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Case Report

Hydrocephalus in children – A rare case of pineal cavernoma and literature review

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ABSTRACT

Background: Cavernous malformations prevalence ranges from 0.4 to 0.6% and accounts for 5–15% of all central nervous system vascular malformations. Pineal cavernomas constitute <1% of all locations published in the literature, with a total of 26 cases reported, only 5 regarding the pediatric population until 2020. Overall annual hemorrhage rate is 2.4%. Symptoms are often due to hydrocephalus and intracranial hypertension.

Case Description: We report a case of a 5-year-old child with visual disturbances, headache, and progressive neurologic deterioration. MR showed a lesion in the pineal region and triventricular hydrocephalus. She was submitted to endoscopic third ventriculostomy and total excision of the lesion by the infratentorial supracerebellar approach a few days later. Histopathological examination confirmed a pineal cavernous malformation. The patient returned to her normal life without any neurologic deficit and a normal development.

Conclusion: The ideal treatment is primary lesion removal; however, due to the infrequency and because it is a curable lesion, studies seeking to deepen the knowledge of this disease are considered relevant.

Keywords: Cavernous malformation, Hydrocephalus, Pediatric, Pineal, Vascular disorders

INTRODUCTION

Cavernous malformations (CMs) prevalence ranges from 0.4% to 0.6%[9] and accounts for 5–15% of all central nervous system vascular malformations, mostly in the supratentorial compartment.[3,9]

Location in the pineal region is particularly uncommon,[2] constituting <1% of all locations published in the literature. As described in Table 1, there are only 26 cases reported until 2020 in all the population.[4,14,16] The first of those reported in 1961, with a subtotal surgical resection and postoperative hemorrhage, resulting in the patient's death.

The literature about the natural history of CMs in the pediatric population is limited, but the development of CMs appears to increase with age, reaching a plateau in late adolescence.[1]

Pineal cavernomas are even more rare in the pediatric population – the youngest patient was 4 weeks old[7] with a total of five cases in the pediatric population until 2020.[8]

Cavernous malformations are clusters of dilated sinusoidal channels lined by a single layer of endothelium and lack of smooth muscle or normal brain tissue, thus classified as angiographically occult.[12,17]

The etiology of these malformations is widely discussed, but there is evidence of autosomal dominant inheritance located on chromosome 7. Radiotherapy is a known risk factor.[2] The overall annual hemorrhage rate is 2.4% per patient and year, with prior hemorrhage and female sex increasing the risk of subsequent hemorrhage.[6]

The symptoms are often due to hydrocephalus and intracranial hypertension.[14] Due to the risks inherent to approach the pineal region, some of these rare lesions were managed mistakenly with empiric radiation, without a diagnosis.[17]

CASE REPORT

We report a case of a 5-year-old child sent from an African country who presented visual disturbances and headache with a few months of development, progressive deterioration, and abrupt onset of vomiting and irritability.

The child was evacuated to Portugal with a CT-scan that showed a triventricular hydrocephalus, an MR being immediately performed. This examination showed a lesion in the pineal region and triventricular hydrocephalus, as described in Figure 1. The differential diagnosis for this lesion included a germ cell tumor, a neoplastic lesion of the pineal gland parenchyma, a glial series tumor, or a vascular malformation.

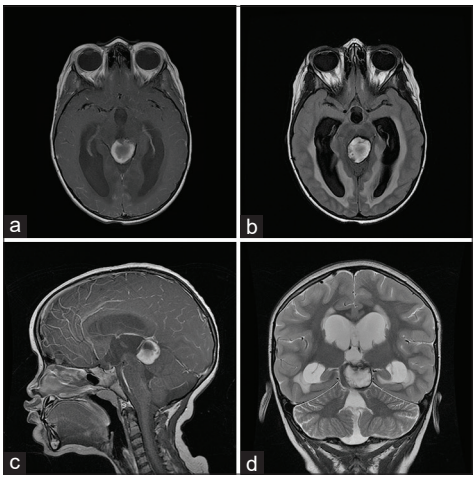

Figure 1: (a) Preoperative axial T1 ponderation – bleeding centered in the region of quadrigeminal lamina/pineal region associated with small round heterogeneous on his left anterior aspect with triventricular hydrocephalus, (b) axial FLAIR showing active hydrocephalus with ependimary transudation, (c) sagittal T1 gadolinium showing pineal lesion with Sylvius aqueduct obstruction, (d) T2 coronal showing pineal lesion with consequent hydrocephalus, the core of the lesion is surrounded by a halo of lower intensity due to hemosiderin.

The patient was immediately submitted to endoscopic third ventriculostomy with a resolution of hydrocephalus. She later underwent suboccipital craniotomy and total excision of the lesion by the infratentorial supracerebellar approach a few days after [Figure 2].

Glial tissue fragments with recent hemorrhage, dilated sinusoidal channels, and infiltration by histiocytes containing hemosiderin confirmed the suspicious diagnosis considering the MR characteristic of pineal cavernous malformation [Figure 3].

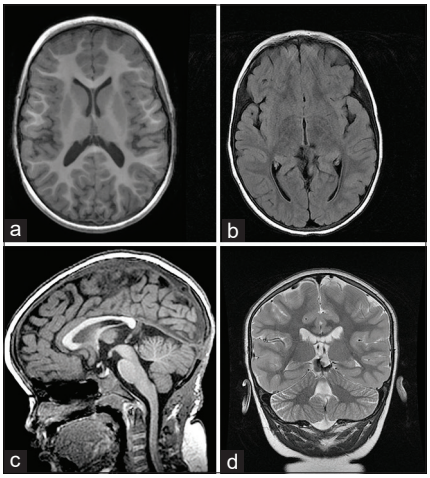

Figure 2: (a) Axial T1 2 months after endoscopic ventriculostomy and total excision of the lesion of the pineal region, (b) axial FLAIR showing nondilated temporal horns, (c) T1 sagittal showing repermeability of Sylvius aqueduct, and (d) T2 coronal showing a small blood residue, no mass effect and without hydrocephalus signals.

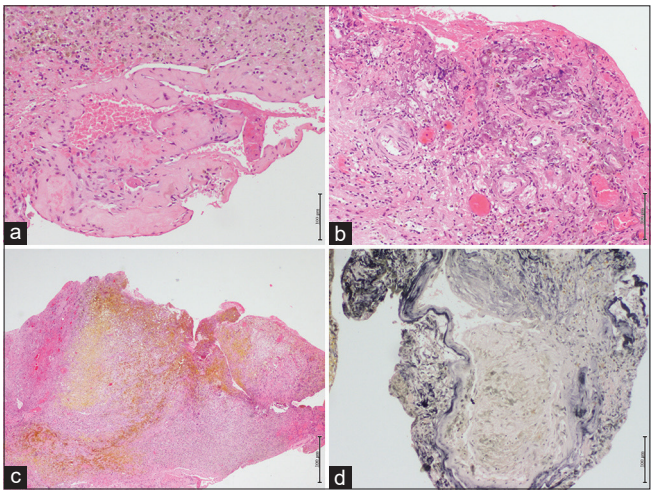

Figure 3: (a) Hematoxylin-eosin shows an agglomerate of vessels of different sizes and thin walls, (b) hematoxylin-eosin shows vessels surrounded blood and histiocytic infiltrate with hemosiderin pigment, (c) hematoxylin-eosin with signs of old hemorrhage, and (d) Verhoeff coloration showing elastic fibers in black.

Table 1: A review of all the cases reported since the first pineal cavernous malformation in 1961.

Author	Age	Gen	ICH	Parinaud	Ocular sympt	Other sympt	Familiar history	Angio	CT	MR	Radio	Surgery	Out	Histology
Miller, 1961	35	F	+	-	+	SAH	-	NR	NR	NR	+	NR	Dead	CM
Mirecka, 1965	3,5	M	+	-	+	Hemiparesis	-	NR	NR	NR	NR	NR	Dead	Mixed Hemangioma
Clark 1970	27	M	+	-	+	Psychomot	+	NR	NR	NR	NR	NR	Dead	CM
Hubschman 1976	45	M	+	+	+	Lethargy	-	N	Hiper	NR	NR	RT	Cure	CM
Vaquero 1980	18	F	-	-	+	Amenorrhea	-	N	Hiper, C-	NR	NR	RT	Cure	CM
Vaquero, 1980	22	M	+	-	-	-	-	NR	Hiper, C+	NR	+	RT	Dead	CM
Sonntag, 1981	3 w	F	+	+	+	-	-	N	Hiper, C+	NR	NR	RT	Cure	Mixed Hemangioma
Fukui, 1983	22	M	+	+	+	DI	-	N	Hiper, C+	NR	+	RT	Cure	CM
Combelles, 1983	39	F	-	-	+	Hemiparesis	+	NR	Hiper, C+	NR	+	Biopsy	Dead	CM
Mazza 1989	<18	?	?	-	?	?	+	?	?	NR	?	?	?	?
Donati 1992	12	M	+	-	-	-	-	NR	Hiper, C-	Mixed	NR	NR	Good	?
Slavin 1994	23	F	+	-	+	-	-	N	Hiper, C+	Mixed, C+	NR	RT	Good	CM
Lombardi 1996	19	F	+	+	+	-	-	Abnormal vein	Hiper, C-	Iso (T1), hipo (T2), C+	NR	RT	Cure	CM
Lombardi 1996	58	F	-	-	+	-	-	N	Iso	Mixed, hipo perilesional (T1)	NR	RT	Cure	CM
Lombardi 1996	73	F	+	+	+	-	-	Abnormal vein	Hiper	Mixed, hipo perilesional (T1)	NR	NR	Good	?
Vishteh 1998	31	F	+	+	+	-	-	NR	NR	Hipo (T1) Hiper/ hipo (T2), C+	NR	RT	Cure	CM
Muzumdar 2000	45	M	+	+	+	Cerebellar signs	-	N	Hiper	Iso-hiper (T1) Hipo perilesional (T1) C+	NR	RT	Cure	CM
Kobayashi 2001	11	F	+	+	+	Lethargy	-	N	Iso	Hiper (T1) Mixed (T2) ring C+	NR	RT	Cure	CM
Vhora 2001	55	F	+	-	+	-	-	N	Hiper, C+	NR	NR	RT	Good	CM
Kim 2005	42	F	+	+	+	-	-	N	Hiper	Hiper-iso (T1) Hipo (T2)	NR	RT	Cure	CM
Kim 2005	37	F	+	-	+	-	-	N	Hiper	Mixed	NR	RT	Good	CM

(Contd...)

Table 1: *(Continued).*

Author	Age	Gen	ICH	Parinaud	Ocular sympt	Other sympt	Familiar history	Angio	CT	MR	Radio	Surgery	Out	Histology
Chamadoira 2010	57	F	+	+	+	-	-	N	Hiper	Hiper (T1), Hiper-iso (T2) hipo ring	NR	RT	Cure	CM
Hernesniemi 2011	80	M	-	-	-	Vertigo	-	N	NR	NR	-	RT	Cure	CM
Hernesniemi 2014	33	M	+	+	+	-	-	N	NR	NR	NR	RT	Good	CM
Chenisz 2015	67	F	+	-	+	Ataxia	-	NR	Hiper	Hiper (T1)	NR	RT	Cure	CM
Ogura 2017	47	F	+	+	+	-	-	-	Hiper	Hiper (T1), Hiper-iso (T2) hipo ring	-	RT	Cure	CM
Current case	5	F	+	+	+	-	-	NR	Hiper	Hiper (T1), Hiper-iso (T2) hipo ring	NR	RT	Cure	CM

ICH: Intracranial hypertension, MR: Magnetic resonance, N: No, NR: Nonreported, Out: Outcome, Radio: Radiotherapy, RT: Total resection. Angio: Angiography, CM: Cavernous malformation, CT: Computed tomography, Gen: Gender

Postoperatively, the patient presented Parinaud syndrome, which disappeared 3 months after surgery. The patient returned to her daily life without any neurologic deficit and a normal development.

DISCUSSION

Symptoms and clinical approaches to cavernous malformations depend on lesion characteristics and location. Supratentorial lesions most commonly present seizures, headache, or focal neurological deficits; however, many patients are asymptomatic.[9]

Of the total cavernous malformations, 9–35% occur in the brainstem,[7] the probability of pineal gland region involvement being <1%.[2]

Since pineal cavernous malformations were difficult to diagnose before the introduction of MR, some incidences of this disorder were erroneously treated with irradiation, before surgical resection was performed.[5,13]

Cavernomas are best appreciated on T2-weighted spin-echo MR, demonstrating a classic signature of "popcorn" or "berry" appearance, surrounded by a ring of low signal intensity due to hemosiderin.[11]

Cavernous angioma located in the pineal gland is usually accompanied by supratentorial hydrocephalus, with dilation of lateral and third ventricles, the fourth ventricle being preserved. About 75% of patients had partial or complete Parinaud's syndrome after treatment of pineal region lesions.[10]

In our case, the patient presented hydrocephalus and hemorrhage in the pineal region, tumors being the most common cause for the latter.[11]

These malformations are very rare in the pediatric population – the youngest patient being 4 weeks old,[7] the oldest 80 years old, and a total of only five cases in the pediatric population until 2020. Familiar occurrence is known to follow an autosomal dominant pattern of inheritance, with variable penetrance. Our patient had no family history of vascular malformations.

The clinical manifestations related to the disease vary; a common manifestation is the sign of Parinaud, characterized mainly by difficulty in eye movement, superiorly; others may be the signs of intracranial hypertension, headache, visual disturbances, gait ataxia, and changes in circadian rhythm. In addition to these, there are still signs of hemiparesis, hemi-hypoesthesia, diabetes insipidus, and amenorrhea, with high prolactin, neuroendocrine disorders occurring mainly due to damage to the hypothalamus region by distention of the third ventricle floor.[15]

Recent studies[5,9,11] recommend surgical intervention in the case of clinically significant hemorrhage or progressive neurological deterioration, or if the MR imaging reveals suspicion of pineal cavernous malformation.

Direct microsurgical excision of the vascular malformation is the standard treatment and when completely removed, the risk of further growth or hemorrhage of cavernous malformations is very low,[16] and there is a high rate of cure and good outcome.[15]

The approaches that are generally used to access the pineal region are supracerebellar infratentorial and occipital transtentorial.[8,13,16] Whatever approach is selected, skillful and clean microneurosurgery preserving the normal anatomy is imperative during pineal region operations.[16]

Standard management options for CCMs classically include observation and surgical removal.[8] Asymptomatic lesions are generally treated conservatively. Surgery is indicated for accessible symptomatic lesions. Complete resection eliminates the risk of hemorrhage from that particular lesion but may lead to neurological morbidity, especially for CCMs located in the eloquent cortex or brainstem. A timely approach is important to minimize treatment morbidity.[8]

Procedures to treat hydrocephalus, such as endoscopic ventriculostomy, should be performed if definitive treatment cannot be carried out immediately.[16]

Timely microsurgery should be considered several weeks after hemorrhage, to allow perilesional swelling to subside, as well as hematoma contents to evolve and soften, so when there is an emergency situation with hydrocephalus, an ETV should be performed immediately, as well as surgery a few days later.[8]

The role of radiosurgery in the treatment of cavernous malformations remains controversial. Stereotaxic radiosurgery has been advocated for the treatment of small cavernous angiomas in critical areas. A significant decrease in the bleeding rate has been found after 3 years of treatment.[9] Long-term studies with careful clinical and imaging follow-up are necessary to establish the true role of radiosurgery in the treatment of this condition.

CONCLUSION

The ideal treatment is primary lesion removal;[16] however, due to the infrequency and because it is a curable lesion, studies seeking to deepen the knowledge of this disease are considered relevant.[6]

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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