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A case of Foster Kennedy Syndrome (FKS) with Increase Intracranial Pressure (ICP) Attributed to Cerebral Venous Sinus Thrombosis (CVST)

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Here we present a case of Foster Kennedy-like syndrome, in which meningioma compressed one optic nerve and cerebral venous sinus thrombosis impair cerebral venous drainage inducing intracranial hypertension, and papilledema in the other eye. This case emphasis the importance of considering of CVST as underlying pathogenesis of raised ICP in absence of cerebral edema or obstructed hydrocephalus.

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A Case of Foster Kennedy Syndrome (FKS) with Increase Intracranial Pressure (ICP) Attributed to Cerebral Venous Sinus Thrombosis (CVST)

Madihah ALhubayshia[α] & Tahir Obeidb [σ]

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I. Introduction

The Foster Kennedy syndrome consists of optic disc pallor in one eye, optic disc edema in the other eye, and reduced olfaction caused by space-occupying lesion. [1, 2]

Classically, The ophthalmological sign of Foster Kennedy syndrome (FKS) produced by direct compression of the ipsilateral optic nerve by basal frontal lobe, olfactory groove or sphenoid wing meningioma leading to optic atrophy with concomitant contralateral optic disc swelling secondary to raised (ICP) caused by the mass effect of the tumor. [1-6] Anosmia results from direct compression of the olfactory nerve.[1, 2]

We describe a case report regarding the different pathogenic mechanisms, for which increased intracranial pressure (ICP) resulting from CVST. This is to suggest that not all FKS cases have the same underlying pathogenesis.

Our aim is to document this unique association and to draw attention on the importance of its presence. This is because the management in such situation advocates the use of anticoagulation.

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II. Case Study

27 year old female referred to King AbdulAziz Medical City (KAAMC), Jeddah in 2004 with one week history of headache, diplopia and visual deterioration in both eyes, particularly in the left eye. She had occasional vomiting with no nausea. She had no symptoms to suggest underlying systemic diseases.

Examination showed visual acuity of 20/30 in the left eye and no light perception in the right eye. A right relative afferent pupillary defect was present with optic disc edema in the left eye, **(fig.1.b)** and an atrophic right optic disc **(fig.1.a)**

Slit lamp examination normal for both eyes.

There was left 6thcranial nerve palsy. Humphrey perimeter of the left eye revealed a superior nerve fiber bundle defect **(fig.2.)**. The rest examination was normal.

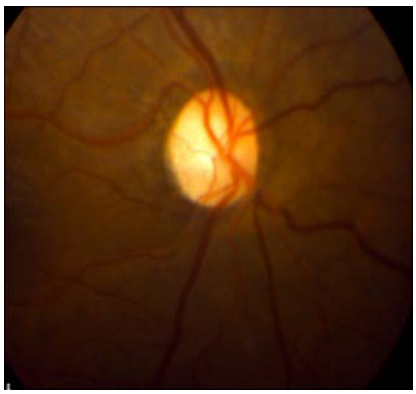

Figure 1.a : Atrophy of the right optic disc

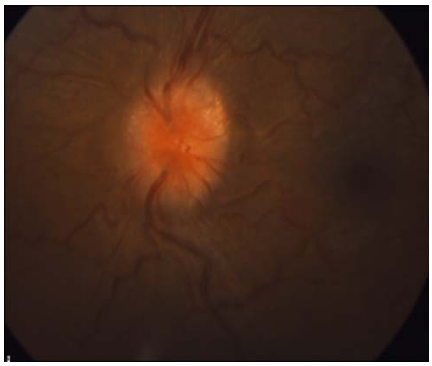

Figure 1.b : Edema of the left optic disc

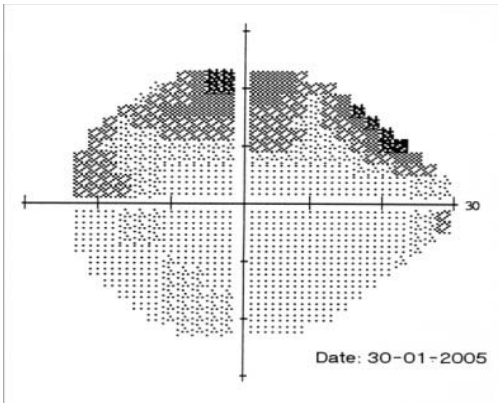

Figure 2 : Humphrey perimeter of the left eye disclosed an superior nerve fiber bundle defect

CT scan of the brain showed hyperostosis in right sphenoidal and right Occipitoparietal bone **(fig.3)**.

MRI/MRV**(fig.4)** showed right sphenoidal wing strongly enhanced mass that consistent with the diagnosis of meningiomatosis. It extended from the right orbital apex through the optic canal and into the intracranial space. MRV study showed thrombosis of the posterior third of the superior sagittal sinus and the right sigmoidal sinus **(fig.5)**

Diagnostic Angiogram showed tow tumor blushes highly suggestive of meningiomas **(fig.6)**

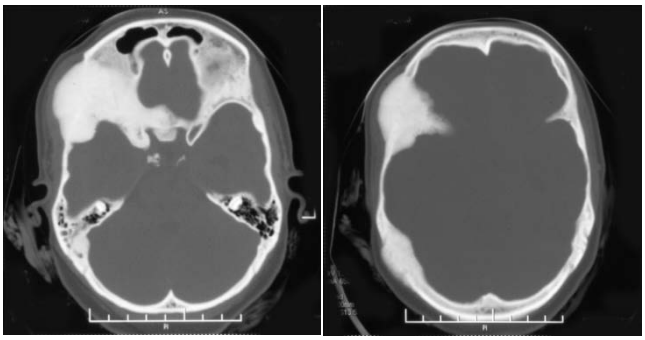

Figure 3 : CT scan of the brain bone window showed Hyperostosis in right sphenoidal and right Occipitoparietal bone

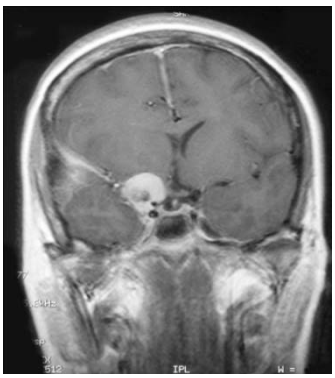

Figure 4 : MRI/ T1 with contrast Showing Right sphenoidal Wing strongly enhanced mass

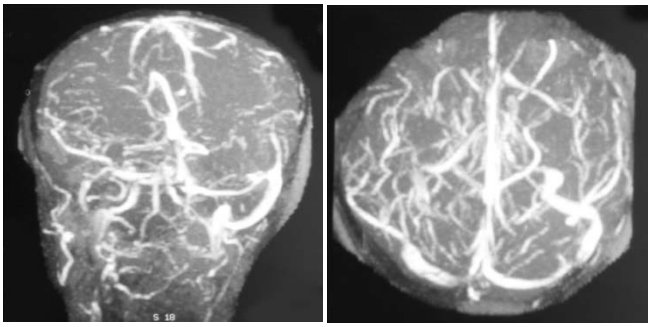

Figure 5 : MRVthrombosis of the posterior third of the SSS

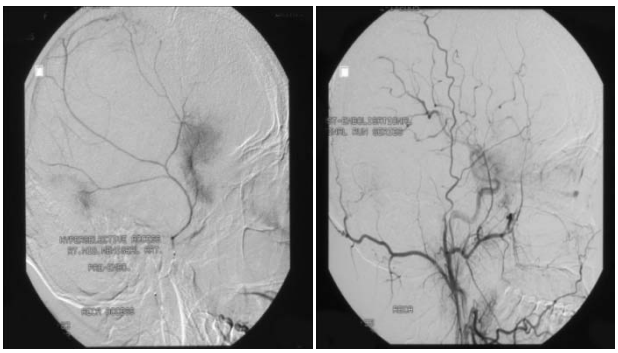

Figure 6 : Angiogram showed tow tumor blushes highly suggestive of meningiomas

The patient was treated with enoxoparine 1mg/kg subcutaneous BID, acetazolamid 500mg BID.

As vision of the left eye continued to deteriorate, the decision was taken to relief the pressure surgically and the patient underwent lumboperitoneal shunt, CSF examination done intra operatively showed opening pressure of 450 mm with normal compositions and negative cultures for bacterial and fungal infections, there was no evidence of malignant cell.

Other diagnostic tests including complete blood count, complete metabolic panel, erythrocyte sedimentation rate, c-reactive protein, and were unremarkable.

Post operatively, The patient received warfarin with therapeutic INR (2-3).

One month later the papilledema and visual field had improved on the left eye, and visual acuity was stable.

After 5 years, the patient vision stabilized and developed no shunt complication.

III. Discussion

The mechanism and underlying pathology of ophthalmic feature of the FKS is variable. The classical pattern is caused by unilateral direct compression of the optic nerve fiber by meningioma and secondary increased intracranial pressure (ICP) causing papilledema of the contralateral eye. [1,2,4] Other mechanism have been suggested to explain the underlying pathology due to bilateral direct optic nerve fiber compression with normal ICP while atrophic

changes resulting from asymmetrical compression of both optic nerve by tumor. [7]

It may also results from chronic increased ICP which initially gives bilateral papelledema, with one optic disc subsequently developing pallor as a result of axonal death while the other optic disc remains swollen. [7]

This case of FKS meningioma cause direct compression of the right optic nerve, and presence of CVST impair cerebral venous drainage that round the other optic nerve by thrombus in which CSF is at a higher pressure than normal and which becomes responsible for papilloedema on the opposite side.

The treatment of visual loss due to Papilledem depends largely on the underlying pathology hence it is important to look for CVST when obvious brain edema and obstructed hydrocephalus are lacking, Prompt use of anticoagulation and cerebrospinal fluid diversion is critical in prevented visual loss in our case.

There is a previous report a patient with FKS in which meningiomas compressed the superior sagittal sinus to block cerebral venous drainage causes increased intracranial pressure, and papilledema in the other eye.[8] Our case differ from this patient by having CVST.

We may postulate that some of the previous cases of FKS may have CVST as a cause of ophthalmic feature of this syndrome rather than a direct compression and/or an intracranial hypertension while MRI, MRV were lacking it emphasizes the importance of looking for this association as adding anticoagulation among other measures.

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