

carcinoma grade III with < 1mm(close)margins staged PT1 N0 M0. The immunohistochemistry revealed the negative expression of ER and PR assays. **Methodology:** In December 2022, he received adjuvant radiation to the tumor bed (66 GY) in 33 fractions over 6 weeks based on the VMAT technique. 12-month follow-up, the patient showed no evidence of local or regional disease recurrence or distant metastasis. **Results:** Radical surgery, followed by adjuvant radiotherapy, should be considered the standard of care for a patient, with significant improvement in 5-year locoregional control. and in general, salivary gland neoplasms respond poorly to chemotherapy and are currently indicated only for palliative sitting. More prospective data is needed to establish a role for hormonal therapy and molecularly targeted therapies. **Conclusion:** CXPA is an uncommonly aggressive malignancy that, if associated with regional metastasis, invariably leads to mortality. Total resection of the tumor, followed by adjuvant radiotherapy, should be considered the standard of care for a patient with significantly improved 5-year locoregional control. Early and prompt diagnosis, followed by aggressive surgical intervention and adjuvant radiotherapy for patients with carcinoma ex pleomorphic adenoma, can enhance their survival rates.

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AGGRESSIVE SALVAGE THERAPY OF OLFACTORY NEUROBLASTOMA CASE REPORT EXPERIENCE

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Objective: Olfactory neuroblastoma (ONB) is a rare malignant neoplasm arising from the olfactory neuroepithelium. It accounts for 3–5% of all nasal and Sinonasal malignancies, with an incidence of approximately 0.4 cases per million. A complete surgical resection of tumor followed by a full course of radiotherapy, is considered the treatment modality of choice for most ONBs. We aim to assess the impact of aggressive salvage radiotherapy in olfactory neuroblastoma on local recurrence and overall survival. **Case report:** A 41-year-old Libyan female presented in 2020 with a mass in the right nasal cavity that caused persistent nasal congestion with intermittent epistaxis over one year ago. Histopathological characteristics and immunohistochemical findings of the biopsy confirmed an olfactory neuroblastoma grade III, Radiological imaging evaluation revealed group B stage, and an incomplete excision was done, followed by radical radiotherapy with 70 GY in 35 fractions over 5 weeks to the residual disease. **Methodology:** Imaging follow-up for three years up to February 2024 shows no signs of local recurrence or distant metastasis. **Results:** Although multi-disciplinary care is required, surgical treatment alone is effective for low-grade tumors with free margins. Adjuvant radiation is used for low-grade tumors with close margins, residual disease, or recurrent disease, and for all high-grade cancers. The poor

prognosis associated with high-grade tumors may also mandate the addition of chemotherapy. Because recurrence can occur after 5 or even 10 years, aggressive management and long-term follow-up are mandatory. **Conclusion:** Multimodal therapy, including post-operative radiotherapy of high-grade incompletely resected ONB, with precise treatment planning based on CT simulation, could achieve an excellent local control rate with acceptable toxicity and reasonable overall survival for patients with ONB. Still, the rarity of the disease makes it difficult to draw definitive conclusions about the role of systemic treatment in induction and concomitant settings.

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PP 18

SEVERE CONGENITAL NEUTROPENIA WITH GLUCOSE-6-PHOSPHATASE CATALYTIC SUBUNIT 3 (G6PC3) DEFICIENCY OR DURSUN SYNDROME DIAGNOSED AT ADULTHOOD

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Case report: Severe congenital neutropenia is rare and usually diagnosed at childhood. G6PC3 deficiency emerge by mutation in glucose metabolism controlling genes as a syndromic variant. We here present a young adult case with unexplained neutropenia after kidney transplantation for FMF related AA amyloidosis. He had facial dysmorphism, growth retardation, and atrial septal defect. Parents were relatives and he had recurrent infection history. Genetic screening revealed G6PC3 gene mutation in patient.

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PP 19

THE RARITY OF PRIMARY CUTANEOUS MALIGNANT MELANOMA OF THE BREAST REQUIRES SPECIAL CONSIDERATION IN THE MANAGEMENT.

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Objective: Cutaneous malignant melanoma of the breast is a rare tumor, accounting for less than 5% of all malignant melanomas, Surgical resection is the commonly adopted treatment method for malignant melanoma, supplemented by chemotherapy, radiotherapy, and immunotherapy treatments, resulting in a comprehensive treatment strategy. We aim to assess the efficacy of adjuvant radiotherapy in managing cutaneous