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Original Article

The clinical spectrum and management outcome of adult-onset aqueductal stenosis: Insight from South-West Nigeria

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ABSTRACT

Background: Adult-onset aqueductal stenosis (AOAS) is an uncommon cause of hydrocephalus in adults. Its etiopathogenesis is poorly understood, with various proposed mechanisms, spectrum of presentation, and management. Very little has been reported on this anomaly in African literature. This study aimed to describe the pattern of presentation, clinical spectrum, and early outcomes following the shunt procedure in our population practice setting.

Methods: This was a retrospective case series of patients with non-tumoral AOAS managed between 2008 and 2023 in a tertiary center in South-West Nigeria. Relevant demographic, clinical-radiologic, and outcome data were retrieved and analyzed.

Results: There were seven males and one female. Their age ranged from 18 to 50 years. The duration of symptoms ranged from 3 to 120 months. All patients admitted having headaches; however, visual deterioration (n = 5) was the most common presenting symptom. Features of endocrinopathy (n = 1), cerebellar dysfunction (n = 2), cognitive deficit (n = 2), spontaneous cerebrospinal fluid leak (n = 1), and sphincteric dysfunction (n = 1) were also observed. The mean Evan's index at the presentation was 0.43. Ventriculoperitoneal shunt (VPS) insertion was performed in seven patients with good outcomes. One patient opted for a referral. One patient had shunt revision 6 years later on account of shunt disconnection.

Conclusion: AOAS is an infrequent cause of hydrocephalus in our setting, with most patients seeking medical consultation following visual deterioration. Proper evaluation of adults with long-standing headaches and associated features is advocated to avert total visual loss in this subgroup. VPS insertion is a viable treatment option with a good outcome.

Keywords: Adult-onset aqueductal stenosis, Africa, Profile, Ventriculoperitoneal shunt

INTRODUCTION

Aqueductal stenosis (AS) is a common cause of obstructive hydrocephalus, especially in neonates and infants. It is characterized by a narrowed cerebral aqueduct and tri-ventriculomegaly (of the lateral and third ventricles).[2] While it is commonly diagnosed in children, adult-onset presentation is rare and accounts for approximately 10% of hydrocephalus cases in this subgroup.[1] Histologically, nontumoral AS is classified as stenosis, forking, septum formation, and

gliosis.[2] Adult-onset AS (AOAS) may manifest with features of increased intracranial pressure (ICP); however, unusual presentations have been reported, including Parkinson-like features, hypopituitarism, cranial neuropathies, hearing loss, psychiatric disorders, and spontaneous cerebrospinal fluid (CSF) leaks.[11,13] Treatment modalities are shunt diversion and endoscopic third ventriculostomy (ETV).[14] At present, there is no high-level evidence to determine the criterion standard for care.[5] Our understanding of AOAS emanated mainly from a few cohort studies and case reports conducted in Western countries. Studies specific to AOAS from Africa appear very scanty, and an electronic search yielded one.[4,10,13,14] In a series from Egypt, Mohammed *et al.* reported on 25 patients with late-onset AS. However, some of the patients included in the study were within the pediatric age range (5–64 years), and no subgroup analysis of clinical features and outcomes was performed for those within the adult age range.[10] This study was to describe the clinical spectrum and outcomes of managing adults with nontumoral AS at our institution. A relevant literature review was also presented, emphasizing the clinical profile and proposed etiopathogenesis.

MATERIALS AND METHODS

This is a single institutional retrospective review of cases of AOAS managed at our tertiary institution between 2008 and 2023. Only patients aged ≥18 years with non-tumoral AS were included in the study. Patients with a history of cranial surgery or those diagnosed with hydrocephalus during infancy were excluded from the study. All patients underwent detailed neurological examinations and neuro-ophthalmic evaluations. AS in the absence of any extrinsic cause was confirmed using brain magnetic resonance imaging (MRI) studies. Patients' neuroimages were analyzed using RadiAnt DICOM Viewer (version 2024.1, Beta). The third ventricular width was defined as the maximum interthalamic distance at the level of the foramen of Monro. Relevant demographic, clinical-radiologic, and management outcome data were retrieved in a predesigned form using Microsoft Excel. These variables were analyzed using IBM Statistical Package for the Social Sciences version 27, and the mean, median, and frequency of various variables were presented. Following ventricular-peritoneal shunt insertion, the assessment of clinical features was classified as resolution (complete remission of clinical feature[s]), improved, and stable (no improvement or deterioration). The outcome was good if ≥50% of the patient's clinical features improved and poor if <50% of clinical improvement.

RESULTS

Within the review period, 63 patients presented with adult-onset hydrocephalus. However, only 8 (12.7%) met the selection criteria. There were seven males and one female. The median age at presentation was 29 years. The median duration of symptoms was 9 months. Based on the Fukuhara classification, two had subacute symptomatology, while the others had chronic symptomatology. While all patients admitted having headaches, visual deterioration (n = 5) was the most common reason for presentation. The mean Evan's index and maximum third ventricular width at presentation were 0.43 (range 0.40–0.48) and 2.04 cm (range: 1.28 cm–3.44 cm), respectively. Visual acuity and fundoscopy were normal in three cases [Serial Nos. 2, 8, and 6, Table 1]. The abnormal fundoscopic findings were secondary optic atrophy. Two patients admitted having prior mild head injuries. None of the patients had any other associated congenital anomalies.

Seven patients had ventriculoperitoneal shunt (VPS) insertion, the treatment modality available in our institution. The remaining one opted for a referral. The intraoperative findings were clear CSF under high pressure during ventricular cannulation. All CSF cultured from intraoperative specimens returned negative. Headache was the first symptom to subside following CSF diversion, and all admitted to complete resolution. All patients who had shunt insertion had good outcomes, with one patient [serial No. 4, Table 1] developing a VPS malfunction 6 years later requiring shunt revision. The mean follow-up period was 18 months (2–73 months). Further details are provided in Table 1. Figures 1 and 2 are representative brain MRI scans of patients that we managed. Figure 3 was the cranial computed tomography scan image delineating the bony defect in our patient with CSF rhinorrhea.

DISCUSSION

Our series included eight patients with non-tumoral AOAS. Although retrospective, with a small sample size and short follow-up, we present early comprehensive data on this anomaly in our subpopulation to aid in promoting awareness of this condition. A male preponderance was observed in our series, similar to that of Tisell *et al*., Locatelli *et al*., and Montemurro *et al*.[8,11,15] In contrast, Fukuhara and Luciano noted a female preponderance.[4] Martinoni *et al.* reported an equal sex distribution.[9] While there appears to be a male trend in the available reports, AOAS has no clear-cut sex predilection.[16] Its proposed that multifactorial etiologies may explain this observation.

Whether AOAS is primarily idiopathic, due to a primary insult in adulthood, or a delayed presentation of congenital AS remains unclear. Kita *et al.* reported a case of late-onset idiopathic AS in a 17-year-old male with an earlier normal ventricular system from previous MRI scans performed in childhood.[6] In contrast, Oi *et al.* posited that AOAS may be due to decompensated long-standing ventriculomegaly that is likely not detected in childhood.[13]

Table 1: Clinical spectrum and outcome.

Serial number	Age at presentation (years)	Sex	Duration of symptoms (months)	Presenting complaint	Other symptoms	Signs	Outcome (time of assessment, post-op)
1	18	Male	11	Visual impairment	Headache, gait abnormality, vertigo	Ataxic gait	Improved vision Resolution of other symptoms. (3 months)
2	19	Male	5	CSF rhinorrhea	Headache	Reservoir sign	Resolution of headache with reduced CSF rhinorrhea* (2 weeks)
3	23	Female	36	Visual impairment	Headache, amenorrhea, urinary incontinence, cold intolerance	Right abducens nerve palsy	Improved vision Resolution of other symptoms (1 month)
4	35	Male	7	Visual impairment	Headache, dizziness		Visual impairment remained stable. Resolution of other symptoms (1 year)
5	46	Male	8	Visual impairment	Headache, lower limb weakness, and memory impairment	Hemiparesis (MRC grade 4)	Resolution of headache and hemiparesis, improved memory. Visual impairment remained stable (2 months)
6	50	Male	120	Headache	Memory impairment	Horizontal Nystagmus, fine tremors	Opted for referral
7	36	Male	10	Visual impairment	Headache		Resolved headache, visual impairment stable (3 months)
8	19	Male	3	Headache			Resolved (1 week)

*Planned for surgical repair, Op: Operative, CSF: Cerebrospinal fluid, MRC: Medical Research Council

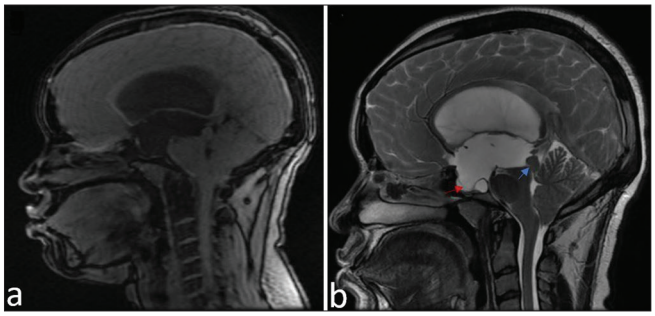

Figure 1: (a) T1 sequence depicting triventriculomegaly with stenosis of the aqueduct and small fourth ventricle and (b) T2 sequence depicting the downward bowing of the third ventricle with an enlarged and empty sella (indicated by the red arrow). The site of aqueductal stenosis is indicated with a blue arrow.

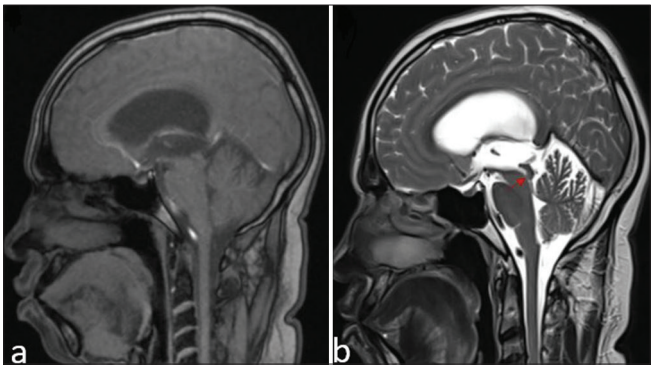

Figure 2: (a) T1 sequence depicting triventriculomegaly with a small fourth ventricle and stenosed aqueduct. (b) T2 sequence showing the area of stenosis (indicated by the red arrow).

Similar to the neuroimaging findings in three of our patients [Figures 1 and 3], they opined that the observed expanded or empty sella turcica is a pointer of long-standing ventriculomegaly.[13] The chronicity of the symptoms observed in our series (3–120 months) suggests a long-standing pathology. However, only one patient's symptoms predated adulthood [serial No. 1, Table 1].

Pyogenic ventriculitis and intraventricular hemorrhage can result in secondary stenosis of the aqueduct through gliosis due to inflammation and occasionally from direct blockage with fibrinopurulent debris or clot, respectively.[2,14] None of the patients in our cohort had a history suggestive of this. Various mechanisms have been postulated on the role of head trauma. It is thought to be an inciting factor through the early-

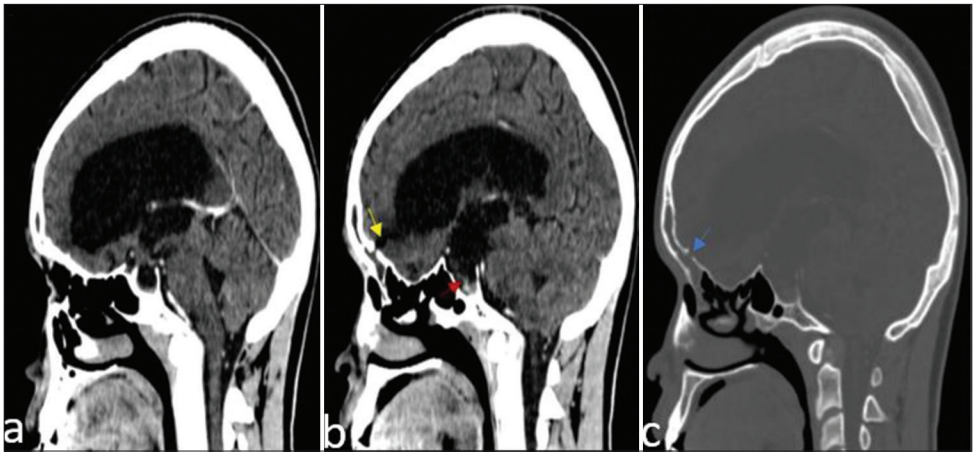

Figure 3: (a) Triventriculomegaly with stenosed aqueduct and small fourth ventricle. (b) Note the enlarged and empty sella (indicated by the red arrow) and pulsion diverticulum (indicated by the yellow arrow) of the frontal horn of the lateral ventricle communicating with (c) the defect on the posterior wall of the frontal sinus (indicated by the blue arrow). Fluid collection (likely cerebrospinal fluid) is noted within the frontal sinus.

mentioned sequelae of intraventricular hemorrhage or an aggravating factor in those with subclinical or compensated AS-associated hydrocephalus with no functional reserve to alteration in CSF hydrodynamics.[3,12,14] Two patients in our cohort admitted to having had a prior mild head injury, which they reported as trivial with no neurological deficit. Based on the preceding discussion, AOAS may be due to an interplay of the various stated factors.

All of our patients presented with symptoms of increased ICP. This was corroborated by neuroimaging features and intraoperative findings of the CSF at high pressure following ventricular cannulation. In addition, features suggestive of endocrinopathy, cerebellar dysfunction, and cognitive and sphincteric dysfunction were observed. Other researchers have reported similar presentation patterns.[4,11,15] The primary ischemic effects of ventricular dilatation on the paraventricular white matter and periaqueductal gray matter, as well as the secondary pressure effect on eloquent neural structures, are responsible for these clinical features.[1,3]

Headache in AOAS is attributed to the effect of raised ICP due to CSF obstruction. Similar to the findings by Tisell *et al*., headache was also the most common symptom in our series and resolved completely following CSF diversion.[15] The total resolution gives credence to its earlier stated pathophysiology. In contrast, none of our patients reported resolution of visual deficit; however, two patients had improved vision, while in the other three, visual deterioration was stabilized. This could be attributed to varying degrees of optic atrophy at presentation. Locatelli *et al*. and Montemurro *et al*. reported gait disturbance as the most common presenting symptom in their cohorts, whereas Tisell *et al*. reported postural disturbances.[8,11,15] All of our patients had preceding headaches, with most (n = 5, 62.5%) seeking medical care following poor vision.

Rarely, following long-standing ICP and trans-mantle pulsatile stress in AS. There is progressive thinning of the ventricular wall with the formation of a pulsion diverticulum commonly observed on the lateral ventricles. This may rupture spontaneously, resulting in a ventriculostomy and a reduction in ICP.[2,12] This phenomenon was well illustrated in one of our patients with CSF rhinorrhea from a spontaneous lateral ventriculostomy that communicated with the frontal sinus through its posterior wall defect [Figure 3]. The possible causes of this bony defect are progressive bony erosion from long-standing intracranial hypertension and a fracture from a mild head injury that he sustained 5 years ago. In such cases, secondary surgical repair of the CSF fistula is warranted following CSF diversion.[2,12] In our case, the patient opted for discharge following the reduction in CSF rhinorrhea despite counseling for definitive CSF fistula repair. He is currently being followed up on and planning for the second procedure when he gives consent.

Although ETV is generally recommended in the management of AOAS.[11,16] At present, VPS is the treatment modality available in our institution and was performed on seven patients with good outcomes, with one requiring a revision 6 years later. To date, no randomized controlled trials have compared the efficacy of ETV and VPS in AOAS management.[5] It has been widely reported that VPS compared favorably with ETV, results in quicker resolution of symptoms, ventriculomegaly, and brain compliance and has a lower failure rate. However, VPS is associated with more complications than ETV, mainly due to infection and shunt malfunction.[5,7,14] While the shunt procedure is readily available and has general applicability, ETV is limited by the CSF absorptive capacity of the arachnoid granulations.[14] Moreso, with the advent of improved shunt systems, further comparative studies are needed in the AOAS setting.

CONCLUSION

AOAS is an infrequent cause of hydrocephalus in our setting, with most patients seeking medical consultation following visual deterioration. Its multifactorial etiopathogenesis and varying clinical spectrum could make early diagnosis elusive. Proper evaluation of adults with long-standing headaches and associated features is advocated to avert total visual loss in this subgroup. VPS insertion is a viable treatment option with a good outcome.

Ethical approval

The Institutional Review Board approval is not required as it is retrospective study.

Declaration of patient consent

Patient's consent was not required as there are no patients in this study.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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