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Aneurysm of the vein of galen presenting with proptosis

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Summary

A case of proptosis, arterio — venous malformation and aneurysm of the Vein of Galen in a 12 year old schoolboy is presented. This is in addition to the literature of the rare aneurysm involving the Great Vein of Galen. This is the first time this syndrome will be described in association with ophthalmic presentation.

It is however remarkable that with such a huge sized aneurysm and A-V malformation, the patient did not have any other neurological deficit except visual field loss. Perhaps the previous series had undetectable visual field loss which was missed as patients might not have complained of visual symptoms.

Résumé

Il est ici présenté un cas de proptose, de la malformation artério-veineuse et de l'anéurysme de la rane de Galen chez un écolier de 12 ans. Ceci vient en addition a la littérature sur l'anérysme rare de la Grande veine de Galen. C'est la première folo que ce syndrome sera decrit en association avec sa présentation ophthalmique.

Il est cependant remarquable qu'en dépit d' un anérysme aussi grand et d' une malformation du type A-V, le patient ne connait pas d' autre carence neurologique en dehors d' une perte de vision. Il se pait que les séries précédantes aient au une perte de vision non décelable qui n' a pas été remarqué puisque le patient ne s'est pas plaint des symptômes de vision.

Introduction

Very little has been written about aneurysms involving the Vein of Galen, especially in this environment. However, the few cases reported have identical characteristics, e.g. a palpable skull defect through which a pulsatile mass is palpable and a machinery bruit heard over the mass radiating to the neck. It is usually a neurological syndrome, as such, features raenging from convulsions, aphasia, stupor, head enlargement, engorged scalp and facial veins are not unusual. Cardio-pulmunary problems are also well recognised (Tables 1a and 1b). The angiographic findings are classical. Aneurysmal dilation of the vein in its position is prominent. The straight and lateral sinuses may be enlarged and tortous. The feeder vessels include the posterior cerebral, the vertebral and the basilar, the aneurysm drains via the dilated straight sinus.

Table 1a: Aneurysm of the vein of Galen in 37 patients reported in the literature Russell and Newton (1964)

Sex	
Male	23
Female	10
Unknown	4
Age	
Newborn	11
1 - 6 months	4
7 - 12 months	5
13 months - 2 years	5
3 - 5 years	3
6 - 10 years	0
11 - 20 years	3
21 - 30 years	2
31 - 40 years	1
41 - 50 years	1
50 years	0
Unknown	2

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Table 1b: Aneurysm of the vein of Galen in 37 patients reported in the literature Russell and Newton (1964

Presenting Features	
Prominent superficial veins	13
Respiratory distress	11
Cranial bruit	9
Heart failure	9
Craniomegaly	8
Cyanotic episodes	8
Convulsions	7
Arteries Supplying Aneurysm	
Posterior cerebral	23
Superior cerebellar	3
Vertebral	3
Arterior Choroidal	3
Basilar	3
Pericallosal	3
Middle Cerebral	1

Case Report

A 12 year old school boy presented with a 5 year history of a pulsatile defect in the left occipital region of his skull and a 3 month history of proptosis of the right eye. This was associated with generalised headaches. The father denied any visible congenital anomaly and trauma.

General examination revealed an otherwise normal and intelligent young boy who likened his symptom to 'a clock working behind his left ear'. Head circumference was 56cm. Vision in both eyes was normal and examination of the eyes revealed a grossly normal left eye while the right upper and lower lids were swollen with visibly dilated, engorged and tortous vessels. There was moderate proptosis and lateral displacement of the right eyeball (Fig. 1). Exophthalmometric reading was 23mm on the right and 18mm on the left at 100mm base reading. The right orbit was full with no definite palpable mass. There was no difficulty at retropulsion. The right conjunctiva was moderately hyperaemic with dilated vessels typical of arteriovenous malformation. Funduscopy revealed moderately engorged and tortous veins in the right eye. An audible bruit was present over the right orbit. Field studies show moderate constriction in both eyes but worse on the right.

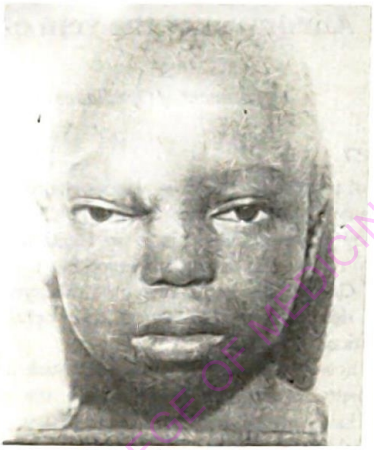

Fig. 1: 12 year-old boy with mild proptosis of the right eye. Note dilated veins in the right upper and lower eyelids

Based on these findings, it was not difficult to make a diagnosis of aneurysm of branches of the vertebral or basilar vessels. Right carotid angiography, showed generalised enlargement and marked tortousity of the carotids and its branches (Fig. 2). Vertebral angiography showed marked aneurysmal dilation of the Vein of Galen with the vertebral posterior cerebral and the basilar being the feeder vessels (Fig. 3).

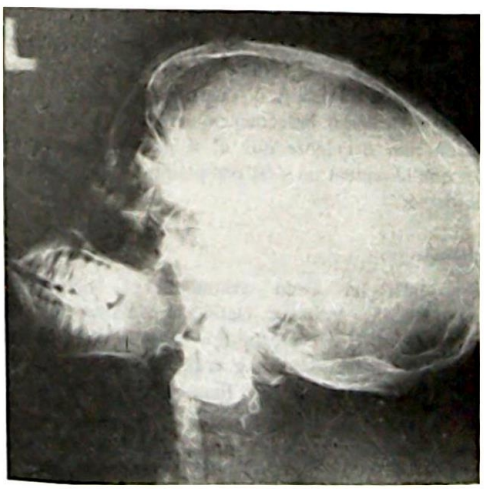

Fig. 2: Left carotid angiogram. Note generalised enlargement and marked tortousity of the carotids and its branches

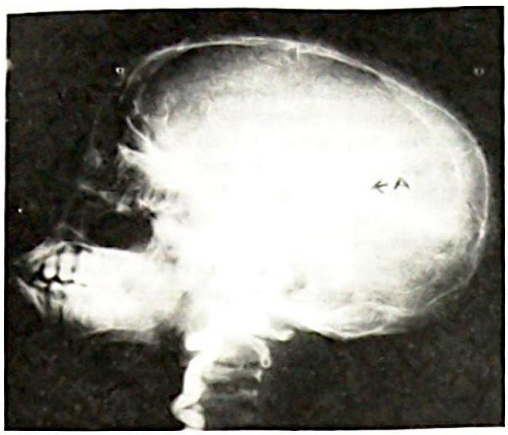

Fig. 3: Left vertebral angiogram. Note marked enlargement and tortousity of the vertebral artery, and aneurysmal dilation of the vein of Galen (A)

Discussion

To date, not many of A-V malformation involving the Great Vein of Galen have been documented. Odeku *et al* [1] described a case complicated by chronic subdural abscess. Russel and Newton[2] found only 37 cases in their series in 1964. Claireux and Newton tabled 22 cases from the literature. Seven of these had cardiac involvement while 3 out of these 7, had frank cardiac failure. Glatt and Rowe[3] reported cerebral A-V malformation associated with congestive cardiac failure in 2 neonates.

These authors were able to identify most of the characteristics of A-V malformation and aneurysms involving the Vein of Galen.

The following were highlighted.

1. A male to female ratio of about 2:1
2. High peri-natal and neonatal mortality rate
3. Death from cardiac decompensation at adolescence
4. Reduced life expectancy.

Litvak *et al* [4] described 3 types of midline A-V anomalies involving the Vein of Galen.

1. Singular dilation of true aneurysms of the Vein of Galen
2. Recemose conglomeration of deep cerebral vessels
3. Transitional types of midline A-V Shunts.

Boldrey and Miller (1949), French and Peyton (1954) are advocates of ligation of feeding arteries followed by excision of lesion when singular dilation of vein was present. Popper and Avman[5] favoured complete removal of lesion where aneurysm derived supply from carotid and vertebrobasilar sources. Gagmon and Boileau considered 'clipping' nine vessels located at different frontal plains near the midline. This formidable effort will probably be fruitless and at best questionable when one considers the attendant complications. It is our candid opinion that our patient was thoughtfully considered along this line before the patient defaulted. The presence of proptosis is explained by generalised dilation of veins which is not uncommon as part of this entity, widespread in the central nervous system in particular. It is however remarkable that with such a huge size of aneurysm and A-V fistula the patient did not have any other neurological deficit except visual field loss. This is due to the close relationship of these vessels with the visual pathway. Perhaps the previous series had undetectable visual field loss which was missed as patients did not have visual symptoms. I therefore suggest full visual assessment of all patients presenting with this entity with or without ophthalmic features, as this will also help in the following up of these patients.

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