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Robotic Assisted Resection of a Nasopharyngeal Teratoma in a New-Born: A Case Report

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Abstract

Teratomas in the head and neck region are rare embryonal neoplasms consisting of tissues derived from all three blastodermic layers. Typically, these lesions have a heterogenous histological appearance. Head and neck location of a teratoma may be associated with an impressive clinical presentation caused by upper airway obstruction. The first line treatment for teratoma is surgical excision. However, especially in the head and neck region, total removal can be impossible due to difficult access and location of the lesion. We present the first case reported in literature of a nasopharyngeal teratoma in a new-born which was completely removed by using the da Vinci robot. Adhesion to the vomer and skull base was noted on MRI. Nevertheless, total excision of the mass was confirmed on histology. The infant was discharged after 2 days without any signs of upper airway obstruction.

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Introduction

Teratomas are embryonal neoplasms consisting of tissues derived from all three blastodermic layers (ectoderm, endoderm and mesoderm). They occur most commonly in the sacrococcygeal region (incidence of approximately 1 in 27,000). Teratomas in the head and neck region represent only 3% of all teratomas [1-3]. Although considered a benign lesion, a teratoma in a neonate can be associated with a considerable morbidity and even mortality. Fetal hydrops and premature delivery can result from the volume of the tumor (respiratory distress because of partial or complete airway obstruction and dysphagia, facial disfigurement or orbital involvement) [4]. The most common location for teratomas in the aerodigestive tract mucosa is the nasopharynx. Other less commonly involved sites include the oral cavity (tonsils, tongue, palate), sinonasal cavity, the ear and the temporal bone [5]. Teratomas originate from totipotential germ cells which fail to migrate appropriately during development and escape the influence of their regional primary organizers [5]. In the head and neck region, the abnormal descent of Rathke's pouch endoderm and neuroectoderm with trapped elements of adjacent endoderm and mesoderm may form a nasopharyngeal teratoma [6]. Ultrasound and Magnetic Resonance Imaging (MRI) are the primary imaging techniques used to evaluate neonatal teratomas, both in the pre- and post-natal period. In some cases, a high-resolution modern ultrasound technique allows for direct visualization of large teratomas of the head and neck on routine prenatal ultrasound studies. However, often only secondary findings, such as polyhydramnios, are seen. The identification of calcifications within the lesion suggests the diagnosis of teratoma over other neonatal masses of the head and neck, such as a Lymphatic Malformation (LM), and may be easier to identify on ultrasound than on MRI [6]. Surgical excision is the treatment of choice for teratomas because they do not have intracranial extension, being encapsulated or pseudo encapsulated. Yet, nasopharyngeal masses are often difficult to remove

completely due to the difficult access and location [6].

Case Presentation

A one-day-old male neonate was admitted to the Neonatal Intensive Care Unit (NICU) because of respiratory distress noticed immediately after birth. The baby was born by normal vaginal delivery in a peripheral hospital at 36 weeks of gestation. Birth weight was 2,980 g with APGAR score of 7/4/5 with apnea and cyanosis several minutes after birth. Prenatal ultrasound was unremarkable. No sign of polyhydramnios was noted. Specific medical history of the parents reveals on maternal side (G3P3A0) presence of sickle cell anemia (4 family members positive) and deep venous thrombosis after second part. Because of severe respiratory distress postpartum, caused by upper airway obstruction at the level of the nasopharynx which made it impossible to insert a nasotracheal tube, the infant had to be intubated orally and was transferred to the NICU department. Because extubation on the second day was unsuccessful with recurrent distress symptoms, further investigations were required. Flexible nasal endoscopy revealed a mass in the nasopharynx extending into the oropharynx and obstructing the upper airway. Due to agitated state, the new-born had to be maintained under sedation during examination.

Investigations

Chest radiography and echocardiography showed no abnormalities. Magnetic resonance imaging demonstrated a pedunculated soft tissue mass with both fatty and multi-cystic components occupying the nasopharynx extending inferiorly into the oropharynx. No adhesion is suspected to the nasopharyngeal walls, but attachment to the vomer and/or skull base is suspected (Figure 1). It measured approximately 1.6 cm anteroposterior × 1.9 cm laterolateral × 3.6 cm craniocaudal diameter (Figure 2). Differential diagnosis was made between a teratoma and a dermoid cyst.

Genetic testing was also performed by SNP (Single Nucleotide Polymorphism) array to detect pathologic deletions or duplications, but no anomalies were detected in current case.

Surgical management

Given the dimensions of the nasopharyngeal mass, extubation was deemed impossible and in order to avoid a tracheostomy, a surgical intervention was scheduled at earliest convenience and after counselling of the parents. A surgical intervention was scheduled on day five. During the pre-operative planning, it was decided to use a transpalatal approach and to employ the da Vinci robot and its angled instruments. We expected this would allow for a total removal of the mass, which was protruding behind the soft palate with suspected adhesion to the vomer and skull base. Moreover, the da Vinci robot would allow for delicate soft tissue dissection of the palatal structures and provide an adequate access to the tumoral borders. Intraoperatively a mass arising from the nasopharyngeal area was observed protruding from behind the soft palate and resting on the base of the tongue. The orotracheal tube was fixed at the midline of the lower lip and a Dingman retractor was placed to access the oral cavity. A dilute solution of adrenaline (1/200000) was infiltrated into the palate. The soft palate was excised in the midline to expose the tumor. Unfortunately, this was not enough to access the base of the tumor. Consequently, the intra-oral incision of the soft palate was continued anteriorly, up to the half of the hard palate, exposing the palatal shelves and posterior nasal spine (PNS). The PNS and part of the posterior palatal bone was then removed using a round diamond bur up to the vomer. The posterior part of the vomer was partially removed by a roungeur. At this moment, the da Vinci robot was installed. The robot-guided instruments were used to resect the base of the tumor posterior toits attachment to the

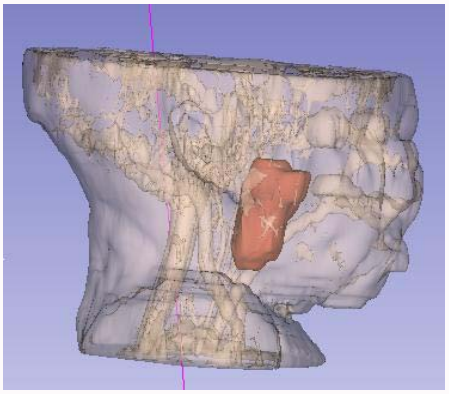

Figure 2: 3D reconstruction of magnetic resonance imaging showing nasopharyngeal mass extending to the oropharyngeal cavity.

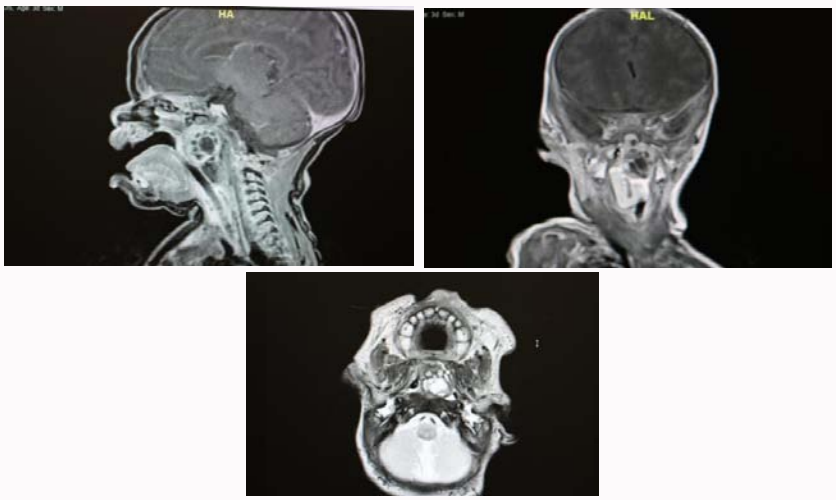

Figure 1: Magnetic resonance imaging showing nasopharyngeal mass extending to the oropharyngeal cavity**.** A) Sagittal T2-weight shows both fatty and multi-cystic components. Adhesion to the vomer and skull base is suspected. B) Coronal T2-weight view shows extension to the oral cavity. C) Axial T2 weight view.

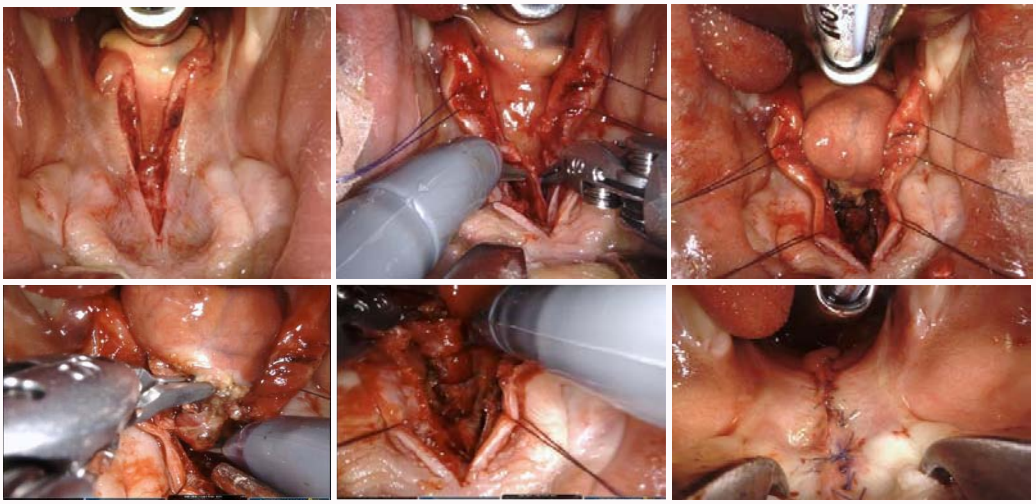

Figure 3: Intra-oral view using da Vinci robot to access and remove teratoma**.** A) Transpalatal incision and dissection of levator veli palatini to access the nasopharyngeal base of the tumor. B) Dissection of the adhesion at the os vomer. C-D) Visualization of the neck of the teratoma and removal of the cystic component. E) Further dissection of the attachment to the skull base was performed assisted by the magnified view from the da Vinci robot. F) Adequate closure of nasal mucosa, restitution of the levator veli palatini muscle and the oral mucosa could be performed due to detailed prior dissection.

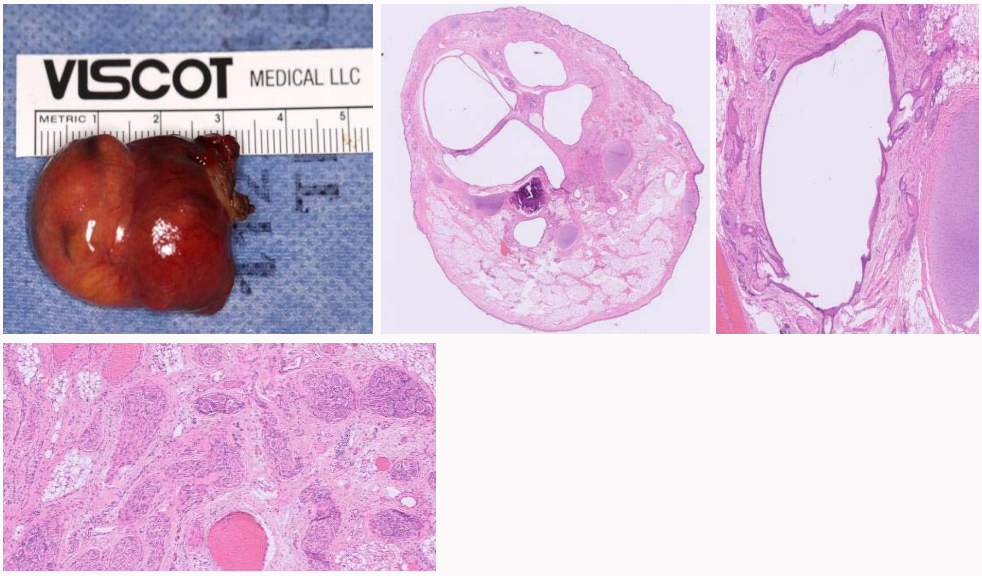

Figure 4: Histological properties of teratoma. A) Macroscopic visualization of the teratoma with a cranio-caudal length of 31 mm. B) Hematoxylin and Eosin (HE, x2 mm). Overview of the tumor with partially cystic component, cartilage and bone. C) HE (x200 µm). The cystic component has a well differentiated squamous epithelium with adnexal structures (hair follicles, sebaceous glands). D) HE (x100 µm). Small to medium-sized nerve bundles, some with ganglion cells, in a background of fat and blood vessels.

vomer and the skull base. Subsequently, the remaining part of the tumor was peeled off its attachment to the skull base using the Da Vinci robotic system (Figure 3A-3F). After complete removal, the mass was sent for histopathological examination. The soft palate was then reconstructed in layers. The surgical procedure took 2 hours and 20 minutes. The infant was transferred back to the NICU for postoperative management and could be extubated on the second postoperative day. He was discharged from the hospital with nasogastric tube feeding for another two weeks. Follow-up in the outpatient clinic, four weeks after surgery showed a small fistula of the soft palate without clinical implications. This fistula was no longer present by the age of three months.

Histology

Histological examination revealed a partial multilocular cystic structure which consisted of a pseudo multi-layered cylindrical ciliated epithelium covering the wall of the cyst and transitioning locally to a squamous epithelium. Focal presence of cartilage and calcifications was found, surrounded by fatty tissue, vascular tissue, nerve bundles and glial tissue. Based upon these histopathological features, a diagnosis of a mature mixed teratoma was established (Figure 4). No atypical cytonuclear or cellular structures were found. Examination of the surgical margins confirmed a complete removal of the mass.

Discussion

To the best of our knowledge, we present the first case in literature of a pre-term new-born with a nasopharyngeal teratoma, which was undetected on prenatal ultrasound, successfully operated using the da Vinci robot before the age of 1 week. Neonatal head and neck

teratomas are a very rare clinical entity with only a few hundred cases reported in literature. The largest series on head and neck teratomas was published by Brodsky et al. [6]. These masses most commonly arise from the midline or lateral nasopharyngeal wall and may result in life threatening upper airway obstruction [7,8]. Several cases reported associated abnormalities including palatal clefts, cardiac problems, microcephaly and atresia of the common carotid [7]. In the case presented, no associated abnormalities were detected and SNP micro-array was normal. Modern imaging techniques such as high-resolution ultrasound and MRI and plays a crucial role in early and prenatal detection of these lesions. The presence of intralesional calcifications can help to differentiate from other nasopharyngeal masses including intranasal glioma, meningoencephalocele, encephalocele, congenital rhabdomyosarcoma, hemangioma, neurofibromatosis and lymphatic malformation [6,7]. Imaging also plays a key role in treatment decision making. Magnetic resonance imaging may allow to investigate the relationship of the mass with adjacent vascular structures and can provide information on intracranial extension [9]. In approximately 80% of the cases, teratomas are benign lesions and congenital teratomas are usual mature in nature. However, rare cases of malignant variants are described and occur most frequently in adults and children. The malignancy cannot be equated with the degree of immaturity of the tissue elements [8]. Some isolated cases of malignant head and neck variants have also been described in neonates [9,10]. Severe upper airway obstruction caused by the teratoma may result in perinatal death. This is especially the case when the lesion has not been detected prenatally. Prenatal diagnosis therefore plays an important prognostic factor in survival. In cases where a difficult airway is anticipated, an EXIT procedure should be considered and timely referral to a tertiary center is recommended [6]. The EXIT procedure consists of a hysterotomy, delivery of the neonate and immediate orotracheal intubation while uteroplacental perfusion is maintained. Whereas surgical resection is the treatment of choice, complete removal of nasopharyngeal teratomas may be difficult to accomplish. Parajuli et al. [11] described a transoral technique assisted by nasal endoscopy to remove a similar lesion, but encountered difficulties in excising all the margins near the ostium of the Eustachian tube as well as pharyngeal extension. At our center, we have an eight-year experience in robotic assisted palatoplasty (TORCS). We therefore assumed that the best exposure and an adequate range of motion could be achieved using the Dingman mouth gag in combination with the 3-dimensional 30° endoscope and 8 mm instruments for working behind the soft palate deep into the nasopharynx and the skull base. The 30° angled high magnification 3-dimensional camera optics permitted optimal visualization of all anatomic landmarks, which facilitated the careful identification and dissection of the base of the tumor. The high-resolution 3-D imaging compatibility of the robotic system capitalizes on the importance of visual cues and provides excellent compensation for the lack of touch [12]. To conclude, transpalatal access and the use of angled instruments and high-resolution visualization with a da Vinci robot allowed for a complete removal of a life threatening nasopharyngeal teratoma in a five-day old infant. The preoperative and postoperative course was uneventful.

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