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Saints and pseudo-Seizures - A Christmas story

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It was a few weeks before the Christmas of 1951, and all through the Neuro the surgeons were busily stirring. Dr. Penfield operated on a young man (P.S.) sent from Paris by Dr. Henry Hécaen because of seizures for thirteen years. Herbert Jasper had found epileptic spiking in the anterior temporal region. At operation spikes were recorded from the temporal cortex and also from the amygdala, from which stimulation evoked one of the patient's typical attacks of confusion and automatic movement for which he had no recall afterwards (Fig. 1). During the temporal lobe excision an unexpected low grade glioma was also removed. (Today this would have been detected on MRI - see Neuro-Image for February 1989). Before operation the young man's seizures were vivid - he would describe colored lights - purple, violet, blue and yellow - and at night, green stars. In other attacks he heard bells or would hum a tune. In still another, he was convinced that he was dead and in heaven - he saw violet and blue saints and began to pray over and over again. When questioned later, he seemed to recall this as not just a dream, but the real thing.

His post-operative course, as surgeons say, was uneventful. Everyone was pleased that he had no more of these fearful attacks and elaborate hallucinations. It was nearing Christmas and he was eagerly making plans to return with his father to France.

But before he left hospital, a curious and distressing episode occurred. The Neuro then, perhaps more than at any time in its history, was jammed with patients. The Military Annex of some 30 beds had been demolished to make way for the McConnell Pavilion and its eighty new beds, still under construction. T o accommodate the heavy admissions, especially the

(suite à la page 8)

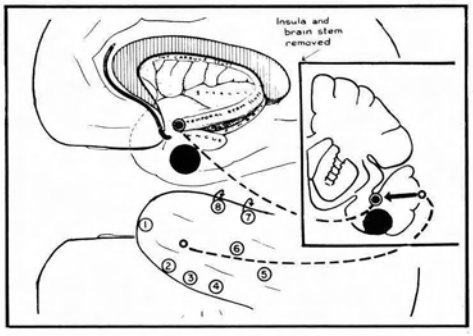

Figure 1 - Depth stimulation at operation in the anterior and lateral part of the amygdala producing automatism in case P.S. The solid circles indicate the site of the small glioma.

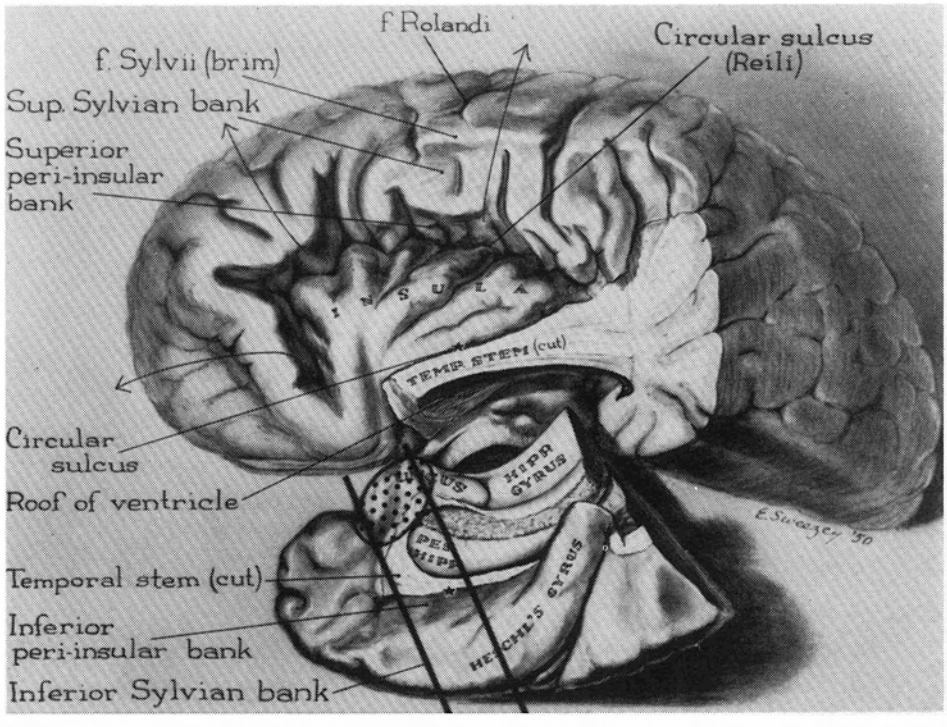

Figure 2 - The temporal lobe dissected and laid aside shows the position of the amygdala (stippled) between the temporal stem and the uncus at the tip of the temporal horn of the ventricle. (Both figures modified from Feindel and Penfield, 1954).

Cauda Equina Syndrome Complicating Ankylosing Spondylitis: Clinical, CT and MR Features

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Introduction

Ankylosing spondylitis involves primarily the spine and sacroiliac joints. The cauda equina syndrome (CES) is a late neurologic complication of the disease. We report a patient with a long history of ankylosing spondylitis who developed CES. The clinical history and the computed tomography and magnetic resonance findings are discussed.

Case report

A 52 year-old man had a 28 year history of ankylosing spondylitis. During the last 10 years before admission, he progressively developed a neurogenic bladder and constipation. He also had impotence, hypoesthesia over the sacral dermatomas and paresthesias over both legs.

On physical examination, there were severe limitations of movements at the neck, trunk and lower limbs. There was cutaneous loss and muscle weakness in the distribution of L4, L5 and sacral roots. Both ankle jerks were absent while knee reflexes were increased.

Conventional radiographs of the spine showed complete fusion from C2 to the sacrum and bony ankylosis at both sacroiliac joints. Computed Tomography of the lumbar spine without contrast (Fig. 1) showed erosions of the laminae and spinous processes. Magnetic resonance imaging (Fig. 2) showed dilatation of the spinal canal from L1 to L5 with dural ectasia and posterior diverticula filled with CSF.

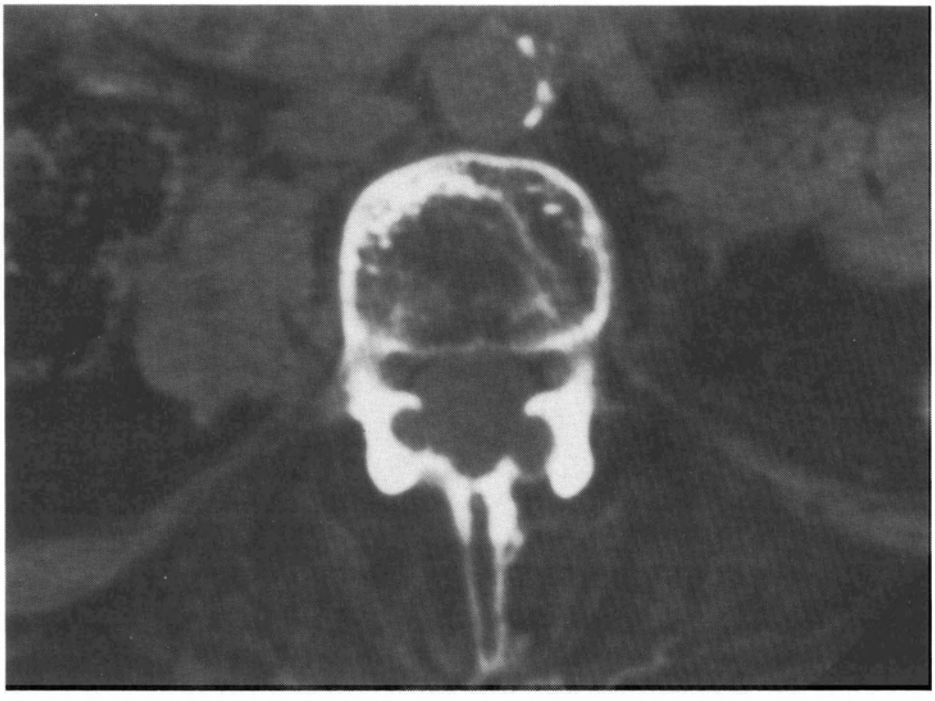

Figure 1 - Axial CT scan at L4 shows multiple erosions of the laminae and spinous process. The spinal canal has a homogeneous CSF density.

Discussion

Cauda equina syndrome (CES) is a rare, late neurologic complication of ankylosing spondylitis. It usually happens at a time when the disease appears to be in a quiescent stage both in terms of symptoms and laboratory tests. There is usually a long time interval between onset of ankylosing spondylitis and that of CES. Bartleson et al (1) reported on 14 patients where this time interval was 35 years. Most of their patients had completely fused spines by the time symptoms referable to CES appeared. Urinary complaints are usually prominent and include urgency, frequency, decreased force of the urinary stream and incontinence, usually overflow in type (1,2,3). This may progress to neurogenic bladder. Patients frequently present with incontinence of stool or constipation and impotence.

Pain in the legs, feet or rectum may also be present. There is usually symmetrical bilateral cutaneous sensory loss in lumbosacral dermatomas at and below L5. Superficial pain, temperature and touch sensations are most likely affected. Lower limb weakness in the distribution of L5 and sacral roots is usually not a prominent feature. The lower limb reflexes may be reduced or absent. Rarely, they are increased.

The Computed Tomography (CT) findings have been described previously (1-4). There is irregular dilatation of the lumbosacral spinal canal with asymmetric erosions of the inner surfaces of laminae and spinous processes at multiple levels. CT following myelography shows diverticula that fill with contrast and extend posteriorly towards the laminae and spinous processes and laterally through intervertebral foramina.

Abello et al (5) and Rubenstein et al (6) have described the MR appearance of CES complicating ankylosing spondylitis. The findings in their patients were identical to those in our patient. There is moderate to severe ectasia of the dural sac with multifocal dorsal sacculations filled with CSF and consistent with diverticula. The pathogenesis of these findings is unknown. Bartleson (1) suggested that arachnoiditis was the initial insult which subsequently became inactive. In addition, ankylosing spondylitis is associated with atrophy of the peridural tissues and adherence of the dura to periosteum (1). These pathological changes presumably reduce the elasticity and compliance of the dural sac and impair its ability to dampen brief CSF pressure fluctuations. Over a course of years, this mechanism would cause the formation of slowly enlarging arachnoid diverticula and secondary erosions of the laminae and spinous processes. The CSF pressure fluctuations could also be responsible for the thinning and demyelination of nerve roots of the cauda equina which have been observed. As attractive as this hypothesis may be, the occurence of arachnoiditis as the primary event has not been proven. Operative findings have usually shown the dura and arachnoid to be neither inflamed nor thickened. In addition, CSF in patients with CES is normal except for an occasional mild increase in protein levels. The cauda equina deficits are not totally explained by the dorsal arachnoid diverticula. Some diverticula may extend into the intervertebal foramina and compress the exiting nerve roots. Intradural cysts have been described in CES complicating ankylosing spondylitis and these may also cause nerve root compression. Finally, ischemic and immunologic processes have been suggested to explain thinning and demyelination of the nerve roots but evidence supporting these hypotheses are lacking at this time (2,3).

In conclusion, the cauda equina syndrome in a well-known long term complication of ankylosing spondylitis. It is associated with definite morphological changes that can be particularly well demonstrated by MR without intrathecal injection of contrast material.

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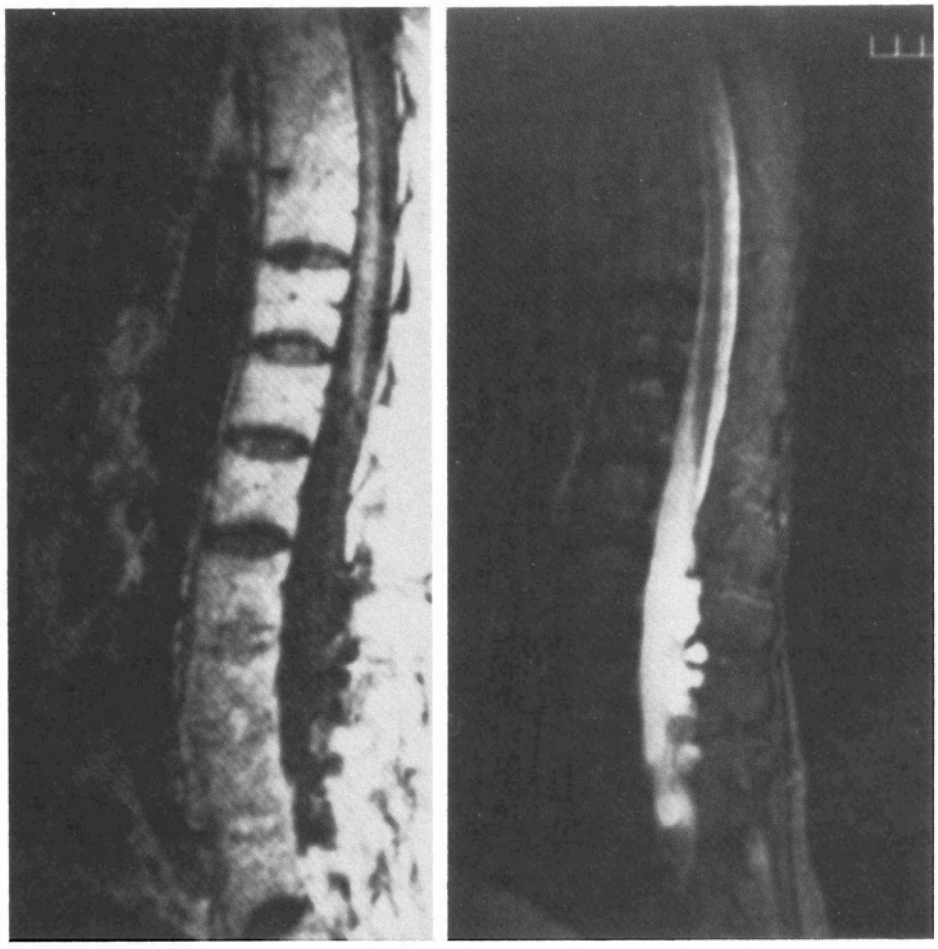

Figure 2 - Sagittal T1-weighted (A) and T2-weighted (B) images show evidence of dural ectasia with posteriorly oriented diverticula. These diverticula are filled by CSF as demonstrated by a signal intensity similar to that of the dural sac.

Role of MRI in temporal lobe seizures: localization of pathology and anatomical verification of surgical resections

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In a recent analysis based on the MNI surgical series of patients treated for temporal lobe epilepsy, we found that two samplings, of 100 cases each, showed the same percentage of successful seizure control (2 to 24 year follow-up) when the anterior mesial temporal resection included only the amygdala and part of the pes of the hippocampus, as compared to a similar resection that included the amygdala but, in addition, half or more of the hippocampus (1,2).

The rationale for focussing on the removal of the amygdala derives from our original findings (3) where the amygdala was identified from electrographic and stimulation findings at operation as the structure most commonly giving rise to seizures with automatism and amnesia. Our subsequent results in many hundreds of patients have confirmed this view (4).

The sparing of the hippocampus, especially on the dominant side, avoids impairment of certain aspects of memory function as detailed by careful neuropsychological evaluation by Milner and associates before and after operation and at longer term follow-up (5).

Magnetic Resonance Imaging (MRI) has proved invaluable in supporting the implication of the mesial temporal region, and especially the amygdala, in temporal lobe seizures with automatism; focal structural lesions, such as indolent hamartomas, low grade tumors (such as gangliogliomas) and venous angiomas are well demonstrated on MRI (some 30% are missed or poorly visualized on CT scans) (6). Moreover, about half of the remaining patients have shown on MRI examination (in 3 planes and with appropriate scanning parameters) evidence of a signal change in the amygdala and anterior hippocampus that correlates on pathological examination with gliosis and neuronal loss (7).

The precise extent of the excision in the treatment of temporal lobe seizures can be identified by MRI so that the results from different neurosurgical clinics can now be compared more usefully. For example, the completeness of resection of the amygdala with sparing of the hippocampus can be readily confirmed anatomically on post-operative MRI (Fig. 1,2,3). This will undoubtedly improve the critical evaluation of post-surgical results in relation to the different patterns of surgical resection, whether this involves (as in many patients of the MNI series) mainly anterior cortical resection and resection of the amygdala, with sparing of the hippocampus, or the standardized block resection popularized by Falconer et al. (8) where a larger extent of the hippocampus is included (but probably with subtotal removal of the amygdala), or the operation with no cortical excision but with removal of the amygdala combined with the hippocampus as initiated by Niedermeyer (9) and more recently adapted by Yasargil and Wieser (10) using selective EEG techniques and microsurgical dissection.

Precise definition of the anatomy of surgical removal in such patients by MRI also provides more exact pre- and post-operative correlation with EEG findings, neuropsychological assessments, and psychiatric

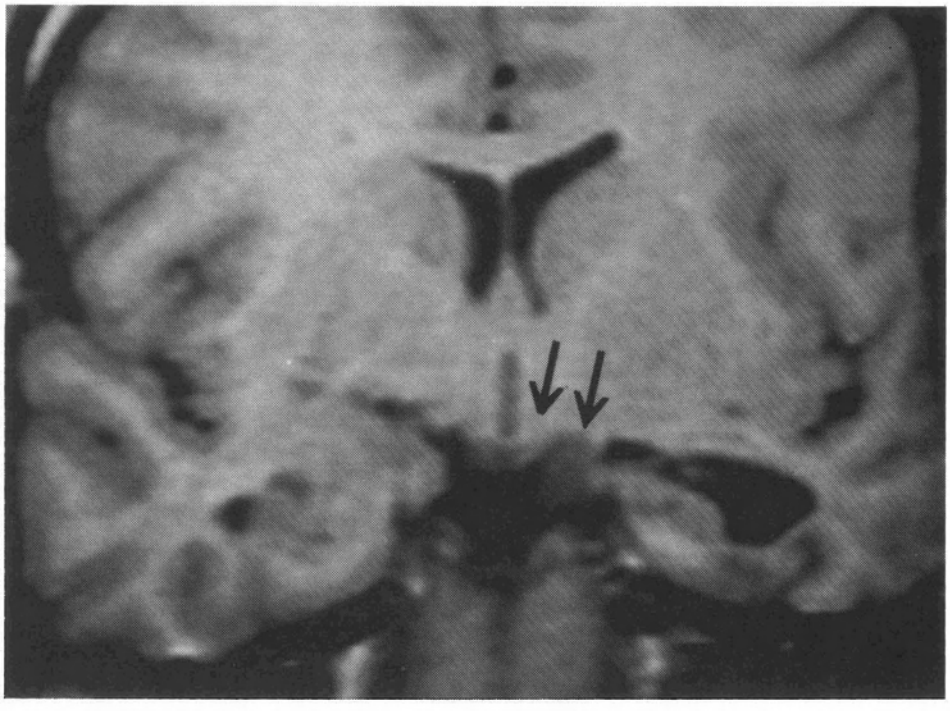

Figure 1 - Coronal MRI showing resection (arrows) of the posterior mesial part of the amygdala.

evaluation, as well as, of course, with the most crucial feature, the results of control of seizures as determined from long term follow-up studies (11).

NOTES:

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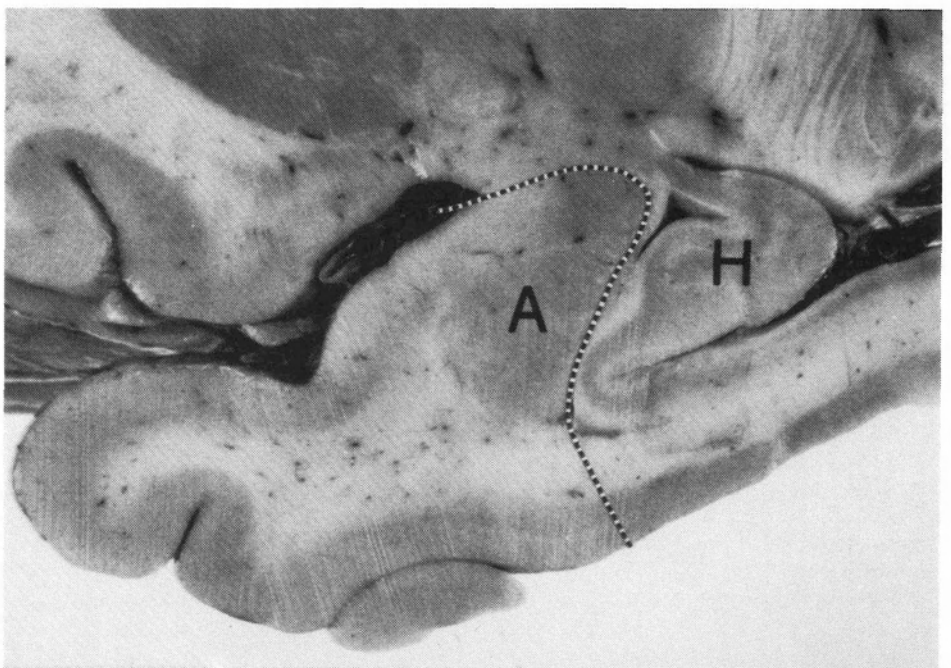

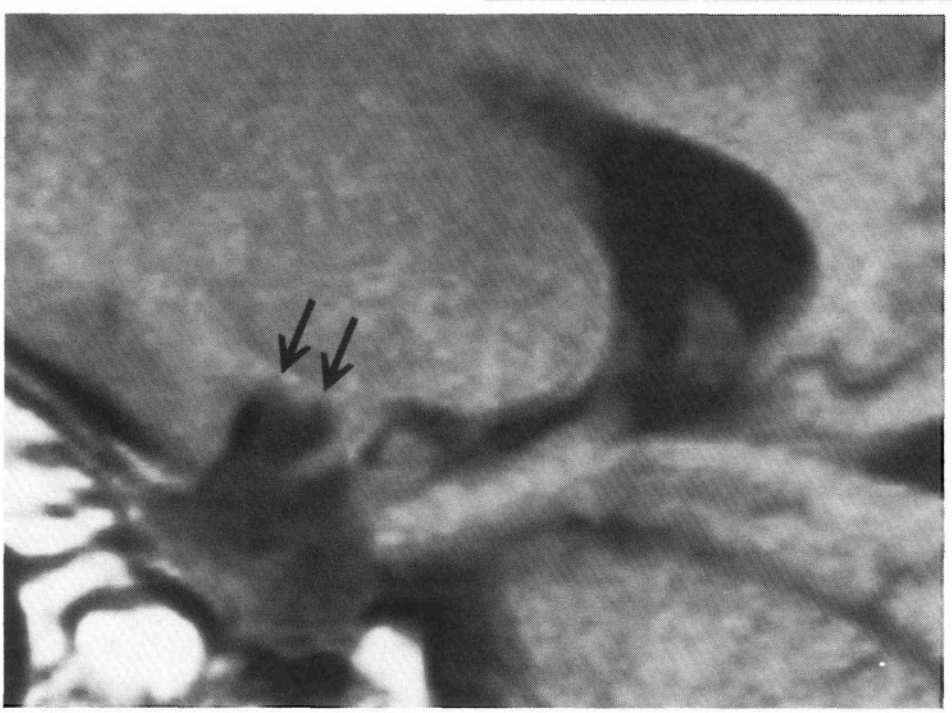

Figure 2 - Sagittal section through the amygdala and hippocampus, separated by the slit of the temporal horn of the ventricle. Line indicates margin of excision.

Figure 3 - Sagittal MRI clearly identifying the resection of superior mesial part of the amygdala (arrows).

An unusual presentation of Wegener's Granulomatosis: Headache and Diplopia

STEPHEN Z GRAHOVAC

Wegener's granulomatosis is an uncommon systemic disease of unknown etiology which may on occasion present with neurologic complications. A case presenting itself in an unusual fashion is illustrated.

Case report

This 74 year old gentleman presented to hospital with a one week history of severe squeezing headache centered above the right orbit. Four days prior to admission diplopia developed and progressed the next day to visual blurring in the right eye. One day prior to admission a severe right ptosis developed.

One year previously similar headaches had been present on the left side, associated with night sweats and were thought to be cluster headaches. At that time the ESR was 45 and a temporal artery biopsy had shown an adventitial lymphocytic infiltrate.

On examination there was a right afferent pupillary defect, a right 6th nerve palsy, a right 3rd nerve palsy sparing the pupil and a right central scotoma as well as ptosis.

CT of the head showed severe sinusitis with total opacification of the sphenoid sinus and slight bulging of the cavernous sinus (Fig. 1 and 2). A chest X-ray demonstrated multiple pulmonary nodular infiltrates with evidence of cavitation in some. An MR examination followed which confirmed the disease within the sphenoid sinus and demonstrated contiguous disease within the right cavernous sinus (Fig. 3 and 4). An open biopsy confirmed diagnosis of Wegener's granulomatosis.

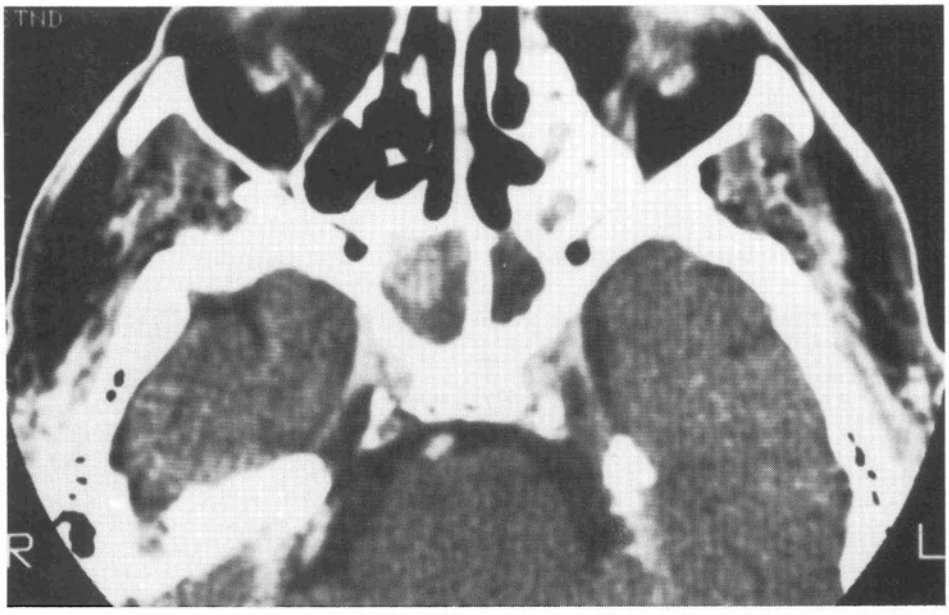

Figure 1 - CT scan (enhanced) - evidence of soft tissue density filling the cavernous sinus.

Figure 2 - CT scan (enhanced - coronal reconstruction) sphenoid sinus disease with fullness of right cavernous sinus.

Discussion

Wegener's granulomatosis is a multisystem disease with a distinct clinical pattern, characterized by granulomatous vasculitis of the upper and lower respiratory tracts together with glomerulonephritis. Untreated, the disease is rapidly fatal, secondary to renal involvement; however, more recently a more benigh form "limited Wegener's granulomatosis" (1) has been described in which no renal involvement is seen.

Although the etiology of this disease is unknown, it is generally considered a hypersensitivity disorder because of the granulomata, vessel inflammation, glomerulonephritis and circulating or deposited immune complexes, which are often seen.

The mean age of presentation varies according to the study, from 43.6 years (2) to 46.9 years (3). The male to female ratio is 16 to 1 (2). Most patients initially presented with upper airway illnesses, including severe and persistent rhinorrhea, sinus drainage, mucosal ulcerations and pain (2) Pulmonary infiltrates shown on chest X-rays in 71% of patients at the time of the presentation led to diagnosis (2). Non-renal manifestations almost invariably precede functional renal disease. Only 11% of patients presented with functional renal disease initially (2).

The predominant organ system involved in Wegener's granulomatosis is the lungs (94%), manifested on X-ray as bilateral nodular infiltrates with a tendency to cavitate (2). The paranasal sinuses are involved in about 90% of patients (2), with a granulomatous vasculitis in most cases. Erosions of the sinus walls is