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NEUROLOGICAL PICTURE

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Fast-growing adenoid cystic carcinoma: a neurotropic malignancy

An 81-year-old man presented to the neurology department with a 2-month history of diplopia and recent onset of left-sided ptosis. He had also had pain in the left eye and left nasal congestion for the past month.

Two years ago, he had presented to a different department with a 6-month history of left upper lip and cheek paraesthesia, which was still present. At that time, a small (1×0.5 cm) mobile lump was found in the left upper lip and excised. Histological examination confirmed a benign muco-coele retained in the salivary gland. This was insufficient to explain the continuing symptoms of paraesthesia and therefore the brain and orbits were subjected to magnetic resonance imaging (MRI). No abnormality was found (fig 1).

On presentation to the neurology department, fullness of the left orbit and left ptosis and restriction of left eye movements in all directions were observed. In addition, pinprick sensation was reduced in the maxillary division of the left trigeminal nerve. An urgent MRI scan was repeated 12 months after the first (fig 2A,B). This showed an advanced soft-tissue mass filling the left maxillary sinus, with tumour margins beyond the medial and lateral borders and extension from the hard palate to the cavernous sinus. Biopsy results showed proliferating epithelial cells with "honeycomb" spaces, consistent with adenoid cystic carcinoma.

Adenoid cystic carcinoma is a rare malignant neoplasm arising from the epithelial cells of secretory glands, most commonly the salivary glands.¹ This is of interest to neurologists, as contiguous perineural invasion is a hallmark of adenoid cystic carcinoma and partly explains its tendency to spread. It surrounds and invades nerve sheaths, causing cranial or peripheral neuropathy, a feature possibly mediated via expression of surface cell adhesion molecules.² Presentation is late as growth is often slow, but it can occasionally be rapid. It was so in this case, potentially with an indolent initial phase that was not apparent on MRI. Treatment focuses on surgical excision with concomitant radiotherapy, but mortality is high from multiple tumour recurrence and late distant metastases.

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Figure 1 T2 axial scan showing minor inflammatory changes in the left antrum (arrow).

Figure 2 T1 axial (A) precontrast and (B) postcontrast scans, showing mixed-signal mass in and extending beyond the left antrum (arrow).

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