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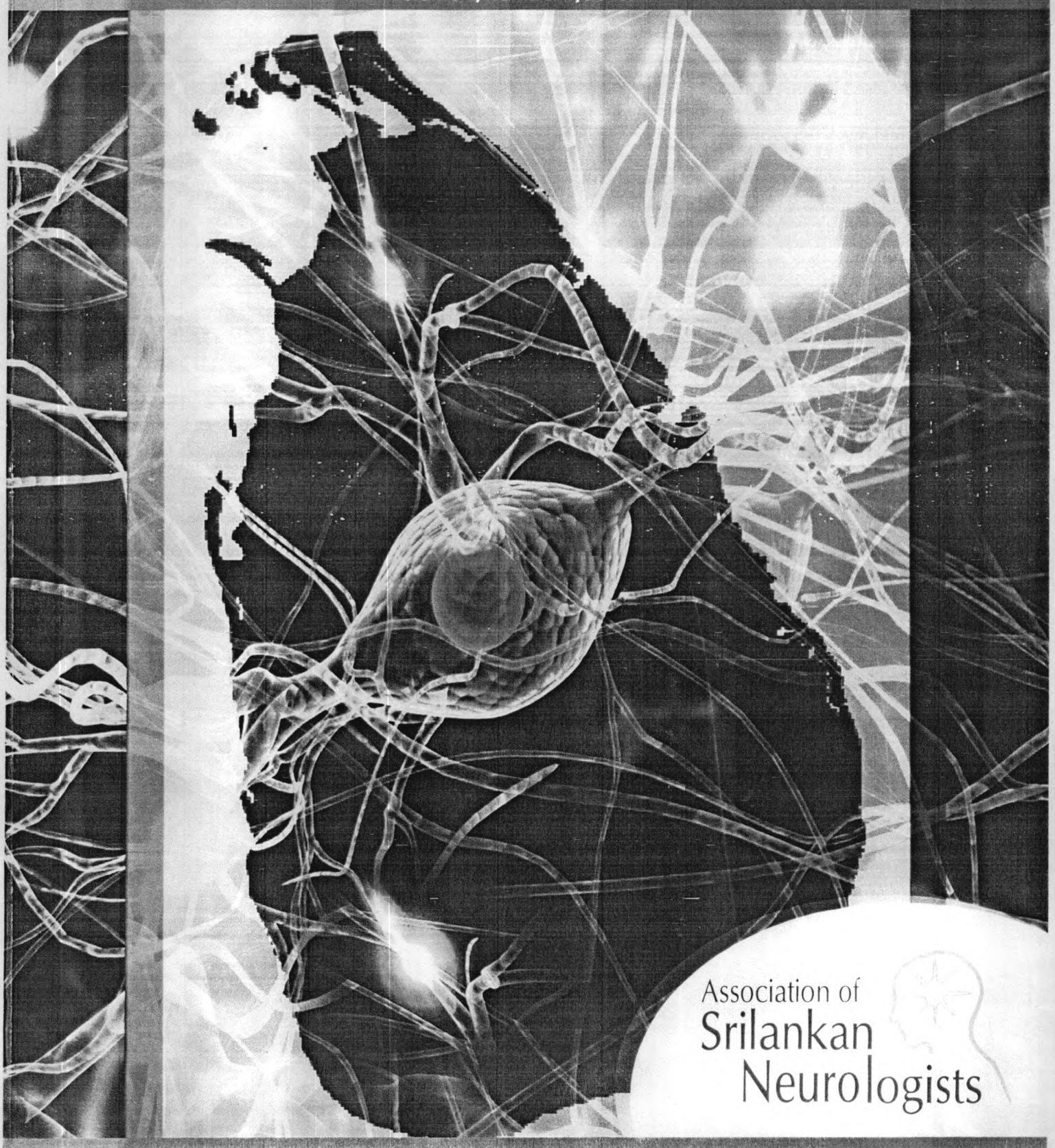

Association of Srilankan Neurologists

Isolated sphenoidal fungal sinusitis in a patient with ulcerative colitis on steroids

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Background

Isolated sphenoid sinus lesions are rare, and account for 1-2.7% of all paranasal sinus lesions[1]. These lesions are sphenoid cyst, sphenoid sinusitis, fungal disease, inverted papilloma, sphenochoanal polyp, foreign body, malignant tumors. Isolated sphenoidal sinusitis can be bacterial or fungal and the incidence of fungal infection is about 25-30% of all and this is categorized as non-invasive, invasive indolent and fulminant[2]. Noninvasive fungal sinusitis usually involves only one sinus and out of all para-nasal sinuses, commonest to be involved is the maxillary sinus and the commonest organism being aspergillus[3]. Isolated sphenoidal sinus fungal disease is rare as a result of the unfavorable anatomic location of the sphenoids and decreased nasal airflow in that region[4]. This disease is more common in immune-compromised patients, but there are cases reported in immune-competent individuals as well[5].

Since isolated sphenoidal fungal sinusitis is rare, and also because it presents with a range of non specific symptoms, such as headache, visual symptoms or cranial nerve palsies, the diagnosis can be easily missed if not for a higher degree of suspicion. The diagnosis of isolated sphenoidal fungal sinusitis usually requires advanced imaging because, nasal endoscopy and sinus X-ray can be normal in these patients. Due to all these reasons, the literature reveals, that the diagnosis of fungal sinusitis in sphenoid sinus is often delayed, where the patients suffer for years with headache or deteriorate with complications.

We report a case of isolated sphenoidal sinus fungal sinusitis, in a patient with ulcerative colitis in remission, presenting with severe unilateral headache and facial numbness leading to a diagnostic dilemma in the initial stage. This elaborates the importance of a high degree of suspicion of this rare disease in this type of a clinical presentation, to image early and prevent further serious complications.

Case presentation

A 43-year-old female presented with a severe unbearable right frontal headache for two weeks duration. It had been persistent and progressive, associated with right orbital pain and facial numbness. She complained of photophobia, and the headache was not associated with tearing or vomiting. The pain did not resolve with simple conventional analgesics. She had no similar episodes in the past. She had no features suggestive of sinusitis such as nasal blockage, discharge or a history of sinusitis. In hospital she continued to have episodic excruciating right sided headache, with a persistent background dull headache. She had been diagnosed with ulcerative colitis 10 yrs back, now in remission and also having sero-negative arthritis for which she is on long term treatment with a small dose of oral prednisolone and sulfasalazine.

On examination, she had no sinus tenderness or intranasal abnormalities. Pupils were normal and there was no papilloedema. But on neurological examination, there was a marked sensory loss over the area of right maxillary sensory division, where she complained of numbness. Other divisions of fifth cranial nerve were found to be normal along with the motor component. Corneal reflex was present and all other cranial nerves were normal. On eye screening, intra ocular pressures of both eyes were normal.

The initial differential diagnosis considered for her presentation were severe migraine, pseudo tumor cerebri, space occupying lesion, or venous sinus thrombosis. Her blood counts showed an elevated WBC ($12x10^3$/uL) and a high ESR (91mm/1st hour). But CRP was normal. Non contrast CT of brain did not reveal any specific abnormality. Considering severe sinusitis as a possibility for unilateral headache, even without other features of sinusitis, an x-ray sinus view was done and was normal. Nasal endoscopy performed was also normal. Negative results of initial procedures and CT imaging, created a diagnostic dilemma at this point.

Therefore a contrast enhanced CT scan of sinuses was performed with a high degree of suspicion because of severe persistant unilateral pain with trigeminal nerve involvement. It revealed, probable sphenoidal sinusitis. MRI scan of brain with skull base, with infra-temporal views was performed and this confirmed right sided isolated sphenoid sinusitis (Figure).

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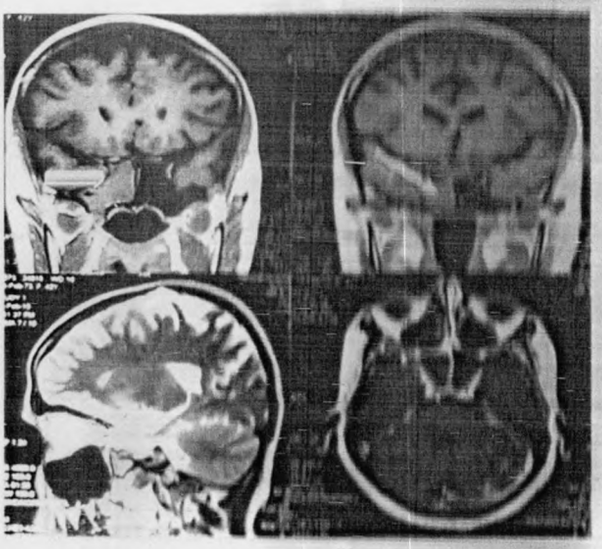

Figure. MRI showing the right sphenoid sinus opaque indicated by arrow

She underwent functional endoscopic sinus surgery where a freely movable mass was identified in sphenoid sinus suggestive of a fungal growth and it was completely removed. Histo-pathological diagnosis confirmed fungal sinusitis with fungal hyphae in tissue. Patient completely recovered after surgery without any complications, even without systemic antifungal therapy. Now, at five months of follow up, she has no headache and the numbness of face had resolved over one month following surgery.

Discussion

Isolated sphenoidal fungal sinusitis is a rare clinical entity and is often misdiagnosed due to lack of specific features in the presence of a non-resolving headache. Headache is the most common presentation of this disease and is mostly deep seated and retro-orbital. The mucous membrane of the sphenoid sinus receives sensory innervation by the posterior ethmoidal nerves (branch of the ophthalmic nerve), and post-ganglionic parasympathetic fibers of the facial nerve that synapsed at the pterygopalatine ganglion which control secretion of mucus. Retro orbital pain is likely as a result of this innervation[1]. But other variants of headache such as vertex, referred occipital or diffuse are commonly seen[3].

Our patient had a deep right sided frontal headache which was throbbing in nature and in the background of dull persistent headache. Other symptoms of isolated sphenoidal fungal sinusitis include visual disturbances including unilateral vision loss, diplopia, blurring of vision due to involvement of optic nerve. Also nasal obstruction, rhinorrhea, hyposmiaare some other features reported[1]. Involvement of cranial nerves especially ophthalmic and maxillary branches of trigeminal nerve that was evident in our patient had been reported in 4 patients in a case series of 15 patients[2].

Depending on the severity of the fungal infection the treatment methods vary. Our patient was falling into non-invasive fungal sinusitis, where hyphae were seen in histology but the fungal culture was negative. She had complete cure of headache and numbness after endoscopic nasal surgery. Therefore she was not treated with systemic anti fungal medications.

The sphenoid sinus is anatomically closely related to a set of vital structures including the dura, pituitary, optic nerve, pterygoid canal and nerve, internal carotid artery and the cavernous sinus with its associated cranial nerves (III, IV, V1, V2, and VI). Therefore, early diagnosis and management of sphenoid sinus lesions is crucial to prevent devastating complications that can occur due to involvement of these structures.

Conclusion

Isolated sphenoidal sinus fungal sinusitis is rare and that should be considered with priority in patients presenting with severe deep seated headache, even if the usual features of sinusitis are absent, with normal initial brain imaging and nasal endoscopy. This case emphasizes the importance of awareness of this rare clinical entity and the need for appropriate imaging.

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