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A CASE OF GIANT INTRACRANIAL ANEURYSM AND SYSTEMIC LUPUS ERYTHEMATOSUS: AN INCIDENTAL OR AN ACCIDENTAL FINDING?

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ABSTRACT

A twelve year old girl, an established case of systemic lupus erythematosus, was referred to us suffering from diplopia and nasal speech for a period of 3 months. Neurological examination was conducted which revealed a right-sided sixth nerve palsy and a pulsatile nasopharyngeal mass. Neurodiagnostic imaging studies confirmed a giant intracavernous internal carotid aneurysm. The malady of the aneurysm and its attendant neurological symptoms responded to the surgical procedure.

Keywords: Giant Aneurysm, Systemic Lupus Erythematosus.

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INTRODUCTION

Systemic lupus erythematosus (SLE) is a generalized disease process involving collagen tissue throughout the body.[1] Pathologic manifestations of SLE include changes in medium and small-sized blood vessels harboring lesions of different stages of development. Involvement of the neuraxis is uncommon, but neurologic problems are encountered in 75% of patients.[2-4] Systemic collagen vascular diseases such as SLE or polyarteritis nodosa can also produce subarachnoid hemorrhage. These conditions sometimes give rise to aneurysmal dilatation of intracranial vessels.[5]

We describe a patient with established SLE who developed a cerebral aneurysm exhibiting an unusually rapid rate of expansion.

The purpose of this report is to emphasize the fact that in patients with connective tissue disorders intracranial aneurysms displaying an unusually fast growth and their attendant complications can of course be averted by timely surgical intervention.

Case report

The patient, a 12 year old girl, had been followed at Dr. Ahari Children's Hospital from April 1992 till January 1995. Her initial complaints were fever and polyarthritis. She had a butterfly rash on the face, and both knees and ankle joints showed arthritic changes. Her clinical picture was highly compatible with SLE as confirmed by laboratory studies. Positive antinuclear and anti-DNA (over 100 IU) antibodies, a 2+ proteinuria, microscopic hematuria and 24 hour urinary excretion of 450 mg protein were the laboratory findings obtained. C3 and CH_{50} were less than 10 and 20% respectively, whereas C4 was 1. A renal biopsy specimen showed minimal vascular changes.

A year later (1993), she developed diplopia. MRI, MRA and cerebral angiography revealed a giant intracavernous aneurysm of the right carotid artery (Fig. 1).

In January 1995, she was referred to us with complaints of nasal speech and blurred vision. On examination, she had a right-sided sixth nerve palsy and a pulsating midline nasopharyngeal spherical mass with a diameter of about 5cm. CT scan demonstrated a nonhomogenous mass in the region of the right cavernous sinus eroding the sphenoid bone and extending from the nasopharynx to the suprasellar region. It expanded to the midline and posterior fossa eroding the petrous bone. A repeat angiogram confirmed

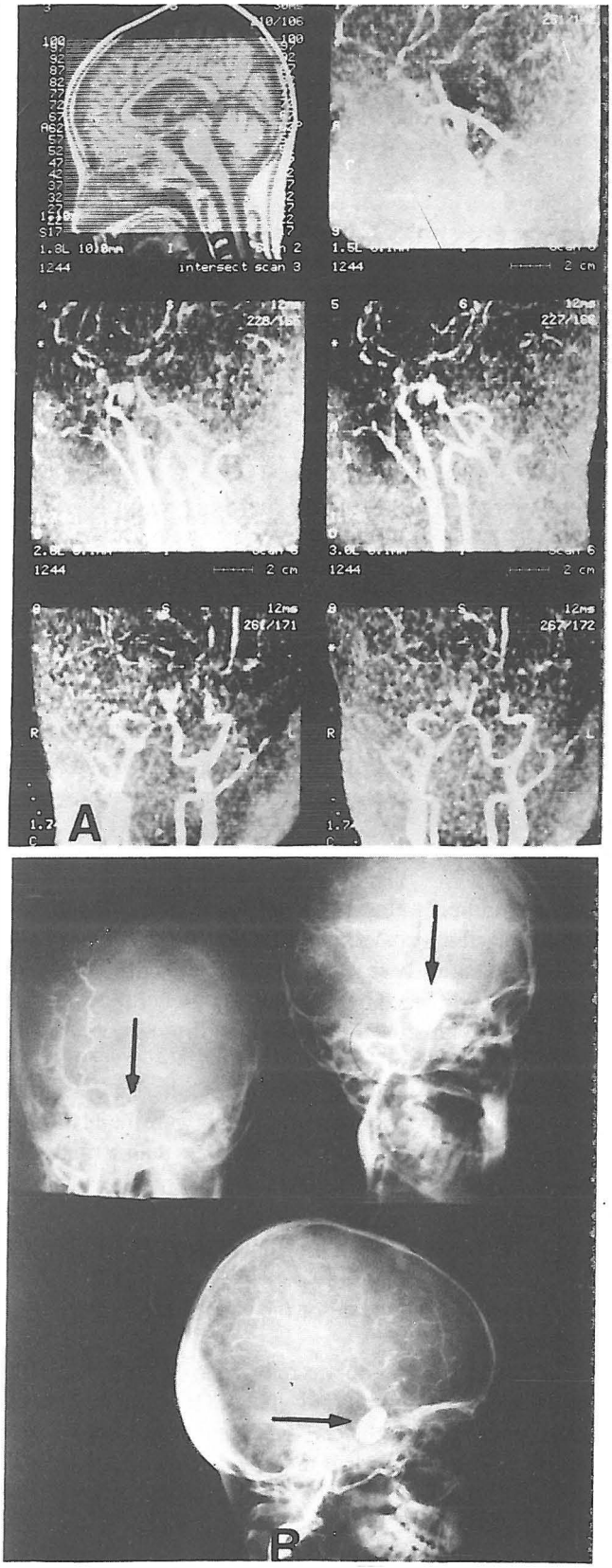

Fig. 1. Preoperative magnetic resonance angiogram (A) and conventional angiogram (B) displaying the aneurysm (arrow).

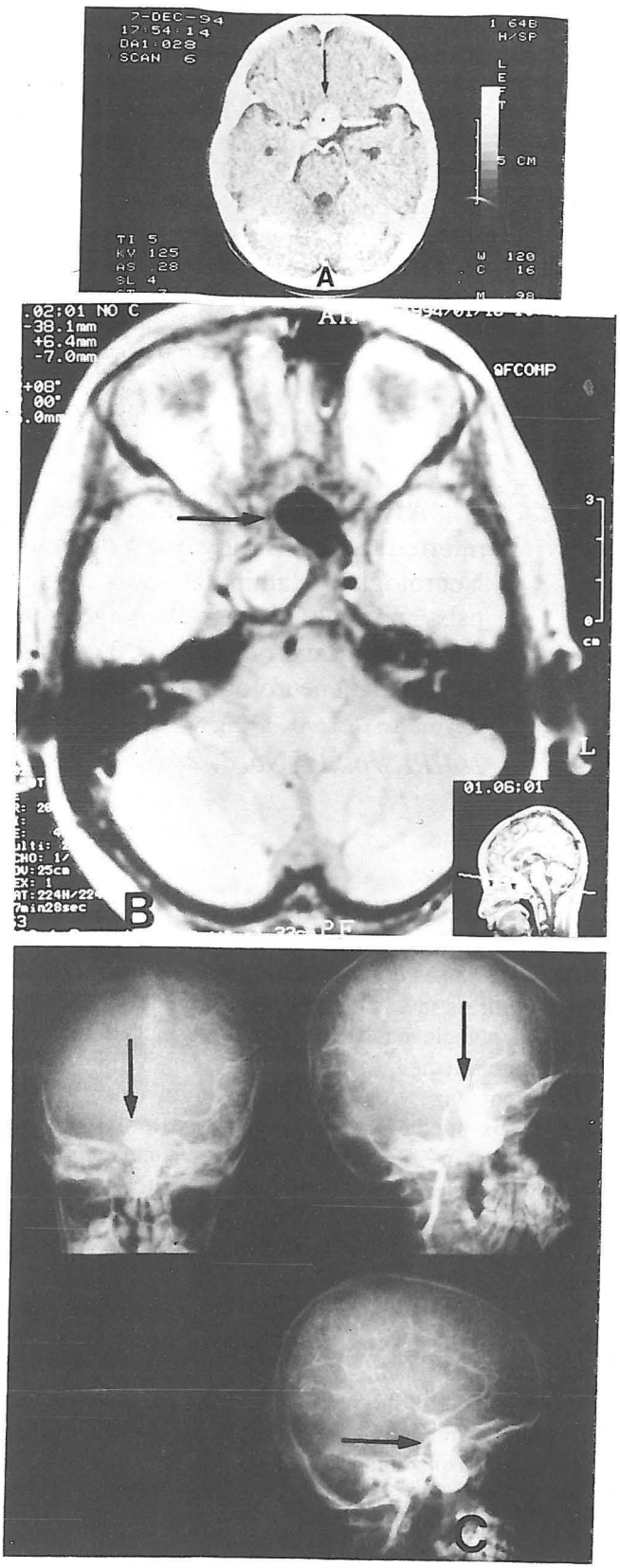

Fig. 2. CT scan (A), MRI (B) and repeat angiogram (C) showing an expanded aneurysm (arrow).

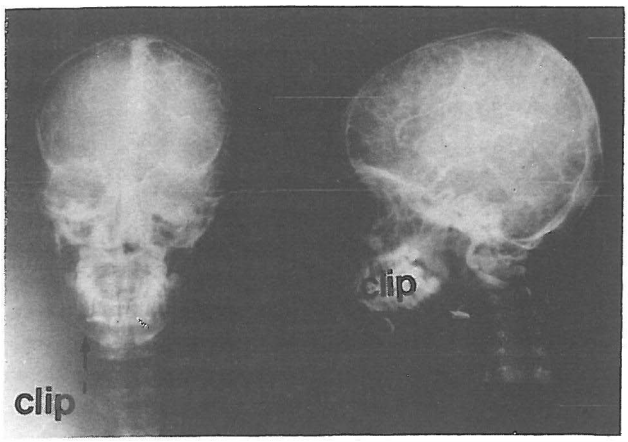

Fig. 3. Postoperative angiogram with no evidence of the aneurysm. Adequate cross-circulation of both hemispheres being reflected after ligation of the right carotid artery in the neck.

the earlier established aneurysm which had grown out of proportion (Fig. 2).

Bilateral carotid artery angiographic studies revealed that the left carotid artery had cross circulation for both cerebral hemispheres and as such the right internal carotid artery was ligated in the neck 3cm distal to the bifurcation of the right common carotid artery.

Postoperative angiogram failed to detect the aneurysm, establishing the adequacy of the planned surgical procedure (Fig. 3).

DISCUSSION

Cerebral arteritis secondary to SLE primarily involves small vessels less than 2 mm in diameter.[6,7] Nevertheless, a case of large vessel CNS vasculitis has also been described in a patient with SLE, reflecting an unusual pathologic finding.[8]

Autopsy findings in SLE patients support the theory that the pathogenesis of cerebral aneurysms is acquired rather than congenital.[4,9] There seems to be an increased prevalence of aneurysms diagnosed in patients suffering from vasculitis and connective tissue disorders compared to the overall general population. Nonetheless, because of the sporadic reports of cerebral aneurysms in patients with lupus erythematosus, the relationship between cerebral aneurysms and SLE is still unclear[1] and the association is possibly coincidental.[7] Likewise, although several cases of subarachnoid hemorrhage caused by berry aneurysms have been described in SLE,[10,11] it is not known whether these lesions are more common in those with SLE than in the general population.[11] Some, however, claim that the incidence of subarachnoid hemorrhage due to berry aneurysm is higher in SLE than in the general population.[5]

Cases of cerebral or spinal aneurysms associated with SLE are being increasingly reported, although formerly these two diseases were usually only found to be associated at biopsy[8] and the dilemma as to why some and not all patients with SLE develop neurological involvement remains unsolved. The pathological findings of isolated infarcts and plugging of small vessels with hyaline material thought to be caused by vasculitis have been observed at autopsy, but these studies do not add much to the understanding of the pathogenesis.[4]

Although at least 23 cases of SLE-associated aneurysms have been reported, this association is rare and possibly coincidental,[1] but the following points merit attention and may indicate that this association may be incidental and not an accidental happening:

1. Formation and growth of saccular aneurysms are acquired.[7]

2. Fibrinoid degeneration of collagen and destruction of medial smooth muscles and elastic fibers in SLE may facilitate aneurysmal development.[6]

3. In the reported cases, there was a tendency for cerebral aneurysms of unusual sites to be accompanied with vasculitis. Aneurysms at the sites of frequent occurrence had no evidence of vasculitis.[6]

Some look with skepticism over the established notion and belief that cerebral aneurysms are more common in those with SLE than in the general population and regard this association to be a chance finding. However, we feel that patients with SLE are especially prone to aneurysmal formation because the arterial wall becomes unexpectedly fragile. On a limited number of 23 cases of aneurysm in SLE, it is rather difficult to establish with certainty as to whether this association is accidental or incidental, but the pathological changes observed in the vessels of patients with SLE strongly support the assumption that these changes could most probably lead to aneurysmal dilatation of intracranial vessels.

Although cases of cerebral or spinal aneurysms associated with SLE have been reported,[4,5,8] this giant intracranial aneurysm seems unique. The protracted illness of the patient enabled us to follow the natural course of an aneurysm based on a connective tissue disorder in a patient with SLE. The unusually rapid rate of growth of this aneurysm has brought us to the conclusion that prompt intervention in SLE patients harboring giant intracranial aneurysms is highly indispensible to prevent catastrophic complications.

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