Intraoral Approach for a Congenital Teratoma in the Orbit Extending into the Pterygopalatine Fossa

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Summary: This article reports an intraoral approach for a congenital mature teratoma in the orbit, extending into the pterygopalatine fossa and the zygoma. The operation took place on the 56th postnatal day due to gradual proptosis of the left eye from the socket. The entire orbital tumor could be surgically removed by the pressure of the tumor. This case suggests that orbital tumors expanding into pterygopalatine fossa can be removed by the intraoral approach because the bone of infants is soft. The eyeball of the patient was temporarily depressed after the operation, but that was improved by tissue remodeling with time. Therefore, it is recommended that reconstructive surgery for an infant should be performed at the age of 12 months or older. (Plast Reconstr Surg Glob Open 2020;8:e3238; doi: 10.1097/GOX.0000000000003238; Published online 30 November 2020.)

Termas most commonly occur in the sacrococcygeal region, genital organs, and the retroperitoneum, whereas the frequency of head and neck teratomas is 2%–5%—in particular that of the orbit is <1%. The location of the teratoma corresponds to the embryonic resting sites of primordial totipotential germ cells. Surgical approaches to the orbit and pterygopalatine fossa reported so far include skin incision and osteotomy. In this report, we investigated the surgical approach for a congenital mature teratoma in the orbit, extending into the pterygopalatine fossa.

CASE REPORT

A 43-day-old boy born after a normal pregnancy was examined at the department of plastic and reconstructive surgery due to swelling of the left maxillary region and proptosis of the left eyeball. Magnetic resonance imaging (MRI) findings at 53 days of age revealed a 5 × 3.5 × 2.7 cm solid tumor, which was partly cystic in the left orbital expanding into the pterygopalatine fossa through the inferior orbital fissure. A computed tomography (CT) scan performed at 53 days of age showed an expansion of the inferior orbital fissure and zygomatic arch (Fig. 1). So we chose the intraoral approach. Serum α-fetoprotein (αFP) was not elevated at 1171 ng/ml. An immature teratoma was suspected because of its rapid increase in size and based on MRI findings. Therefore, an earlier surgery was performed on the patient 3 days from the first visit.

A part of the mass in the pterygopalatine fossa was excised through an intraoral approach (Fig. 2), and a part of orbital mass was excised under a microscope using a similar approach through the inferior orbital fissure (Fig. 3). Because the inferior orbital fissure and zygomatic arch are expanded by the tumor, we did not perform maxillary antrostomy or opt for excision of the inferior orbital fissure to reach the orbital tumor. Moreover, the excision was complicated. A drain tube was inserted through the cheek skin and into the space created from the tumor excision, and no reconstructive surgery was required for the space.

Cryosectioning and subsequent pathological examination of the tumor occupying the orbit and pterygopalatine fossa revealed that the tumor was a mature teratoma. Most neonatal teratomas consist of ectodermal and mesodermal component, and teratomas are histologically classified as mature or immature. Most pediatric teratomas are mature and benign.

The patient had an eyeball depression after the operation, but this depression improved, and the patient’s

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maxillary region became symmetric over the next 2 months. An MRI scan performed at 5 months after operation revealed that there was no tumor recurrence, zygoma remodeling, and the space that had been created from the tumor excision was filled with soft tissue (Fig. 4). The patient’s condition was stable over the next 10 months.

**DISCUSSION**

This article reports an intraoral approach for treating a congenital mature teratoma in the orbit, extending into the
fossa approach or the Weber-Fergusson incision. 7, 8 For our excision of this type of tumor involves the infratemporal orbit expanding into the pterygopalatine fossa is rare, and approach. 5 Surgical approaches available for the orbit are combined endonasal and sublabial endoscopic transmaxillary approach, and recently a less-invasive route, which combined (transfacial) or lateral/posterolateral (transcranial) approach for the resection of the orbit and pterygopalatine fossa and the zygoma remodeling during the neonatal period. The entire tumor could be removed through an intraoral approach alone, as the bone was so soft, and the inferior orbital fissure and zygomatic arch were expanded by the tumor. It is suggested that the improvement of the patient’s eyeball depression after the operation was the result of facial bone and soft tissue remodeling, which occurs only in the infant. Therefore it is recommended that reconstructive surgery should be performed in children 12 months or older.

**CONCLUSIONS**

This article reports an intraoral approach for a congenital mature teratoma in the orbit, extending into the pterygopalatine fossa and the zygoma remodeling during the neonatal period. There are 2 important points to note. First, the entire tumor could be removed through an intraoral approach because an infant’s bone is so soft that the inferior orbital fissure was expanded by the tumor. Second, it is possible that our patient’s eyeball depression after the operation can be improved as a result of facial bone and soft tissue remodeling, which can occur only in an infant.

Thus far, surgical approaches available for the pterygopalatine fossa include the traditional anterior approach (transfacial) or lateral/posterolateral (transcranial) approach, and recently a less-invasive route, which combined endonasal and sublabial endoscopic transmaxillary approach. 5 Surgical approaches available for the orbit are transconjunctival (anterior), transcranial (superior), transmaxillary (inferior), lateral orbitotomy (lateral), and endoscopically endonasal (medial) approaches. 6 A tumor in the orbit expanding into the pterygopalatine fossa is rare, and excision of this type of tumor involves the infratemporal fossa approach or the Weber-Fergusson incision. 7, 8 For our case, the left inferior orbital fissure was 21.5 mm, which is larger than the right 13.8 mm to the expanding cyst, and the left bony orbital volume was 175.5 mm; 2 which is also larger than the right 53.4 mm. 2 It was therefore thought that the entire tumor could be removed through an intraoral approach alone. Therefore, we recommend the intraoral approach for the resection of the orbit and pterygopalatine fossa tumor when the bone is so soft and the inferior orbital fissure and zygomatic arch are expanded by the tumor.

In facial bone and soft tissue remodeling process in the infant due to several facts, 1 of them is the functional matrix theory. 9 In the orbit sagittal and vertical directions, respectively, 56.3% and 61.5% of the growth increase occurs during the first 12 months; therefore, it is thought that high plasticity of the tissue during this period affects craniofacial remodeling. 10 In this case, the patient was 2 months old and recovery of facial symmetry after the operation suggests that remodeling of the patient’s tissue had occurred. Therefore it is recommended to consider the remodeling of an infant’s tissue, and reconstructive surgery of infants should be performed at the age of 12 months or older.

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**Fig. 4. Post MRI T2 FatSat (5 months after operation) of the patient. There is no tumor recurrence and the space after resection is filled with soft tissue.**