Recurrent Meningioma Presenting as a Parotid Swelling

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Mr W H, aged 56

History: The patient noticed a prominent right eye in 1950. Two years later, a meningioma of the outer third of the right sphenoidal ridge was excised. Portions invading the temporalis muscle, cavernous sinus and right orbit were left behind, and because of this radiotherapy was given. He remained well, but in 1953 noticed a pea-sized swelling in the right cheek. In November 1968 he had a grand mal attack, followed by a craniotomy and excision of recurrent intracranial meningioma.

March 1970: He was seen with a swelling in the anterior part of the right parotid region and cheek which had gradually increased in size and was now 8 cm in diameter.

Investigations: Skull X-rays and tomograms demonstrated a right temporal decompression. The roof and lateral wall of the right orbit had also been partially removed. There was irregular bone destruction in the roof and lateral wall of the right maxillary antrum extending into the adjacent lateral orbital margin, and a soft-tissue mass in the lateral half of the antrum.

External carotid angiography showed abnormal circulation from the maxillary artery in the right antrum, and the lower and outer orbit, extending into the cheek. Internal carotid angiography demonstrated abnormal circulation in the orbit from the ophthalmic artery; this area of abnormal circulation was in continuity with the area supplied by the external carotid artery.

Operation (25.7.70): The tumour was excised from the right maxillary antrum and cheek through a standard right maxillectomy incision, after preliminary ligation of the right external carotid artery. A large soft encapsulated tumour was found in the soft tissues of the cheek, continuous through a defect in the anterolateral wall of the antrum with the soft-tissue mass within the antrum. It also extended beneath the zygomatic arch to the temporal fossa in dumb-bell fashion, but the whole tumour mass was easily dissected away. There was no apparent intracranial extension through the posterior wall of the maxillary antrum. The resulting cavity was packed and the skin closed, the pack being removed six days later. The patient made an uneventful recovery and has remained well.

Histology: Sections showed typical transitional meningioma with cellular whorls, but in one area the tumour was infiltrating muscle.

Discussion

Forty cases of intracranial meningioma with extracranial dissemination have been published in the literature, and the subject has been recently reviewed by Shuangshoti *et al.* (1970), who described one of the cases. The commonest mode of spread is by the venous system to the lungs, liver, pleura and lymph nodes. The suggestion that previous bony decompression might aid dissemination of the tumour (Russell & Rubinstein 1959) may be applicable to the local spread in this case. The present patient had had a craniotomy in the past, but is unique in that the tumour had spread locally into the maxillary antrum and cheek. The angiographic demonstration that at least some of the blood supply of the tumour was derived from the internal carotid system suggested a spread from within the cranium, although there was no direct evidence at operation that the tumour had emerged directly from the cranial cavity.

The radiological details of this case will be described elsewhere by Dr Brian Kendall, who was responsible for the radiological investigations.

REFERENCES
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Sarcoidosis and Amyloidosis

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Mr J B, aged 30. Machine operator

History: Pneumonia in 1961. 1962: basal bronchiectasis diagnosed at mass X-ray unit. 1969: presented with bilateral loin pain and pyrexia.

On examination: Anæmic. Coarse basal crepitations. Not œdematous or hypertensive. Hepatosplenomegaly. Heavy proteinuria. Urine microscopy: a few red and white blood cells; no casts.

Investigations: Hb 10 g/100 ml. Marrow iron deficient. No excess of plasma cells. No evidence of hypersplenism. Blood urea 84 mg/100 ml. Creatinine clearance 15 ml/min. Serum proteins: albumin 2 g/100 ml; raised IgG and IgA globulins. Proteinuria: 10–20 g/24 h; no Bence-Jones proteinuria. Maximum serum calcium 6·2 mN

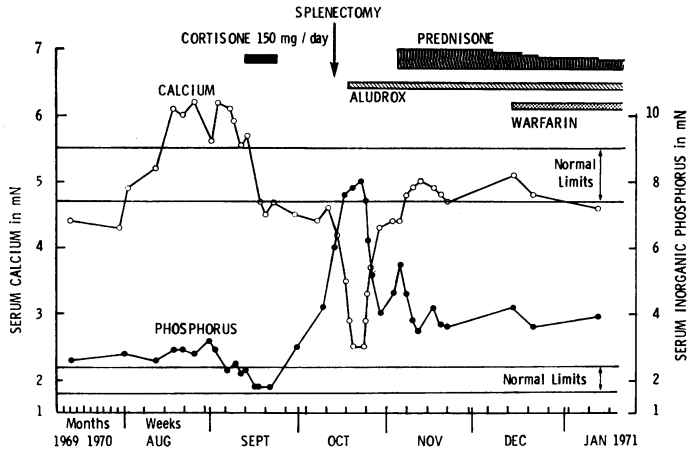

Fig 1 *Calcium metabolism chart*

(12·4 mg/100 ml), suppressed by cortisone acetate 150 mg/day for 10 days (Fig 1). IVP: large kidneys (14·5 and 14·8 cm) with calyceal distortion and cysts. Alkaline phosphatase 40 K-A units. 5-nucleotidase 73 i.u. Liver biopsy: granulomata around portal tracts; extensive amyloid deposits. Kveim test positive. No AFB in urine, sputum, liver biopsy or marrow. Mantoux test (1:100), toxoplasma dye test, brucella antibody test – all negative. Chest X-ray: basal bronchiectasis. No hilar lymphadenopathy. Sputum infected with pseudomonas or proteus.

Treatment: A provisional diagnosis of bronchiectasis with secondary hepatic and renal amyloidosis was made. He was thought also to have sarcoidosis, but a reticulosis could have explained his hepatosplenomegaly, amyloidosis and hypercalcæmia. A diagnostic laparotomy and splenectomy were performed.

A large sarcoid spleen was found with deposits in lymph nodes and liver. No evidence of a lymphoma. A renal biopsy showed extensive amyloid but no sarcoid deposits. He has received anticoagulant therapy because of the risk of renal vein thrombosis in patients with renal amyloid.

Post-operatively, a reversible period of renal failure was treated with aluminium hydroxide gel to help reduce his serum phosphorus. Subsequently, a steroid trial produced no improvement in renal function but prevented any further hypercalcæmia (Fig 1).

Comment

The absence of any association between sarcoidosis and amyloidosis has been emphasized by H A Azar (1968, *Pathol. Ann.* **3**, 105). It is possible that some immunological abnormality links the two diseases, but a coincidental association seems an equally valid explanation.

Professor J Hobbs (*Westminster Hospital, London SW1*) said that amyloid appeared to be derived largely from the variable portions of the light chains (V_L) of immunoglobulin molecules. It seemed that primitive (e.g. myeloma) plasma cells were more likely to produce V_L fragments, especially those cells producing Bence-Jones proteins. Since sarcoidosis itself seemed to represent some imbalance between humoral and cellular immune mechanisms, it was conceivable that some of this patient's plasma cells had gone astray and had produced V_L amyloid precursors. He had seen sarcoid-like lesions together with amyloid in only one other patient, who had also had hypogammaglobulinæmia. In that case the sarcoid had shown as microgranulomata, all of much the same size, differing from the present case.

Whereas cytotoxic treatment might prevent the formation of new deranged plasma cells producing V_L fragments, i.e. might arrest further deposition of amyloid, he was not convinced that amyloid could regress to an extent visible in repeated biopsies. Corticosteroid treatment was known to abrogate the marginal cuff of new potential antibody-forming cells surrounding germinal centres, i.e. could prevent a primary antibody response. This very action allowed the marginal area to be further populated from the underlying secondary response cells in existing germinal centres, i.e. prednisone alone would encourage long-standing amyloid-precursor-forming cells to multiply and produce V_L fragments. This hypothesis was in accord with the observed worsening of amyloidosis on prednisone alone. Prednisone combined with a cytotoxic drug (e.g. cyclophosphamide) might encourage precursor cells to divide and thereby become more susceptible to the cytotoxic drug. Since cytotoxic drugs carried a mutagenic risk, they should only be used in patients who would otherwise die. If one waited until the renal disease advanced to a stage where the proteinuria lessened due to the impairment of glomerular filtration but not until the patient was too uræmic, cytotoxic treatment could then arrest further amyloid for 1–3 years or more, during which time the patient might be quite comfortable, without a full-blown nephrotic syndrome.