times a day. Within a few hours she noticed improvement. She was able to perform her usual activities and stated that she felt 80% to 90% better. She started a 5 day course of acetazolamide at the onset of each exacerbation.

While taking acetazolamide her speech was no different from the postmenstrual state. Opoculosus was absent. The myoclonic jerks were infrequent at rest and were markedly reduced in amplitude and incidence during intentional motion. Efficacy continued for five consecutive cycles without side effects. A double-blind placebo trial during the next menstrual period was aborted on the second day as she became bedridden with myoclonus. Several hours after starting acetazolamide she improved. The next menstrual cycle went smoothly with this medication. Valproic acid serum levels were unchanged with the addition of acetazolamide.

Catamenial exacerbation of action myoclonus has been described. The mechanism is not apparent. The patient previously described underwent bilateral oophorectomy and worsened. She improved with very high dose conjugated estrogen (Premarin 6-25 mg/day). Progesterone and acetazolamide were not administered. Declining progesterone levels prior to menstruation are known to increase seizure activity in some epileptics. Whether this plays a role in action myoclonus is uncertain.

Acetazolamide has failed to improve myoclonus in patients without catamenial exacerbations. However, it does elevate brain anticonvulsant concentrations, brain carbon dioxide levels and gamma amino butyric acid, all of which have antiepileptic effects. The impact on serotonin is unknown. The mechanism of acetazolamide in this patient is likely to be by direct effect on the central nervous system or tissue anticonvulsant concentrations. Antagonism of hormones or another systemic physiologic change is also possible.

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References


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Enophthalmos and metastatic carcinoma of the breast

Sir: Enophthalmos is an uncommon finding in neurological practice. It may be seen following orbital trauma, in patients with Horner’s syndrome, facial hemiatrophy and in a small proportion of patients with orbital metastases from carcinoma of the breast. The latter association is recognised in ophthalmological practice but has received little attention in the neurological literature. We describe a 66-year-old woman in whom enophthalmos was the presenting feature of a breast carcinoma.

A 66-year-old woman developed ptosis, enophthalmos, and diplopia 3 years before her present admission. At the same time the symptoms first appeared, computed tomography of the head and orbits revealed abnormal tissue behind the left eye. At another hospital a biopsy was taken from this area (via a left frontal approach) but the tissue obtained only revealed non specific changes and a definite diagnosis was not possible. Two years later she developed progressive weakness of both legs which appeared to respond to a course of methyl prednisolone. Over the months prior to her present admission she had become aware of blurred vision in the right eye. Vision in the left eye remained normal. Her general health was otherwise satisfactory and there was no significant past or family history. Examination revealed a left frontal craniotomy defect, the site of the previous orbital biopsy. Higher mental functions and speech were normal. She was able to stand only with support and to walk with the aid of a frame. The corrected visual acuities were VAR 6/18 and VAL 6/9. Colour vision was impaired in the right eye and there was a right relative afferent pupillary defect. The right optic disc was grossly swollen and there were haemorrhages and exudates. The left disc was normal. The right blind spot was markedly enlarged but the left visual field was full. There was left enophthalmos with narrowing of the palpebral fissure. Abduction and vertical movements of the left eye were restricted but a full range of ocular movement was present on the right. There was a mild facial diplegia. The remainder of the cranial nerves were normal. There was global wasting of the muscles of both legs. Upper limb strength was normal and reflexes were present. There was truncal weakness and moderate to severe global leg weakness particularly distally. Deep tendon reflexes were absent in the legs. The plantar responses were absent. Vibration was not perceived in the legs and the sense of joint position was
impaired in the toes. In the outer half of the right breast there was nodular mass measuring 2 cm in diameter with a nodule in the right axilla. Electromyography showed gross neurogenic changes in the leg muscles with a normal sural nerve conduction velocity consistent with a radiculopathy. Computed tomography of the orbits (fig) revealed abnormally enhancing tissue throughout the left orbit with retraction of the globe. This tissue extended back into the region of the cavernous sinus. On the right there was thickening of the optic nerve sheath, sclera and the distal half of the right medial rectus. Again the abnormal tissue extended intracranially and beyond the dorum sellae. The cerebrospinal fluid contained 12 white cells, protein 2.28 g/l, glucose 4.0 mmol/L (blood glucose 6.9 mmol/L), and a number of malignant cells with features consistent with adenocarcinoma. A breast biopsy revealed a lobular carcinoma.

At the time this patient with a carcinoma of the breast presented to us she had the clinical and radiological picture of enophthalmos associated with abnormal enhancing tissue obliterating the orbital space on the left and infiltrating the right optic nerve. The abnormal tissue extended intracranially. In addition she had the clinical and electromyographic features of a polyradiculopathy. The finding of malignant cells in the cerebrospinal fluid makes it probable that all these features were due to metastases from the breast carcinoma and it seems likely that the enophthalmos was the first manifestation of metastases from the breast carcinoma. Enophthalmos is recognised as occurring in a small number of patients with metastatic breast carcinoma of the scirrhus type. The mechanism responsible for enophthalmos in this situation is thought to be progressive fibrosis and shrinkage of the orbital contents, with retraction of the globe.

Although metastases from breast carcinoma are one of the most common secondary tumours to involve the orbit in females, it is unusual for this tumour to present with orbital metastases. Unexplained progressive enophthalmos should always raise the possibility of a breast neoplasm, even in males.

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We thank Professor W1 McDonald for permission to report this case.

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Pseudomonas oestis causing cranial nerve palsies

Sir: Invasive pseudomonal infection of the skull base is well documented in the otorharyngologic literature. Some neurological sequelae have been reported, but as far as we are aware these are not well known to neurologists. It is a serious disease carrying a high mortality if treatment is delayed. We report a patient who was referred to a neurological department with head pain and who subsequently developed multiple cranial nerve palsies owing to such a pseudomonal infection.

Four months before his admission to this hospital, a previously healthy 74-year-old man developed pain in his left ear associated with some discharge from that ear. He was treated with ampicillin and initially improved but three weeks later his symptoms recurred associated with left parietal headache. A swab taken from his left ear at this time grew Pseudomonas aeruginosa. He had a left mastoidectomy followed by a 3 day course of intravenous gentamicin and piperacillin. Biopsy from the mastoid cavity revealed granulation tissue only. He continued to experience severe and persisting pain in the left temporoparietal region and lost one stone in weight in the month prior to his admission. His urine at this stage was free from glucose. On admission to this department he was apyrexial, he had a healed left mastoidectomy scar and left-sided conductive deafness. Haemoglobin, white cell count, electrolytes, liver function tests, serum and urine electrophoresis, VDRL, radiographs of chest, skull and temporal bones, computed tomographic (CT) scan of the brain and skull base and cerebrospinal fluid were all normal. The erythrocyte sedimentation rate (ESR) was elevated at 90 mm/h and a random blood glucose was 18 mmol/L. Swabs taken from the left external auditory canal grew Pseudomonas aeruginosa; a Te bone scan revealed markedly increased uptake in the left temporal bone. Biopsy specimens of the post natal space revealed granulation tissue only.

Shortly thereafter, the patient developed a partial left VIth nerve palsy, followed a few days later by the involvement of the left VIIth, IXth, Xth and XIth cranial nerves, and a blocked left nostril from which there was a bloody discharge. Repeat plain radiographs of the skull base did not show any significant change; CT scans however showed a small area of low attenuation on the left side of the brain stem at the level of the ponto-medullary junction.

His diabetes was controlled initially with insulin and subsequently with diet alone. He was started on a prolonged course of intravenous azlocillin and netilmicin and dosage was adjusted to give optimal blood levels. His pain resolved within a few days and his nostril cleared. At the end of seven weeks of antibiotic treatment his cranial nerve palsies had improved and his ESR was 30 mm/h. Three months after the completion of treatment he had no pain or neurological deficit.

The persistent growth of pseudomonas from the ear, the bone scan appearances and his diabetic status led to the diagnosis of an invasive pseudomonal infection of the petrous bone and skull base. "Malignant" or "invasive" external otitis was first described by Chandler in 1968 and is consistently due to a pseudomonal infection. Almost all reported patients have been elderly men with long established diabetes who have a persistent otitis externa which spreads to involve the pinna, skull base, temporal bone and surrounding soft tissue.

The present patient demonstrates that the initial development and progress of petrous oestis may be insidious and may occur in a previously healthy individual without previous evidence of diabetes. In this patient, the pseudomonas oestis probably spread from the mastoid cavity even though a satisfactory mastoidectomy had been performed. The initial course of antibiotics, however, had been too short to eradicate residual infection.

The VIIth cranial nerve is the most commonly affected and may occur without evidence of bony destruction. Chandler suggested that the facial nerve is involved outside the temporal bone along its course.

Letters
Enophthalmos and metastatic carcinoma of the breast.

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