



## Embryonal rhabdomyosarcoma of the tongue in pediatric patient: a case report

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## Tongue rhabdomyosarcoma in pediatric patients: Background

- In children, 35% of rhabdomyosarcomas (RMS) located in the head and neck
- Etiology unknown, however, observed risk factors: families with low socioeconomic status,
   exposure to radiation in utero, maternal use of recreational drugs
- RMS of the tongue
  - Extremely rare cases
  - Most common type embryonal
  - Clinical features of RMSs vary according to the site and stage
  - Generally, a painless mass without associated symptoms is the main presentation of tongue RMSs



Cureus. 2022 Oct; 14(10): e30820.

Published online 2022 Oct 29. doi: 10.7759/cureus.30820

PMCID: PMC9621720

PMID: 36337785

#### Epithelioid Type Tongue Rhabdomyosarcoma in a Pediatric Patient: A Case Report With Literature Review

Monitoring Editor: Alexander Muacevic and John R Adler

Baraa I Awad, 12,3 Dania A Kouther, 1,2 and Mohammed Al-Garni 1,2,3

- 8-year-old patient
- Resection of the tongue with left neck dissection from level I to level III lymph nodes
- Primary closure of the defect
- Adjuvant chemotherapy and radiotherapy



Review > Pediatr Hematol Oncol. 2011 Feb;28(1):60-4. doi: 10.3109/08880011003749678.

Epub 2010 May 14.

### Rhabdomyosarcoma of the tongue: report of a case and review of the literature

Rejin Kebudi <sup>1</sup>, Gül Nihal Ozdemir

Affiliations + expand

PMID: 20469976 DOI: 10.3109/08880011003749678

- 2-year-old boy
- Surgery (tumor resection with primary closure of the defect) and chemotherapy





#### Pathology - Research and Practice

Volume 200, Issues 7-8, 20 August 2004, Pages 537-543



TEACHING CASE

# Spindle cell rhabdomyosarcoma of the tongue in an infant: a case report with emphasis on differential diagnosis of childhood spindle cell lesions

Anita Gupta b, John Maddalozzo a, Thanda Win Htin b, Anil Shah a, Pauline M Chou b

- Congenital lesion of right side of the tongue and floor of the mouth
- 3-day-old neonate girl
- Surgery and chemotherapy



#### **Ewing sarcoma of the tongue**



Oral Oncology Volume 128, May 2022, 105818



Primary Ewing's sarcoma (ES)/peripheral primitive neuroectodermal tumor (pPNET) of the tongue in a child: A case report

Yuanyuan Wu a, Aihong Mao b, Jun Wang a 🙎 🖂

Braz J Otorhinolaryngol. 2020 Dec; 86(Suppl 1): 26–29. Published online 2017 May 2. doi: 10.1016/j.bjorl.2017.04.001

Trans-oral robotic surgery for a Ewing's sarcoma of tongue in a pediatric patient: a

case report

Cirurgia robótica transoral para sarcoma de Ewing da língua em um paciente pediátrico: relato de caso

Frank Rikki Canevari, a Filippo Montevecchi, b Stefania Galla, a Raffaele Sorrentino, a Claudio Vicini, b and Federico Sireci<sup>c,\*</sup>



PMCID: PMC9422648

PMID: 28571929



#### Patient information: Demographic data

- Male
- Czech nationality
- Born 27<sup>th</sup> February 2021
- Parents: Mother hypothyreosisFather healthy
- Gravidity: maternal diabetes, childbirth in 39<sup>th</sup> week, cloudy amniotic fluid, normal adaptation



#### Symptoms and medical history

- Congenital asymmetry of the tongue, no other symptoms
- No comorbidities, normal evolution
- First examination (ENT) 03/2021, MRI: evaluated and treated as a hemangioma
- Therapy:
  - Propranolol (10/2021 05/2022)
- MRI 10/2022: progression of the size
- Biopsy 11/2022 (ENT): embryonal RMS

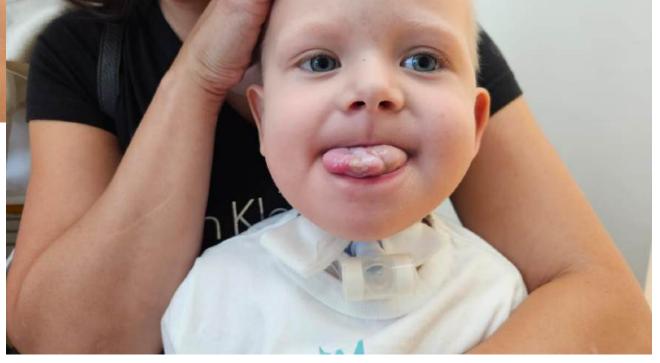


#### **Treatment: ENT and oncology**

- 11/2023 bone marrow aspiration, tracheostomy, venous port insertion
- 11/2023 MRI susp.metastatic process in right lung
- Since 11/2023 chemotherapy
  - 7 blocks of IVA (ifosfamide, vincristine, actinomycin) 11/2023 04/2023, then local progression of tumor
    - Infectious complication (dermatitis, enterocolitis, RS virus, influenza)
  - 2 blocks of VIrT (vincristine, irinotecan, temozolomid) 05 06/2023
  - 2 blocks of Topotecan/Carboplatin and Doxorubicin/Carboplatin









#### **Surgical treatment**

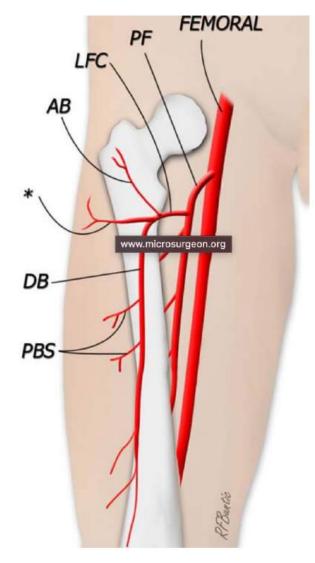
- 24<sup>th</sup> of July 2023 resection of the tumor and primary reconstruction with the ALT flap
- ENT: lymph nodes extirpation, preparation of the vessels VJI and ACC and branches,
   resection of the tumor (transorally) tumor exofytic, bounded, resectable
- Plastic surgery: reconstruction of the defect with ALT flap consisted of skin and fascia,
   6x5cm, 2 perforators (according to pre-operative duplex sonography), pedicle length 5cm,
   end to end anastomosis with superior thyreoid vessels, periphery of the flap thinned to Scarp fascia, modeling of the flap
- Duration: 8 hours



#### Antero-lateral thigh (ALT) flap

- Tissue: A skin, fat and fascia flap it can be thinned free of fascia. Or the skin and fat can be removed to make it a thin fascial flap. It can be made up to 8 x 25 centimeters, or larger if the donor area is grafted.
- Innervation: Yes lateral femoral cutaneous nerve of the thigh.
- Blood supply: Descending branch of lateral femoral circumflex artery.
- Artery: 1.5 to 2.5 millimeters
- Vein(s): Slightly larger than artery when taken to the origin.
- Pedicle length: Up to 7 centimeters or longer, depending on how the flap is designed and where the perforator(s) enter the flap.



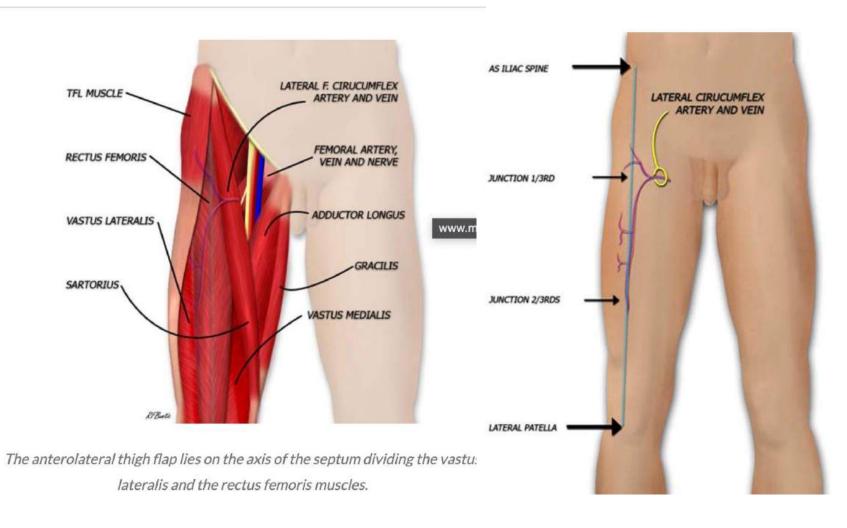


Vascular anatomy of the lateral thigh. The ALT flap is nourished by perforating branches (PBS) from the descending branch (DB) of the lateral femoral circumflex (LFC) vessels.

(PF) profunda femoral

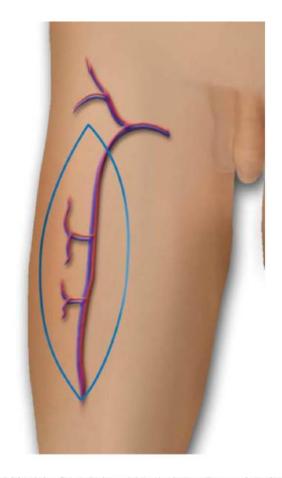
(AB) ascending branch

(\*\*) perforator through TFL muscle to skin.

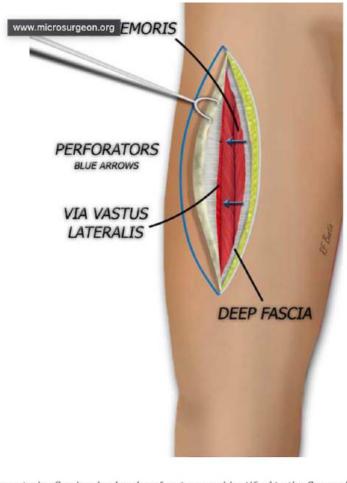


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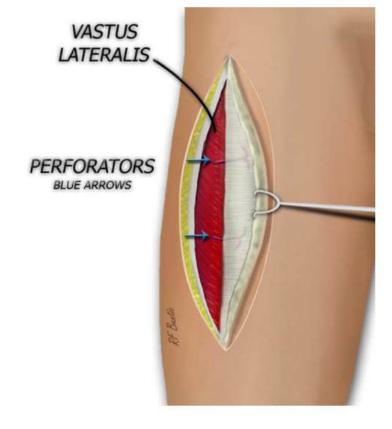




The maximum width of the flap is judge with a pinch test. Donor sites that can not be primarily closed are skin grafted.



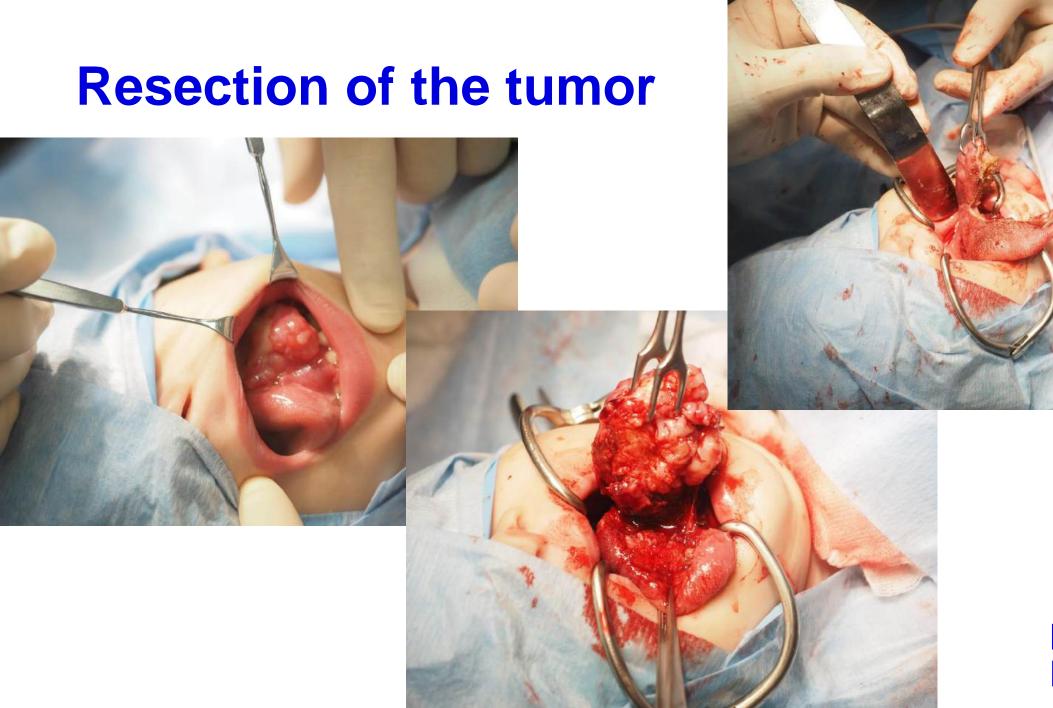
The anterior flap is raised and perforators are identified to the flap and spared.



The posterior flap is elevated and the perforators to the flap are surrounded. The dominant perforator(s) is chosen.

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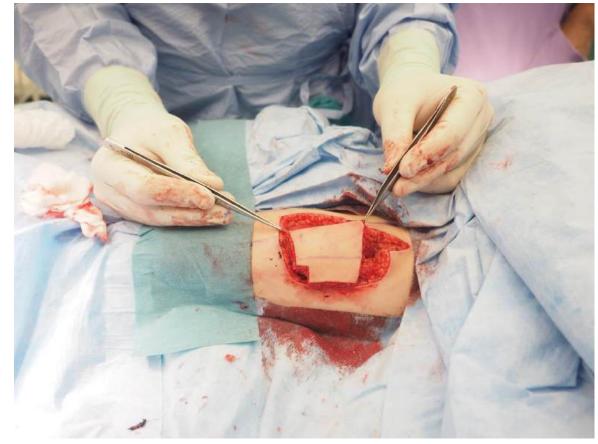






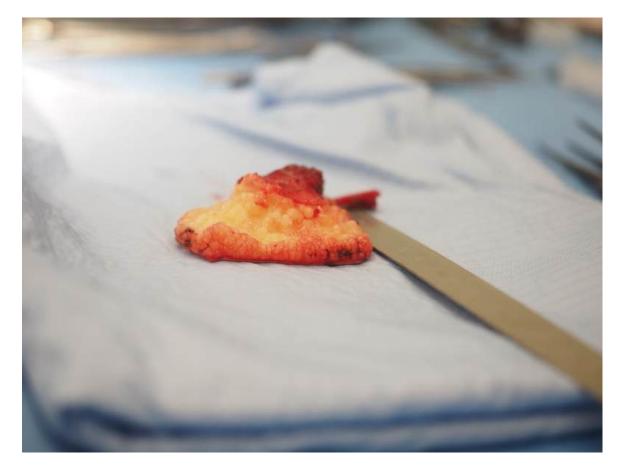
#### Reconstruction with the ALT flap

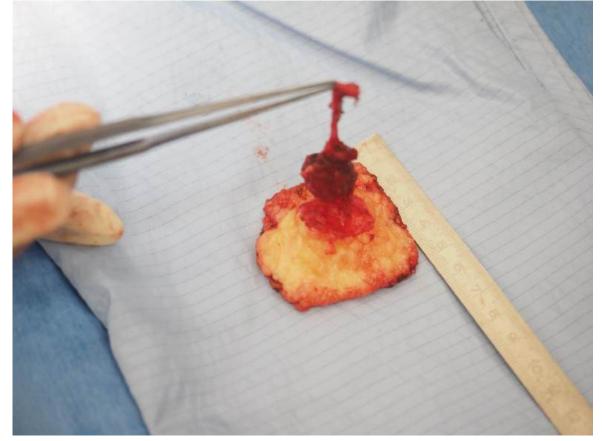






#### Reconstruction with the ALT flap







#### The result





#### Postoperative recovery

- Observation at the Department of pediatric anesthesiology and intensive care
   1 week
- Heparin 1000 IU continually 24 hours
- Antibiotics
- Antiedematous therapy (dexamethasone)
- Complication: dehiscence of the suture on the apex of the tongue due to rupture of stitch, resuture in analgosedation



#### Follow up and outcomes

- Continuation with chemotherapy
- Control MRI 18<sup>th</sup> February 2024 no residual or recurrent tumor
- Plan to remove tracheostomy, PEG and central venous catheter
- No other complications with the flap



#### **Conclusions**

- First case of the rhabdomyosarcoma of the tongue treated with tumor resection and reconstruction of the defect with ALT flap
- ALT flap is a perfect option for reconstruction in the oral cavity in pediatric patients

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#### Thank you for your attention!



