

Background. Foster Kennedy syndrome (FKS) is described as ipsilateral optic atrophy and contralateral papilledema from an intracranial mass. FKS is uncommon manifestation of Neurofibromatosis type 2 (NF2), which is generally presented with hearing loss and tinnitus.

Case report. In this report we present a 26-year-old female with the atypical presentation of NF2. First symptoms were progressive vision loss and cognitive dysfunction. Ophthalmological examination revealed right-sided papilledema and left-sided optic atrophy. Magnetic resonance imaging (MRI) of the brain revealed bilateral vestibulocochlear schwannoma and three intracranial meningiomas, involving the parafalcine region and the olfactory groove. Whole spine MRI showed one intramedullary tumor at C1-C2 level, multiple spinal canal nodules in cervico-dorsal regions and one Th12-L2 extramedullary tumor. Based on clinical and imaging findings the diagnosis of neurofibromatosis type 2 was established. The patient underwent surgical resection of giant parasagittal meningioma, subtotal resection of the olfactory groove meningioma and total resection of Th12-L2 meningioma. Six months after brain surgery, she underwent Gamma knife radiosurgery for remnant frontobasal meningioma and for both vestibulocochlear schwannomas. Despite the combined treatment of intracranial lesions, only an insignificant vision improvement was achieved.

Conclusions. FKS can be the presenting symptom of NF2. Early detection and treatment of ophthalmologic manifestations of NF2 may prevent amblyopia development.

Key words: Foster Kennedy syndrome, neurofibromatosis type 2, intracranial meningioma, intramedullary tumor, extramedullary tumor

DEPARTMENT OF NEUROLOGY no.1

22. INTRACEREBRAL HEMORRHAGE IN A PATIENT WITH MOYAMOYA SYNDROME: CASE REPORT

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Background. Moyamoya disease is a cerebrovascular disease that is characterized by bilateral chronic and progressive stenosis or occlusion of the arteries around the circle of Willis with development of collateral circulation, of unknown etiology. It has a high incidence in Japan and Asian population, with fewer cases described in Europe. Similar angiographic findings can be seen in patients with other medical conditions that are described as Moyamoya syndrome. Main clinical features include transient ischemic attacks, ischemic strokes, and hemorrhagic strokes.

Case report. We describe a 38-year-old female patient that presented with an intracerebral hemorrhage with a typical location for hypertensive bleeds. She had no vascular risk factors, but a high normal blood pressure (140/90 mmHg), and elevated ESR. A magnetic resonance angiography showed occlusion of internal carotid artery with development of collateral cerebral circulation on the side of the bleeding. Unilateral affection and elevated ESR were more characteristic for a moyamoya syndrome within a systemic disease.

Conclusions. Despite a typical hypertensive location of the bleeding, vascular imaging is warranted in all patients with intracerebral bleedings to evaluate for atypical etiologies. Our case represents a patient that might benefit from revascularization surgery in the context of multifactorial risk factor control.

Key words: Moyamoya syndrome, stroke, hemorrhage, intracerebral, collateral flow.

23. POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME MIMICKING