CASE REPORT

Surgical resection of invasive adenoid cystic carcinoma of the lacrimal gland and wound closure using a vertical rectus abdominis myocutaneous free flap

João Paulo Andrade,¹ Sergio Figueiredo,¹ Julio Matias,² Ana Catarina Almeida³

¹Department of Neurosurgery, CHLO, Lisboa, Portugal

²Department of Plastic Surgery, CHLO, Lisboa, Portugal

³Department of Ophthalmology, Hospital Egas Moniz, Lisbon, Portugal

Correspondence to
Dr João Paulo Andrade,
joapaaa@gmail.com

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SUMMARY

A 64-year-old man presented with a 3-month history of recurrent conjunctivitis. He was evaluated by an ophthalmologist and submitted to a CT scan that revealed an intraconic mass with invasion of the lateral orbital wall. He was operated, the mass was completely removed (with preservation of the intraorbital structures) and the lateral orbital wall rebuilt. The histopathological analysis revealed an adenoid cystic carcinoma of the lacrimal gland. 4 months later a painful recurrence of the lesion was diagnosed with invasion of the orbital roof and eyelids. After a multidisciplinary discussion and request from the patient, an exenteration of the orbit and removal of the lateral and superior orbital wall and dura mater was performed with the objective of a total resection. The wound and orbit were closed with a vertical rectus abdominis myocutaneous free flap to ensure closure.

BACKGROUND

We believe this is an important case to report because of the challenge in choosing the best therapeutic strategy taking the patient's wishes into account. We also wanted to point out the need and importance of multidisciplinary teams for the best treatment of patients with diseases affecting multiple organs, not only in preoperative meetings and also in the operating room.

CASE PRESENTATION

A 64-year-old man presented with recurrent episodes of tearing and conjunctival hyperaemia for

3 months that were interpreted as conjunctivitis. He had a history of pulmonary tuberculosis and gastro-oesophageal reflux which was controlled by a proton-pump inhibitor. He was a non-smoker with no family history of cancer. Two months later he reported of periorbital swelling and xerophthalmia. At that time the neurological examination revealed a limitation in the elevation of the right eyelid, limitation in the upward and lateral gaze of the right eye and double vision in those positions. A CT scan and the MRI were performed and an intraorbital lesion was diagnosed that invaded the lateral orbital wall and slightly compressed the optic nerve (figure 1).

The case was evaluated in the neurosurgical and ophthalmology departments and we decided to remove the lesion trying to preserve the optic nerve. A lateral orbitectomy was performed with 'en-bloc' removal of the lesion and external orbital wall. After removal, the lateral and part of the superior orbital wall were reconstructed with titanium mesh and methyl methacrylate (figure 2). The patient was discharged 5 days postoperatively. On the first follow-up consultation the patient had an improvement in eye movements and could see clearly with his right eye. A slight limitation in the

Figure 1 MRI showing the optic nerve compression by the tumour.

Figure 2 CT scan image of the lateral orbital wall reconstruction.

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Figure 3 Intraorbital recurrence of the tumour.

Figure 4 Histology slide with the characteristic small bland myoepithelial cells with scant cytoplasm and dark compact angular nuclei surrounded by pseudoglandular spaces (red arrow).

elevation of the eyelid persisted. The histopathology revealed an adenoid cystic carcinoma of the lacrimal gland (figure 3) and he started radiation therapy.

Four months later he started to feel pain in the right eye and eyelids. On inspection the skin of the eyelid had a thick and irregular texture.

A CT scan was performed and showed a recurrence of the tumour in the orbital apex and infiltration of the eyelids and part of the superior orbital wall (figure 4). The case was again evaluated by the radiation therapists, oncologists, neurosurgeons, ophthalmologists and plastic surgeons and, since it implies a better prognosis, a radical removal of the orbit and its superior wall was proposed. The patient also wanted to have the surgery if it meant prolonging his life.

TREATMENT

Before the surgery the infiltrated skin was marked and the incision was performed with a 10 mm margin (figures 5 and 6). An enucleation was performed and after that an exenteration.

Figure 5 Preoperative marking of the facial skin incision.

Figure 6 Preoperative marking of the abdominal incision.

The infiltrated bone on the orbital roof was also removed as well as the dura mater of the anterior skull base (figure 7). All the bone was cauterised (figure 8). A duroplasty was performed. A vertical rectus abdominis myocutaneous free flap was prepared. After the removal of the orbit and tumour the flap was placed over the wound, the facial vessels were dissected and a microvascular anastomosis was performed. The skin was closed with running nylon stitches (figure 9).

OUTCOME AND FOLLOW-UP

During the postoperative period the patient had no symptoms. The wound showed good cicatrisation and vascularisation. There were no signs of cerebrospinal fluid (CSF) fistula.

On the 13th day postoperative the patient was discharged from hospital. He started radiation therapy and so far no recurrence of the tumour was noted.

DISCUSSION

Lacrimal gland tumours are rare with a reported incidence of one per 1 000 000.¹ Adenoid cystic carcinoma of the lacrimal gland is the most common malignant tumour of the lacrimal gland (~20–30%).^{2 3} There is no gender preference described in the literature and most cases present at around 50 years.^{1 2} The age of diagnosis has also been described as an important factor for disease-specific survival and overall survival.²

Malignant tumours of the lacrimal gland present more often with orbital and periorbital pain, lacrimation and visual

Figure 7 Exenteration of the orbit.

Figure 8 Orbital cavity after cauterisation.

Figure 9 Skin defect covered with the flap.

changes. In this case the most common symptoms were not the first which may have delayed the diagnosis.¹

As described in the literature the initial lesion appeared round on CT and appeared to invade the lateral orbital wall which is according to the characteristics found in other records.^{1 3}

Initially, the therapeutic approach was focused on a complete removal of the tumour with preservation of the orbital structures as described in some case reports.^{2 4} However, a T3 or T4 classification has been associated with worse prognosis⁵ and once a

recurrence was noted with bony and skin invasion the orbital structures as well as the bone and surrounding tissues had to be excised in order to improve the prognosis and disease-free survival.⁶

At this point the decision to perform an exenteration was difficult but was readily approved by the patient once the prognosis^{7 8} and low tendency to metastasise⁹ were explained.

Before the second surgery, by predicting the large skin and bony defect after surgery, a decision was made to close the opening^{10–14} with a free tissue flap in order to prevent post-operative complications such as possible CSF fistula. This approach was also described by one centre.¹⁵

After surgery the patient underwent radiotherapy and there has been no recurrence.¹⁶

This was a difficult case to solve and it was only possible to do so with a multidisciplinary approach^{17 18} and special care to the patient expectations.

However, due to the small number of cases in each centre there is the need for more studies (namely multicentre) to better understand the behaviour of these tumours and how to treat them.

Learning points

- ▶ Lacrimal gland tumours are very rare and the adenoid cystic subtype is the more aggressive one.
- ▶ The therapeutic approach should be decided by multidisciplinary teams to predict and prevent complications and morbidity.
- ▶ In such cases the patients' opinion is very important when it comes to decision-making.

Competing interests None declared.

Patient consent Obtained.

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