Rare disease

A dry, droopy and dilated eye: a neuro-ophthalmic case with correlation of signs and anatomical tumour spread

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Summary

Adenoid cystic carcinoma (ACC) has a propensity for perineural tumour spread. This report concerns a 51-year-old woman who presented with a unilateral red eye with reduced vision, ptosis and a non-reactive pupil. This patient had a maxillary sinus ACC with perineural spread, leading to the neuro-ophthalmic signs. She is currently being treated with radiotherapy.

BACKGROUND

This case is an unusual presentation of a relatively uncommon disease. The findings were not consistent with one anatomical location for a lesion, but after imaging and biopsy it became clear that one disease was causing multiple nerve disruptions.

CASE PRESENTATION

A 51-year-old woman presented with a 2-week history of blurred vision and redness in the left eye. Her general practitioner noticed that she had a fixed dilated pupil with partial left ptosis. She had a background history of trigeminal neuralgia. An urgent referral to neurology was made.

Neurology organised an urgent CT scan for a suspected pupil involving partial third nerve palsy. The CT angiogram was reported as normal and she was referred for ophthalmological assessment.

The patient gave a history of gradual deterioration and redness but no irritation of her left eye over several weeks. She had been unable to cry on the left side for several weeks and the left side of her face did not turn red or sweat during exertion. She had left visual acuity of 4/12, pin-holing to 4/6. She had normal colour vision and extraocular muscles. She had 2 mm of left ptosis with a larger left pupil that was non-reactive to light. Her cornea and the rest of the fifth cranial nerve had reduced sensation. She had significant superficial punctuate erosion on the cornea (see figure 1A and

B). At this point it appeared that she had a Horner's syndrome but with a dilated pupil as well as altered lacrimation and trigeminal nerve dysfunction.

INVESTIGATIONS

A CT of the head and angiogram was initially reported as normal.

An MRI showed an enhancing mass involving the left cavernous sinus, the pterygopalatine and sphenopalatine fossae, the trigeminal nerve, and the facial nerve to the genu. This was reported as a possible schwannoma. There was also a mass at the left maxillary sinus (see figure 2 A, B and C).

A biopsy of the maxillary mass showed adenoid cystic carcinoma (ACC).

DIFFERENTIAL DIAGNOSIS

- ▶ Schwannoma or meningioma
- ▶ Perineural spread of a squamous cell carcinoma.

TREATMENT

The patient is currently undergoing radiotherapy.

OUTCOME AND FOLLOW-UP

Unfortunately, the patient has had problems with neurotrophic ulcers, although this had settled on regular lubrication ointments at her last ophthalmology follow-up visit.

Figure 1 (A) Photograph of the patient showing anisocoria and ptosis. (B) Photograph of the patient showing corneal superficial punctuate erosions.

Figure 2 (A) Transverse MRI showing the tumour in the pterygopalatine fossa. (B) Transverse MRI showing the tumour in the trigeminal nerve and pressing on the carotid artery. (C) Transverse MRI showing the tumour in the infraorbital nerve.

DISCUSSION

ACC most commonly arises in the salivary glands including the parotid, submandibular, lacrimal and minor salivary glands. It has also been found in prostate, breast, bronchial mucosa, facial sinuses, intestinal and genital tracts and skin.^{1–4} It has three growth patterns: cribriform, tubular and solid. Predominant solid tumours are associated with the highest metastases and perineural spread.¹

ACC is a malignant tumour involving the exocrine glands.

It has a slow growth rate, with frequent perineural and metastatic spread. ACC perineural tumour spread from branches of the trigeminal nerve to invade the cavernous sinus is commonly reported.^{2 5–7}

ACC often has non-specific initial symptoms including non-specific facial pain and non-specific headache. Perineural spread along the trigeminal nerve may be misdiagnosed as trigeminal neuralgia.² Presentation of a partial cavernous sinus syndrome or an orbital apex syndrome is common.^{2 8} Later symptoms include diplopia, epistaxis, sinusitis, Bell's palsy, lacrimal gland swelling and proptosis.^{1 2 9} Presentation with Horner's syndrome is uncommon.¹ An Adie tonic

pupil followed by an orbital apex syndrome has been reported.^{1 2 10 11} The time between the onset of symptoms and diagnosis ranges from 6 months to 6 years.^{2 8}

Radical surgical resection followed by radiotherapy is considered the best treatment for ACC, but surgical margins often involve critical structures due to perineural tumour spread.² Therefore, incomplete resection is common with local recurrence within 3 years of the initial treatment.² In patients with complete resection of the tumour, the presence of distant metastases is present in 39% of cases.²

ACC is radiosensitive so tumour size and symptoms can be reduced, but it is not radiocurable as 93% of patients have recurrence within 5 years of radiotherapy.²

As in our case, ACC may mimic schwannoma or meningioma, MRIs were misinterpreted in 31.2% of cases.^{2 12}

Features associated with increased sensitivity and specificity of detection of perineural spread are:

- Increase in the nerve size or replacement of perineural fat with tumour
- Widening or excessive enhancement within the pterygopalatine fossa

- ▶ vidian canal
- ▶ foramen rotundum
- ▶ ovale
- ▶ Expansion or abnormal enhancement of the cavernous sinus.²

In this patient, the trigeminal neuralgia is explained by tumour spread from the maxillary antrum along the maxillary nerve to the pterygopalatine fossa and trigeminal ganglion.^{7 13}

The facial flushing can be accounted for by tumour infiltration of the postganglionic sympathetic fibres that supply the facial sweat glands and travel via the trigeminal nerve branches.^{14 15} The cavernous plexus supplies branches to the third nerve via the ophthalmic artery which supply the Muller's superior palpebral muscles.¹⁴

The ptosis can be explained by the tumour located where the internal carotid plexus forms lateral to the artery or by retrograde tumour spread from the pterygopalatine ganglion.¹⁴ The dilated pupil could be due to interruption of the pre or post-ganglionic parasympathetic fibres at the ciliary ganglion¹⁰ due to tumour spread from the pterygopalatine ganglion via the zygomaticotemporal nerve to the orbit¹⁴ or be damaged at the cavernous sinus.¹⁵ The lack of lacrimation can be explained by interruption of the parasympathetic supply to the lacrimal gland at the pterygopalatine ganglion, the deep petrosal nerve at the foramen lacerum or the vidian nerve in the vidian canal.^{14 15}

This case describes ophthalmic signs and symptoms resulting from perineural spread of a rare tumour. It shows how history and examination findings can be closely related to anatomical pathology.

Learning points

- ▶ Adenoid cystic carcinoma often presents with a non-specific insidious onset with slow growth.
- ▶ Perineural tumour spread can be missed on imaging.
- ▶ Perineural tumour infiltration should be a differential diagnosis for unusual neurological presentations.
- ▶ Examination findings can be closely related to anatomical pathology.

Competing interests None.

Patient consent Obtained.

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