CASE REPORT

Sight preserving orbital decompression: a novel multidisciplinary approach to managing severe proptosis in neurofibromatosis type 2

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SUMMARY

We describe the importance of collaboration between multiple surgical specialties in managing a complex case of sight-threatening severe proptosis in a young woman with type 2 neurofibromatosis (NF2) complicated by pre-existing contralateral blindness. Trans-nasal and lateral orbital surgical approaches were aided by stereotactic navigation to debulk a large frontal/sphenoid wing meningioma, which had been exerting pressure onto the right globe and optic nerve. The patient made an excellent postoperative recovery along with preserved residual visual acuity, normal neurology and a good aesthetic outcome.

BACKGROUND

Neurofibromatosis type 2 (NF2) is a rare and disabling condition with a poor prognosis. Despite its benign histological nature, this and other, intracranial tumours can grow and exert a pressure effect onto important surrounding anatomical structures leading to cortical blindness, hearing loss, increased intracranial pressures and even death. Vital anatomical structures, contained within the cranial vault, can often make surgical resection of these tumours complex and challenging.

This case report demonstrates an example of a young woman with NF2 receiving successful sight-preserving orbital decompression surgery, via a coordinated multidisciplinary team approach. Her successful surgery highlights that three wall orbital decompression can be achieved trans-nasally and trans-orbitally, without the need to perform a craniotomy, substantially reducing surgical risk, operating time and time to recovery.

Figure 1 Clinical photograph taken 3 months prior to surgery showing right-sided proptosis and chemosis.

Figure 2 Clinical photograph taken 3 months prior to surgery showing right-sided proptosis and chemosis. The patient was unable to obtain full closure of her right eyelids at this stage.

CASE PRESENTATION

A 24-year-old Greek woman presented with severe proptosis and chemosis of her right eye from a rapidly expanding frontal/sphenoid wing meningioma (figures 1 and 2). Her management was transferred from Greece to a London hospital in an effort to reduce her severe proptosis and preserve visual acuity. She had pre-existing contralateral blindness, with only perception of light in the affected eye. She had an 11-month history of worsening frontal headaches and decreased visual acuity in the affected right eye. MRI identified a frontal meningioma in July 2016 measuring 38 mm by 27 mm.

At the time of presentation in London in May 2017, this had considerably increased in size with noticeably enlarged proptosis and chemosis. On examination, she demonstrated right-sided hearing loss and cortical blindness of the left eye. Visual acuity was perception of light in the affected, proptotic right eye.

Her medical history is extensive. She has suffered multiple sequelae of NF2 including bilateral vestibular schwannomas causing unilateral right-sided sensorineural hearing loss, multiple cranial and spinal meningiomas and previous facial reconstructive surgery. She has undergone numerous courses of radiotherapy, targeting intracranial meningiomas and a tonsillectomy and tympanotomy for otitis media with effusion. She also suffered a mandibular rhabdomyosarcoma at 4 months of age. This was successfully treated and has remained in remission following a course of chemotherapy.

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Figure 3 Anterior perioperative view showing severe right-sided chemosis and proptosis. A temporary tarsorrhaphy had been carried out at this stage.

INVESTIGATIONS

In 2009, CT and MRI scans in Greece revealed multiple intracranial and spinal lesions. Her diagnosis of NF2 was later confirmed via multiplex ligation probe amplification genetic testing. CT and MRI imaging scans were performed again, prior to surgery, to assess the size and nature of her intracranial lesions.

TREATMENT

Considerable planning and communication took place to organise surgery and ensure all required specialities were present. Multiple specialities were called on to tackle the complexity of this case. Oculoplastic, ENT, craniofacial and neurosurgical specialists played equal parts in the delivery of care.

A four-stage surgical procedure lasting 7 hours was carried out as follows:

1. Oculoplastic surgeons decompressed the chemosis, and conjunctival flaps were raised and sutured temporarily as corneal protection (figure 3 and 4).
2. ENT surgeons endoscopically debulked the medial aspect of the right orbit via a trans-nasal approach using a Medtronic Fusion ENT surgical navigational system (product no 9733560), linked to the patients' recent CT scan. There was evidence of previous partial right medial orbital wall decompression, which obscured the surgical field on the right hand side. This necessitated a left-sided endoscopic approach with partial removal of the septum superiorly to provide direct access to the right orbital apex. Decompression of the orbital apex took place and was uncomplicated.
3. Craniofacial surgeons used a bicoronal incision with preservation of a pericranial flap for access to the superior and lateral aspects of the right orbit. The temporalis

Figure 4 Lateral perioperative view showing the extent and progression of chemosis and proptosis over figures 1 and 2, taken only 3 months previously.

Figure 5 Preoperative CT imaging demonstrating extensive right-sided proptosis.

remnants were turned down, and bone drilled away from the right temporal fossa and lateral orbital wall. The orbital rim was also removed on this side to permit further decompression of the globe.

Neurosurgeons later accessed the orbital apex, and removed bone from the lateral aspect of the right orbital canal. The tumour was debulked from the roof of the orbit, with no cranial fenestration. This created a significantly larger orbital volume and reduction of the globe proptosis. The conjunctival sutures were removed, and temporary tarsorrhaphies performed as a means of protection (figure 5) (figure 6).

4. Frontal bossing of pathologically affected outer cortical bone was recontoured and multiple specimens were sent for tissue analysis.

OUTCOME AND FOLLOW-UP

Postoperatively, the patient remained intubated for 72 hours and was transferred to the ITU. She was weaned from the ventilator, and made an uncomplicated recovery.

Within 4 days she was mobile, eating and drinking and reporting no discomfort.

Tarsorrhaphy sutures remained in situ for 12 days and the patient was discharged back to Greece 13 days following surgery. On removal of the tarsorrhaphy sutures, the globe was lying

Figure 6 Postoperative CT imaging showing a reduction in excess right orbital soft tissue and removal of bone from the lateral orbital wall.

Figure 7 Anterior view 12 days postoperatively showing markedly reduced right-sided proptosis and chemosis.

passively within the orbit, with minimal effort required for eye closure. Visual acuity was as preoperatively and will continue to be monitored closely for improvement (figure 7) (figure 8).

She will be followed up in a joint craniofacial neurofibromatosis clinic in London 6 weeks postoperatively.

Figure 8 Lateral view 12 days postoperatively showing markedly reduced right-sided proptosis and chemosis.

Unfortunately due to the nature of her disease, overall prognosis remains poor.

DISCUSSION

NF2 is an autosomal dominant disease with a birth incidence of approximately 1 in 33 000.¹ A tumour suppressor gene found on chromosome 22q12 undergoes inactivating mutations leading to the development of tumours within the central nervous system.² Intracranial vestibular schwannomas and meningiomas form the main traits of this condition.³

Lesions involving the orbit may require sight preserving orbital decompression surgery and this can be achieved through a variety of different approaches. Endoscopic trans-nasal decompression is considered a safe and effective method of inferior aspect sphenoorbital meningiomas.⁴ However for this case, due to the size and position of the right frontal meningioma a trans-nasal resection would have achieved insufficient orbital decompression.

To our knowledge, this is the first recorded case of three-walled orbital decompression achieved via simultaneous, trans-nasal and lateral orbital approaches. By avoiding performing a craniotomy, surgical risks such as bleeding, CSF leaks and meningitis were reduced.

Reich *et al* states there is no current specific gold standard for endoscopic approaches to orbital decompression. Despite this, a majority of surveyed American Society of Ophthalmic Plastic and Reconstructive Surgery members prefer combined approaches to single wall approaches.⁵

Patient's perspective

As for the feedback we are very pleased of the communication prior, during and after the surgery with the respective teams. The doctors were very careful and showed the utmost interest in the case. We were constantly being informed about her situation and the process of her recovery. The nursing staff executed perfectly the treating process and were very careful and helpful as well as friendly.

We are very pleased and want to thank the ward for providing a single room since we had communications problems because of lack of vision and hearing.

Furthermore, since we came back to Greece, whenever it was needed to communicate with the doctors back in England the responses were instant and very helpful.

Last but not least we are grateful to everyone for ensuring she was not in pain during the entire time in the hospital as well as when we left by providing us with the proper medication.

(Brother and translator for the patient)

Learning points

- ▶ A large degree of orbital decompression can be achieved via trans-nasal and lateral orbital surgical approaches, reducing the need to perform a craniotomy and its associated risks.
- ▶ Vital to the success of this complex procedure is the equal input from multiple specialties. Oculoplastic, ENT, craniofacial and neurosurgical specialties all provided valuable information and skills in the delivery of care to this patient.
- ▶ Surgical navigational systems, like the one used in this case, can prove a valuable tool for accessing deep orbital tumours, trans-nasally.

Contributors GHL gathered the information, patient consent and drafted all sections of the report. SJE helped write the treatment section especially regarding craniofacial surgical input and follow-up. JH edited and helped to write the introduction, summary and background sections. NO edited and helped write the ENT surgical management and discussion sections.

Competing interests None declared.

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