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Original article

Oropharyngeal teratomas in newborns: Management and outcome

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ABSTRACT

Objectives: Congenital teratomas of the oropharyngeal cavity are extremely rare and are associated with a high neonatal mortality rate due to severe airway obstruction. Management has been improved with progress in antenatal diagnosis. The authors describe this progress in the light of a series of 4 cases and a review of the literature.
Methods: The medical charts of four neonates treated in the department since 1995 were reviewed. The following criteria were studied: age at diagnosis, clinical and radiological features of the tumour, management at birth and outcome.
Results: All four cases occurred in female neonates with an antenatal diagnosis in two cases, allowing preparation for endoscopy in the delivery room in one case and an EXIT procedure in the other case. Three neonates had to be intubated in the delivery room. Imaging showed invasion of the infratemporal fossa in 3 of the 4 cases. Surgical resection via various approaches to the infratemporal fossa was complete in every case. Adjuvant chemotherapy was administered in one case.
Conclusion: Surgery for these mostly benign tumours is very challenging and requires a multidisciplinary team. Perinatal planning allows appropriate management at birth, decreasing the risk of airway obstruction. Surgery is the mainstay of treatment of teratomas.

1. Introduction

Teratomas are the most common congenital tumour (25–35%) [1] with an incidence of 1/4000 births [2]. Eighty percent of these tumours are situated in sacrococcygeal and gonadal sites. Teratomas of the head and neck represent 5 to 15% of all sites, predominantly involving the nasopharynx and neck [3], while oropharyngeal tumours are exceptional (2%). Systematic antenatal ultrasound allows early diagnosis of these tumours and planning of appropriate management. The objective of this review was to report the long-term outcome of 3 infants treated in our department [4] and to describe the management of a recent case treated after birth by the EXIT procedure.

2. Clinical cases

2.1. Case 1

F. was a baby girl born prematurely in 1995 at 32 weeks of amenorrhoea (WA). Emergency caesarean section was necessary due to severe maternal pre-eclampsia. The baby was immediately transferred to the neonatology unit with rapidly resolving airway obstruction. However, airway obstruction gradually returned and examination demonstrated an oropharyngeal mass with retropalatal extension. Intubation was required at 3 weeks of life for airway obstruction. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a tumour arising from the soft palate with a heterogeneous appearance comprising calcifications and cysts. The lesion extended to the left infratemporal fossa with a fat density intracranial extension. Alpha-fetoprotein (AFP) assay was normal for age (1700 ng/ml) (Table 1 [5]). Endoscopy under general anaesthesia allowed biopsy of the mass and relief of airway obstruction. The lesion was implanted posteriorly to the tonsil, on the posterior pillar and dorsal surface of the soft palate. Histological examination revealed glial heterotopia suggesting teratoma with a probable immature contingent. Surgical resection was performed at the age of 2 months. The infratemporal contingent was resected via a superior transmandibular approach with resection of the coronoid process (according to Shaheen's description [6]) and the oropharyngeal nodule was resected via an intraoral approach. Histological examination revealed a diagnosis of mature teratoma based on analysis of the epithelial, nerve, muscle, bone and cartilage contingents. Chronic Eustachian tube insufficiency subsequently required insertion of a tympanostomy tube in the left ear, followed by treatment of cholesteatoma. This child did not present

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any soft palate insufficiency. Trismus due to temporomandibular ankylosis required two surgical procedures. The child presented minimal persistent mandibular laterognathia. This patient, now a 14-year-old adolescent, has no signs of recurrence on clinical and MRI follow-up.

2.2. Case 2

S. was a baby girl born at term in 1996. Antenatal ultrasound revealed a very large nasopharyngeal mass from the second trimester of pregnancy. Severe airway obstruction due to an oropharyngeal tumour required intubation at birth by an otorhinolaryngologist present in the delivery room. CT and MRI imaging also revealed extension to the right infratemporal fossa and extrameningeal intracranial extension to the temporal fossa (Fig. 1A). AFP assay was normal for age (5000 ng/ml) (Table 1 [5]). Endoscopy under general anaesthesia, performed at 9 days of life, revealed an irregular, hard, whitish oropharyngeal mass arising from the free edge of the soft palate. Resection of this pharyngeal mass allowed extubation and histological examination revealed a diagnosis of mature teratoma. At the age of 2 months, surgical resection was planned in collaboration with a neurosurgeon. Pterional craniotomy allowed resection of the superior part of the tumour via the zygomatic arch as far as the infratemporal fossa and the soft palate portion of the tumour was resected via an intraoral approach. Histological examination confirmed the diagnosis of mature teratoma. At the age of 3 months, the child presented a temporomalar mass. MRI demonstrated recurrence in the infratemporal fossa extending to the floor of the middle cranial fossa and middle ear. Surgical revision comprised a type C infratemporal approach (according to the description of Fisch [7]) and temporal craniectomy. Histological examination of the lesion confirmed the diagnosis of mature teratoma. AFP on postoperative day 7 was normal. The child was reoperated at the age of 3 years for residual cholesteatoma after exclusion of the ear. At the age of 14 years, she is still regularly reviewed and presents no clinical or radiological signs of recurrence with a normal AFP. She presents right mandibular hypoplasia with limited mouth opening (Fig. 1B), but normal facial movements.

2.3. Case 3

E. was a baby girl born at term in 1998. She was immediately transferred to neonatal intensive care because of airway obstruction that was relieved after expelling a pink tumour from the mouth, but which rapidly relapsed on deglutition. A stay suture placed in the tip of the tumour prevented subsequent episodes of asphyxiation. This soft, pink lesion was implanted on the tonsil and extended to the soft palate. MRI did not reveal any signs of extension of this heterogeneous fat density lesion. Complete resection was performed on the first day of life with removal of the tonsil and the soft palate implantation. The lesion measured 5 cm long and 1.5 cm in its largest diameter. Histological examination revealed a diagnosis of mature teratoma. No clinical and radiological recurrence was observed at the age of one year. The child's family subsequently left the region and this child has been lost to follow-up.

Table 1
Normal alpha-fetoprotein (AFP) levels in neonates and infants (ng/ml) [5].

Age	Number of children	Mean
Premature	11	134,734 ± 41,444
Neonate	55	48,406 ± 34,718
Birth – 2 weeks	16	33,113 ± 32,503
2 weeks – 1 months	43	9452 ± 12,610
1 months	12	2654 ± 3080
2 months	40	323 ± 278
3 months	5	88 ± 87
4 months	31	74 ± 56
5 months	6	46.5 ± 19
6 months	9	12.5 ± 9.8
7 months	5	9.7 ± 7.1
8 months	3	8.5 ± 5.5
> 8 months	–	8.5 ± 5.5

2.4. Case 4

G. was a baby girl born at term in 2009 by the EXIT procedure. Antenatal ultrasound was requested at the 17th week of amenorrhoea (WA) to document a triple test with abnormal elevation of AFP and revealed a left lateral cervical foetal cyst measuring 6 mm in diameter. Regular ultrasound and MRI surveillance diagnosed progressive enlargement of a heterogeneous, polycystic neck mass associated with a solid intraoral mass. At 36 WA, the cystic swelling measured 67 × 66 × 40 mm on MRI, but no deviation of the trachea and oesophagus was observed (Fig. 2A). Delivery

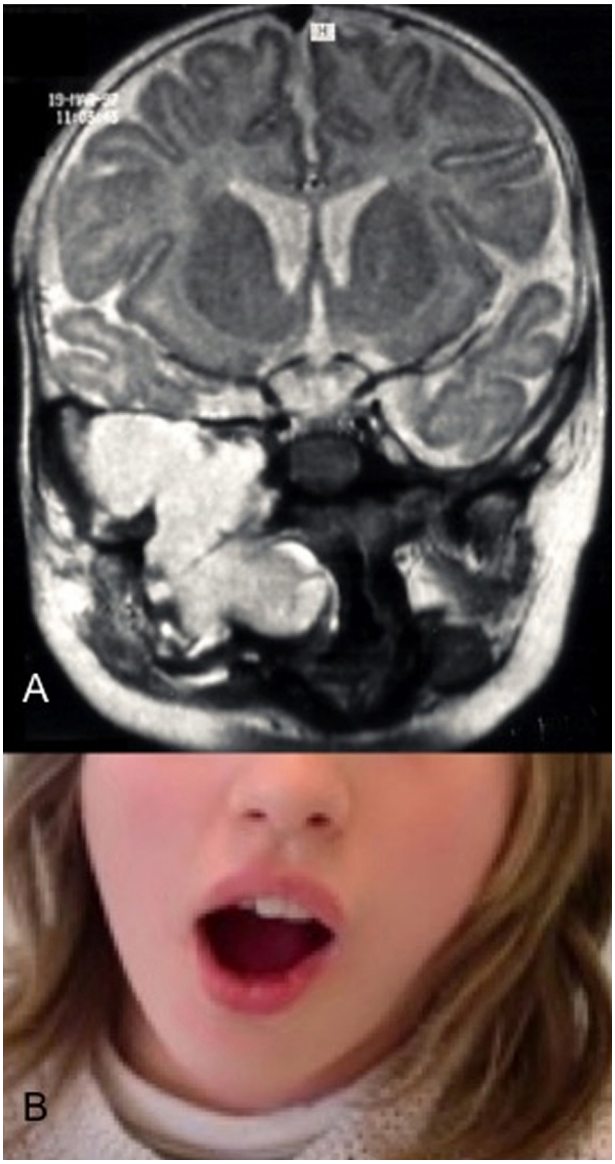

Fig. 1. A. MRI, T2-weighted sequence in a neonate (case 2). The oropharyngeal teratoma has a heterogeneous, polycystic appearance, invading the right infratemporal fossa with intracranial and extrameningeal extension. B. Clinical appearance (case 2) of the child at the age of 14 years with right lateral mandibular deviation and limited mouth opening.