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A CASE REPORT: SUPERIOR ORBITAL FISSURE SYNDROME

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ABSTRACT
Background: Superior orbital fissure syndrome (also known as Rochen-Duvigneaud syndrome) is a collection of symptoms caused by compression of structures anterior to the orbital cusp. The fissure orbitae superior separates the major and minor wings of the sphenoid bone and transmits cranial nerves III, IV, VI, the first branch of the fifth nerve, and sympathetic fibers. Superior orbital fissure syndrome is rare. The most common cause of superior orbital fissure syndrome is now known to be trauma, occurring in 0.3-0.8% of patients.
Case Presentation: A 49-year-old Malay woman presented with difficulty moving her right eye since 19 days ago. She experienced progressive weakness in both lower limbs, initially feeling numbness in her right foot followed by increasing paralysis that limited movement to only toe movements. Additional complaints included double vision and drooping eyelids on the left side. Contrast-enhanced MRI scans of the head were conducted which found no intracerebral abnormalities suggestive of stroke/infection/tumor/AVM bleeding. Ventricular systems appeared symmetrical without hydrocephalus shifts or other structural anomalies. MRI orbits confirmed no signs indicative of pathology affecting ocular structures directly related to suspected syndromes including superior orbital fissure syndrome.
Conclusion: This patient experiences various symptoms in the eyes and nervous system which can be interconnected with each other. However, the underlying etiology has not been discovered because several examinations have obtained normal results. So further exploration is needed to find and understand the cause of the symptoms experienced in this patient
Keywords: *Superior Orbital Fissure Syndrome, Ophthalmoplegia, Cranial Nerve Palsy*

BACKGROUND

Superior orbital fissure syndrome (also known as Rochen-Duvigneaud syndrome) is a collection of symptoms caused by compression of structures anterior to the orbital cusp. Anatomically, the superior orbital fissure separates the greater and lesser ala of the sphenoid bone and transmits cranial nerves III, IV, VI, the first branch of nerve V and sympathetic nerve fibers. The complex and dense anatomy of the superior orbital fissure results in a characteristic pattern of cranial nerve, pupillary, and extraocular findings.[1-3]

Superior orbital fissure syndrome is rare, and most of the literature consists of case reports and case series. The most common cause of superior orbital fissure syndrome is now known to be trauma, occurring in 0.3-0.8% of patients.[3-5] Other causes include neoplasms (especially lymphoma and rhabdomyosarcoma), infections (such as meningitis), syphilis, sinusitis, herpes zoster, inflammatory (such as Lupus, sarcoidosis, Tolosa-Hunt syndrome) and vascular phenomena (such as carotid-cavernous fistula, retro-orbital hematoma, and carotid aneurysm), or idiopathic. Additionally, the only reported risk factor for superior orbital fissure syndrome is a pre-existing narrow superior orbital fissure.[5-8]

The diagnosis of superior orbital fissure syndrome is suspected on clinical grounds. Common clinical findings in superior orbital fissure syndrome include several symptoms such as ophthalmoplegia, ptosis, proptosis, pupillary dilatation, lacrimal hyposecretion and loss of corneal reflex. Management of superior orbital fissure syndrome depends on the etiology. Reported treatment options include medical and surgical intervention, as well as observation alone. Complete

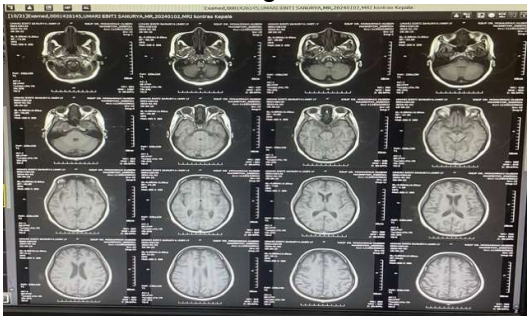
recovery of all nerves has been reported in 24-40% of patients who received steroid treatment compared with 21.4% in patients who did not receive steroid treatment. Recovery usually extends over several months with steady progress within 6 months.[5,9]

In this report, we will examine the patient's history, physical examination findings, diagnostic test results, and the treatment plan implemented. It is hoped that all this information will contribute to further understanding of superior orbital fissure syndrome, while emphasizing the importance of a multidisciplinary approach in responding to such cases. Thus, it is hoped that this case report will make a positive contribution to clinical practice and the development of medical knowledge about this condition.

CASE PRESENTATION

A woman, 49 years old, was treated by the Neurology Department of Mohammad Hoesin Hospital and consulted at the Neuro Ophthalmology subdivision on January, 3rd 2024 reporting symptoms of her right eye was difficult to move since 19 days before admission to the hospital. The patient acknowledged experiencing gradual weakness in the lower extremities, initially feels numbness in the right leg followed by weakness. The patient also complained of double vision and drooping of the left eyelid.

Following the completion of the anamnesis, an ophthalmological examination was conducted, which revealed a right ocular movement deficit and hypotropia. Right palpebral measurement on Margin to Reflex Distance 1 (MRD1) 5 mm, Margin to Reflex Distance 1 (MRD2) 6 mm, and Palpebral Fissure Height (PFH) 11mm. But on the left eye MRD 1 1 mm, MRD 2 5 mm, and PFH 6 mm. The right cornea sensitivity test was hypoesthesia.



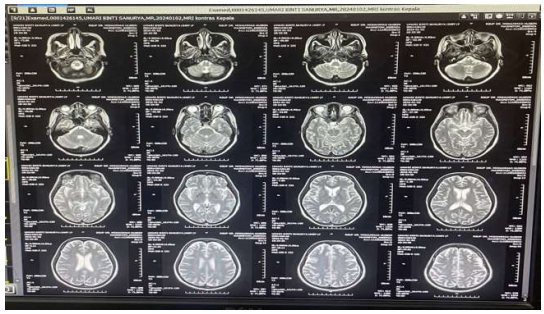


Figure 1. MRI Examination of the patient showed Normal Results.

Radiological examinations have been carried out on patients such as orbital Magnetic Resonance Imaging (MRI), contrast head MRI and whole spine MRI. But all the examinations showed normal results. CSF examination was also carried out, but the results also showed no signs of malignancy.

Figure 2. Binocular photo of the patient's show hypotropia in the right eye and ptosis in the left eye.

DISCUSSION

From the results of the history, it was found that the patient's symptoms indicated superior orbital fissure syndrome (SFOS). SFOS is a rare symptom complex. SOFS occurs due to compression of the superior orbital fissure compartment. The clinical picture can vary from partial to complete neurological sequelae depending on the degree of injury. Clinical findings of this syndrome include upper eyelid ptosis (due to sympathetic denervation of the Mueller muscle or involvement of CN III resulting in paralysis of the levator palpebra superioris

muscle); ophthalmoplegia (due to paralysis of CN III, IV and VI); mydriasis, loss of accommodation reflex, loss of direct pupillary reflex (due to involvement of CN III and accompanying parasympathetic fibers, resulting in paralysis of the ciliary muscle); loss of sensation in the eyeball area, upper eyelid and forehead area as well as loss of corneal reflex (due to CN V1 paralysis) and proptosis (due to loss of extraocular muscle tone). Some of the symptoms found in patients are ophthalmoplegia, mydriasis, proptosis, and loss of sensation in the corneal reflex area of the eye. The patient did not find optic nerve involvement so that suspicion of orbital apex syndrome could be ruled out.

The patient is then planned to undergo an orbital MRI examination to look for the underlying etiology of the patient's symptoms. Previously, an MRI examination with contrast had also been carried out from the neurology department. However, from the expert results, both examinations showed that there were no abnormalities such as lesions in the brain parenchyma or signs of infarction, SOL, or AVM. However, these results still need to be examined again because the neurology department suspects that there are several tumors in the head. Because the patient was suspected of having an intracranial mass, the neurology department consulted the neurosurgery department for a possible biopsy of the mass.

The patient also underwent other examinations from the neurology department such as an MRI Spine examination, cerebrospinal fluid and cytology examination for the cerebrospinal fluid. From examination of the cerebrospinal fluid, it was concluded that cytoalbumin dissociation occurred. Then, for the cytology examination. The literature states that cytoalbumin dissociation can occur due to several things, namely the production or release of intrathecal proteins such as IgG and myelin basic protein, blood-brain barrier dysfunction in meningeal or parameningeal inflammation, blood-nerve barrier dysfunction in neuropathy, CSF absorption in spinal compression, or decreased CSF flow. So, the possibility of infection or inflammation cannot be ruled out. The results of the whole spine MRI examination showed indications of malignancy, so suspicion of a tumor mass or malignancy could not be ruled out.

This patient experiences various symptoms in the eyes and nervous system which can be interconnected with each other. However, the underlying etiology has not been discovered because several examinations have obtained normal results. So further exploration is needed to find and understand the cause of the symptoms experienced in this patient.

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