lesions. Sagittal T1-weighted image of spinal cord (g) shows multiple enhancing lesions in the subarachnoid space.

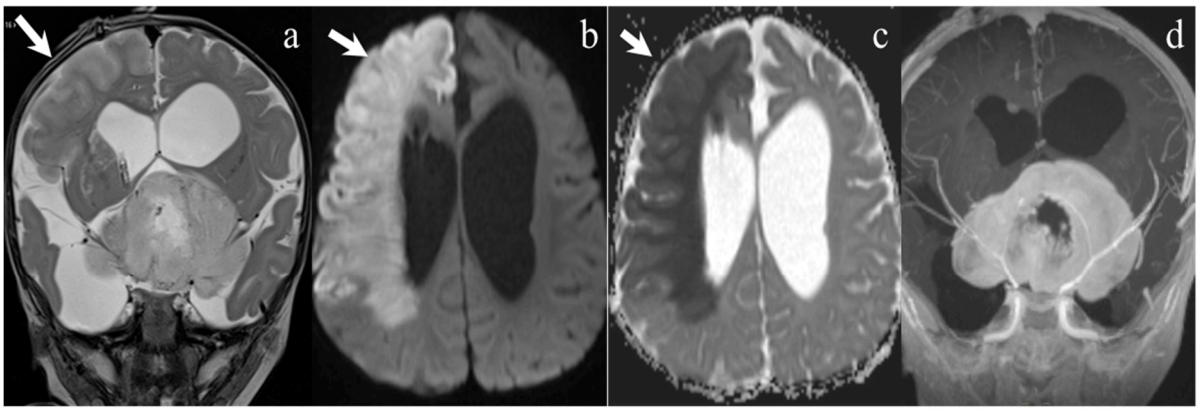

Fig.2: MR coronal T2-weighted (a), DWI (b) and ADC map (c) images show the right fronto-parietal subacute hyschemia (arrows). Coronal TOF multiparametric reconstraction shows encasement and of the Circle of Willis.

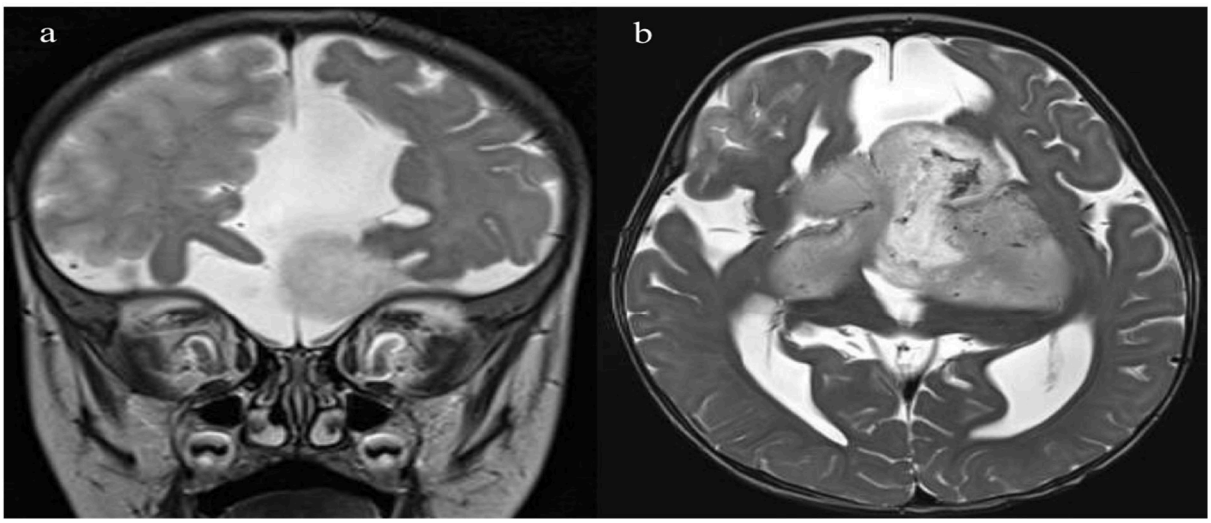

Fig.3: MR coronal (a) and axial (b) T2-weighted images show a perilesional enlargement of cerebrospinal fluid spaces, hypoplasia of the frontal lobe and falx cerebri, remodeling of anterior bone skull fossa with distortion of olfactory traits and scalloping of the adjecent bone.

DISCUSSION

Recently in the literature, PMA have been further studied with regard to their occurence, clinical and radiological features and treatment strategy [6]. PMA is considered a tumor of pediatric population and tend to affect younger age group as compared to PA [7, 8]. A recent study, Alkonyi B. et al. analyzed a population group of 15 children and confirm the youger age at tumor diagnosis compared to PAs. Their median age was 19 months, in accord to current literature (18 months) [9,10]. PMAs are mainly pediatric tumor, but reports have indicated that PMA can occur also in adolescents and adults. In their study, L.L.Linscott et al. revealed a larger age range with a mean age at presentation of 7 years. One third of their patients were adolescents and young adults [11].

PMA histology consists of a markedly myxoid matrix, with small, compact, piloid and higly monomorphous cells. Tumor cells are often located radially around vessels in a pattern that resembles the perivascular rosettes found in ependimomas [Burger PC 2005].

In 2013 Bhargava D. et coll reviewed the histology of all cases coded as Astrocytoma in the histopathology archivies from 1990 to 2003 and only the cases coded as PA or PMA were included. A population group of 91 patients (range 1-17 years) were analyzed, first study about the incidence of this tumor entity. Their results show that 9 patients had histological features of PMA (8 children and 1 adults). PMAs constitute about 10% of tumour previously diagnosed as PAs. The mean age at diagnosis was 3.33 years. In this study, the difference compare to data from literature may due to European setting of the study population based on geographic and cultural variation in disease demographics [6].

PMAs show a predilection for the hypothalamic/chiasmatic region [8, 9], nearly 2/3rds, but may occur anywhere along the neuraxis. L.L.Linscott et al. found that in almost half of patients PMA lesion was located outside this typical region (parietal lobe, temporal lobe, cerebellum, basal ganglia and fourth ventricle). The tumor in the adult age is mostly located in the hypothalamo-chiasmatic region [6, 11].

PMA has been shown to have a more aggressive behaviour with shorter volume doubling time [12] and worse prognosis as a result of higher recurrence rates, more frequent cerebral spinal fluid (CSF) dissemination and difficult in resection [13] compare to PA. CSF dissemination for PA was reported but it is rare [8].

Alkonyi B. et show a significantly higher MRI evidence of CSF dissemination at the time of diagnosis compared to PAs. Patients with PMA show shorter progression-free survival and higher local recurrence than those with PAs [9].

No difference in clinical presentation at diagnosis between the PMAs and PAs. Headaches was the most common symptom, followed by ataxic gait, nausea, diplopia and seizures. In the younger population are recorded developmental delay, failure to thrive, irritability, altered level of consciousness, feeding difficulty, vomiting, hemiparesis, and increasing head circumference [6].

Radiological features of PMAs reported in the previous studies are similar in the description. MRI is routinely performed to delineate the CNS neoplasm findings. This entity is well circumscribed, isointense on T1-weighted images and hyperintense on T2-weighted images, may have solid or cystic components without evidence of peritumoral edema or parenchymal infiltration and an usually homogeneous enhancement after the injection of gadolinium. Hydrocephalus was noted in 20% of cases. Khanani et al.[14] hightlighted that radiological features in their case resemble the findings of Tihan et al.[1] and Arslanoglu et al.[8], who described typical pilomyxoid tumors as enhancing lesions with central cystic component.

Nabavizadeh SA. et al. [13] found that PMA can be differentiated from PA with high degree of accuracy using Arterial Spin Labeling (ASL) perfusion imaging. In addition their results did not show any difference in diffusion imaging ADC values between PA and PMA, in keeping with previous studies which demonstrated that

neither of these tumors show restricted diffusion [10,11].No singular neuroimaging feature can actually reliably diagnose PMA.Our case was radiologically characterized by a polylobulated lesion with well circumscribed margins, dysomogeneous hypointense on T1-weighted images, hyperyntense on T2-weighted images. After injection of gadolinium, the lesion showed an heterogeneous markedly enhancement of the solid component with a central areas of hypointensity in T1, probably do to necrosis based on DWI features. It was observed a tentorial dural and lateral ventricles ependyma enhancement. An encasement of the Circle of Willis was found and on the right emisfere, in the fronto-parietal and basal ganglia region, ischemic appearance of the brain parenchyma was detected. They were also present perilesional enlargement of cerebrospinal fluid spaces and hypoplasia of the frontal lobes. These last finding are suggesting for slow growth development or hypothesis of a congenital lesion. To the best of our knowledge, there are no other reports of PMA with similar radiological findings in infants younger than 1 year.In the literature, patients with low-grade astrocytomas rarely present spontaneous intracranial hemorrhage, and only few authors have described bleeding associated with pilocytic astrocytoma [15]. High-grade gliomas and metastatic lesions bleed from involvement of neoangiogenesis vessels and tumor invasion of vessels. However, the mechanisms underlying hemorrhage in low-grade gliomas remain unclear. Two case of pilomyxoid astrocytoma presenting with intracranial hemorrhage have been reported [15,16]. Hamada et al. [15] described a case initially admitted with a suprasellar and third ventricle lesion characterized by acute intracranial hemorrhage, and a second fatal hemorrhage 4 months after the first operation. Gottfried et al. [16] report a case of acute spontaneous hemorrhage associated with a pilomyxoid astrocytoma in a 24-year-old man, unusual age onset for PMA. The location of the lesion induced to consider in the differential diagnosis hypotesis a glioma related to Neurofibromatosis type 1 (NF1). Alteration in the NF1 gene can lead a wide range of complications, including the development of benign and malignant neoplasms. Intracranial tumors are the bigger group of NF1-associated neoplasms and the age range more affected is under age of 10 years. In the literature are reported 3 cases of PMAs patient with NF1 [11,14]. All patients presented with PMA in the optic pathway/hypothalamus/third ventricle region. It is reported a rare case of PMA associated with Williams Syndrome in a 3 years old boy. The tumor location was the cerebellum. Williams syndrome apparently is not correlated with a predisposition for tumor, but several patients have developed malignancy in childhood and younger adults. The tumors was limited to lymphomas and gliomas [17]. It is also reported a patient with Noonan Syndrome that develop an optic pathway pilomyxoid astrocytoma [18].

CONCLUSION

PMA was previously considered as a more aggressive entity compare to PA occurring in youger children. **R**ecent studies have shown extensive histological and genetic overlap between PMA and PA with less certainty that the pilomyxoid variant always follows a more aggressive course than a more classic appearing suprasellar pilocytic astrocytoma.

Our case show a PMAs with the epicentre in the suprasellar region, most common location of this entity, in a 8-month-old boy with leptomeningeal dissemination. The very young age, the associated radiological feature of perilesional enlargement of cerebrospinal fluid spaces and hypoplasia of the frontal lobe suggest to consider the hypothesis of a congenital origin. Further studies should investigate this correlation.

REFERENCES

1. Tihan T, Fisher PG, Kepner JL, Godfraind C, McComb RD, Goldthwaite PT, Burger PC Pediatric astrocytomas with monomorphous pilomyxoid features and a less favorable outcome. J Neuropathol Exp Neurol, 1999; 58:1061-1068

2. Janisch W, Schreiber D, Martin H, Gerlach H: Diencephalic pilocytic astrocytoma with clinical onset in infancy. Biological behavior and pathomorphological findings in 11 children. Zentralbl Allg Pathol, 1985; 130:31-43, 1985
3. Brat DJ, Scheithauer BW, Fuller GN, Tihan T Newly codified glial neoplasms of the 2007 WHO Classification of Tumours of the Central Nervous System: angiocentric glioma, pilomyxoid astrocytoma and pituicytoma. Brain Pathol, 2007; 17(3):319–324
4. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, et al. The 2007 WHO Classification of Tumours of the Central Nervous System. Acta Neuropatho,l 2007; 114:97–109
5. Okano A, Oya S, Fujisawa N, Tsuchiya T, Indo M, Nakamura T, Soo Chang H, Matsui T Significance of radical resection for pilomyxoid astrocytoma of the cerebellum: a case report and review of the literature. Childs Nerv Syst, 2013; 29:1375-1379
6. Bhargava D, Sinha P, Chumas P, Al-Tamimi Y, Shivane A, Chakrabarty , Surash S, Novegno F, Crimmins D, Tyagi AK.. Occurrence and distribution of pilomyxoid astocytoma. Br J Neurosurg, 2013; 27:413-418
7. Mahore A1, Kammar A, Dange N, Epari S, Goel A. Diencephalic juvenile pilomyxoid astrocytoma with leptomeningeal dissemination. , 2011;21(2):222-5.
8. Arslanoglu A, Cirak B, HOrska A, Okoh J, Tihan T, Aronson L, Avellino AM, Burger PC, Yousem DM. MR Imaging characteristics of Pilomyxoid Astrocytomas. AJNR, 2003; 24:1906-1908
9. Alkonyi B, Nowak J, Gnekow AK, Pietsch T, Warmuth-Metz M. Differential imaging characteristics and dissemination potential of pilomyxoid astrocytomas versus pilocytic astrocytomas. Neuroradiology, 2015; 57:625-638
10. Komotar RJ, Zacharia BE, Sughrue ME et al. Magnetic resonance imaging characteristics of pilomyxoid astrocytoma. Neurol Res, 2008; 30(9):945–951
11. Linscott LL, Osborn AG, Blaser S, Castillo M, Hewlett RH, Wieselthaler N, Chin SS, Krakenes J, Hedlund GL, Sutton CL. Pilomyxoid Astrocytoma: Expanding the imaging spectrum AJNR, 2008; 29:1861-1866
12. Lee IH, Kim JH, Suh YL et al. Imaging characteristics of pilomyxoid astrocytomas in comparison with pilocytic astrocytomas. Eur J Radiol, 2011; 79(2):311-316
13. Nabavizadeh SA, Assadsangabi R, Hajmomenian M, Santi M, Vossough A. High accuracy of arterial spin labeling perfusion imaging in differentiation of pilomyxoid from pilocytic astrocytoma.. Neuroradiology. 2015; May;57(5):527-33
14. Khanani MF1, Hawkins C, Shroff M, Dirks P, Capra M, Burger PC, Bouffet E. Pilomyxoid astrocytoma in a patient with neurofibromatosis.Pediatr Blood Cancer. 2006 Mar;46(3):377-80.
15. Hamada H, Kurimoto M, Hayashi N, Nagai S, Kurosaki K, Nomoto K, Kanegane H, Nomura K, Endo S. Pilomyxoid astrocytoma in a patient presenting with fatal hemorrhage. Case report. J Neurosurg Pediatr. 2008;1(3):244-6. Review.
16. Gottfried ON1, Fults DW, Townsend JJ, Couldwell WT. Spontaneous hemorrhage associated with a pilomyxoid astrocytoma. Case report. J Neurosurg. 2003; 99(2):416-20.
17. Chonan M1, Kanamori M1, Kumabe T2, Saito R1, Watanabe M3, Tominaga T1. Pilomyxoid astrocytoma of the cerebellum with Williams syndrome: a case report. Childs Nerv Syst. 2013;29(7):1211-4. 2013 Apr 20.
18. Nair S1, Fort JA, Yachnis AT, Williams CA. Optic nerve pilomyxoid astrocytoma in a patient with Noonan syndrome. Pediatr Blood Cancer. 2015;62(6):1084-6

How to cite this article:
Chiara Carducci, Giovanna Stefania Colafati, Mariangela Marras, Matteo Di Nardo, Antonella Cacchione, Ioan Paul Voicu, Andrea Carai, Angela Mastronuzzi. *Suprasellar Pilomyxoid Astrocytoma With Leptomeningeal Dissemination In An Infant: A Case Report And Review Of The Literature.* Br J Bio Med Res , Vol.02, Issue 02, Pg.310-316, March-April 2018. ISSN:2456-9739 Cross Ref DOI : https://doi.org/10.24942/bjbmr.2018.214

Source of Support: Nil **Conflict of Interest:** None declared.