Case Report

SAGE Open Medical Case Reports

SAGE Open Medical Case Reports
Volume 6: 1–4

DOI: 10.1177/2050313X18761310
journals.sagepub.com/home/sco
SAGE

Intraorbital ancient pilocytic astrocytoma of the optic nerve in neurofibromatosis type 1 patient presenting with sudden ocular pain

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Abstract

Optic nerve pilocytic astrocytoma is an uncommon but well-known entity; however, intraorbital ancient pilocytic astrocytoma of the optic nerve is extremely rarely reported. To our knowledge, this is the first detailed description regarding the intraorbital ancient pilocytic astrocytoma, reported in available English literature, to date. We presented an extremely unusual neurofibromatosis type 1 case of a 17-year-old male's sudden ocular pain secondary to intraorbital pilocytic astrocytoma of the optic nerve with markedly cystic degeneration, fluid production, and hemorrhage, due to ancient and possibly ruptured glioma. Future prospective studies are required to validate the significance of intraorbital ancient pilocytic astrocytoma arising from the optic nerve and the close correlation with ruptured cystic degeneration and ocular pain, after collecting and investigating a larger number of its cases examined.

Keywords

Pilocytic astrocytoma, ancient, degeneration, optic nerve, ocular pain

Date received: 8 November 2017; accepted: 1 February 2018

Introduction

Optic nerve pilocytic astrocytoma (PA) is an uncommon but well-known entity;[1,2] however, intraorbital ancient PA of the nerve is extremely rarely reported, composed predominantly of spindle-shaped, bipolar astrocyte-like cells with prominent degenerative features including cystic changes, extracellular deposits, or calcification.[1] To our knowledge, this is the first detailed description regarding the intraorbital ancient PA, reported in available English literature, to date.

Case presentation

A 17-year-old male patient with neurofibromatosis (NF) type 1 and mental retardation presented with sudden ocular pain, during the ophthalmologic follow-up of neovascular glaucoma and vitreous hemorrhage in the left eye. Ten months before his emergency surgery for enucleation of left eyeball, the crescent-like intraorbital solid content measured approximately 20 mm × 10 mm in the posterior eyeball and generated a low (grayish) signal intensity on T2-weighted magnetic resonance imaging (MRI) (Figure 1(a)). There were neither apparent signs of optic nerve or surrounding orbital tissue involvement, nor any intracranial tumor lesions (Figure 1(a)). On gross examination of the surgical specimen, the sagittal cut surface of the left eyeball characteristically displayed an oval, well-demarcated nodular and solid lesion, yellow-whitish in color, measuring approximately 10 mm × 5 mm in diameter and likely floating in the abundant bloody to serous fluid, coexisted with a subsequent cystic component (Figure 1(b)). A gross connection between its solid tumor part and the optic nerve was not evident (Figure 1(b)). On its scanning magnification, this intraorbital tumor was found to contain a large volume of eosinophilic serous

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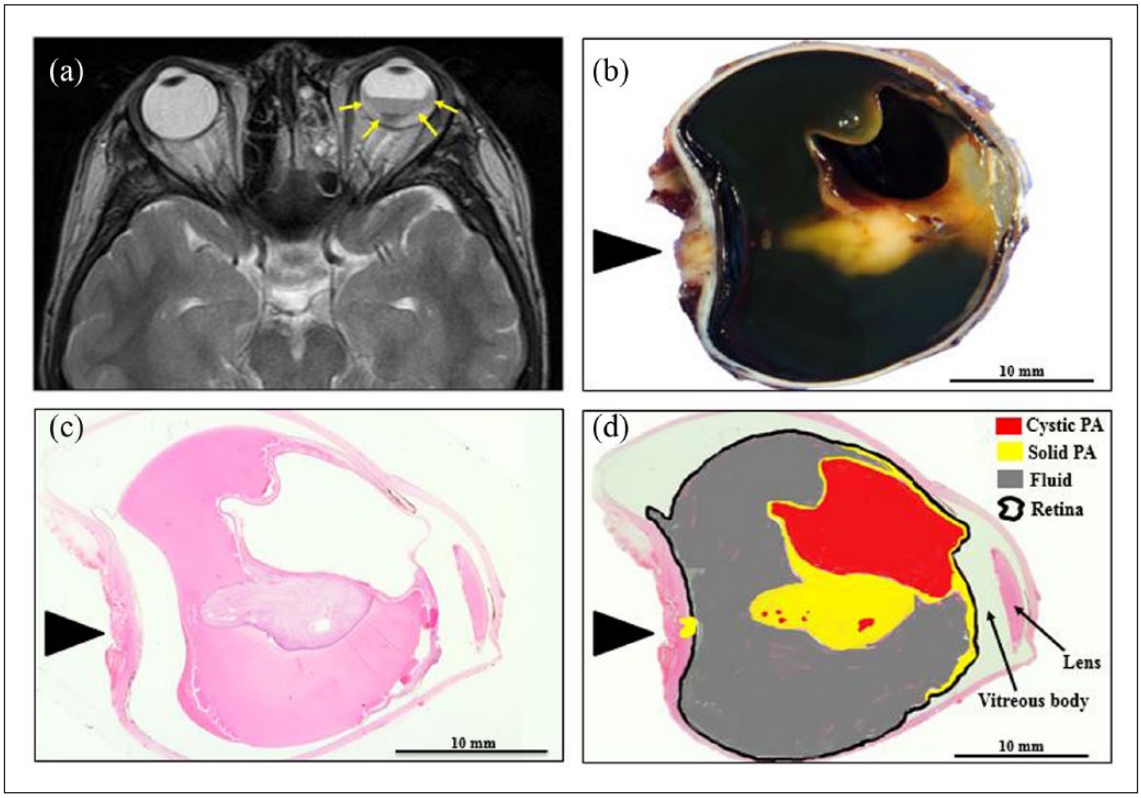

Figure 1. Imaging, gross and microscopic examination, and schema of the enucleated eyeball containing the intraorbital ancient PA arising from the optic nerve. (a) T2-weighted MRI scanning 10 months before his emergency surgery showed that the crescent-like intraorbital solid content measured approximately 20 mm × 10 mm in the left posterior eyeball (arrows), generating a low (grayish) signal intensity in marked contrast to a high (white) signal intensity of the vitreous body. There were neither apparent signs of optic nerve or surrounding orbital tissue involvement, nor any intracranial tumor lesions. (b) The enucleation of the left eyeball was performed, and a gross examination of the sagittal cut surface revealed an oval, well-demarcated nodular and solid lesion, yellow-whitish in color, measuring approximately 10 mm × 5 mm in diameter and likely floating in the abundant bloody to serous fluid, coexisted with a subsequent cystic component. A gross connection between its solid tumor part and the optic nerve (arrowhead) was not evident. Bar = 10 mm. (c) On scanning magnification of the surgical specimen (H&E stain), this intraorbital tumor contained a large amount of eosinophilic serous fluid admixed with a number of red blood cells, displaying a likely floating and well-circumscribed but distorted nodule, uniquely accompanied by frequent, large to small cystic degeneration. Retina was mostly detached and overtly separated by this intraorbital ancient PA arising from the optic nerve (arrowhead). Bar = 10 mm. (d) The schema of its ancient optic nerve PA components (cystic PA, red; solid PA, yellow; and fluid, gray) was shown, involving the orbital portion of the optic nerve (arrowhead), detaching/separating the retina (bold black line), and likely demonstrating a sequential degeneration with possible rupture. This intraorbital ancient PA strongly compressed the pre-existing vitreous body, resulting in severe lens luxation; however, there was no apparent infiltrative growth fashion. Bar = 10 mm (H&E stain).

fluid with a number of red blood cells and showed a well-circumscribed but distorted nodule, uniquely accompanied by frequent, large to small cystic degeneration (Figure 1(c)). Resection was diagnosed as complete. After the thorough histopathological examination, retina was mostly detached and overtly separated by this intraorbital ancient PA of the optic nerve (Figure 1(c) and (d)). The intraorbital ancient PA strongly compressed the vitreous body, resulting in severe lens luxation (Figure 1(c) and (d)). The schema of each cystic or solid PA component was shown, involving the orbital portion of the nerve, and likely demonstrating a sequential degeneration with possible rupture (Figure 1(d)).

Microscopic findings showed that the solid PA component was noted in the optic nerve and that this solid part arising from the optic nerve uniquely permeated the retina and was protruded from the orbital portion of the nerve into the eyeball (Figure 2(a)). Foci of arachnoidal hyperplasia around the optic nerve were absent (Figure 2(a)). Medium-power view revealed that the tumor predominantly comprised a cellular to acellular (biphasic) proliferation of characteristic spindle-shaped, bipolar astrocyte-like cells having mildly enlarged, pleomorphic nuclei and elongated, hair-like processes with very few mitotic figures (Figure 2(b) and (c)), associated frequently with cystic and hyalinized degeneration and eosinophilic fluid production (Figure 2(b)). Those pilocytic cells appeared as palisading features around cystic spaces or blood vessels (Figure 2(b)). Rosenthal fibers were rarely seen, but foci of other degenerative changes

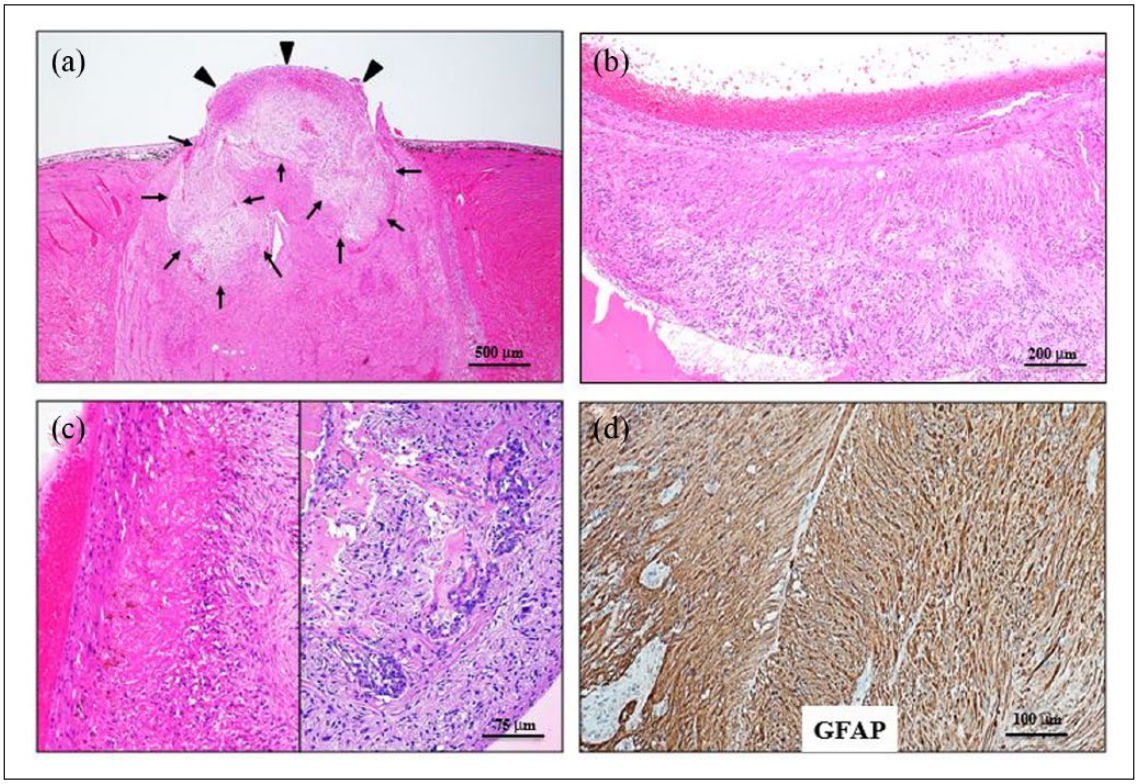

Figure 2. The histopathological and immunohistochemical examination of the intraorbital ancient PA arising from the optic nerve in the enucleated eyeball. (a) Under a low-power view (H&E stain), the solid PA component was also identified in the optic nerve (arrows), and this solid part arising from the optic nerve uniquely permeated the retina and was protruded from the orbital portion of the nerve into the eyeball (arrowheads). Foci of arachnoidal hyperplasia around the optic nerve were absent. Bar = 500 µm. (b) Microscopic findings of medium-power view showed that the PA predominantly composed of a cellular to acellular proliferation of characteristic spindle-shaped, bipolar astrocyte-like cells having mildly enlarged, pleomorphic nuclei and elongated, hair-like processes with very few mitotic figures in a biphasic fashion, associated frequently with cystic and hyalinized degeneration (top and left bottom) and eosinophilic fluid production (left bottom). Those pilocytic cells of PA appeared as palisading features around cystic spaces or blood vessels. Bar = 200 µm (H&E stain). (c) A high-power view (H&E stain) uniquely detected scattered coagulative tumor necrosis with a pseudopalisading feature (left), and glomeruloid microvascular proliferation (right) in this intraorbital ancient PA of the optic nerve, reminiscent of high-grade gliomas. Bar = 25 µm. (d) In immunohistochemistry, those pilocytic tumor cells were diffusely and strongly positive for GFAP. Bar = 100 µm.

including calcification or fibrosis were evident. Interestingly, a high-power view detected scattered coagulative tumor necrosis with pseudopalisading features, and glomeruloid microvascular proliferation, reminiscent of high-grade gliomas (Figure 2(c)). However, there was no apparent infiltrative growth fashion. Immunohistochemically, these neoplastic cells were diffusely and strongly positive for glial fibrillary acidic protein (GFAP, diluted 1:40; Nichirei Bioscience, Tokyo, Japan; Figure 2(d)), whereas completely negative for epithelial membrane antigen (EMA, diluted 1:1; Dako Cytomation Co., Glostrup, Denmark), isocitrate dehydrogenase 1 (IDH-1, diluted 1:100; Dianova, Lyngby, Denmark), and p53 (diluted 1:1000; Dako). In addition, the Ki67 (MIB-1, diluted 1:1; Dako) labeling index was much less than 2.2% in the proliferating tumor cells. All of the immunohistochemical stainings were conducted using the Dako Envision kit in accordance with the manufacturer's instructions. Overall, these features could be diagnosed as optic nerve glioma, named intraorbital ancient PA of the optic nerve. To date, after approximately 7 months of postoperative follow-up, the patient remains well without recurrence.

Discussion

It is very likely that the current case is clinicopathologically remarkable for two reasons at least: first, in case of the present histopathological findings for pseudopalisading necrosis and glomeruloid microvascular proliferation, we pathologists should consider the very rare possibility of glioblastoma arising from the optic nerve.[2] Despite that, we must be aware that optic nerve ancient PA can be easily distinguished from high-grade glioma, by clinicopathological features of neither infiltrative/invasive growth pattern nor increase in mitotic rate and MIB-1 index, as shown here. In addition, nuclear pleomorphism and/or cystic degeneration

are strongly manifested by the slowly growing and long-standing nature of these tumors. Second, our case unusually presented sudden ocular pain, secondary to frequent cystic degeneration, fluid production, and hemorrhage, due to ancient and possibly ruptured PA. In fact, this patient is much older than reported, since optic nerve gliomas tend to occur in children with NF type 1 (mean, 4.9 years).[1,3] Furthermore, follow-up MRI inconsistently showed no overt degenerative features, such as cysts or fluids, even 10 months before his surgery. Although most juvenile PAs are indolent, growing very slowly,[1,2] in cases with a strong clinical suspicion of intraorbital ancient PA, it should be raised to alert the ophthalmologists to the emergency surgery, at the very least. Nevertheless, it would be intriguing to assess the significance of unique clinicopathological findings and the close correlation with ruptured cystic degeneration and ocular pain on future larger studies.

Conclusion

We herein reported a very unique surgical case of intraorbital ancient PA of the optic nerve, associated with ruptured cystic degeneration and sudden ocular pain, in a NF type 1 patient. The present case report should interest the scientific community, taken together with new clinicopathological findings, even though the ancient changes of this glioma are histopathologically a well-known feature.

Acknowledgements

The authors thank Nobuhiro Kuroiwa, MD (Opthalmologist), Kei Matsuo (Technician), Orie Iwaya (Technician), and Yoshie Jitoho (Technician) for their expert technical assistance and helpful comments. S.Y. and H.L.M. contributed to study concepts and design. S.Y. and H.L.M. contributed to data acquisition. S.Y., H.L.M., S.N., N.K., and A.T. contributed to data analysis and interpretation. S.Y. contributed to manuscript preparation. S.Y., H.L.M., and A.T. contributed to manuscript review. All authors read and approved the final manuscript.

Availability of data and materials

The dataset supporting the findings and conclusions of this case report is included within the article.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

Funding

The author(s) disclosed receipt of the following financial support for the research, authorship, and/or publication of this article: This work was partly supported by a grant from the Kanagawa Medical University Fund, Ishikawa, & Kodama Memorial Fund for Medical Research, Kagoshima, Japan (to S.Y.).

Informed consent

Written informed consent was obtained from the legally authorized representative of the minor patient for the publication of this case report.

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