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Difficulties in differentiation of Parry–Romberg syndrome, unilateral facial sclerodermia, and Rasmussen syndrome

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Abstract *Introduction:* Parry–Romberg syndrome (progressive facial hemiatrophy) is a unilateral, slowly progressive atrophy affecting the skin, subcutaneous tissues, muscles, and bones. The relationship between Parry–Romberg syndrome and connective tissue disorders, especially scleroderma en coup de sabre, is still unclear. The neurological symptoms, including epilepsy, migraine, and brain lesion, on neuroimaging may be similar. Rasmussen encephalitis (RE) is connected with chronic inflammation and damage of one hemisphere. Clinically, it is manifested by epileptic partial seizures and unilateral neurological symptoms. *Case Report:* The authors present the case of a 10-year-

old girl with features suggestive of RE, with refractory partial motor dextrolateral seizures followed by development of hemiparesis and with progressive intellectual deterioration. At the age of 2 years, some changes on the left part of the face typical of Parry–Romberg syndrome or a linear form of scleroderma were noticed. *Discussion:* The authors discussed the difficulties in differential diagnosis in that patient. The presented girl constitute the case from the borderline zone of the aforementioned disorders.

Keywords Progressive facial hemiatrophy · Scleroderma en coup de sabre · Rasmussen encephalitis

Introduction

Parry–Romberg syndrome is characterized by progressive unilateral facial atrophy involving soft tissues (skin, subcutaneous tissue, and muscles) and adjacent bones. The onset of the disease is usually observed in the first or second decade of life. The disease at the beginning is slowly progressive, and then, the pathological process usually stops. Frequently, in Parry–Romberg syndrome, some neurological symptoms occur, of which the focal epilepsy is the most common one.

Very similar in the clinical picture of symptoms and course of the disease in Parry–Romberg syndrome, Rasmussen encephalitis (RE) and linear scleroderma (“en coup de sabre”) makes a true diagnostic dilemma. In Parry–Romberg syndrome and scleroderma, progressive unilateral atrophy

of the brain structures (homolateral to the changes in the face), epilepsy, progressive intellectual deterioration, and hemiparesis may speak for the common autoimmunological pathogenesis of these diseases. Patients with Parry–Romberg syndrome and scleroderma may show also pathological changes in the brain which require differentiation with RE.

In RE, the first symptom is drug-resistant focal epilepsy, which appears usually at about the fifth year of age. About 20% of children have partial seizures. In the course of the disease, hemiparesis occurs. The etiology is probably heterogeneous. In some patients, there are evidences of herpetic or cytomegalic infection. In some cases, antibodies against glutamergic GluR3 receptors are detected. Intrathecal production of immunoglobulin (Ig)G was also described.



Fig. 1 a–b The patient at the age of 10 years

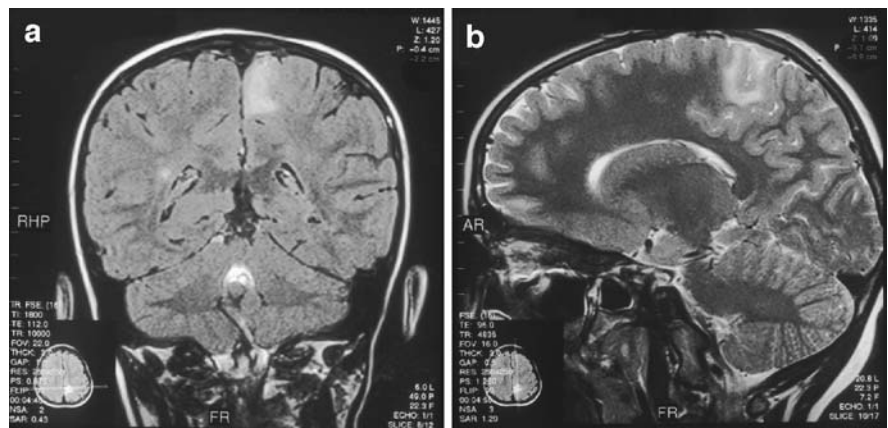
Case report

A girl 10 years of age, born to healthy, nonconsanguineous parents, was referred to our hospital because of intractable

epilepsy. The family history was noncontributory. The girl was born before term by spontaneous delivery at 8 months' twin gestation (as a first twin), with birth weight of 2,250 g, birth length of 49 cm, and an Apgar score of 8 out of 9 points at 1 and 5 min after birth, respectively. The patient achieved developmental milestones without delay. At approximately 2 years of age, the parents noticed asymmetry of the face, with slowly progressive atrophy of the facial muscles in the left suborbital region (not preceded by induration of the skin lesion). Her first focal dextrolateral seizures appeared at the age of 6 years. The child developed recurrent motor seizures and needed drug adjustments many times. During the following 3 years, the epileptic seizures progressed until seizure control could not be achieved by pharmacological treatment (including steroids and Ig). Because of continuous epileptic activity and a right hemiparesis, she started to use a wheelchair. The first magnetic resonance imaging (MRI) scans performed at the age of 6 years showed an SE/T2 i PD hyperintensive lesion in the left pericentral lobulus of unknown etiology. A repeat MRI, performed 1 year after, displayed some resolution of the abovementioned lesion.

At the age of 9 years, the patient underwent surgery for intractable focal motor seizures. The cortical resection in the part of frontal lobe and MSTs (multiple subpial resections) in the precentral gyrus and Broca's area were performed. The transient significant reduction of seizures was obtained. Upon histopathological examination, improper "architecture" of brain cortex, improper vascular network in the cortex, and strongly glial fibrillary acidic protein (GFAP)-positive "feltwork" of processes in subpial zone (Chaslin's gliosis) were found. In the white matter, some single "misplaced" ganglion cells were present (heterotopia). Also, a few "balloon-like" (although not typical) cells were scattered across the cortex. The improper vascularity did not, however, conform anything that could be described as an "angioma." Rather, the conspicuous capillaries showed features of a sort of hypertrophy and more ramifications than normal. There were no morphological features of inflammatory process; in particular, there were

Fig. 2 MR frontal (a, on the right) and sagittal (b, on the left) image of a 10-year-old girl showing the focal lesion in a left parietal region



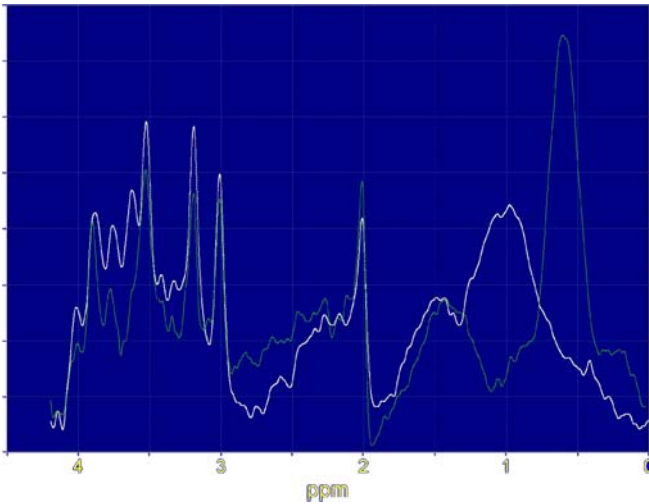


Fig. 3 ^1H MRI spectroscopy in a 10-year-old girl

no lymphocytic or other infiltrations that were either perivascular, intraparenchymatous, or within the arachnoid.

On admission at the age of 10 years, upon physical examination, atrophy of the left side of the face was noted (Fig. 1a,b).

Neurological examination showed right hemiparesis. IQ was equal to 48 (moderate mental retardation). Routine laboratory studies and a comprehensive autoantibody screen [antimitochondrial antibody (AMA), antismooth muscle antibody (ASMA), antiparietal cell antibody (APCA), and antinuclear antibody (ANA)] were negative. Electrocardiography, echocardiography, abdominal sonography, and a chest X-ray did not yield abnormalities. Cerebrospinal fluid (CSF) examination showed 11 lymphocytes per cubic millimeter, with normal protein and glucose. The cultures were negative. Intrathecal production of IgG was detected. Anti-IgM *Borrelia burgdorferi* antibodies at the titer of 9.1 base-band unit (BBU)/ml (IgM-positive >11.0 BBU/ml, IgM-negative <9.0 BBU/ml) in serum were present. The ophthalmologic examination did not discover any abnormalities.

Electroencephalogram (EEG) tracing showed general paroxysmal changes with variable hemisphere predominance. The structure and intensity signal of the soft tissues of the face, sinus, and orbits were correct in an MRI picture. An MRI of the head revealed minor atrophy of the brain parenchyma in the left frontoparietal region, asymmetry of the sulci, hypoxic–ischemic lesion in the left pa-

rietooccipital region, and change of hyperintensity signal in the occipital horn region (Fig. 2).

In angiomagnetic resonance of the head, circulation in the brain arteries and vascularization of the brain were normal. ^1H MR spectroscopy showed a decreased level of *N*-acetylaspartate (NAA)–total creatine (tCr), presence of intensive signal in the range of 0.8–1.8 ppm, where lactate and free lipids are located (Fig. 3, Table 1). All the changes in the signal of the metabolites were more pronounced on the left side. Single-photon emission computed tomography (SPECT) displayed radiomarker accumulation loss in the left parietotemporal and frontal regions.

During the diagnostic workup neuroinfection (including *Toxoplasma gondii*, *Cytomegalovirus*, *Herpes simplex*, and *Listeria monocytogenes* infections), subacute sclerosing panencephalitis (SSPE), some neurometabolic diseases (aminoacidopathies, congenital organic acidurias, lysosomal storage, mitochondrial and peroxisomal disorders, urea cycle disorders, and congenital disorders of glycosylation) were excluded.

Our patient is presently 10 years of age, the facial lesions are less conspicuous, and the atrophy does not progress. She has about 40–50 motor seizures a day and is prepared for vagus nerve stimulator implantation. The whole clinical picture spoke for Parry–Romberg syndrome or scleroderma en coup de sabre. As soon as the patient's condition becomes stationary, it is necessary to perform a skin and muscle biopsy. At present, the parents of the girl do not agree to taking the skin biopsy.

Discussion

Progressive facial hemiatrophy is a sporadic, self-limited, and heterogeneous disorder characterized by localized atrophy of the skin and subcutaneous tissues on one side of the face [2, 4–6, 19, 21, 22, 27]. It has been speculated that PHF, or localized scleroderma, results from disturbed activity of peripheral sympathetic nervous system or trigeminal nerve abnormalities. The suggested causes of PFH include local facial trauma, hyper- or hypoactivity of the sympathetic nervous system, abnormality of the trigeminal nerve, and bacterial or viral infection, including *B. burgdorferi* [4, 5]. Progressive hemifacial atrophy may be associated with local alopecia and pigmentary changes of the skin. The onset is usually insidious, and the disease progresses over 2–20 years and then becomes stationary.

Table 1 Values of the area signal ratio of the basic metabolites in ^1H MRI spectroscopy

	NAA/tCr	Cho/tCr	Cho/NAA	ml/tCr	Lac/NAA	Lip/NAA	Glc/tCr	Glx/tCr
VOI1, left hemisphere, lobulus	0.72	1.04	1.43	0.78	1.02	7.22	0.35	1.76
VOI2, right hemisphere	0.74	0.91	1.22	0.80	0.66	4.60	0.37	1.77
Population average values	1.53	0.98	0.64	0.70	0.00	0.00	0.30	2.10
Standard deviation	0.23	0.20	0.13	0.18			0.14	0.41

Cho Choline, Lac lactate, Lip lipid, Glc glucose, Glx glutamine–glutamate

The disease may be associated with some neurological abnormalities in about 10% of cases; among them, epilepsy is the most common one [4, 5, 19, 21]. Considerable controversy remains regarding whether Parry–Romberg syndrome is a distinct disorder or a form of linear scleroderma, especially the involuntary form (after regression of sclerosis) [2, 7, 17]. There are closely related disorders that shows a striking similarity and may have some distinguish features Table 2 [1, 2, 4–7, 11, 13, 16, 17, 21–23, 26].

In the internet survey performed by Stone, 205 patients with progressive facial hemiatrophy were collected; among them, 80% were women. Eleven percent of them reported epilepsy, 52% migraine or facial pain, 46% eye/vision problems, and 46% depression and anxiety. Some described a coexistence with the other autoimmune disorders such as vitiligo (17%), thyroid disease (10%), systemic sclerosis (5%), inflammatory bowel disease (5%), rheu-

Table 2 Comparison of PHF and focal scleroderma

Disorder	Parry–Romberg syndrome	Scleroderma en coup de sabre
Facial involvement	<p>Unilateral atrophy of the face</p> <p>Absent or minimal previous signs of induration or inflammation</p> <p>Cutaneous atrophy (the involved skin is thin and soft, hair is normal, and sclerosis is absent)</p> <p>Associated with dysplasia of underlying bone, skull, tongue, gingiva, and palate unilaterally</p>	<p>Unilateral band-like sclerotic changes on the frontoparietal area, usually preceded by induration of the skin</p> <p>Usually does not extend below the eyebrow</p> <p>Cutaneous sclerosis (the involved skin area is hard, depressed, hyperpigmented, shiny, and devoid of hair)</p> <p>Frequently causing marked deformities and contractures</p> <p>Progressive softening of lesions takes place over time</p>
Neurological symptoms	<p>Vision disturbances</p> <p>Headaches</p> <p>Epilepsy</p> <p>Vertigo</p> <p>Ipsilateral paresis of nervus facialis</p> <p>Trigeminal neuralgia</p> <p>Cognitive abnormalities</p>	<p>Epilepsy</p> <p>Headaches</p> <p>Ocular symptoms</p>
MRI picture	<p>Predominantly ipsilateral high-signal white matter lesions [28]</p> <p>As described above plus ill-defined sulci and ventricular dilatation [31]</p> <p>Ipsilateral cortical thickening, gyral enhancement, blurring of the gray–white matter interface, leptomeningeal enhancement, cystic infarcts with foci of encephalomalacia [6]</p> <p>Ipsilateral meningeal and basal ganglia lesions [33]</p> <p>Unilateral focal infarction in the corpus callosum, diffuse white matter changes, leptomeningeal enhancement [34]</p> <p>Bilateral intracranial aneurysms, carotis dissection, arteriovenous malformation [21]</p> <p>Hamartomas [5]</p>	<p>Ipsilateral intracerebral calcification alone or with migration abnormality and dysmyelination [29, 30]</p> <p>Hyperintense, mainly ipsilateral cortical and subcortical lesions [32]</p> <p>Remitting and relapsing lesions [predominantly ipsilateral involvement of the gray matter and superficial and deep white matter [23]</p> <p>Contralateral frontal brain lesion [16]</p>
Intracranial neuropathology	<p>Leptomeningeal fibrosis, degenerative cortical changes, microvascular malformations [35, 36]</p> <p>Ipsilateral gray and white matter, perivascular lymphocytic infiltration with pial and glial proliferation [38]</p>	<p>Perivascular infiltrates, neuronal loss, gliosis suggesting chronic inflammatory process [37]</p> <p>Leptomeningeal band-like sclerosis, thickened walls of the blood vessels with surrounding gliosis, intraparenchymal calcification [29]</p> <p>Focal inflammatory changes in the form of low grade vasculitis with focal cerebral necrosis [23]</p>

matoid arthritis (4%), ankylosis spondylitis (2%), lupus erythematosus (2%), and multiple sclerosis (MS) [24].

Scleroderma en coup de sabre denotes the linear variant of localized scleroderma on the face and skull. Variants of localized scleroderma are classified by Tuffanelli [13]:

1. Localized scleroderma
 - (a) Morphea (plaque type; guttate; profound, nodular, and bullous atrophoderma of Passini and Pierini)
 - (b) Linear scleroderma (“en coup de sabre”: with facial atrophy (Parry–Romberg syndrome); acral; with arthritis; myositis and growth defects of bone; melorheostotic)
 - (c) Generalized morphea (pansclerotic; edematous; with lichen sclerosis)
2. Localized scleroderma as a component of an overlap syndrome
 - (a) Mixed connective tissue disease
 - (b) Undifferentiated connective tissue disease
 - (c) Sclerodermatomyositis
3. Localized scleroderma as a component of eosinophilic fasciitis (Schulman syndrome)

As mentioned above, some authors even suggest to include Parry–Romberg syndrome to a spectrum of linear scleroderma [2, 13, 17].

Linear scleroderma en coup de sabre is associated with neurological complications: epilepsy, headaches, ipsilateral intracerebral lesions, and ocular symptoms (ptosis, uveitis, iridocyclitis, enophthalmus, exophthalmus, pseudooculomotor palsy, and retrobulbar pain) [2, 7, 9, 11, 13, 16, 23]. This disease of unknown origin is characterized by fibrosis of connective tissue and usually involves the area of the scalp lesions. Linear atrophy is probably connected with the sympathetic nervous fibers of nervus trigeminus injury.

The scleroderma en coup de sabre and PFH are overlapping and sometimes coexisting conditions [2, 17]. There are no reliable criteria to differentiate Parry–Romberg syndrome from scleroderma. Some papers describe patients with scleroderma en coup de sabre converting with time to PFH [17]. Most likely, the time we see the patient is of great importance when making the correct diagnosis. Both the central nervous system (CNS) abnormalities and neurological symptoms are very diverse.

Rasmussen encephalitis is a rare childhood disease characterized by a progressive destruction of the single hemisphere associated with chronic inflammation [8, 10, 14, 15]. This disorder usually begins in the first decade of life, with 85% of occurrences before the age of 10 years [14]. Children with RE manifest unilateral neurologic symptoms such as intractable partial epileptic seizures (in about one half of the patients as an *epilepsia partialis continua*), language deficits, cognitive difficulties, hemiparesis, hemianopia, and hemisensory loss, which worsen over time.

Neuroimaging reveals progressive unihemispheric atrophy. Also, a few patients with an otherwise similar to RE syndrome occurring in adults and adolescents have been described. Clinically, they show some important differences from the children with respect to the time of onset, a more benign course, and increased incidence of occipital lobe involvement or even bilateral involvement [25].

The chronic inflammatory process appears to begin in the frontotemporal regions [10, 15]. Some studies on RE have detected genetic material from cytomegalovirus, Epstein–Barr, herpes simplex I virus by polymerase chain reaction (PCR) technique and in situ hybridization. Recent works suggest that Rasmussen syndrome is an autoimmune disorder; one of the most important and pathogenic autoantigens is a glutamate receptor, GluR3, in the mammalian cortex (target antigens are absent outside the CNS) [8]. According to O’Hara et al., the ligand-binding domains of glutamate receptors are structurally similar to bacterial periplasmic amino acid binding proteins. So maybe, the precipitating illness or head injury may be a trigger factor initiating an immune-mediated process [10].

In 10% of the patients with RE, focal neuronal migration and vascular malformation are present. A histopathologic examination of the affected brain hemisphere reveals changes ranging from active inflammation process to chronic disease [18].

Does Rasmussen syndrome come in the differential diagnosis of this case? Despite the fact that the patient has a structurally normal brain, on neuroimaging, the clinical picture supports the hypothesis of RE. The theory of RE onset is also a possibility.

The quest for the mechanisms explaining the coexistence of hemifacial atrophy and ipsilateral brain pathology (e.g., in RE form) quite naturally may lead to exploration of the embryological data with the hope that there might be clues [3]. In this context, especially interesting are cells forming the cranial part of neural crest since their progeny and derivatives not only form the ganglia of cranial nerves and parasympathetic ganglia but also many other types of cell, among them facial and skull bones and even dermis, fat, and smooth muscles of the skin. Cranial neural crest cells contribute also in the formation of meninges. Therefore, one cannot exclude that it is the maldevelopment or some kind of an insult affecting the part of the neural crest in one side of the head that may account for the “mixed” unilateral syndromes, like Parry–Romberg, scleroderma, and (probably secondarily) RE.

In the case of our patient, the problem has arisen as to whether this case belongs to scleroderma or Parry–Romberg syndrome and whether there is any relationship between them and the Rasmussen syndrome. The suspicion of RE was based on a clinical picture—mainly the *epilepsia partialis continua* and hemiparesis. However, the results of the ancillary tests and also the histopathology of the brain tissues were not entirely conclusive, although in the case of histopathological examination, it seemed to

exclude RE, at least at this stage of disease. On the other hand, the local IgG production in the cerebrospinal fluid, seen in PFH, focal scleroderma, and in our patient, indicated an active inflammatory reaction in the CNS. Of the patients with PFH, 30–40% have changes typical for morphea or linear scleroderma localized outside the face [2, 7, 12, 17]. There are also no typical abnormalities for PHF and scleroderma based on MRI scans or neuropathology. According to recent reports, scleroderma en coup de sabre, PHF, and RE may share the same etiology. The oligoclonal response, the presence of autoantibodies, and reaction to immunosuppressive therapy or plasmapheresis support the immune-mediated process. Also, in all three conditions, the inflammation mechanism is postulated. The underlying pathology is not clear because of scarcity of neuropathologic examinations. Also, the connection between CNS involvement, PHF, and scleroderma relies mainly on functional examination.

One may presume that the presented case is the next-in-line example speaking for common or similar pathogenesis of all “hemilateral” malformation neurological syndromes, which probably include facial scleroderma, Parry–Romberg syndrome, and supposedly, Rasmussen syndrome [3]. On the other hand, this problem is controversial since the unilateral character of these disorders is not adequately explained by autoimmune theory. Also, the facial hemiatrophy was not present at birth, which is against the malformation pathology. It is quite possible that RE, with its characteristic unilaterality, occurs when the infection (probably viral) affects the child with focally disturbed cytoarchitecture of brain cortex, especially with concomitant

improper vascularity [20]. This may result in a “leaky” blood–brain barrier (BBB). Such a leaky BBB may be permissive, for example, for autoantibodies against brain tissue components (that may be a byproduct of an inflammation outside the brain) if not for the viruses themselves. One may speculate that in the case presented here, and likewise in similar cases, RE, even if it has not started already, may occur in the future as a most worrisome sequel of focal cortical maldevelopment and superimposed inflammation. Although the problem of Parry–Romberg syndrome differing from scleroderma is still debated, we consider these diseases as parts of a clinicopathological spectrum that also includes cases with both diseases’ features [2, 12, 17].

Regardless of time and our efforts, the pathogenesis of many disorders is not fully discovered. The example for that is RE—one of the most mysterious and devastating epileptic syndromes [8]:

It is highly probable that in all these cases there was located damage to the brain.... The question of the nature of the disease process is much more difficult... in all cases the illness [sic] developed little by little once it had developed persisted for a very long time so we can postulate only a chronic processes here... Thus, of the chronic processes, encephalitis with transition to secondary hardening of the brain, or sclerosis cerebri, is almost the only possibility.

(Kozewnikow A. *A peculiar type of cortical epilepsy*, 1894).

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