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A Rare Case of Parotid Gland Carcinosarcoma

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Abstract
Carcinosarcoma of the parotid gland is considered as a rare true malignant mixed tumor within both the stromal and epithelial components. We report the clinical case of a 37-year-old patient, with no previous history, who is followed up for a tumor of the right parotid revealed by a fast-growing neck mass on regard of the parotid region. After a parotidectomy with a cervical lymph node dissection, we retained the diagnosis of a carcinosarcoma probably developed on pleomorphic adenoma. We then, performed an adjuvant radiotherapy.

Keywords: Parotid Gland, Carcinosarcoma, Radiotherapy, IMRT

1. Introduction
Carcinosarcoma is considered as true malignant mixed tumor that represents less than 2% of all mixed tumors of salivary glands. It is a rare type of neoplasm within both the stromal and epithelial components [1]. It has accounted for only 0.04%-0.2% of all salivary tumors [2].
Carcinosarcomas have been described in several organs, but very few cases of carcinosarcomas of the salivary glands have been reported, the first case has been described by Kirklin in 1951 [3].
We present a new case of parotid gland carcinosarcoma. A review of described cases of salivary gland carcinosarcoma has been presented and treatment discussed.

2. Medical Observation
The patient was 37 years old, father of one child, who had been experiencing for 2 months the apparition of a fast-growing neck mass on regard of the right parotid region. No pain or contralateral cervical abnormality have been identified.
The patient had an MRI that showed a right parotid mass measured in axial section at 59 x 52 mm and extended over a height of 48 mm. This mass was mostly superficial, its deep pole protrudes medially 5 mm beyond the mandibular plane towards the deep part. This lesion presents with a high mixed signal and is decreased in T1 before injection. In T2 it is observed to contain innumerable logettes with a liquid level of hemorrhagic type. The mass includes a nodular component of very diminished signal in T2, 24 mm in diameter, at the level of its lower and lateral pole, recent hemorrhage. On perfusion,we see a vascularization in the tissue area without typical profile of cystadenolymphoma or malignant salivary tumor. No cervical adenomegaly in the field of exploration were seen (Fig 1).
A biopsy had been performed that showed no malignancy. The patient subsequently then had a parotidectomy with a cervical lymph node dissection was indicated. Anatomopathological and immunohistochemical studies concluded that the profile was in accordance with a carcinosarcoma probably developed on pleomorphic adenoma. The resection was incomplete. No nodes of the lymph node resection were positive.
Then we performed a positron emission tomography scan that had shown a fixation in the parotidectomy's bed (Fig 2).
After that the case was discussed in a multidisciplinary consultation meeting and it was decided to perform an adjuvant radiotherapy treatment.
Using intensity modulated radiation therapy technique with integrated boost, it was possible to deliver a total dose of 55.5 Gy on the right parotid lodge as well as the probable microscopic extensions with a prophylactic dose on the homolateral cervical lymph nodes at a rate of 30 fractions of 1.85 Gy, 5 sessions per week, and a boost of up to 69 Gy on the parotid lodge at a rate of 30 fractions of 2.3 Gy, 5 sessions per week (Fig 3). The patient finished recently his radiation treatment, the tolerance was globally correct, he had a cutaneous toxicity and dysphagia. He will be seen for follow-up within one month.

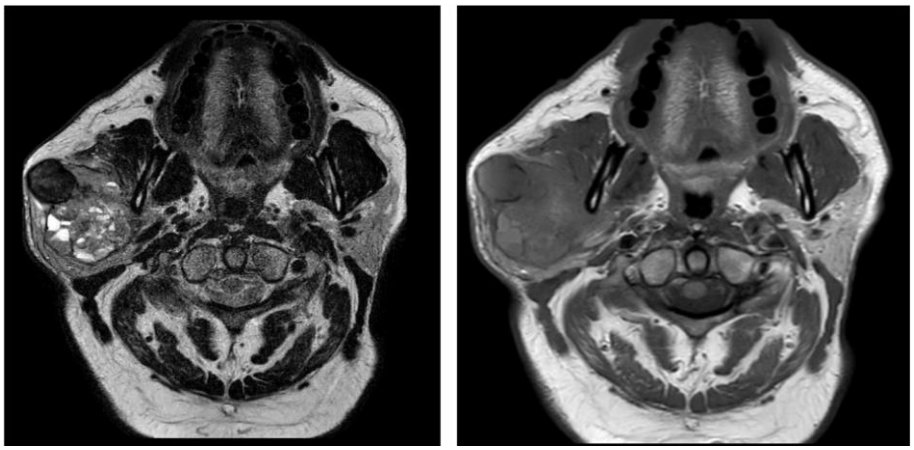

Fig 1: MRI axial view of the initial parotid gland tumor

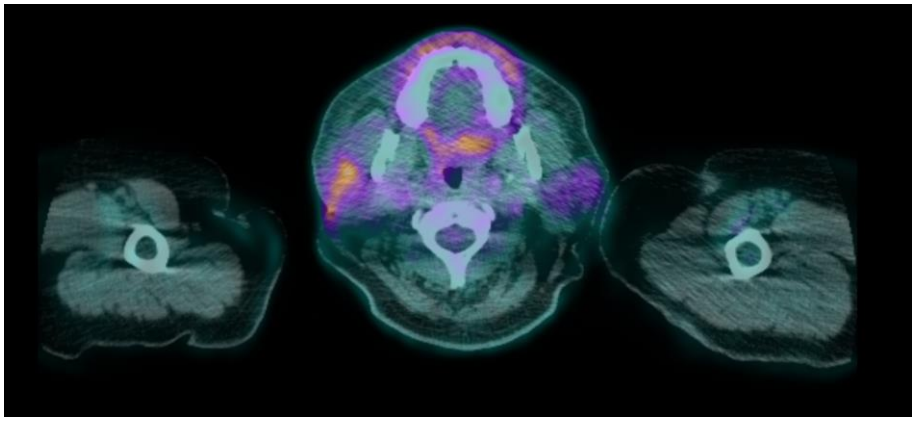

Fig 2: Positron emission tomography scan axial view aspect of the residual disease after performed surgery

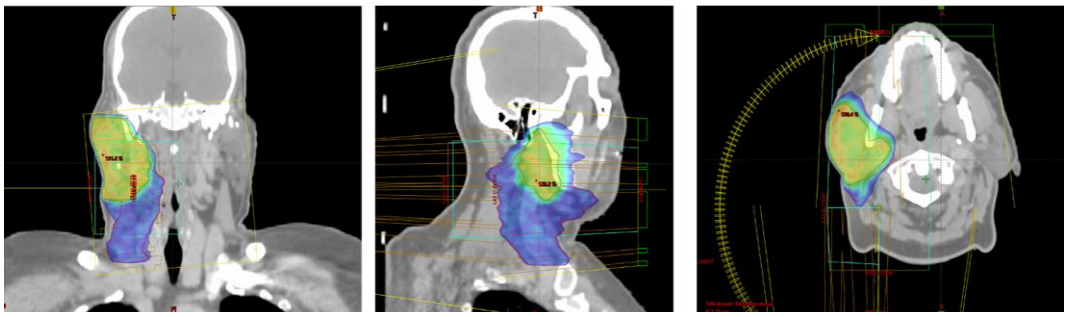

Fig 3: Dose distribution, isodose 95 on axial, coronal and sagittal view

3. Discussion

Carcinosarcoma is a very rare histological subtype tumor of the salivary gland containing both malignant epithelial and mesenchymal elements [4]. These mixed tumors can occur de novo or next to the transformation of a pre-existing pleomorphic adenoma [5]. Representing only 0.04%-0.2% of all salivary tumors, the parotid gland being the most reported affected site of all salivary glands [6].

As it had happened with our patient, the apparition of a fast-growing neck mass regarding the parotid region was the most common symptom clinical [7].

MRI is the gold standard and the imaging of choice to analyse parotid gland lesions. It provides a better soft tissue resolution and ability to detect deep lobe involvement as well as perineural invasion. MRI is also more accurate than CT in predicting a histological diagnosis [8].

Carcinosarcoma is aggressive and survival rates depends some prognostic factors as the extent of invasion, grade of tumor, and regional lymph node spread or distant metastases [9]. Treatment may integrate surgery, radiation and chemotherapy. Unfortunately, due to the rarity of salivary glands carcinosarcoma, a well-established therapeutic approach is lacking.

According to a study published in the Annals of Surgical Oncology, surgery is the primary treatment for parotid gland sarcoma, but radiation therapy is often used in combination with surgery or as an alternative when surgery is not possible. Chemotherapy is generally reserved for advanced cases of the disease [10]. Maximal safe tumor resection continues to be the primary treatment modality, positive margin defined as disease present within 1 mm of the final resected edge [11]. Surgery should be a total resection of the

tumor, the parotid gland and proliferated tissue or lymph nodes.

Radiation therapy can be an effective treatment option for parotid gland sarcoma, particularly when the tumor is inoperable or the patient is not a good candidate for surgery. Radiation therapy (RT) is commonly indicated for positive surgical margins or tumors with close (<1 cm) margins with other high-risk features, even if we don't have enough evidence bases of the benefit of RT. Indeed, there are not systematic reviews or meta-analysis on the role of RT in parotid gland sarcomas. Although, for soft tissue sarcomas of the extremities, it has been shown that RT increases rates of local control in prospective randomized trials [12]. In case of epithelial malignant neoplasms of the parotid gland, adjuvant RT may be indicated for high histological grade, microscopic or macroscopic residual disease, perineural or lymphovascular invasion, T3-4 stage disease and/or positive lymph nodes [13].

Intensity modulated radiation therapy (IMRT) is the reference technique, proton therapy is recommended for pediatric cases [14].

The typical dose of radiation used in the treatment of parotid gland sarcoma ranges from 60 to 70 Gy delivered in daily fractions over a period of 6 to 7 weeks [15]. Regarding the histological subtype, the delivered dose can vary. In cases of complete resection, the prescribed dose can go from 45 to 54 Gy. Meanwhile, the total dose can reach 70 Gy in case of a residual disease after surgery or undefined sarcoma. Beside exceptions, there is no interest in prophylactic nodes RT; nodes RT is only indicated in case of macroscopic positive nodes [16, 17].

Overall, the dose and volume of radiation treatment for parotid gland sarcoma will depend on a variety of factors, including the size and location of the tumor, the patient's overall health, and the goals of treatment.

The role of chemotherapy in the management of parotid gland carcinosarcoma remains unclear. Although chemotherapy has been used in the treatment of various types of sarcomas, its effectiveness in treating carcinosarcoma is still not well-defined. The limited number of case reports and small series that have evaluated the role of chemotherapy in the management of parotid gland carcinosarcoma have produced inconsistent results. Some studies have reported a benefit from chemotherapy, while others have shown no significant improvement in outcomes.

Chen *et al*. reported a case of a patient with metastatic carcinosarcoma of the parotid gland who was treated with a combination of doxorubicin and ifosfamide chemotherapy. The patient showed a partial response to the treatment, with a reduction in the size of the tumor and the resolution of lung metastases [18]. Another study reported a case of a patient with parotid gland carcinosarcoma who was treated with adjuvant chemotherapy consisting of cisplatin, doxorubicin, and cyclophosphamide. The patient remained disease-free for 5 years following the treatment [19]. However, retrospective analyses of patients with parotid gland carcinosarcoma who were treated with surgery and adjuvant chemotherapy showed no significant improvement in outcomes compared to those who received surgery alone [20]. These studies suggest that the role of chemotherapy in the management of parotid gland carcinosarcoma is not well-established.

Given the aggressive nature of the disease and the high risk of recurrence and metastasis, the use of chemotherapy as an adjuvant therapy may be considered in select cases, common regimens included doxorubicin and Ifosfamid. However, further studies are needed to better define the role of chemotherapy in the management of parotid gland carcinosarcoma.

The prognosis of parotid gland carcinosarcoma is poor due to its aggressive nature and high potential for local recurrence and distant metastasis. The reported 5-year survival rate is less than 30%, and the median survival is approximately 18 months.

4. Conclusion

Parotid gland carcinosarcoma remains a rare and aggressive malignant tumor that is composed of both carcinomatous and sarcomatous components, with a poor prognosis due to the sarcoma component. The diagnosis is carried out based on histopathological and immunohistochemical study and and the treatment is mainly based on surgery. The role of adjuvant radiotherapy or chemotherapy is not clear due to the limited number of cases reported in the litterature. The prognosis is poor, and the reported 5-year survival rate is less than 30%. More investigations should be made so that we can have more evidence based therapeutic strategies.

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