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 dorsum 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 endophthalmos 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 endophthalmos was the first manifestation of metastases from the breast carcinoma. Endophthalmos is recognised as occurring in a small number of patients with metastatic breast carcinoma of the scirrhous type. The mechanism responsible for endophthalmos 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 endophthalmos should always raise the possibility of a breast neoplasm, even in males.

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References


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Pseudomonas osteitis causing cranial nerve palsies

SIR: Invasive pseudomonal infection of the skull base is well documented in the otolaryngological literature.2 Some neurological sequelae have been reported,1,4 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 Tc99 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 VIIth nerve palsy, followed a few days later by the involvement of the left VIIth, IXth, Xth, XIth and XIIth 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 Chandler5 in 1968 and is consistently due to a pseudomonal infection. Almost all reported patients4 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 osteitis may be insidious and may occur in a previously healthy individual without previous evidence of diabetes. In this patient, the pseudomonas osteitis 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. Chandler4 suggested that the facial nerve is involved outside the temporal bone along its course.
in subtemporal soft tissue. According to Doroghazi et al, patients with multiple lower cranial nerve palsies usually have bone erosion around the jugular foramen which suggests that palsies of lower cranial nerves, apart from VII, are due to osteomyelitis of the skull base or an associated extradural abscess or petrosal sinus thrombosis.

According to Mendelson et al, Te99 and Ga67 scans are the most helpful radiological investigations in the diagnosis of invasive external otitis, computed tomography is useful for diagnosing and assessing soft-tissue extension, and Ga67 scans are best for following resolution of infection. The Te99 bone scan in our patient supported the possibility of osteomyelitis, the plain radiographs, tomograms and CT scans having shown no evidence of bone infection.

Patients with multiple lower cranial nerve palsies may require detailed investigation to establish a diagnosis. Pseudomonas infection of the petrous bone and skull base is a curable condition which should be considered particularly if pain is a prominent feature and if there is no evidence of recent otitis externa or mastoiditis.

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References

Nutritional amблиopia in a patient with Crohn’s disease
Sir: Bilateral visual failure associated with centro-caecal scotomata is a rare condition in which the aetiology is often unclear. Two hypotheses are that it is due to exogenous toxins, such as cyanide, or alternatively that it is due to nutritional deficiency. In the latter case many vitamins have been implicated particularly the B group. Many reports describe patients who have had a heavy intake of tobacco and alcohol and have elevated cyanide levels, but the evidence for vitamin deficiency relies largely on cases of malnutrition studied during the second world war in which cyanide levels were not measured and in which dietary deficiencies of substances other than vitamins may have been present.2 3 We report a case of bilateral visual failure associated with night blindness in which the most likely cause was a deficiency of vitamin A and B group vitamins. The vitamin lack was related to longstanding malabsorption due to a short bowel, following surgery for Crohn’s disease.

The patient was a 62-year-old widow who 3 years prior to admission suddenly noticed she could not find her way at night in the dark, particularly out of doors. In time she developed particular difficulty with poorly illuminated parts of the house, such as corners, cupboards and in lighting her oven. In the latter case she had to feel for the flame with her hands to be sure that she had lit the gas. She did not venture out of doors at night for fear of self injury. This complaint persisted unchanged to her admission. Approximately 6 months before admission she noticed the gradual onset of difficulty seeing objects during the day. This difficulty progressed over a 6 months period to the point where she could not see anything directly in front of her, such as what she was cooking in her pots, or notes and coins, nor could she read ordinary print. Her peripheral vision in daylight was normal. She had also noticed that she was unable to discern colours correctly and at times could not see colour at all.

She had a past history of Crohn’s disease diagnosed at laparotomy 9 years earlier when she presented with bowel obstruction. Ileal resection and ascending colectomy was performed at the time. She subsequently was treated with azathioprine and prednisolone. A colo-vaginal fistula healed spontaneously after several months. She remained on the azathioprine and prednisolone up to the time of her admission. Although the disease activity was quiescent, the patient continued to experience steatorrhoea for all that period of time. She passed 7 to 10 stools per day which were pale and greasy. Renal calculi were removed from both kidneys 3 years prior to admission following an episode of haematuria. An oligo-articular arthritis developed 2 years prior to admission and this was successfully treated with non-steroidal anti-inflammatory agents and local steroid injections. She smoked 15 to 20 cigarettes per day and drank alcohol occasionally. She was otherwise asymptomatic, in particular she gave no symptoms of a peripheral neuropathy, myelopathy or other nutritional deficiencies. Medications on admission were azathioprine 75 mg per day, prednisolone 10 mg per day, codeine phosphate 60 mg per day, flurazepam 30 mg per day and hydroxycobalamin 1000 μg monthly intramuscularly.

Ophthalmological examination on admission revealed a visual acuity of N24 and 6/60 in the right eye and N8 and 6/18 in the left for near and distant vision respectively. Colour vision was absent bilaterally; only the first of the Ishihara plates could be read, with difficulty. Visual fields performed by confrontation, Freidman chart and Bjerrum screen revealed bilateral centro-caecal scotomata for red but not for white. The pupils were large and poorly reacting to light. Funduscope examination was normal including the optic discs. The remainder of the neurological and general examination was also normal.

Abnormal results of investigations included the following: an iron deficiency anaemia with a haemoglobin of 9.2g/dl (normal: 12-16g/dl), serum iron of 5μmol/l (15-35μmol/l) and a total iron binding capacity of 74μmol/l (normal: 45-70μmol/l). The film showed burr cells, Howell Jolly bodies, basophilic stippling and occasional nucleated red blood cells. Serum carotene was 0.1μmol/l (normal: 0.9-5.6μmol/l), alkaline phosphatase of 381 iul/l (normal 100-280 iul/l), decreased gamma globulins on electrophoresis and prominent bone resorption on bone biopsy. Barium enema, small bowel enema and large bowel endoscopy revealed shortened ileum, resected ascending colon without blind loops and no active disease. The malabsorption was attributed to short bowel syndrome. Negative investigations included cranial CT scan, CSF examination including cytology, syphils serology, prothrombin time, folate, red blood cell folate, liver function tests including gamma GT, electrolytes, renal function, calcium, phosphate, magnesium, urinary calcium, urinary urate, urinary oxalate, urine microscopy and culture, intravenous pyelogram and skeletal survey for osteomalacia.
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