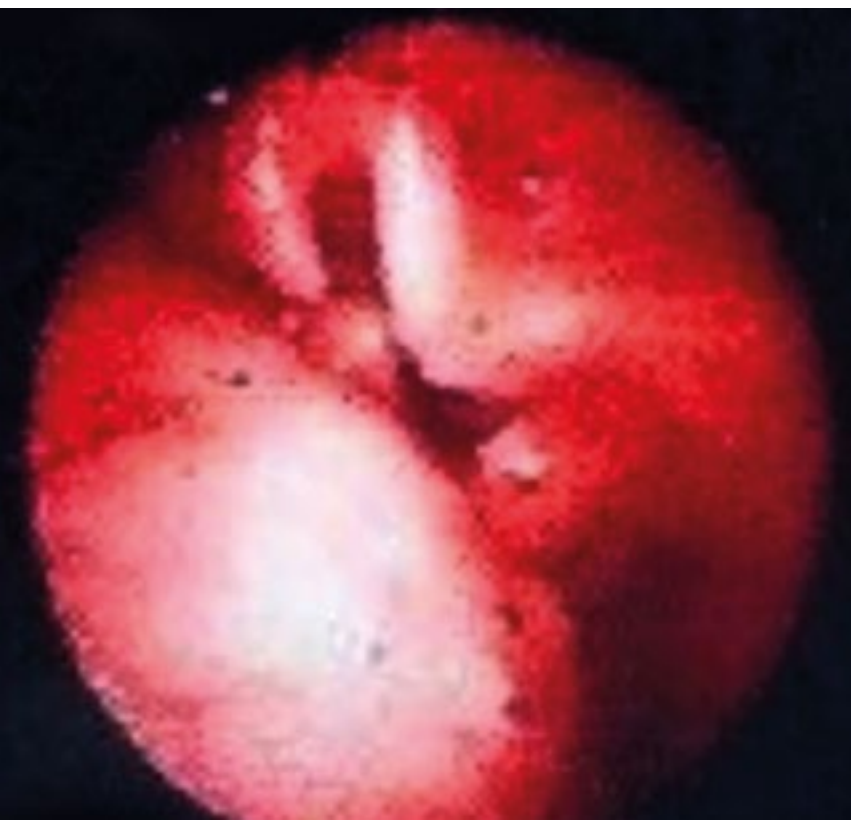


SWISS SOCIETY OF NEONATOLOGY

Winner of the Case of the Year Award 2003

Severe upper airway
obstruction caused by a
congenital cervical tumor –
management and differential
diagnosis

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A male infant was delivered at term after a pregnancy complicated by increasing polyhydramnios first noticed at 32 weeks of gestation. Shortly after delivery, the infant developed respiratory distress, in- and expiratory stridor and pronounced salivation. Several attempts were necessary to pass a nasogastric tube into the stomach, thus ruling out esophageal atresia. Respiratory symptoms were markedly relieved by the application of nasal CPAP (Fig. 1).

When a first bolus feeding lead to immediate aspiration, a flexible laryngo-bronchoscopy was performed. A large pharyngeal tumor obstructing the upper airway was demonstrated. Application of PEEP during laryngo-bronchoscopy stabilized the upper airway sufficiently (Fig. 2).

Ultrasonography followed by MRI including MR angiography showed a solid retropharyngeal and prevertebral cervical mass which was neither connected to the thyroid gland nor did it extend into the spinal canal (Fig. 3-5). The tumor was highly vascularized with calcifications. Urinary secretion of catecholamine-metabolites (VMA, HVMA) was slightly elevated, additional tumor markers (AFP,b-HCG, NSE) and tumor lysis markers were negative.

At surgery a non-infiltrating solid tumor was completely removed. Primary suspicion of a cervical neuroblastoma was confirmed by histology. Molecular genetic analyses

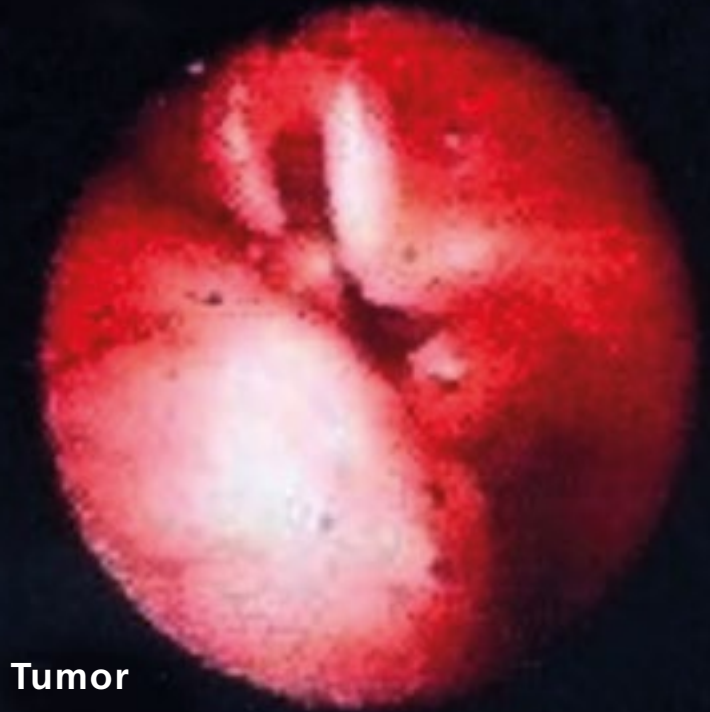
ruled out MYCN oncogene amplification; tumor aneuploidy was demonstrated by DNA flow cytometry. Tumor staging by MIBG scintigraphy and bone marrow examination ruled out disseminated disease, thus defining stage 1 according to the INSS classification (1).



Fig. 1

Patient supported non-invasively on nasal prong CPAP.

Epiglottis



Tumor

Fig. 2

Upper airway endoscopy.

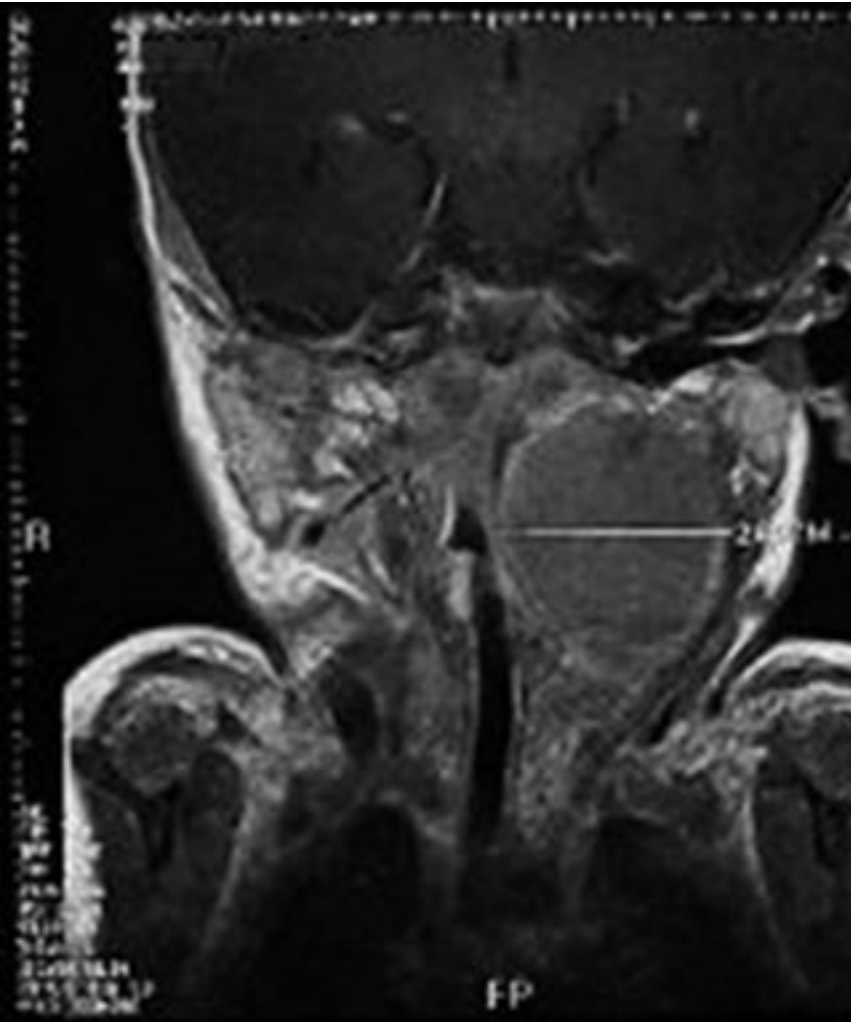


Fig. 3

MRI of head and neck region (coronal view).



Fig. 4

MRI of head and neck region (sagittal view).



Fig. 5

MRI of head and neck region (horizontal view).

DISCUSSION

Congenital cervical masses are rare entities. The differential diagnosis includes vascular tumors, cystic hygromas, branchial cysts, cervical neural tube defects, as well as solid tumors, such as teratomas, neuroblastomas, rhabdoid tumors, or sarcomas. Life-threatening obstruction of the upper airway can occur. When lesions are diagnosed prenatally, a multidisciplinary approach at birth is mandatory. Depending on size or location, cervical masses can lead to diagnostic pitfalls as their clinical symptoms may resemble those of esophageal atresia. Splinting of the upper airway by application of CPAP followed by immediate non-invasive diagnostic procedures might help avoid hazardous maneuvers of more invasive airway management (2).

The retropharynx is a rare location of congenital neuroblastoma (3-5). The prognosis of localized congenital disease is excellent with a disease-free survival of > 95% of affected patients even after incomplete removal of the primary tumor (6,7). Therefore, a radical surgical approach is only indicated when the upper airway is compromised. Chemotherapy is either restricted to disseminated disease or to tumors with non-favorable biological characteristics since spontaneous tumor regression due to cellular differentiation tends to occur during the first year of life in the vast majority of cases.

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