

## A Case of Neonatal Mature Teratoma Transformed to Malignancy in the Neck Extending to the Mouth Floor

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**A case with neonatal teratoma originating from the cervicofacial region which transformed to be malignant during treatment is reported. The case is a full-term baby girl with swallowing difficulty and has a mass at the floor of her mouth with the right neck swelling. The mass was revealed to be multi-cystic and extending deep into the sublingual space and protruding outside. Puncture and marsupialisation of the cyst could not relieve her symptom and the tumor was resected in three occasions and was diagnosed as mature teratoma without malignant component. However, three months after the last resection, the solid right neck mass enlarged rapidly and the serum alpha-fetoprotein level was elevated. Biopsied specimen demonstrated the mass to be germ cell tumor with embryonal carcinoma and yolk sac tumor component. Eight courses of JEB regimen with recurrent mass resection successfully lead to complete regression without compromising patient growth as well as cosmetics. Head and neck teratomas in children are mostly benign amenable to curative excision but its rarity and site and size of the tumor make its treatment challenging. It is important to have multi-disciplinary management for the disease from neonatal period until growth has finished. There exists a relationship between the age at diagnosis and outcome of a patient with teratoma and head and neck teratomas in neonate are mostly benign but should be removed completely as soon as the patient condition is stabilized to reduce the risk of malignant change.**

**Key words:** mature teratoma, neonate, neck, malignant teratoma

### INTRODUCTION

Teratomas are exceptionally rare tumors in the head and neck region. Neonatal teratomas occur in about 1: 20,000-1: 40,000 live births [1, 2] and only 5% are in the region [3]. Congenital teratomas are mostly benign and said to be amenable to curable resection but the site of their origin and their rarity make the treatment challenging [1-4]. Furthermore, most congenital teratomas are mature or immature ones but they can recur as malignant teratoma after initial surgery [5]. In this paper, a case of neonatal teratoma originating from deep in the neck is presented. The patient developed malignant teratoma during treatment but several surgical resections and multi-agent chemotherapy successfully completed regression without compromising patient growth. Appropriate management of this rare entity is discussed with a literature review.

### CASE REPORT

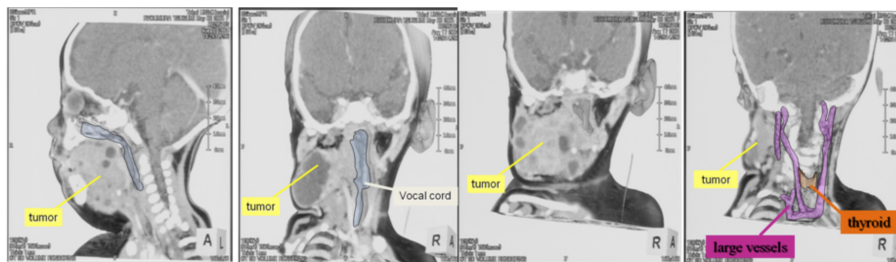
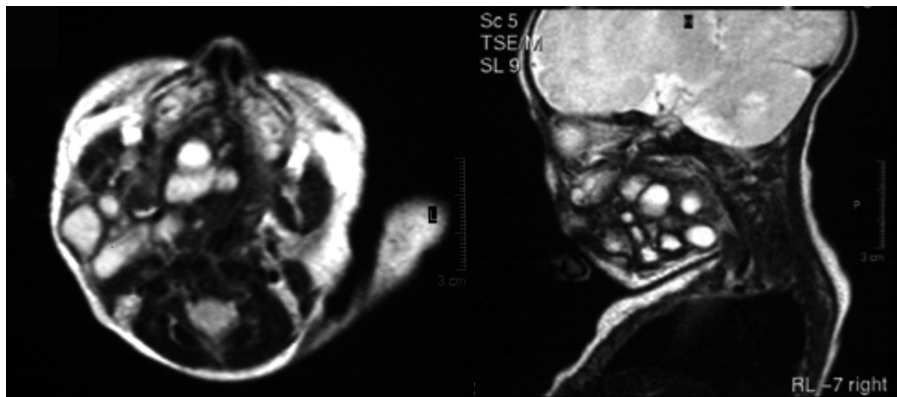
A female newborn baby was born by vaginal delivery at 39 weeks of gestational age with 2650 g of weight. A mass at the floor of the mouth causing the tongue deviate to the left was noticed with the right cervical swelling (Fig. 1) and she was transferred to our NICU. Although she could be fed by milk with some difficulty, the mass was revealed to be multi-cystic which had extended deep into the sublingual space as well as outside of the lateral neck but without

compromising vital structures (Fig. 2). Biopsy done at 30 days of age demonstrated smooth muscle layer with epithelial lining, which suggested enterogenous cyst but no definite pathological diagnosis was made. Her ingestion of milk became more difficult and puncture of the sublingual cyst at 48 days of age followed by marsupialisation of the cyst somewhat relieved her symptom but not satisfactorily. Enlargement of the tumor necessitated neck tumor resection at 4 months of age and histopathologically, the tumor was diagnosed to be mature teratoma (Fig. 3). The resection resulted in adequate weight gain and she was discharged from the hospital. The sublingual tumor, however, caused bleeding at her 10 months of age and the tumor was resected as much as possible when she was 13 months old. Pathological diagnosis was again mature teratoma without malignant component.

However, one month after discharge, the solid right neck mass enlarged rapidly and the serum alpha-fetoprotein (AFP) level was elevated to be 6591.6 ng/ml. Biopsied specimen was demonstrated the mass to be germ cell tumor with embryonal carcinoma and yolk sac tumor component (Fig. 4). Four courses of the JEB regimen according to UKCCSG protocol, which consists of 120 mg/m<sup>2</sup> of etoposide at day 1, 2, 3, 600 mg/m<sup>2</sup> of carboplatin at day 2 and 15 mg/m<sup>2</sup> of bleomycin at day 3, decreased serum AFP to 2.9 ng/ml and remained within normal range. Mass became unpalpable from outside but the multi-cystic



**Fig. 1** Neck tumor extending to the mouth floor.



**Fig. 2** MRI (T2 weighted) shows multi-cystic mass and 3-D CT images show the tumor expanding but without compromising vital structures.

mass was demonstrated to be remained at the floor of the mouth by ultrasonography and MRI. Complete tumor resection was attempted when she was 20 months old but the resected tumor demonstrated only mature component of the teratoma without malignancy.

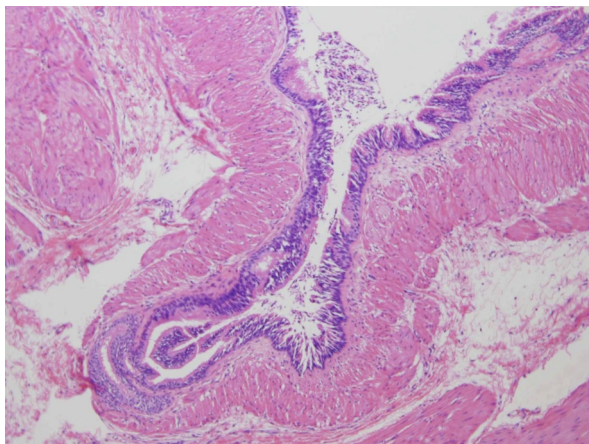
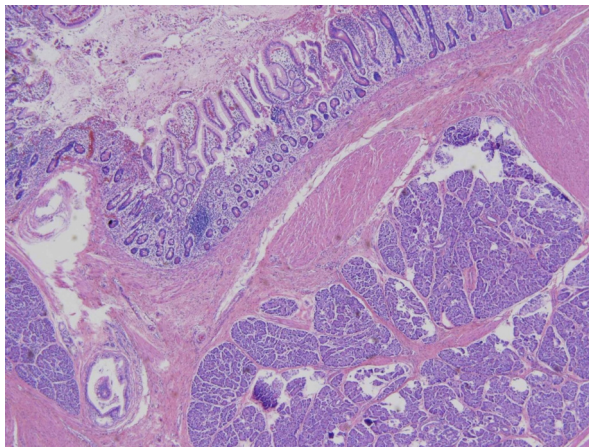
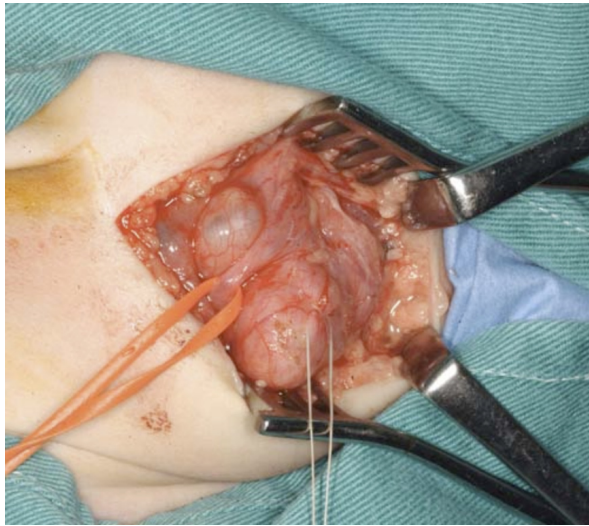
Two months after the resection serum AFP level was elevated again up to 127.9 ng/ml and the small lump was noted at the right submandibular region on palpation and the tumor was resected, which was demonstrated to be recurrence of yolk sac tumor component. Another four cycles of JEB chemotherapy were added and the AFP returned to normal and remained low after completion of therapy (Fig. 5). Her cosmetic

outlook is satisfactory with minimal deviation of the tongue and the mandible at her 3 years of age (Fig. 6). She has been continually cared with other specialist including oro-dental surgeons.

#### DISCUSSION

Teratomas are tumors containing components of all three embryological germ layers originating from multi-potential germ cells. The germ cell tumor can commonly arise from the gonads but the extragonadal teratoma can arise outside the gonads. Sacrococcygeal area is the most common site of the tumor and head and neck are the least one in children consisting only

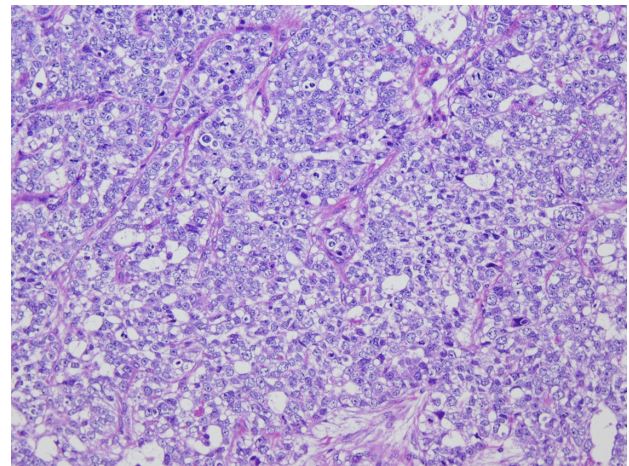
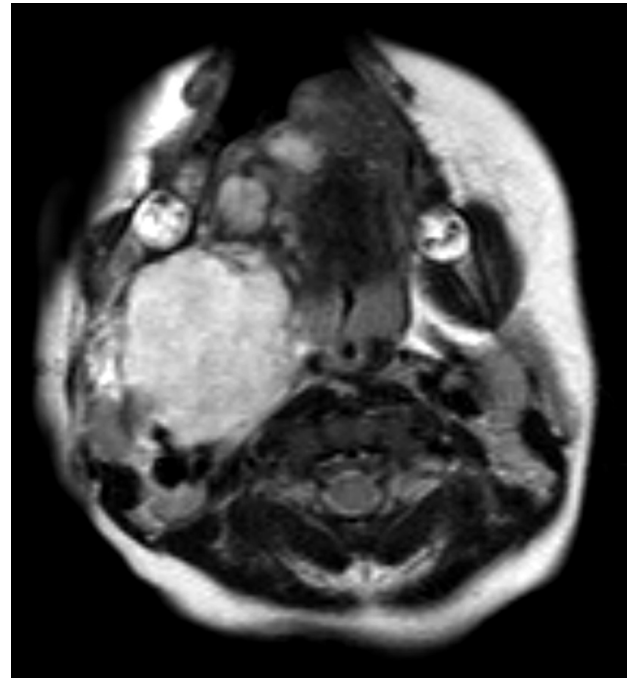




**Fig. 3** Biopsied specimen revealed to be mature teratoma histologically.

5% of the case [3]. Head and neck teratomas in children are mostly benign amenable to curative excision but its rarity and site and size of the tumor make their treatment challenging [1-4].

Prenatal ultrasound scanning is so widely undertaken that it is possible that tumors can be detected prenatally. Some prenatal cases with neck teratoma have been reported to be terminated pregnancy [5]. The natural history of this condition is essential for clinicians to enable informed parental choice. When prenatal diagnosis is made to have cervicofacial tumor, it is mandatory to secure the airway at the delivery



**Fig. 4** MRI of the recurrent tumor and its histology which contains embryonal carcinoma and yolk sac tumor.

room. The management of cervicofacial teratomas in neonates is often complicated and may result in significant morbidity and death. In a Childrens Cancer Group retrospective study that identified twenty neonates with cervicofacial teratomas two died in a delivery room and two required tracheostomy to survive [1]. To secure the compromised airway a multidisciplinary team should be involved for ex utero intrapartum treatment (EXIT) or operation on placental support (OOPS) [6-9]. In our case the airway was not compromised and the milk feeding is possible in spite of the large mass.

Another challenging aspect of the head and neck teratoma was cosmetic result, which is satisfactory in spite of multiple resections and the tumor location in the present case. In the era when anatomical details of the 3D tumor location can be obtained by multimodal imaging techniques, meticulous dissections to conserve the normal components are possible and treatment strategy without jeopardizing the functional and cos-

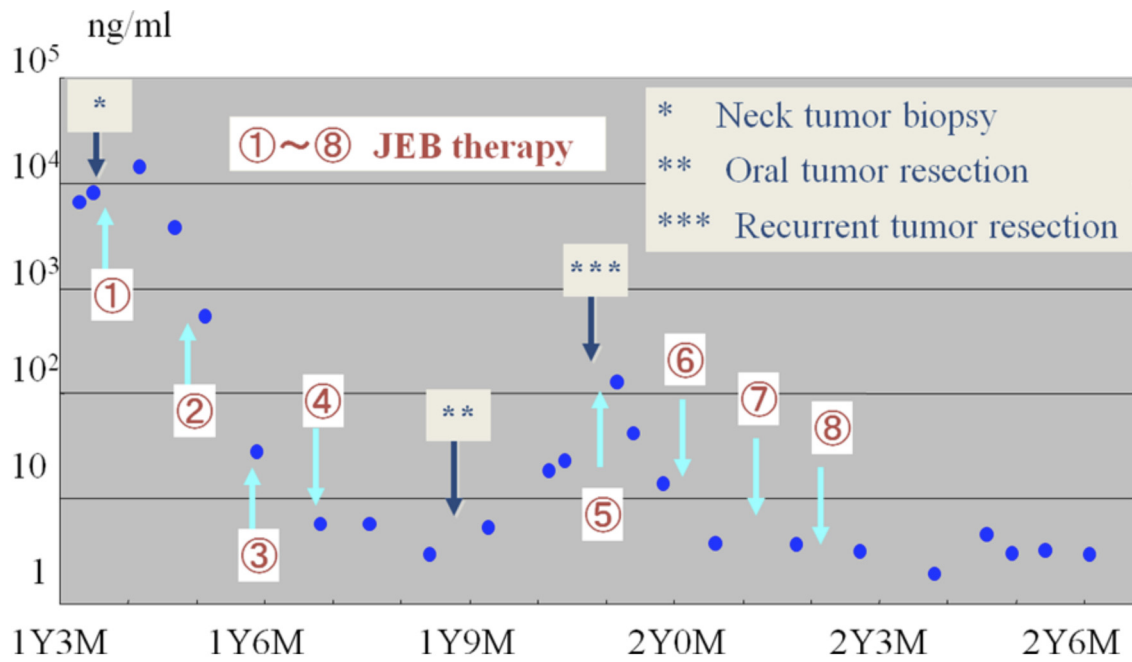


Fig. 5 Treatment after malignant transformation and serum AFP level.



Fig. 6 Patient appearance after completion of therapy at her 3 years of age.

metic outcome should be considered. It is important to have multi-disciplinary management for the disease from neonatal period until growth has finished.

Malignant teratoma is exceptional in perinatal cervical cases. There have been reported malignant teratomas in the head and neck region in neonates [1, 2, 4, 10-13] and Kerner et al reviewed the literature in 1998 and found malignant teratomas have been reported in 9 of 220 cases [4]. According to the extensive review of 534 patients with teratoma by Issacs Jr [3], 10 of the 70 (14%) cervical teratoma cases had metastases to either adjacent lymph nodes or to distant organs but

without malignant behavior except one with yolk sac tumor which is the leading malignant germ cell tumor component.

It is well known that there exists a relationship between the age at diagnosis and outcome of a patient with sacrococcygeal teratoma. The incidence of malignancy in the neonate is approximately 10%, approaching almost 100% by 3 years of age [3, 10, 14, 15]. The yolk sac tumor microfoci are speculated to give rise to malignant relapse after incomplete resection. In the present case, partial resection of the tumor and wait and see attitude must have resulted in malignant



transformation of the microfoci, a situation analogous to sacrococcygeal teratomas. Head and neck teratomas in neonate are mostly benign but should be removed completely as soon as the patient condition is stabilized to reduce the risk of malignant change.

In our case, diagnosis of malignancy was made by rapid growth of the tumor associated with marked elevation of the serum AFP. Subsequent multi-agent chemotherapy along with complete resection of the tumor has led to disease-free state of the patient. Treatment strategies for malignant germ cell tumors in children have changed dramatically over the past few decades after introducing the platinum-based chemotherapy [15, 16]. Children with extensive or invasive disease at diagnosis are treated by a conservative approach with biopsy only. Chemotherapy is highly successful in tumor shrinkage that will allow a precise resection with defined margins. The current regimens include cisplatin, etoposide, and bleomycin (PEB) and the most recent trials have survival rates ranging from 85% to 95% and our used regimen includes carboplatin instead of cisplatin to reduce the known renal toxicity. For those children with a large or persisting mass after chemotherapy, however, an aggressive surgical approach is justified. Residual malignant component can be monitored by serial AFP measurements and complete resection is assured by the decline of AFP to normal after cessation of the chemotherapy.

In conclusion, head and neck teratomas in neonates are mostly benign but their perinatal management and treatment strategy is challenging. A multidisciplinary team should be involved for securing the airway in the delivery room. Once the tumor is suspected to be teratoma, it should be removed completely as soon as the patient condition is stabilized to reduce the risk of malignant change. Surgical resection should be preferably done after the tumor 3D location is elucidated by multimodal imaging techniques to conserve the functional and cosmetic outcome. Malignant transformation of benign teratoma in children is possible but the malignant teratoma can be curable with multi-agent chemotherapy along with complete resection. Residual malignant component can be monitored by serial AFP measurements and complete resection is assured by the decline of AFP to normal after cessation of the chemotherapy. Multi-disciplinary management for the disease should be continued from neonatal period until growth has finished.

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