Giant Intracranial Aneurysm in a Ten-Year-Old Boy with Parry Romberg Syndrome

A Case Report and Literature Review

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Summary

Parry-Romberg syndrome (PRS) is a rare acquired syndrome consisting of progressive hemiatrophy of the face. We present a child with PRS and progressive neurological deficit caused by a giant intracranial aneurysm and reviewed the literature concerning all intracranial abnormalities in patients with PRS. A literature search identified 27 articles reporting on 88 patients with PRS and intracranial abnormalities. Ipsilateral brain calcification and hemiatrophy are the most prominent features on CT scan and hyperintense white matter lesions are most frequently seen on T2-weighted MRI. Although lacking precise prevalence data, intracranial abnormalities are not uncommon in patients with PRS. We found three other PRS patients with intracranial aneurysms. Our case and literature search suggests a possible association between PRS and intracranial aneurysms.

We consider this association important for clinical practice and recommend including intracranial vascular diseases in the differential diagnosis when dealing with a PRS patient with neurological symptoms.

Introduction

Parry-Romberg syndrome (PRS) is an acquired progressive hemiatrophy of the face first reported by Parry in 1825 and later described as a syndrome in 1846 by Romberg, naming it prophoneurosis. In 1871, Eulenburg was the first to use the more precise term progressive facial hemiatrophy. PRS is characterized by unilateral wasting of bone. There seems to be a predilection for the left side of the face. Onset is insidious, often affecting patients during the first or second decade of life, and is usually followed by progression over the first two to 20 years after which stabilization occurs. Patients with earlier onset of the disease are often more severely affected, with involvement of facial cartilage and bone. Later ages of onset rarely seem to occur (range 1-50 years) [1].

Neuropsychiatric problems are common in PRS patients with reported prevalence as high as 50%. In accordance with these findings, several neuroradiological abnormalities have been described in patients with PRS (e.g. cerebral hemiatrophy and calcifications) [2]. By contrast, neurovascular abnormalities are rarely reported in PRS.

This paper describes a ten-year-old boy with progressive neurological symptoms attributed to a partially thrombosed giant intracranial aneurysm and two additional asymptomatic aneurysms.

To our knowledge this is the second report describing an association between PRS syndrome and intracranial aneurysms [3] in childhood. To facilitate the diagnostic work-up of PRS patients with neurological symptoms we

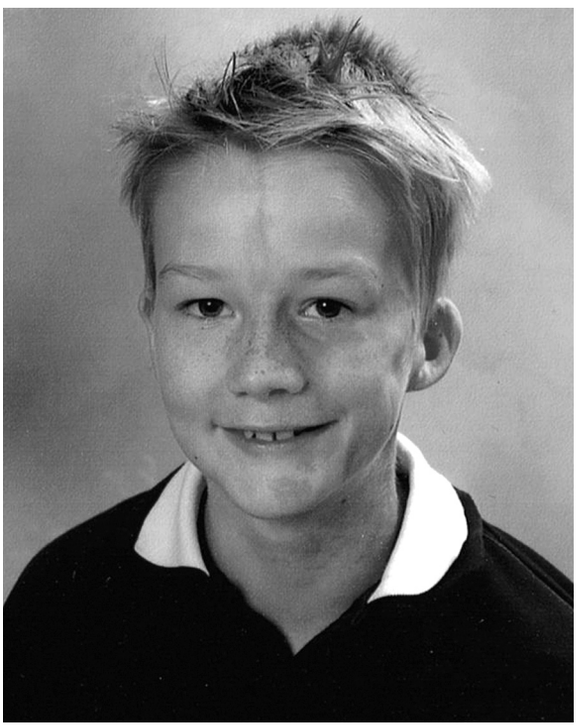

Figure 1 Ten-year-old boy with Parry-Romberg Syndrome. Note the left-sided hemifacial atrophy with wasting of subcutaneous tissues and thinning of the skin. This is more prominent over the forehead resulting in the typical `en coup de sabre` morphology. Hyperpigmentation is present in the region of the second and third division of the trigeminal nerve. (With permission from parents).

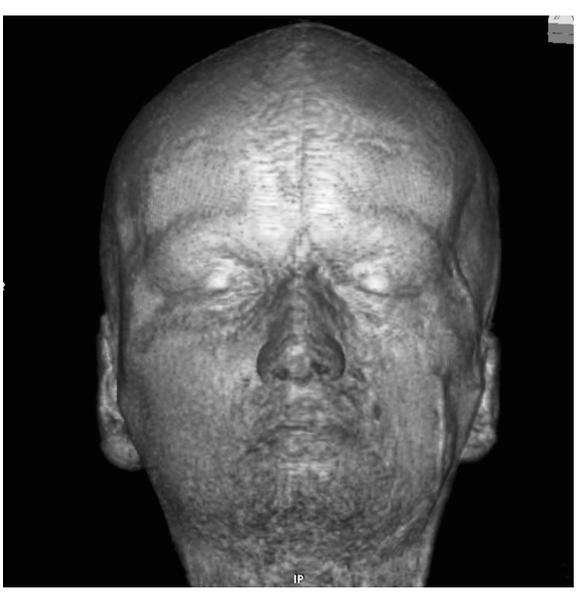

Figure 2 3D volume-rendered MR reconstruction of the face. Note the left-sided atrophy of the subcutaneous and osseous structures most obvious in the temporal fossa, but also visible in the asymmetry of the nostrils and around the orbit. The `en coup de sabre` can also be seen.

reviewed the literature on the presence of intracranial abnormalities (vascular and non-vascular) on neuroimaging in these patients.

Case Report

A ten-year-old boy with Parry-Romberg syndrome was referred to our neurovascular unit for subacute aggravation of his chronic headaches (Figure 1). He was suffering from a progressive headache for over two years. His medical history described a left-sided hemifacial atrophy, first appearing shortly after birth, that seemed to have reached a stable phase over the last couple of years. On physical examination, hemifacial atrophy was noted involving the entire left side of his face, with wasting of subcutaneous tissues and thinning of the skin, which was more prominent over the forehead resulting in the typical `en coup de sabre` morphology. Hyperpigmentation was present in the region of the second and third division of the trigeminal nerve (Figures 1 and 2). Neurological examination showed a disorientated somnolent boy with left temporal visual field deficit and nugal rigidity. Ophthalmologic examination revealed bilateral optic disc pallor without edema indicating longstanding increased intracranial pressure.

Brain computed tomography (CT) showed a large space-occupying lesion (5.9 x 4.2 cm) at the level of the basal ganglia. Heterogeneous densities and calcifications were observed in the wall of this lesion. Mass effect was evident with midline shift and brain stem compression resulting in an obstructive hydrocephalus. We also noted subcortical calcifications in the left frontal lobe. No signs of subarachnoid hemorrhage were present (Figure 2). Magnetic resonance imaging (MRI) showed a large aneurysm of the left distal internal carotid artery with mural thrombus and signs of an acute mural hemorrhage (Figure 3A,B). On digital subtraction angiography (DSA) distal carotid dolichoectasia was shown. Furthermore, two additional aneurysms were discovered. One located at the M1-M2 trifurcation and one of the cavernous portion of the ICA (Figure 4).

After multidisciplinary evaluation, it was decided to treat the giant aneurysm by placing an endovascular stent. Because of the unusual size of the aneurysm, the stent had to be custom-made with a one month delivery time, consequently delaying treatment of the patient in

this stage. One week after admission the patient developed acute deterioration of consciousness, epileptic seizures and paresis of the left leg and paralysis of the left arm. Brain CT showed increased enlargement of the ventricular system, configuration of the aneurysm was not altered. A ventriculoperitoneal drain was placed with subsequent gradual improvement of consciousness, while left hemiparesis persisted. On diffusion weighted MRI, increased signal intensity in the right middle cerebral artery territory, centrum semiovale on both sides, and in the dorsal part of the head of the left caudate nucleus was noted (Figure 5), indicating occurrence of recent ischemia in these areas. A subsequent CT angiography showed gradual narrowing of the right ICA which was most pronounced at the level of the siphon but also present in the distal ICA and M1 and proximal M2 branches (Figure 6). Gradual further deterioration of neurological deficit occurred with development of dysphasia.

Endovascular treatment options were not available at that time, so the patient was transferred to a neurosurgical centre with expertise in performing high-flow bypasses. A high-flow bypass from the external carotid artery to an M3 branch of the middle cerebral artery (MCA) was performed, with ligation of the MCA just after the bifurcation. The aneurysm was debulked to diminish mass effect. After surgery, with a follow-up of six months, the patient almost completely recovered from his neurological deficit.

Search Strategy of Systematic Review of the literature

To identify cases with PRS associated with intracranial abnormalities we performed a literature search using PubMed, Embase and Web of Science (from 1988 up to September 2008) with the following key words: [hemifacial atrophy [OR] Parry-Romberg syndrome] [AND] [intracranial aneurysm [OR] cerebrum [OR] intracranial arterial diseases [OR] neuroimaging [OR] intracranial abnormalities]. Additional articles were found by searching the reference lists of relevant articles. We included all articles reporting patients diagnosed with Parry-Romberg disease and intracranial abnormalities discovered by neuroimaging. Articles in languages other than English were excluded. Two authors (T.B., J.v.B.) independently reviewed the included articles.

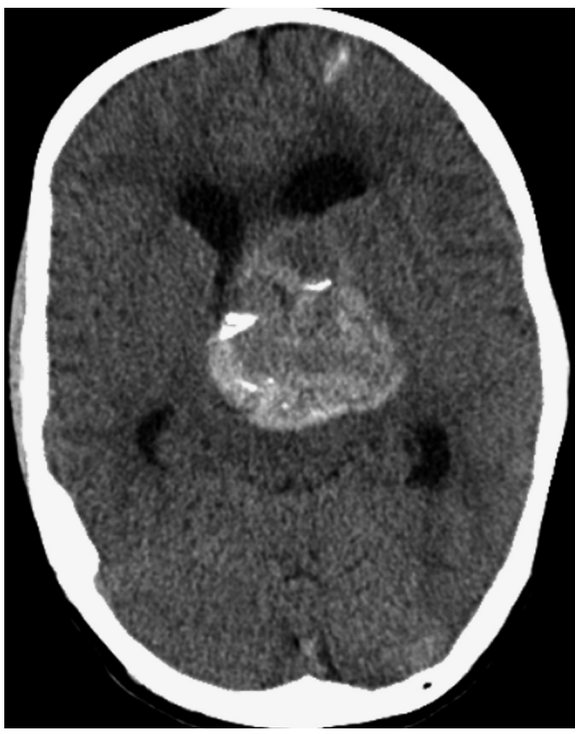

Figure 3 Noncontrast axial CT showing a large space occupying lesion (5.9 x 4.2 cm) at the level of the basal ganglia with evident mass effect causing obstructive hydrocephalus. Different densities and calcifications are seen in the wall of the lesion. Note the subcortical calcification in the left frontal lobe.

Results

We identified 27 articles reporting on intracranial abnormalities in 88 patients with PRS (see Table 1). These articles were mostly case reports and some case series, with ages of onset ranging from neonatal onset up to 50 years. All patients underwent neuroimaging, except for the patients in the study by Tollefson et Al[4] who included both patients with `en coup de sabre` morphea and PRS in their study. Different abnormalities are described by the various authors. Hemispheric atrophy was described in 11 out of 88 patients. On CT ipsilateral calcifications were the most prominent anomaly, described in nine out of 88 patients, including: basal ganglia, cingulated gyrus and frontoparietal parenchyma.

On MRI white matter lesions on T2-weighted images were the most frequent finding, occurring in 31 out of 88 patients. Moko et Al published the largest case series on intracranial MRI appearances in patients with PRS, includ-

Table 1 **Overview of the literature on intracranial abnormalities in patients with PRS between 1988 and 2008.**

Authors	Date	N	Imaging	Abnormalities
Hirata et Al (7)	1988	1	CT CCA	Ipsilateral AVM with calcification pre- and postcentral gyrus Ipsilateral AVM
Fry et Al (18)	1992	6	CT MRI	Cerebral calcifications (N=3) Ipsilateral white matter lesions on T2 (N=5)
Leao et Al (19)	1994	1	CT MRI	Asymmetrical lateral ventricles Ipsilateral atrophy and agenesis head of caudate nucleus
Terstegge et Al (20)	1994	3	MRI	Ipsilateral monoventricular enlargement Meningocortical dysmorphia. White-matter changes
Derex et Al (21)	1995	1	CT + MRI	Ipsilateral porencephaly and cerebral calcifications
Schievink et Al (8)	1995	1	CT MRI CCA	Bilateral middle fossa arachnoid cysts and cerebral atrophy Bilateral atrophy and ipsilateral middle fossa arachnoid cyst Ipsilateral carotid-jugular fistula and carotid dissection
Schievink et Al (3)	1996	1	CT CCA CCA-FU1 CCA-FU2	Ipsilateral calcifications (basal ganglia, wall of the ventricle, parietal lobe) atrophy and giant aneurysm Ipsilateral cavernous giant aneurysm Contralateral cavernous giant aneurysm Ipsilateral posterior cerebral artery aneurysm Ipsilateral distal PCA fusiform aneurysm
Cory et Al (6)	1997	1	CT MRI CCA	Ipsilateral calcifications and hypodensities cingulate gyrus corpus callosum and frontoparietal parenchyma Ipsilateral focal infarctions corpus callosum Diffuse deep and subcortical white matter signal changes Mild cortical thickening Leptomeningeal enhancement Normal
Goldberg et Al (22)	1997	1	MRI	Ipsilateral increased signal surrounding anterior horn lateral ventricles and corpus callosum Diffuse leptomeningeal enhancement
Taylor et Al (23)	1997	1	MRI	Vascular malformation
Woolfenden et Al (9)	1998	1	MRI CCA	Ipsilateral leptomeningeal enhancement, increased T2 signal in the white matter, loss of the normal cortical gyral pattern and enlargement of the ipsilateral ventricle Ipsilateral irregularities of arteries
Miedziak et Al (10)	1998	1	MRI	Ipsilateral white matter lesions on T2 Ipsilateral hypoplastic vertral and carotid artery
Chang et Al (24)	1999	1	MRI	Ipsilateral cerebral atrophy
Yano et Al (25)	2000	1	CT MRI	Ipsilateral calcifications and hypodensities frontoparietal gyri Ipsilateral white matter lesions on T2
Aynaci et Al (26)	2001	1	CT MRI	Ipsilateral calcifications subcortical in the parietal lobe Ipsilateral white matter lesions on T2

Authors	Date	N	Imaging	Abnormalities
DeFelipe et Al (27)	2001	1	SPECT MRI	Ipsilateral hypoperfusion Ipsilateral atrophy and blurring of sulci
Pichiecchio et Al (1)	2002	1	CT + MRI CCA	No intracranial abnormalities Ipsilateral middle cerebral artery aneurysm
Moko et Al (5)	2003	10	MRI	Ipsilateral atrophy or white matter hyperintensity (N=4) Bilateral white matter hyperintensity (N=1)
Shah et Al (15)	2003	1	MRI FDG-PET	Ipsilateral atrophy and hyperintensities on T2 Ipsilateral increased glucose uptake
Blaszczyk et Al (28)	2003	19	MRI SPECT	White matter lesions, vascular malformations, cortex thickening, gyral effacement, cystic infarcts, intracranial calcifications. (N=11) Ipsi- and contralateral hypoperfusion (N=13)
Sathornsumetee et Al (29)	2005	1	MRI	Ipsilateral atrophy
Aktekin et Al (30)	2005	1	MRI	Ipsilateral enlargement of the ventricle and poor demarcation of the white-gray matter
Okumura et Al (31)	2006	1	MR spectr MRI SPECT	Normal Ipsilateral white matter hyperintensity Ipsilateral hyperperfusion
Aoki et Al (11)	2006	1	CT CCA	Mass lesion with partial defect of the petrous bone Ipsilateral carotid artery giant fusiform aneurysm
Tollefson et Al (4)	2007	54	CT MRI	Negative T2 abnormalities
Carreno et Al (32)	2007	1	MRI	Ipsilateral progressive hemispheric atrophy
Moon et Al (33)	2008	1	MRI DTI	Ipsilateral atrophy Fiber derangement

Legend: N= number of patients, CT= Computed Tomography, CCA= Conventional Catheter Angiography, MRI= Magnetic Resonance Imaging, FU= Follow-up, PET= Positron Emission Tomography, SPECT= Single Photon Emission Tomography, spectr= spectroscopy, DTI= Diffusion Tensor Imaging

ing ten patients. Three patients showed central nervous system (CNS) involvement clinically and on MRI. Two patients without clinical CNS involvement showed MR abnormalities. MRI abnormalities consisted either of cerebral atrophy or white matter hyperintensities. Five patients did not show CNS involvement, clinically or on MRI[5]. Functional imaging was performed in sixteen of all reported PRS patients, giving rise to conflicting results. Some authors showed areas of hyperperfusion on single photon emission computed tomography (SPECT) and increased uptake in FDG-positron emission tomography (PET) while other authors depicted areas of hypoperfusion on SPECT in patients with PRS. Details on the different findings are shown in Table 1.

Follow-up observation of imaging findings is limited. Only Cory et Al monitored a child with PRS for nearly two years and demonstrated that cerebral calcifications were relatively unchanged but that the underlying white matter lesions had progressed together with progression of facial atrophy[6]. We specially searched the literature for possible associations between PRS and neurovascular abnormalities. The results are depicted in Table 2. We found one patient with an AVM[7], one patient with a dural

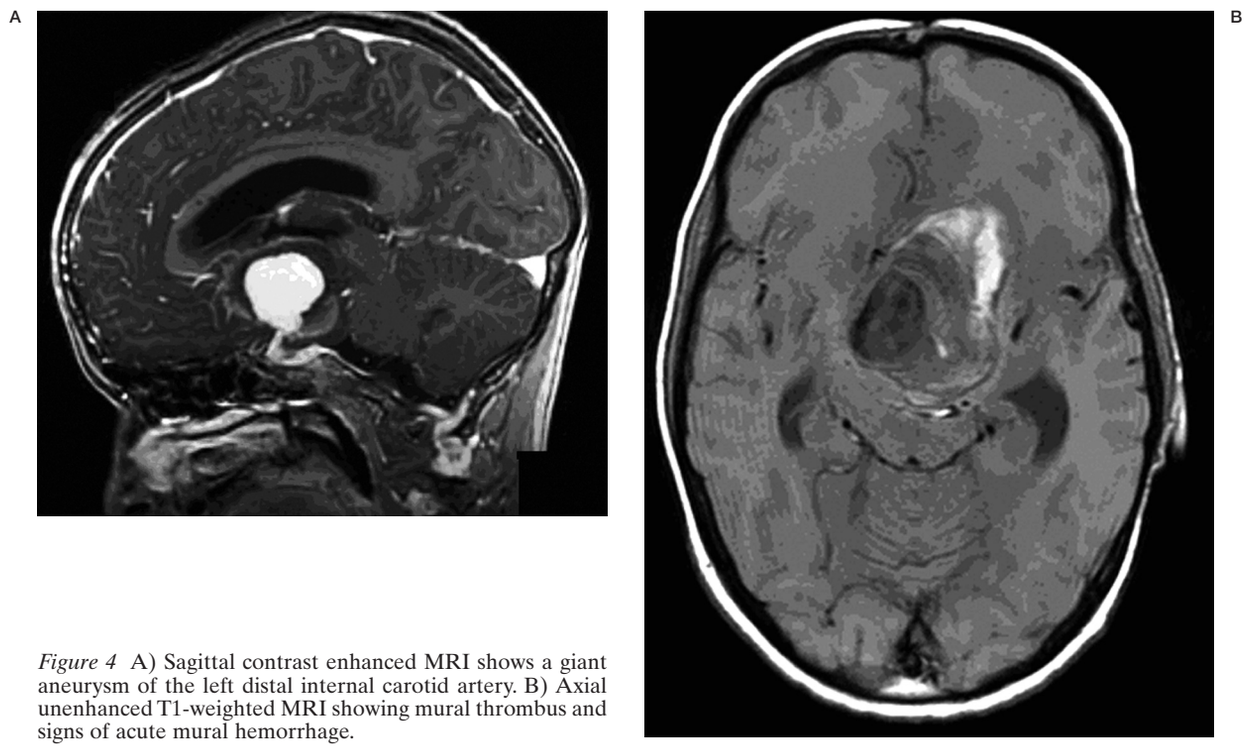

Figure 4 A) Sagittal contrast enhanced MRI shows a giant aneurysm of the left distal internal carotid artery. B) Axial unenhanced T1-weighted MRI showing mural thrombus and signs of acute mural hemorrhage.

fistula [8], two cases with vascular congenital malformations [9,10] and three patients with intracranial aneurysm [1,3,11]. Vascular malformations consisted of one patient with reversible arterial caliber changes and one patient with an ipsilateral hypoplastic verbral and carotid artery [9,10].

Schievink et Al described a child with PRS and multiple intracranial aneurysms. This child was treated for a giant aneurysm of the left cavernous carotid artery at the age of five years. Treatment consisted of carotid ligation in the neck and a superficial temporal artery-middle cerebral artery bypass. At the age of 12 the patient was similarly treated for a giant aneurysm of the cavernous carotid artery on the opposite site, which had progressed from a previously noted small dilatation. At age 21 the patient underwent endovascular treatment for a de novo saccular aneurysm of the left posterior cerebral artery at the P1-P2 junction and a fusiform aneurysm of the distal left posterior cerebral artery [3]. Pichiecchio et Al published the case of 32-year-old woman with PRS who had migraine and an intracranial aneurysm [1]. More recently Aoki et Al described the case of a 56-year-old women with a giant fusiform aneurysm extending from the petrous segment of the ACI to the cavernous segment [11].

Discussion

Compared to the healthy population central nervous system involvement seems to occur more often in patients with PRS, most commonly consisting of migraine-type headache, focal epilepsy and trigeminal neuralgia. A recent global internet survey of 205 patients with PRS estimated the central nervous system involvement to be over fifty percent [2]. The presenting symptom in our patient was a slowly progressive daily headache. For more than two years this never led to a diagnostic work-up. Eventually the subacute aggravation of his headache led to the diagnosis. This aggravation was probably due to increased mass effect of the giant aneurysm caused by acute intramural hemorrhage. Another diagnostic problem we faced was the explanation for his secondary deterioration with decreased consciousness and development of left-sided hemiparesis. Occurrence of vasospasm was unlikely since no subarachnoid hemorrhage was present. The hydrocephalus only partially explained his neurological deficit and we believed the left-sided hemiparesis to have a thromboembolic origin. However, because this patient probably suffers from an underlying condition affecting the entire

Table 2: **Overview of literature on intracranial vascular abnormalities in patients with PRS between 1988 and 2008.**

Authors	Date	Sex/Age	Symptoms	Vascular Abnormalities
Hirata et Al (7)	1988	M/39	Epilepsy	Ipsilateral AVM
Schievink et Al (8)	1995	F/45	Pulsatile tinnitus Retroorbital pain	Ipsilateral carotid-jugular fistula and carotid dissection
Schievink et Al (3)	1996	M/5	Diplopia Left-sided ptosis Left-sided headaches Progressive vision loss left eye	Ipsilateral giant aneurysm Ipsilateral cavernous giant aneurysm Contralateral cavernous giant aneurysm Ipsilateral posterior cerebral artery aneurysm Ipsilateral distal PCA fusiform aneurysm
Woolfenden et Al (9)	1998	M/35	Hemiplegic migraine	Ipsilateral reversible arterial caliber changes
Miedziak et Al (10)	1998	F/23	Transient Numbness contralateral upper and lower extremities	Ipsilateral hypoplastic vertebral and carotid artery
Pichiecchio et Al (1)	2002	F/32	Migraine	Ipsilateral middle cerebral artery aneurysm
Aoki et Al (11)	2006	F/56	Facial pain and left-sided headaches	Ipsilateral carotid artery giant fusiform aneurysm

Legend: M = Male, F = Female

cerebral vasculature, other causes for this event could not be excluded. How a thrombus of a giant aneurysm of the left middle cerebral artery could give rise to embolisms in the right hemisphere with sparing of the ipsilateral circulation remains unanswered.

Confronted with two rare disorders in the same child we searched the literature for an association between children with PRS and giant aneurysms, hoping that this would facilitate our decision-making. We found one other child with the same association [3]. Because other abnormalities, apart from the giant aneurysm, were seen on CT, we reviewed the literature published in the last 20 years (1988-2008) reporting on intracranial abnormalities of any sort in patients with PRS. As shown in Table 1, ipsilateral calcifications (both cortical and subcortical) and atrophy on CT, and hyperintense white matter lesions on T2-weighted MRI are the most prominent intracranial features of PRS. Functional imaging yields inconsistent results. Although there are no exact data on the prevalence of PRS in the general population, we believe that based on this review intracranial abnormalities are not uncommon in PRS.

In contrast, intracranial vascular abnormalities are rarely associated with PRS. Our patient

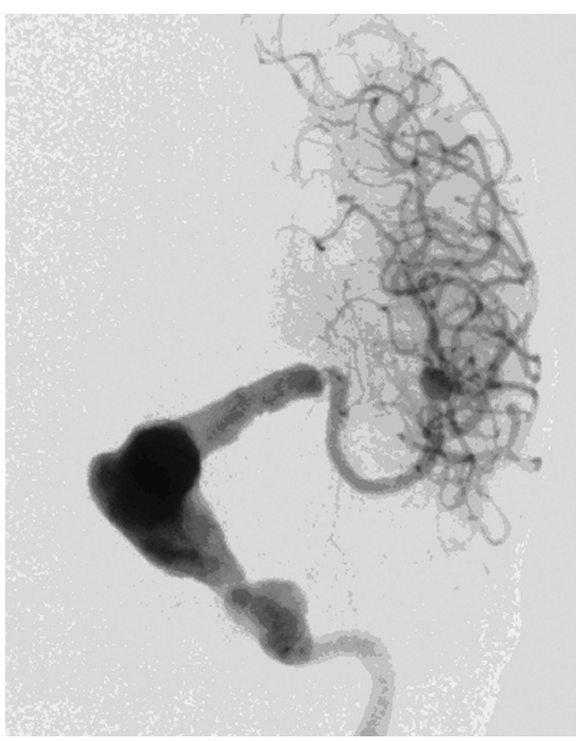

Figure 5 Digital subtraction angiography showing dolichoectasia of the distal carotid artery and a severe junctional stenosis. In addition, aneurysmal dilation of the cavernous portion of the ICA and an aneurysm on the M1-M2 trifurcation were seen.

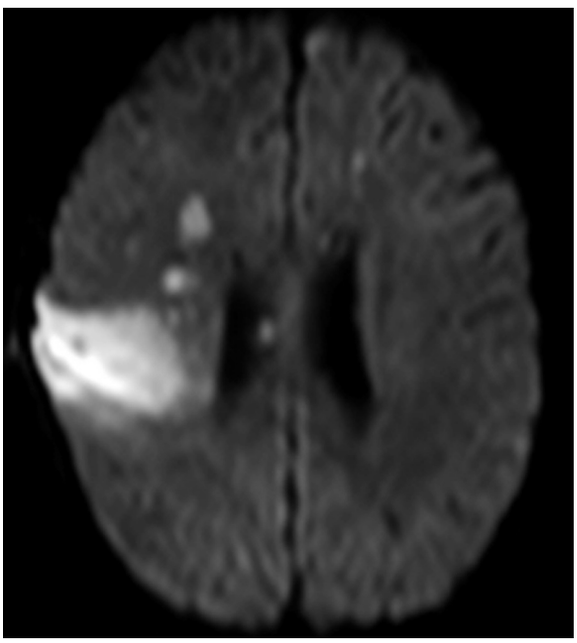

Figure 6 On diffusion weighted MRI, increased signal intensity in the right middle cerebral artery territory, centrum semiovale, and in the dorsal part of the head of the left caudate nucleus were noted suggestive of infarction in the right MCA territory.

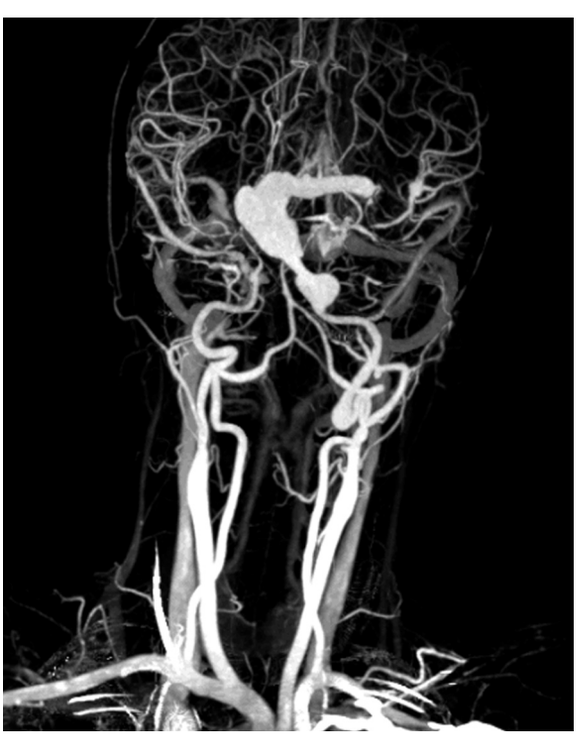

Figure 7 CT-angiography showing narrowing of the right carotid artery most pronounced at the level of the siphon but also present in the distal carotid artery, M1 and proximal M2 branches.

is the second child reported with intracranial aneurysmal disease with an associated Parry-Romberg syndrome. In the adult population we found two cases of PRS with an intracranial aneurysm [1,11]. Other reported neurovascular abnormalities include one AVM [7], one dural fistula [8] and two cases with vascular congenital malformations [9,10].

In recent literature, aneurysms seen in the pediatric population account for around 7% of all aneurysms [12]. Their features differ significantly from the aneurysms in adults especially in their sexual prevalence, location, morphology and etiology. Giant aneurysms are known to be of increased incidence in this group. Recently the Toronto group and the Bicêtre group published two large series on this subject reporting an incidence of giant aneurysms of 29.7% [12,13].

When children present with one or more intracranial aneurysm, a generalized connective tissue disorder is suspected. However, quite often the underlying disease remains unknown despite extensive investigations [14]. In our patient the possible underlying disorder may be PRS, since both entities are extremely rare and a coincidental occurrence seems highly unlikely. Speculating on a common etiology of both disorders is beyond our expertise because both diseases are associated with a wide variety of conditions, such as trauma, infections, connective tissue disorders, familial occurrence, inflammatory diseases, etc.[12,15,16].

Interestingly an Australian group recently described a chronic sympathetic hyperactivity in the areas of distribution of the trigeminal nerve of the affected side of PRS patients [17]. In earlier reports several authors also proposed that cranial vasculitis in PRS is the cause of trigeminal dysfunction [5]. In view of these findings, an inflammatory process as a common etiology for PRS and giant seems logical and reasonable, although the primary trigger remains obscure.

Although very rare, we consider the described association between PRS and giant aneurysm important for clinical practice. When confronted with a patient with PRS presenting with neurological symptoms, we recommend including intracranial vascular diseases in the differential diagnosis. At the moment, we lack the knowledge on precise prevalence data to recommend routine screening for intracranial abnormalities in patients with PRS.

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