

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.

<https://doi.org/10.1016/j.htct.2024.04.039>

PP 17

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.

<https://doi.org/10.1016/j.htct.2024.04.040>

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.

<https://doi.org/10.1016/j.htct.2024.04.041>

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

malignant melanoma of the breast in long-term local and regional control. **Case report:** A 65-year-old Libyan woman was diagnosed with stage III primary cutaneous malignant melanoma of the breast in 2021. She presented with a progressive painless mass of preexisting nevus, which is located on the skin of the upper inner quadrant of her left breast post-wide local excision without ipsilateral regional lymph node sampling. A month later, a regional ipsilateral axillary LN recurrence occurred. Modified radical mastectomy and axillary LN dissection were done. **Methodology:** subsequently, six cycles of chemotherapy were received, followed by 40 GY in 15 fractions of adjuvant radiotherapy to the left chest wall, ipsilateral axilla, and supraclavicular LNs. In November 2022, lung metastasis was identified, and immunotherapy was advised. Subsequent imaging up to January 2024 indicated no local or regional recurrences and a complete disappearance of lung metastasis. **Results:** The rarity of cutaneous malignant melanomas of the breast has made it difficult to evaluate a life-threatening disease in which local recurrence and regional or distant metastasis may develop after surgical removal of MM, which is common. Wide local excision and prophylactic lymphadenectomy, including radical mastectomy, gave the best long-term local and regional control. Internal mammary node Dissections are not indicated; radiotherapy decreases locoregional failure from 30-50 % to 10–20%. **Conclusion:** Given the notable local, regional recurrence, and distal metastasis rate, local radiotherapy and immune checkpoint inhibitors monotherapy could serve as potent adjuvant treatment in metastatic cutaneous breast malignant melanoma.

<https://doi.org/10.1016/j.htct.2024.04.042>

PP 20

EVALUATION OF THE ASSOCIATION OF TUMOR BIOMARKERS WITH CHILDHOOD CANCERS

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Objective: We aimed to investigate the indications for Carbohydrate antigen 19-9 (CA 19-9), carbohydrate antigen 125 (CA-125), carbohydrate antigen 15-3 (CA15-3) and carcinoembryonic antigen (CEA) tumor biomarkers, less commonly used in children, and their association with patients diagnosed with childhood cancers. **Methodology:** The study aimed to include patients aged 0-18 who had CA 19-9, CA-125, CA 15-3 and CEA tumor biomarker assessments at Adana City Training and Research Hospital (ACTRH) between 01.11.2022 and 01.11.2023. CA 19-9, CA-125, CA 15-3 and CEA values were recorded from routinely collected serum/blood samples of the patients. The relationship between tumor biomarkers and patients diagnosed with childhood cancers was evaluated. **Results:** The study included 211 patients. Out of 211 patients, 145 (68.7%) were female, and 66 (31.3%) were male. Malignancy was detected in 35 patients (16.6%). There was no statistically

significant relationship observed between CA 15-3, CA 19-9, and CEA positivity and the detection of malignancy. The respective p-values were found to be (p=0.711, p= 0.533, p=0.573). A statistically significant relationship was observed between CA-125 positivity and the detection of malignancy (p=0.002). **Conclusion:** Tumor markers alone are not sufficient for making a definitive diagnosis or determining treatment decisions. However further comprehensive studies are needed for detection of association conventional tumor markers and childhood cancers.

<https://doi.org/10.1016/j.htct.2024.04.043>

PP 21

THE EFFECT OF FERRITIN LEVEL ON RESPIRATORY FUNCTIONS IN PATIENTS WITH B-THALASSEMIA MAJOR

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Objective: β -thalassemia major (β -TM) is an autosomal recessive disorder caused by mutations in the β -globin gene of hemoglobin. The disease is characterized by splenomegaly due to ineffective erythropoiesis, iron accumulation signs in tissues as a result of increased iron absorption, bone expansion due to increased erythropoietic activity, and decreased tissue oxygenation. One of the effected organ can be the lungs due to excessive iron deposition in these patients. The current study aimed to investigate the effect of serum ferritin level, which is known as a marker of iron accumulation in tissues, on pulmonary function tests (PFT) in patients with β -TM. **Methodology:** Patients aged ≥ 6 years who were regularly followed in the pediatric hematology section of Mersin City Research and Training Hospital with a diagnosis of β -TM were included. All patients received regular blood transfusion and iron chelation therapy. Study participants underwent PFT prior to blood transfusion in the pediatric pulmonology section. **Results:** A total of 43 patients with β -TM were studied. Included patients were divided into two groups according to the mean annual ferritin level; low ferritin group if below 2000 ml/ng and high ferritin group if above 2000 ml/ng. The low ferritin group was consisted of 19 patients and the high ferritin group was consisted of 24 patients. The characteristics of these two groups are shown in Table 1. There were no statistical significance in age, gender, body mass index, age at diagnosis, mean annual hemoglobin, splenectomy, cardiac involvement and oxygen saturation among both groups, but the number of annual transfusion was significantly higher in the high ferritin group than lower ferritin group. When PFT parameters of both groups were compared, FVC (forced vital capacity) was statistically lower in the high ferritin group compared to the low ferritin group. Other parameters