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# Postoperative Intensity Modulated Radiotherapy for Acinic Cell Parotid Carcinoma in Childhood- A Clinical Case from our Practice with Literature Review

**Lena Marinova*, Katia Sergieva, Vaska Vasileva and Nikolay Kahchiev**

Medical Oncology Clinic, Department of Radiation Oncology and Metabolic Brachytherapy, UMHAT "Queen Joanna" Sofia, Bulgaria

**ABSTRACT**

Malignant parotid tumors in childhood are extremely rarely diagnosed diseases. Due to the rare incidence, there is no consensus on their complex treatment, as well as on the application of postoperative radiation therapy (RT). We present a clinical case of acinic cell parotid carcinoma in a 12-year-old girl, against the background of which we emphasize the optimal complex therapeutic approach. Despite radical facial nerve-sparing parotidectomy in pT2N0 acinic cell parotid carcinoma, due to the two pathohistological negative factors/two foci with vascular tumor invasion and the close proximity of the resection line to the preserved facial nerve, we conducted postoperative adjuvant RT.

Due to the risk of late radiation reactions of normal tissues and organs after RT in childhood, intensity modulated radiation therapy (IMRT) with lower radiation doses in the target volumes was performed. Active surveillance and follow-up imaging studies are required for early diagnosis of possible recurrence or distant metastases.

***Corresponding author**

Lena Marinova, Medical Oncology Clinic, Department of Radiation Oncology and Metabolic Brachytherapy, UMHAT "Queen Joanna" Sofia, Bulgaria.

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## Intraduction

Diseases of the salivary glands are rare in infants and children (with the exception of diseases such as parotitis epidemica and cytomegaly) and the therapeutic regimen differs from that in adults [1]. Salivary gland tumors are uncommon in children and adolescents and represent only 0.5% of pediatric malignancies [2]. According to Ussmüller et al. about one half of all juvenile salivary gland tumors may be malignant tumors [3}. They are characterized by diverse histological appearances and variable biological behavior [4]. Due to the epithelial and the non-epithelial histology of the affected organ, many histological types of parotid tumors are possible, although some are rare [5]. They are even rarer in childhood, with less than 5% of these tumors reported in the pediatric population [6]. Due to the rarity of salivary gland malignancies in the pediatric population and lack of large single institution clinical studies, it is challenging to develop a consensus on treatment [2]. In this article, we present an extremely rare childhood acinic cell parotid carcinoma after radical surgery/ parotidectomy with preservation of the facial nerve, in which postoperative radiotherapy (RT) should be evaluated and justified.

## Clinical Case

It concerns a 12-year-old girl. According to data from the mother, in the middle of August 2023, a painless swelling of the anterior region on the right was noticed. After consultation with an ear-nose-throat specialist, antibiotic therapy was prescribed without effect. After consultation with Professor Stanimirov / maxillofacial surgeon / a formation of the right parotid gland was established.

## Local Status

Visible facial asymmetry due to swelling in the olast of the right parotid gland in front of the ear. On palpation - a nodule with a dense elastic consistency, mobile, with a surface size of about 3x2 cm. The child reports that he does not feel pain during palpation or in general. The underlying skin is not involved, without erythema. The remaining salivary glands and neck - no masses, no pathologically enlarged lymph nodes on palpation. Oral cavity - without lesions of the premalignant and malignant spectrum.

## X-Ray of Lungs and Heart

Normal lightening of lung halves without X-ray evidence of infiltrative changes and nodular shadows. Unexpanded hilus shadows. Heart shade normal for age.

## MRI From 09/19/23

In the area of the right parotid gland, a solid formation with a size of 29/25 mm, well separated from the capsule, was found. After administration of intravenous contrast, the formation slightly increased its signal, and small fluid collections were well demarcated laterally and toward the base. The large vessels

are intact. The remaining salivary glands are unchanged. Conclusion- Tumor of the right parotid gland.(Figure 1). After biopsy and MRI on 9/27/23, a well-circumscribed solid mass in the right parotid gland measuring 29/25 mm was diagnosed. Surgical treatment is recommended after parental consent. An operation was performed on 20.10.23.

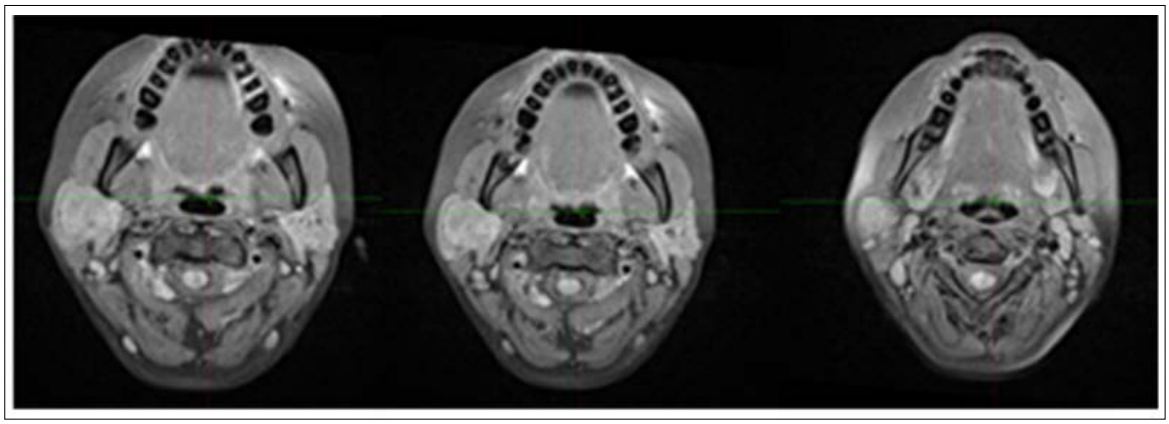

**Figure 1:** MRI from 09/19/23

**Intraoperative**

The tumor is palpated in the deep part of the gland. It develops in the deep part of the gland, filling the retromandibular space and fitting on the masseter muscle. The boundaries of the lesion in depth cannot be assessed by palpation. The posterior border of the dissection is formed in front of and below the ear cartilage. In this place, the bifurcation of the facial nerve is identified in depth. The superior temporofacial branch was traced and its branches appeared uninvolved by the tumor lesion. The lower branch and its ramifications were found to have a greatly thinned and flattened structure, directly lying and adherent to the tumor lying and developing beneath the nerve. The capsule of the tumor is stuck under the branches of the cervicofascial nerve. The nerve was carefully released and his knuckles were preserved. The ramus marginalis mandible appears extremely thin, flattened and literally lies on the pseudocapsule of the tumor mass. It is densely atheromatous and difficult to separate from the tumor wall. In depth, it was found that the tumor pushed the soft tissues and formed a pathological tumor bed. The formation fits on the posterior belly of the digastricus muscle and lies in direct contact on the external carotid artery. A tumor formation of the right parotid gland was removed by the method of parotidectomy with preservation of the facial nerve.

**The Pathohistological Result**

No. Bi 2741/06.11.23 established an acinic cell carcinoma of the right parotid gland with focal infiltration in the parotid parenchyma with two foci of extralesional vascular infiltration. No necrosis, perineural invasion and areas with high-grade morphology are found. In one section, the tumor tissue was less than 1 mm from the periphery of the material. Five lymph nodes with sinus lymphadenitis from the first and second level of the lymph nodes.

**Immunohistochemical Study**

DOG1 - positive reaction in the tumor cells. No metastases were found in the removed five regional lymph nodes.

**Diagnosis**

Acinic cell carcinoma of the parotid gland with focal infiltration in the surrounding parenchyma and focal vascular invasion. – pT2 V1 N0.

The tumor was removed as radically as possible. In connection with the rare diagnosis in childhood, the established pathohistological finding, and apparently two foci of extralesional vascular invasion, the patient is referred for consultation at the pediatric oncohematology clinic to assess the necessary adjuvant postoperative treatment. Due to the two pathohistological negative factors/two foci with vascular tumor invasion and the close proximity of the resection line to the preserved facial nerve, we considered conducting postoperative intensity modulated radiotherapy (IRMT) in the area of the tumor bed up to a total dose of 54 Gy and in the regional lymph nodes up to the third level 50 Gy.

In the area of the right parotid gland, a solid formation with a size of 29/25 mm, well separated from the capsule, was found.

In the Figure 2, we present the preparation for delineating the target volumes after fusion of the MRI image and that of the planning CT. These target volumes are seen in Figure 3. The following Figure 4 presents the IMRT using the VMAT method with two arches and the realized radiation doses in the target volumes CTVp and CTVn, as well as the doses in critical normal tissues and organs.

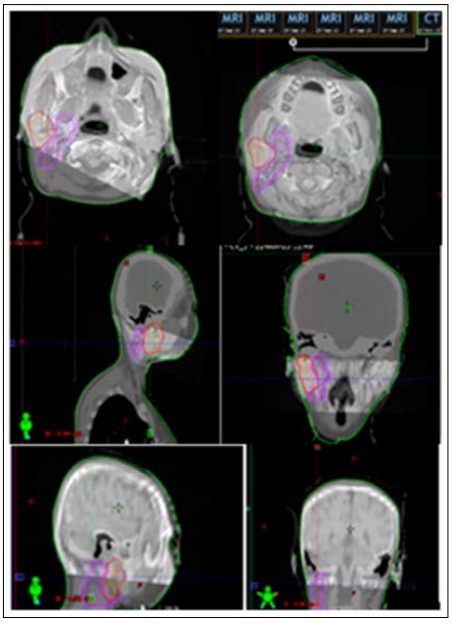


**Figure 2:** Preparation for Delineating the Target Volumes After Fusion of The MRI Image and That of The Planning CT

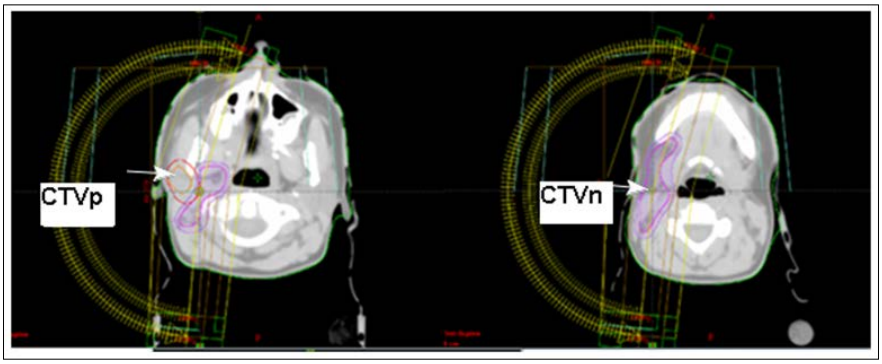


**Figure 3:** Against the Background of Intensity Modulated Radiation Therapy Using the VMAT Method Performed with two Arches, We Present the Target Volumes CTVp and CTVn

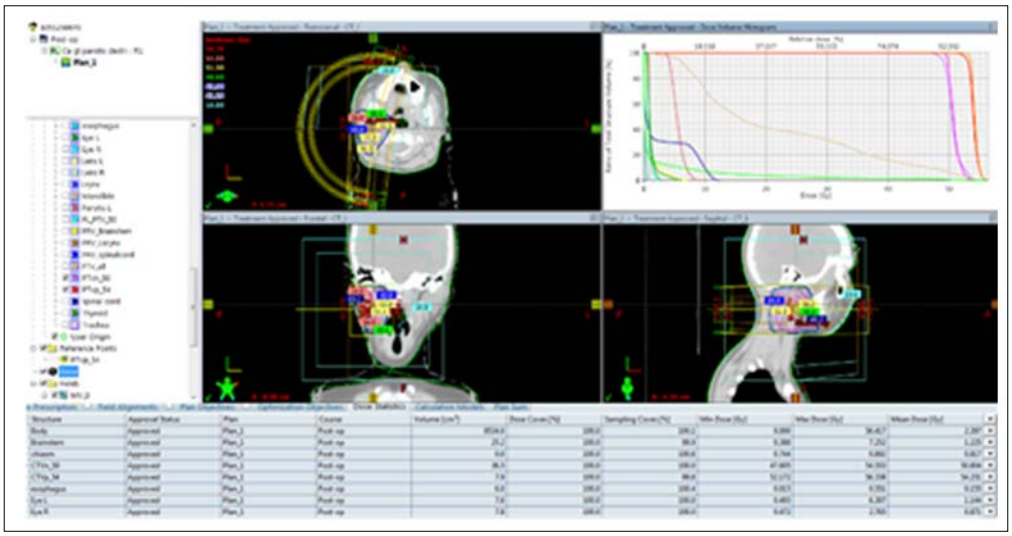

**Figure 4:** IMRT using the VMAT Method with Two Arches and The Realized Radiation Doses in The Target Volumes CTVp and CTVn, as well As The Doses in Critical Normal Tissues and Organs

## Discussion

According to Eneroth C M salivary gland tumors make up 0.3% of all human tumors, and less than 10% of all juvenile head and neck tumors are located in the salivary glands [7]. Approximately 300 cases per year of primary salivary gland malignancy are registered in the United Kingdom, of which fewer than ten occur in children [8]. Acinic cell carcinoma of the head and neck is an uncommon salivary gland cancer, with a reported incidence of less than 1 case per 100 000 population [9]. In childhood, the main group of parotid malignances consists of epithelial tumors, mucoepidermoid carcinoma being the most frequent neoplasm, followed by acinic cell carcinoma, adenoid-cystic carcinoma and adenocarcinoma, together representing > 80% of carcinomas [10-12]. Excretory stem cells give rise to mucoepidermoid and squamous cell carcinomas, while intercalated stem cells can lead to pleomorphic adenomas, adenoid cystic carcinomas, oncocytomas, adenocarcinomas, and acinic cell carcinomas [4].The tumor cell types are numerous, with therapy and prognosis determined by not only the tumor histology, but also location [13].

Computed tomography (CT) or magnetic resonance imaging (MRI) are essential in the evaluation of neoplastic disease, although the child may require sedation or even general anesthesia in order to obtain good images [14]. According to Kashiwagi et al. the imaging aspect seems to depend on histologic characteristics [15]. Low-grade tumors feature a hyperintense area on $T_2$-weighted images, related to abundant mucin secretant cells, and sometimes ill-defined margins, because of peritumoral inflammatory changes. In our clinical case after biopsy and MRI on 9/27/23, a well-circumscribed solid mass in the right parotid gland measuring 29/25 mm was diagnosed (Figure 1). In small, low-grade superficial parotid tumors, a superficial parotidectomy with a margin of at least 1.5 cm may suffice, but otherwise, a total conservative parotidectomy is advised with resection of adjacent neck structures if necessary to achieve an en-bloc resection. A functioning facial nerve should be preserved unless found to be infiltrated with the tumor itself at the time of resection [4]. Surgeons should perform facial nerve preservation in patients with intact preoperative facial nerve function when a dissection plane can be created between the tumor and the nerve [16]. The National Comprehensive Cancer Network guidelines define close margins as less than 5 mm and provide a consensus recommendation for adjuvant radiotherapy in resected salivary gland cancer with close margins [17]. Although margin assessment is subject to several processing and analytic variabilities, including tissue shrinkage and reporting inconsistencies, in resected salivary gland cancer some authors have defined any margin of 1 mm or less as a positive margin [18-20]. Close but negative margins may occur in more than 30% of patients with acinic cell carcinoma of the parotid gland [21]. In the presented clinical case, the tumor was located in the deep part of the parotid gland, surrounded by a pseudocapsule, 1 mm away from the outline of the facial nerve without infiltrating

it. These data give reason to perform a total parotidectomy in which the facial nerve is preserved, an operation that is accepted as applicable for T2 tumors located in the glandular parenchyma of the parotid gland. At the same time, a right-sided dissection of the closest regional cervical lymph nodes of the first and second level was performed. The pathohistological result established an acinic cell carcinoma of the right parotid gland with focal infiltration in the gland parenchyma with two foci of extralesional vascular infiltration. No necrosis, perineural invasion and areas with high-grade morphology are found. In one section, the tumor tissue was less than 1 mm from the periphery of the material. Acinic cell carcinoma of the parotid gland is an extremely rare low-grade malignant neoplasm in childhood [22-24]. In low-risk patients, provided that cancer cells were not detected at the inked resection edge, margins of 1 mm or less were not associated with local recurrence in any patient with or without adjuvant radiotherapy [25]. Postoperative RT should be offered to patients with tumors with the following features: high-grade tumors, positive margins; perineural invasion; lymph node metastases; lymphatic or vascular invasion; and T3-T4 tumors, as well as in patients with tumors with close margins. In postoperative cases, the high-dose target should cover the salivary gland surgical bed and appropriate nodal levels [16]. Stodulski et al. reported on 32 patients with low-grade or intermediate-grade parotid carcinoma with surgical margins ranging from 1 mm to 3 mm treated without adjuvant radiotherapy [20]. In the presence of other histopathologic risk factors, including advanced T category, nodal disease, lymphovascular or perineural invasion, or cancer cells at the inked resection margin, the addition of adjuvant radiotherapy is frequently recommended and may significantly improve locoregional control and survival [26]. In our clinical case the combination of two prognostic negative factors are grounds for postoperative radiotherapy: 1/ From a surgical standpoint - knowing the clinical characteristics of this rare childhood tumor, intraoperatively the tumor is in close proximity to the branches of the facial nerve, which corresponds to a close resection line, given its preservation. 2/ The established two foci of extralesional vascular invasion, to a certain extent, characterize the biology of the tumor, which should be taken into account when evaluating adjuvant treatment/radiotherapy or chemotherapy.. Due to the two pathohistological negative factors, we considered performing postoperative adjuvant RT (Figures 2-4).

## Conclusion

Malignant parotid tumors in childhood are extremely rarely diagnosed diseases. Due to the rare incidence, there is no consensus on their complex treatment, as well as on the application of postoperative RT. The adjuvant postoperative RT should be offered to patients with high-grade tumors, positive margins; perineural invasion; lymph node metastases; lymphatic or vascular invasion; and T3-T4 tumors, as well as in patients with tumors with close margins. Compared to patients in adulthood, due to the risk of late radiation reactions of normal tissues and organs after RT in childhood, intensity modulated radiation therapy (IMRT) with lower radiation doses in the target volumes was performed. Active surveillance and follow-up imaging studies are required for early diagnosis of possible recurrence or distant metastases.

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