CASE REPORT

Odontogenic myxoma involving the orbit in a 3-year-old boy: removal, reconstruction and review of the literature

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SUMMARY

We present a rare case of a 3-year-old boy with an odontogenic myxoma (OM) involving the orbita. Including our case, only nine cases of OM have been reported to involve the eye in children.

There is no gold standard for treatment of OM in children with orbital involvement. The recurrence rate of OM in children seems low, which advocates for less invasive surgery. A gentle resection of the OM was carried out. The floor and medial wall of the orbit was reconstructed immediately using a non-resorbable Medpor implant with passive adaptation. Reconstruction with a Medpor implant in children has rarely been reported in the literature. No clinical or radiological recurrence was observed 24 months after surgical removal, and the patient presented with symmetric appearance and normal vision.

BACKGROUND

Little is known about removal of odontogenic myxomas in relation to the orbit in children and nothing about reconstruction. Since this case is one of only nine reported in the literature, it is important. The reconstruction method is rarely reported in the literature.

CASE PRESENTATION

Introduction

Odontogenic myxoma (OM) of the jaw is the fourth most common odontogenic tumour and represents 3–6% of all odontogenic tumours.¹ It is a benign neoplasm found in the jaw and originates from pulpal ectomesenchymal cells.

The occurrence of OM in children is uncommon and rarely involves the orbit. This case report presents a 3-year-old boy with an invasive OM involving the floor of the orbit and lateral part of the nose and is seen in relation to a developing tooth. Furthermore, a review of the literature regarding OM and the orbit is presented.

Case

A 3-year-old boy was treated in collaboration between the Department of Ear, Nose and Throat Surgery and the Department of Oral and Maxillofacial Surgery, Odense University Hospital, Denmark.

Clinical examination revealed a swelling located in the anterior maxilla, involving the inferior border of the right orbit and the nasolabial fold (figure 1). The swelling was asymptomatic and had

Figure 1 The patient showing a swelling at the right nasolabial fold.

been present for approximately 7 weeks. The patient presented with normal vision, free eye movements and normal dental occlusion. Apart from the swelling, the patient was in good general health.

CT and MRI scans were performed (figures 2 and figure 3). The X-ray images showed a multilocular, 'soap-bubble' lesion separated by bony septa with poorly defined borders (figure 4).¹

Figure 2 MRI of the tumour in relation to the nasal cavity and sinus maxillaris, with signs of invasive growth.

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Figure 3 CT image showing the close relation to developing tooth 13 and the floor of the orbita.

Initially, an incisional biopsy was performed under general anaesthesia. The histological analysis showed loose myxoid stroma with few collagen fibrils, bony trabeculae and reactive

Figure 4 CT image showing the tumour with a multilocular, 'soap-bubble' appearance.

Figure 5 Clinical picture of the tumour after removal.

bone formation. In cancellous bone, the tumour showed signs of being locally invasive. The diagnosis of OM was based on the clinical, radiological and histological findings.

Treatment consisted of resection of the tumour under general anaesthesia. The tumour was approached through an intraoral mucosal incision and a transconjunctival incision. Peroperatively, the tumour showed signs of local invasiveness by infiltrating the surrounding bone, including the floor of the orbit. Furthermore, the tumour was in direct relation to developing tooth 13. The tumour was enucleated ([figure 5](#)), tooth 13 removed, and a gentle resection of 2 mm of the surrounding bone was performed. Postoperatively, the patient had a symmetrical appearance ([figure 6](#)).

The microscopic findings showed features similar to those seen in the previously performed biopsy of our patient and confirmed the diagnosis of OM.

Owing to partial loss of bone in the anterior part of the right orbital floor, reconstruction was performed using a Medpor micro thin-sheet non-resorbable implant (Stryker Corporation, Kalamazoo, Michigan, USA), individually fitted to passive adaptation ([figure 7](#)).

At 3-week postoperative follow-up, the patient presented with normal vision, symptoms were minimal, and the patient complained of only slight discomfort. Objectively, a slight tendency to exophthalmia was registered, which neither the patient nor

Figure 6 Postoperative image of the patient after removal of the tumour.

Figure 7 Peroperative image showing the placement of the Medpor implant in the orbita.

the parents had noticed. There were no signs of ectropion or entropion. The pupillary line was normal.

At follow-up 2 years after the surgery ([figure 8](#)), the patient was in good health with normal vision and unrestricted eye movements. No evidence of exophthalmia or symptoms suggesting infraorbital nerve damage was seen. Follow-up CT scans at 6 and 24 months postoperatively showed no sign of recurrence ([figure 9](#)). A satisfying anatomical position of muscles and the bulb was registered.

Follow-up intervals were every third month for the first year and thereafter every sixth month until 5 years after surgery.

Material and methods

A Medline search (PubMed) was conducted of OM using the following inclusion criteria: orbital involvement, children <13 years, English language, articles published after 1985.

Searching Medline with the term 'Odontogenic Myxoma Children' resulted in 76 hits. In the screening of titles and

abstracts, 47 potential publications were found relevant, and a full-text analysis was performed by one author (TSH). Subsequently, we performed a hand search among these 47 articles, which resulted in finding one additional article. Of the 48 articles, a total of 6 articles were included in the study ([table 1](#)).²⁻⁷

CONCLUSION

Only eight other cases of OM in children with orbital involvement have been presented. We lack data on recurrence rates of OM reported in children. Therapy varies, and there is no gold standard for treatment of odontogenic myxomas in children with orbital involvement. Medpor seems to be a good and obvious choice when reconstruction involves the orbit in children, but may also be considered in adults because of higher recurrence rates and better X-ray diagnostic possibilities following reconstruction.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses included malignant (rhabdomyosarcoma and glioma) and benign lesions (haemangioma, cherubism, Langerhans cell histiocytosis and nodular fasciitis).

OUTCOME AND FOLLOW-UP

At follow-up 2 years after the surgery, the patient was in good health with normal vision and free eye movements. There was no evidence of exophthalmia or symptoms suggesting infraorbital nerve damage.

DISCUSSION

This case is one of only nine reported cases in the literature ([table 1](#)), meaning that OM in relation to the orbita in children is extremely rare. Follow-up of patients in the reviewed articles ranged from 16 months to 14 years. Treatment varied from curettage to resection, but in no article was the range of the resection margin or how the reconstruction was performed mentioned.

The recurrence rate of OM in children seems low,⁵ which advocates for less invasive surgery. In the current case, a 2 mm

Figure 8 Postoperative image of the patient 2 years after removal of the tumour.

Figure 9 CT image 2 years after removal of the tumour.

Table 1 Summary of reported cases of odontogenic myxoma with orbital involvement in children (<13 years)

Reference	Cases, (n)	Gender/age (months)	Treatment	Reconstruction	Follow-up	Recurrence
Chen <i>et al</i> ⁵	1	F/15	Excisional biopsy and curettage	NR	27 months	No
Kansy <i>et al</i> ⁷	2	NR/12	Resection	Not necessary	8 years	No
		NR/11	Resection/partial maxillectomy	Not necessary	24 months	No
Rotenberg <i>et al</i> ⁴	2	F/18	Maxillectomy	NR	14 years	No
		M/16	Resection	NR	7 years	No
Fenton <i>et al</i> ³	1	M/17	Excisional biopsy and partial resection	Not necessary	16 months	No
Wachter <i>et al</i> ²	1	M/19	Resection	NR	2 years	No
Leiberman <i>et al</i> ⁶	1	M/15	Excision	NR (slight erosion...)	NR	NR
Total	8	Average age 15.4 months				

NR, not reported.

resection margin was chosen. Recurrence in children has most often been seen after a mean follow-up period of 24 months.⁵ High recurrence rates have, however, been observed in the adult population (5–10%),^{5,6} but there is a degree of statistical uncertainty regarding these recurrence rates. Surgical techniques and follow-up regimens vary in the literature.

The primary reconstruction in our case was carried out using a Medpor implant (ultra-thin sheet, 0.25 mm thickness). Owing to the availability and ease of use, Medpor is commonly used though for reconstruction after orbital floor fractures in adults^{8,9} and has also been described in the literature for use in children.¹⁰

We chose a Medpor implant because of the smooth surface, which makes it easier to remove than a titanium mesh if recurrence occurs. Furthermore, metal implants do have a tendency to create more radiographic image ‘noise’ than implants made of plastic materials, such as the Medpor implant, which is made of polyethylene. The thickness of the implant is also important. Furthermore, periorbital and intraorbital growth restrictions were considered. Even though roughly 77% of the orbital cavity has been formed by the age of 5,¹¹ the implant chosen must be able to adapt to the growth pattern.

Myxomas are found in the bone, connective tissue and in the myocardium. In the head and neck region, myxomas are rare and mostly found in the jaws. The localisation and the presence of odontogenic epithelium in the tumour or in relation to unerupted teeth suggest that OM is derived from ectomesenchymal tissue.

Since OM is the fourth most common odontogenic tumour, a slowly growing asymptomatic swelling in relation to the

Learning points

- Recurrence rates seem low in children, but the literature is sparse on this matter.
- Reconstruction of the orbita was performed using a Medpor implant. This has not been reported previously.
- This is one of nine reported cases of odontogenic myxoma in relation to the orbita in children.
- Surgical techniques and follow-up regimens vary.

mandible or the maxilla should raise suspicion of OM. Owing to its slow growth, OM can reach a considerable size before becoming symptomatic. In our case, the tumour was asymptomatic, primarily because of its expansion into the maxillary sinus.

Symptoms may include pain, lack of tooth eruption, malocclusion, nasal stenosis, epistaxis, tear secretion and eye infection.

Differential diagnoses included malignant (rhabdomyosarcoma and glioma) and benign lesions (haemangioma, cherubism, Langerhans cell histiocytosis and nodular fasciitis).⁷

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Contributors TSH is the primary and corresponding author. THT planned and carried out the operation. LID, SF and THT reviewed the article.

Competing interests None declared.

Patient consent Obtained.

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Patient's perspective

- *From a telephone conversation with the patient's mother:*
- Immediately after surgery, there were no unusual complaints.
- About 1 year after surgery, my son suddenly experienced a swelling and pain in the right eye and its surroundings. His right eyelid was so swollen that he had difficulty looking out of his eye. Later the same day, he was examined in the emergency room. They did not find anything suspicious, and by that time the eye looked normal again. My son has not had any complaints since then, but the episode made us think about whether the tumour had returned.

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