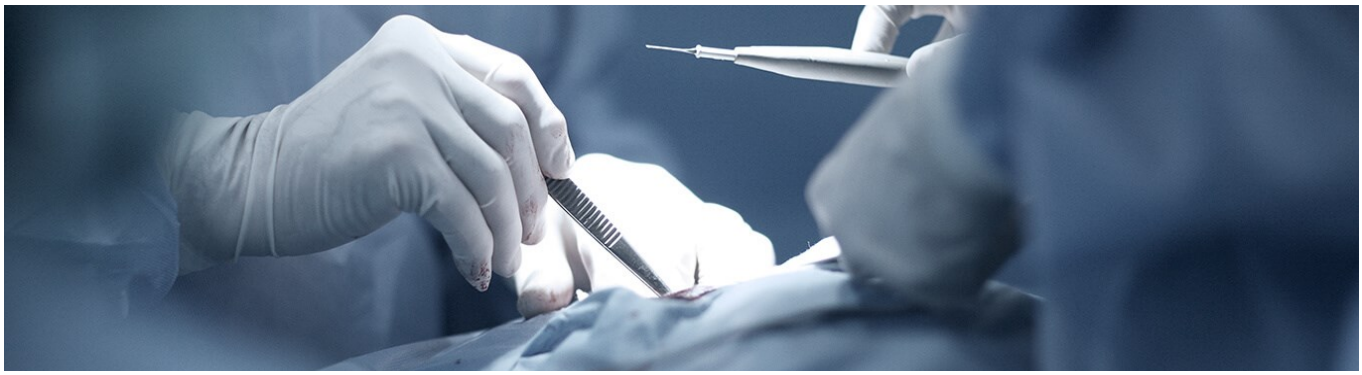
Transmandibulotomy and Transzygoma approach to Rhabdomyosarcoma of the Pterygo-Maxillary space and Pterygoid Fossa: A case report



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Name of the Author (s):

Rahimov Ch.R.[1],
Davudov M.M.[2],
Farzaliyev I.M.[3]
Musaev J.S. [4]

[1] *Professor ,Azerbaiijan Medical Uuniversity, Bakikhanov,23, Baku,Azerbaijan.*

[2,3,4] *Assistant Professor., Azerbaiijan Medical Uuniversity, Bakikhanov,23, Baku, Azerbaijan.*

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INTRODUCTION

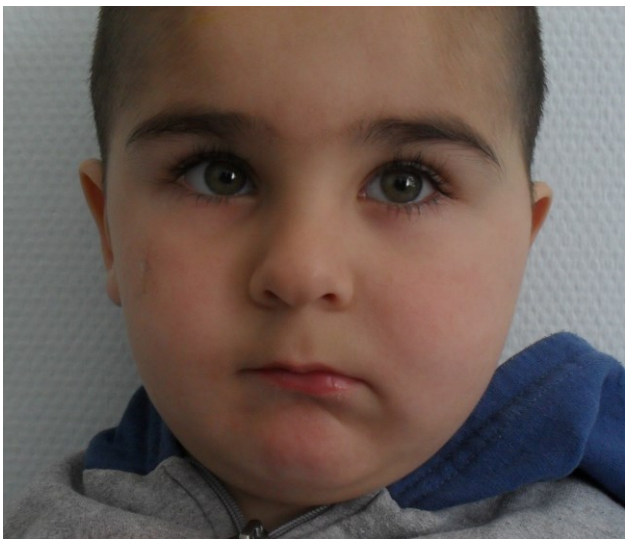
Rhabdomyosarcoma of the head and neck region is a common tumor affecting pediatric population, although in it relatively rare in adults [5]. Statically rhabdomyosarcoma occurs in 84% of cases of all soft tissue sarcomas and in 35-45% of those occurring in the head and neck.

The aim of this article is to describe the clinical case of pleomorphic rhabdomyosarcoma of the child, which was resected by extended transmandibulotomy and transzygoma approach.

I. CASE REPORT.

In September 2011 a 3 years old boy has been admitted to our clinic with complains on dull pain in the region of the left parotid area, weakness and loss of the appetite. His parents report that certain pathology has 20 days history, and were recognized as painless swelling in the region of left parotid area, which prone to enlarge. They also noted that tumor shortly was spread to the oral cavity. No therapy was performed. During clinical investigation painless tumor in the region of left check and zygoma arc occurred. Tumor has smooth surface with clear boundaries. No limitation in opening of the mouth and difficulties in swallowing were noted.

Patient's MRI scan showed tumor to be present in pterygomandibular region with spreading into parapharingeal space (Fig 1,2).



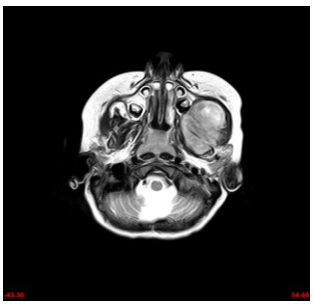


Fig 1. Axial MRI corssection of the tumor

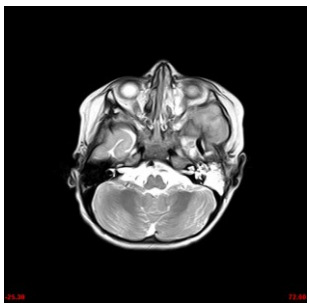


Fig 2. Axial MRI corssection of the tumor

Mandibulotomy approach was chosen as surgical approach. Tumor was resected via this approach with ligation of the external carotid artery prior to resection, in order to prevent massive bleeding while resection (Fig 3,4). Approach was completed with papillae-preserving incision on lingual aspect of the mandible on affected site. Further dissection and elevation of divided mandible helps to visualize inferior compound of the tumor. Tumor was found as mass with a thin well defined capsule. In order to promote complete surgical resection of the tumor in a free margin fashion additional segmental resection of inferior part of zygoma was performed. It makes more complete approach to tumor. The mass was resected within healthy tissue with creating a space in the region of infratemporal and pterygo-maxillary fossa. After resection separated bone fragments was fixed by appropriate 2.0 titanium mini-plates. The mucosa was sutured by vertical matrass sutures, what allowed mucosa to be sutured more atraumatically.

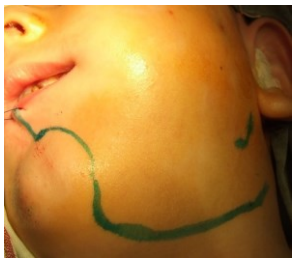


Fig 3. Preoperative marking and carotid flap design.

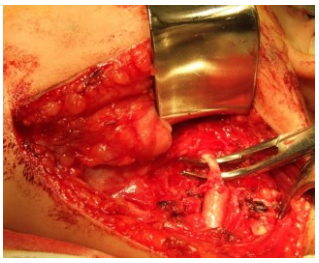


Fig 4.Ligation of external artery.

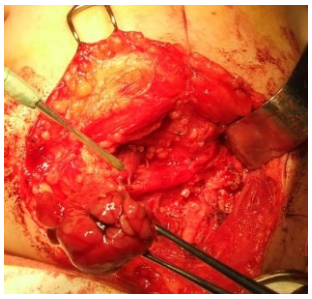


Fig 5. Selective neck dissection.

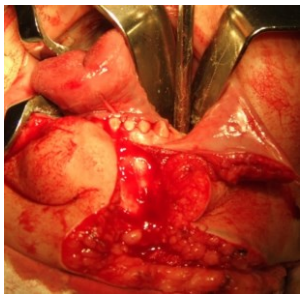


Fig 6. Creation of mandibular-check flap and approaching to the oral cavity.

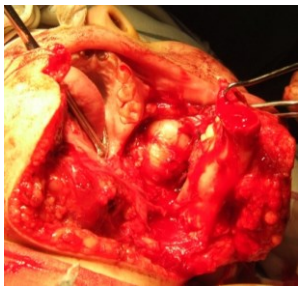


Fig 7. Approaching to the lower pole of the tumor.

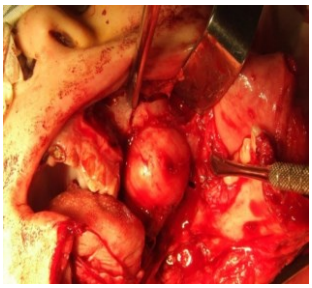


Fig 8. The tumor is completely visualized.

On postoperative follow-up the course seems to be uncomplicated. The wound healed by primary intension healing with no orocutaneous fustule formation.

Histology showed tumor to be primary rhabdomyosarcoma of pterygo-maxillary space. Patient was sent to postoperative chemotherapy course.

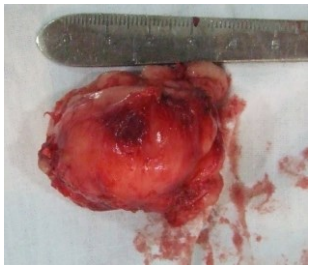


Fig 9. Resected specimen.

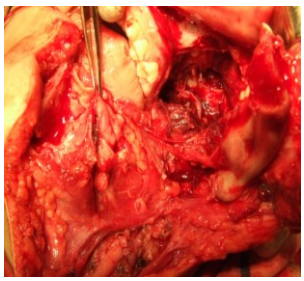


Fig 10. Post-resective site.

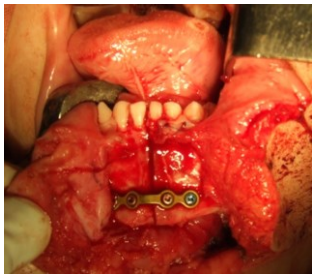


Fig 11. Fixation of the mandible titanium miniplate

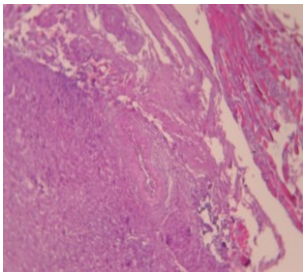


Fig 12. Microscopic view of the by tumor (HE, x100).

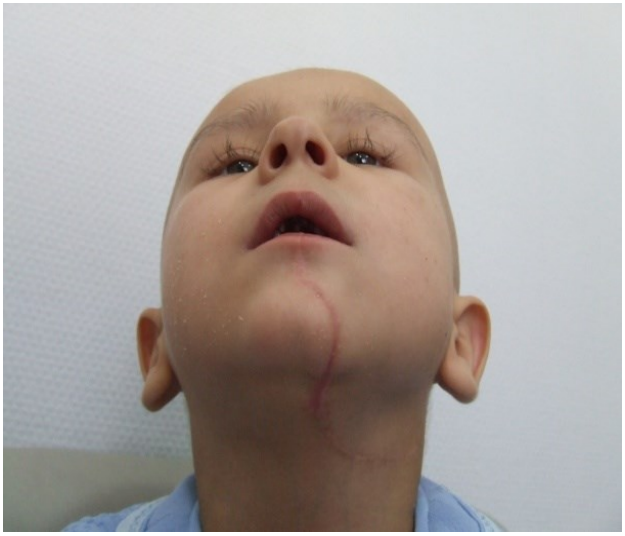


Fig 14. Postoperative view of patient.(6 month after operation and adjuvant chemotherapy)

Two years in 2013 of postoperative follow-up revealed no recurrence occurring within resected site. No violation of mouth opening, mastication and swallowing functions were detected. The function of orbicularis oris wasn't disturbed. The postoperative scar seems to be reasonable from esthetic point of view.

III. DISCUSSION

Rhabdomyosarcoma (RMS) is a malignant tumor that is composed of tissue that exhibits striated muscle differentiation. RMS was first described by Weber in 1854, and accounts for 6% of all malignancies in children under 15 years of age [6]. Peak age for occurrence is between two and six years [11]. It is the most common head and neck soft tissue sarcoma in pediatric patients. Other localization of one could be genitourinary tract, retroperitoneum and, to a lesser extent, the extremities [6].

Rhabdomyosarcomas of the head and neck are divided into two groups according to anatomic, location and propensity for central nervous system invasion [6]:

RMS is the most common pediatric sarcoma, with 35% to 40% arising in the head and neck. Approximately 70% of patients are younger than 12 years of age. In one series, 80% of girls were younger than 4 years and 77% of boys were younger than 5 years of age. Twenty-five percent of head and neck RMS occur in orbit, 50% are parameningeal, and 25% occur at other sites (oral cavity, cheek, parotid, larynx, scalp) [7]. The oral representation of RMS is relatively rare, and the most affected site is the soft palate [8]. Symptoms from the parameningeal tumors are very nonspecific, such as earache, otitis media, stuffy nose, and headache. They are, therefore, frequently diaqnosed late, when skull base invasion and involvement of the cranial nerves and meninges already have occurred.

The RMS is dividing into four types by histological patterns: embryonal, botryoid, alveolar and pleomorphic. The most frequent type is embryonal andit represents more than 70% of all cases [9]. Histologically, embryonal RMS is characterized by a mixture of pleomorphic and skeletal immature muscle cells, the so-called rhabdomyoblasts. These cells have a distinctive eosinphilic-rich cytoplasm and proliferate in a myxoid loose stroma [10].

Histologically, embryonal and botryoid RMS are the most frequent types in the head and neck. These occur in younger children, who have the best proqnosis.alveolar RMS tend to occur in adolescents. The pleomorphic histologic type has the poorest prognosis and occur in adults. Pleomorphic RMS is a most rare type of all RMS of head and neck area. Pleomorphic RMS, a variant most common in adulthood occurs frequently in the extremities and does not involve the head and neck region [13]. In the Interqroup Rhabdomyosarcoma studies (IRS), three related tumors (extraosseous Ewing"s sarcomas, sarcoma of undetermined histology, and undifferentiated sarcoma) also are included and these have very poor survival rates. Prognosis for RMS depends on histologic type, clinical stage, and site. Currently, a site-based TNM staging system is being evaluated agaionts the traditional surgicopatologic clinical grouping system [1].

Table-1 Intergroup Rhabdomyosarcoma Study Tumor Staging

Stage I	Localized disease, completely resected; regional nodes not involved
Stage II A	Grossly resected tumor with microscopic residual disease; no evidence of regional node involvement
Stage II B	Regional disease (extension into adjacent organ or regional nodes); completely resected
Stage II C	Regional disease with involved nodes; grossly resected, but with evidence of microscopic residual disease
Stage III	Gross residual disease after surgery
Stage IV	Metastatic disease at diagnosis

Treatment protocols include multiple modalities: resection, radiation, chemotherapy. Before the IRS, survival with single-modality therapy was 8% to 20%. In IRS-I (trial number one), 3-year relapse-free survival was 91% for orbital, 75% for "other sites", and 46% for parameningeal RMS. If complete resection of the tumor is possible, this gives the best overall survival whencombined with chemotherapy. In parameningeal sites, however, complete resection is rarely possible due to functional and esthetic considerations [2].

Lyos et al. reported 56 rhabdomyosarcomas of the head and neck in children. Complete resection was possible in only three (5.4%) patients, and incomplete resection with microscopic residual disease occurred in 13 (23.2%) patients. Forty patients (71.4%) had grossly positive margins, most having only incisional biopsy. Five-year disease-specific survival was 63% using protocols incorporating radiation and chemotherapy. However, the prognosis for patients with reccurent or metastatic disease was dreadful, with 95% mortality [3,4].

The concept of approaching the retromaxillary area through a mandibulotomy is not new and has been advocated by Conley and Barbosa (11,12). According to Butlin And Spencer, the original concept of division of the mandible using a vertical midline section was first developed by Roux who performed it in 1836 or 1839 [8]. Later in 1844 a similar technique was proposed by Sedillot, but he described a dovetail-shaped cut to add stability to the osteotomy site. This was followed in 1857, 1858 and 1865 by James Syme who described an angular cut to divide the lower jaw. Fiddes (1859) reported his experience with sectioning the mandible in the symphyseal region for the removal of carcinoma of the tongue. In 1861 Billroth suggested a temporary mandibular section for oral tumor ablation. According to Spencer & Cade Kocher (1880, 1902) described his experience with this approach. Wilfred Trotter in 1929 used the mandibular midline split as part of an anterior translingual pharyngotomy for removal of lesions at the base of the tongue, epiglottis and associated glossoepiglottic fold. Sectioning the mandible just anterior to the attachment of the masseter muscle had already received mention by Trotter in 1913 and 1920 .

In 1959 Perzik 22 reported sagittal osteotomy of the mandible at the symphysis together with a median glossotomy to remove an obstructing retropharyngeal tumor. Martin et al. In 1961 reintroduced Trotter's median anterior translingual pharyngotomy and called it median labiomandibular glossotomy.

The infratemporal fossa communicates inferiorly with the neck. If the mandible is laterally retracted and the medial pterygoid muscle is detached from its mandibular attachment the infratemporal space can be reached. This approach provides good control of the vessels and nerves and *en bloc* resection of nasopharynx, posterior maxilla, infratemporal fossa structures, mandibular ramus and parotid gland can be performed. The procedure has been modified by Attia *et al.* to obtain wide field exposure without sacrifice of either mandibular function or the sensory supply of the face and oral cavity.

The concept of access to the infratemporal fossa via an osteotomy of the zygomatic arch has been described in the past by Barbosa, Crockett, Conley, and Samy and Girgis. The basic techniques since have remained the same. There has been renewed interest in this approach by various surgical disciplines to use this as a combined procedure in order to obtain optimal access and visualisation to achieve *en bloc* excision.

IV. CONCLUSION

Rhabdomyosarcomas account for one of the most malignant maxillofacial tumors in children. Management frequently involves multimodality therapy with radiation and chemotherapy in addition to resection with effective approach for possible *en bloc* radical excision of tumours,. Although this strategy has led to increased survival, this has come at the cost of increased morbidity, with facial growth deformities and induced second primary malignancies.

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