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Clinical case discussion on non-melanoma skin cancers

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Presenter: **Dr Carolina Pereira**, Oncology Institute Francisco Gentil, Lisbon, Portugal

Challenges of Palliating, inoperable squamous cell cancer of the scalp , in a patient with Xeroderma Pigmentosum in resource limited setting

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History

- E. W
- 37-year-old female
- Background history of mentally retardation and xeroderma pigmentosum
- Onset of skin freckling and dryness at the age of 5 years
- Presented in early 2020 with a history of rapid expanding ulcer on the right forehead. No associated history of trauma.
- Currently the ulcer is foul smelling and intermittent bloody discharge.

Obs/gynae Hx

Nulliparous

Amenorrhoea for the last 3yrs

Family Social History

- 2nd born in a family of 2
- Sister died at the age of 35 years following a short illness. Cause of death unknown
- No known family history of skin disorder/cancer
- Unemployed single mother is the sole caregiver

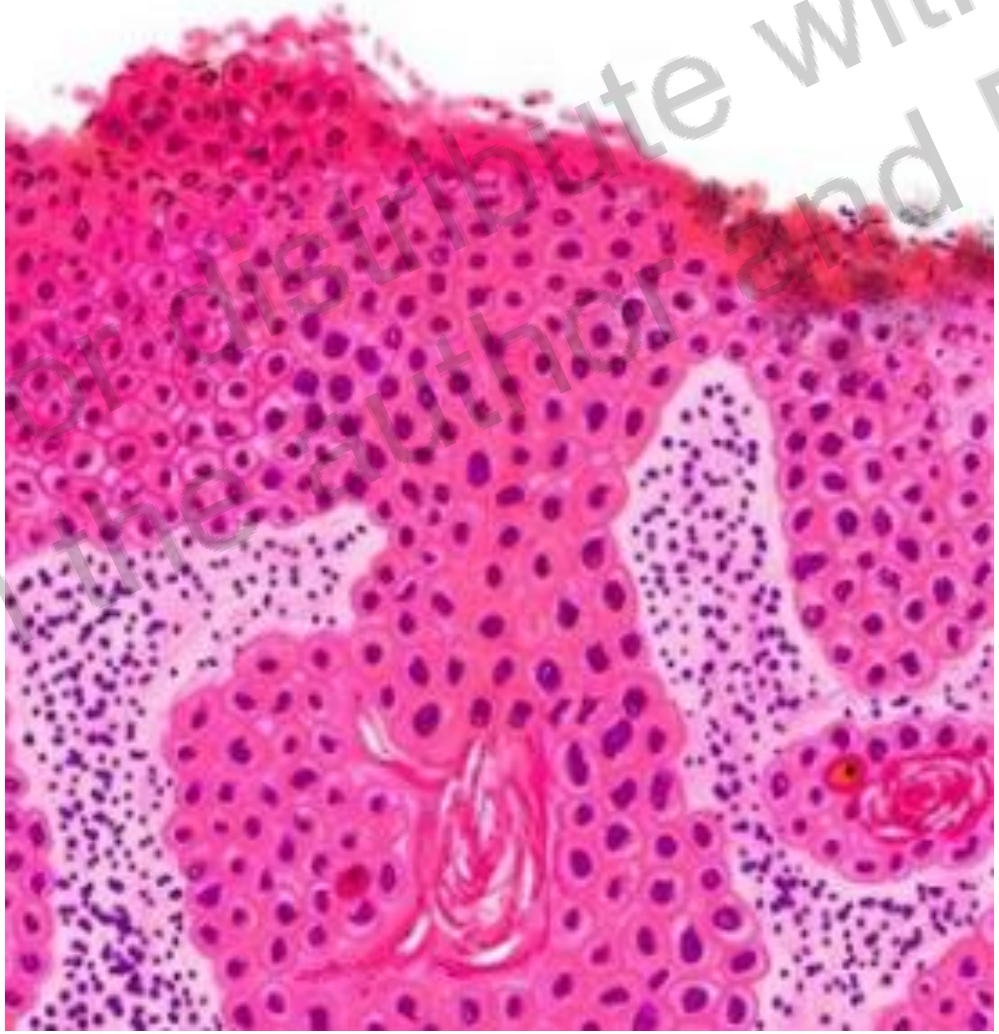
Examination Findings

- Healthy , hyperactive and impulsive behavior. Tendency to self-mutilate
- Right frontal orbital scalp ulcer 10 by 10 cm crossing the midline. Fungating,malodour with serosanguinous discharge.
- Left eye reduced vision .
- Right Bucco- facial and right submandibular adenopathy measures 1cm .
- Oral Cavity showed no lesions.
- SKIN EXAMS: Freckling and scaling of skin, areas of hypo and hyperpigmentation involving the entire body
- Further clinical assessment impeded by patient's mental condition

Investigations - Pathology

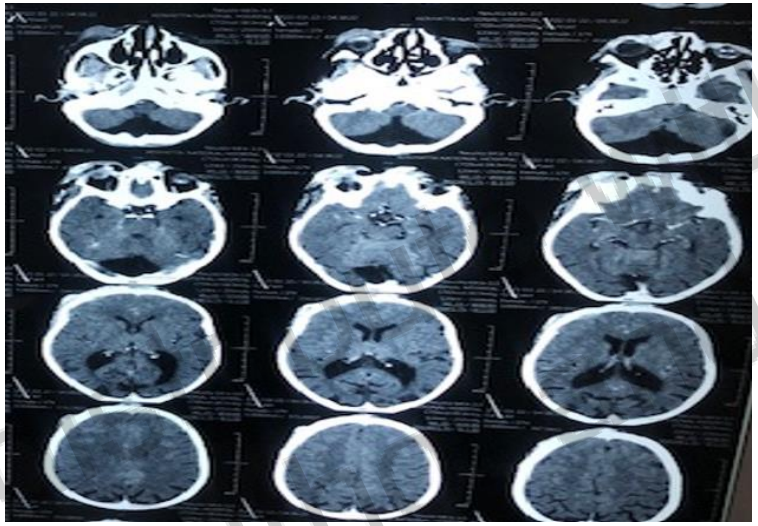
Punch biopsy of scalp lesion (07.12.2021)

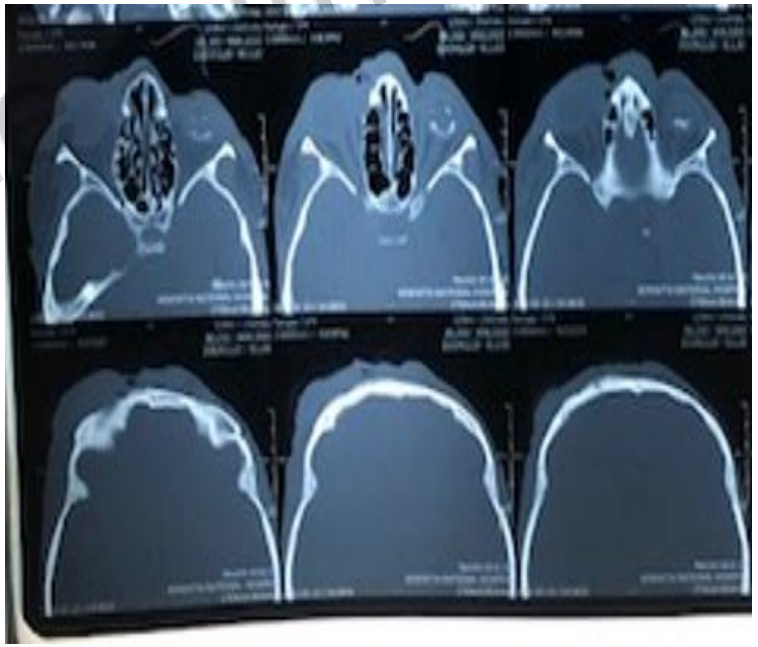
well differentiated squamous cell carcinoma of the skin



CT SCAN OF HEAD : (Patient could not afford MRI)

CT Scan head(22.03.22):
Extensive right frontal and peri-orbital soft tissue mass partially encasing the right orbital globe and crossing the midline with infiltration of underlying skeletal structure and right frontal intra-cranial extension.
Features of brain atropy and ventricular dilatation.





OTHER IMAGINGS

- CT chest/Abdomen (16.03.2022): No evidence of metastatic disease

Management

- Locally advanced locoregional non metastatic cutaneous squamous cell of the head and neck.
- The surgical team unable to offer palliative surgery
- Surgical team referred the patient for palliation with radiation .(Malodor, serosanguinous discharge and fungating)

QUESTIONS FOR DISCUSSIONS

- What are the risk of palliative radiotherapy in XP ?
- Any role of cytotoxic in the background of XP if there is visceral metastasis ?
- This patient has good performance status what is optimal management of such a patient with XP?

Xeroderma pigmentosum

- Genetic disorder autosomal recessive
- Decreased ability to repair DNA damage caused by UV
- Severe sunburnt and hyperpigmentation
- Nervous system problems (hearing loss, poor coordination, loss of intellectual function and seizures) -?unrepaired oxidative damage
- Complications include

— a high risk of skin cancer, with about half having skin cancer by age 10 without preventive efforts
— cataracts
— a higher risk of other cancers such as brain cancers

- The average life expectancy 37 years with no neurological symptoms and 29 years if neurological symptoms are present

Xeroderma pigmentosum

- Paraneoplastic syndromes e.g. hypercalcemia-hyperleukocytosis
- There is no cure for XP; all treatment is symptomatic or preventive
- Skin cancer treatment should follow standard skin cancer guidelines

- **Surgery followed by radiotherapy and chemotherapy (cisplatin)**
- **Neoadjuvant chemotherapy for tumour mass reduction followed by oncological resection surgery**

Kaloga M et al. Squamous Cell Carcinoma in African Children with Xeroderma Pigmentosum: Three Case Reports. Case Rep Dermatol 2016;8:311-318.

- **Chemotherapy consisting of Cisplatin and 5-fluorouracil was started**

Emir S et al. Squamous cell carcinoma associated with Xeroderma pigmentosum: an unusual presentation with a tremendously huge mass over the face and paraneoplastic hypercalcemia-hyperleukocytosis. Turk J Pediatr. 2017;59(6):711-714.

Xeroderma pigmentosum

- **Radiotherapy should be used with caution in XP patients with an anticipated prolonged life expectancy, because the late side effects of ionizing radiation in XP are not well known**

Sakata K at al. Radiation therapy for patients with xeroderma pigmentosum. Radiat Med. 1996;14(2):87-90.

- **RT could be given without serious acute side effects**

Kim R at al. Xeroderma pigmentosum in radiation oncology practice. Int J Radiat Oncol Biol Phys 1982;8:313.

- **Clinical and cellular response to RT is similar in XP as seen with other patients**

Mankada S et al. Radiotherapy as a primary treatment modality for squamous cell carcinoma of tongue in a case of xeroderma pigmentosum. J Curr Oncol 2019;2:29-32

- **The reports of use of radiotherapy as a treatment modality for cutaneous neoplasms in patients with XP are rare. The doses and techniques for radiotherapy are not defined for these patients**

Sahai P at al. Basal cell carcinoma in a child with xeroderma pigmentosum: Clinical response with electron beam radiation therapy. Indian J Dermatol Venereol Leprol 2013;79:533-535

- **Cemiplimab in advanced SCC in XP (following proton treatment)**

Rubatto M et al. Immunotherapy in Xeroderma Pigmentosum: a case of advanced cutaneous squamous cell carcinoma treated with cemiplimab and a literature review. Oncotarget. 2021 May 25;12(11):1116-1121

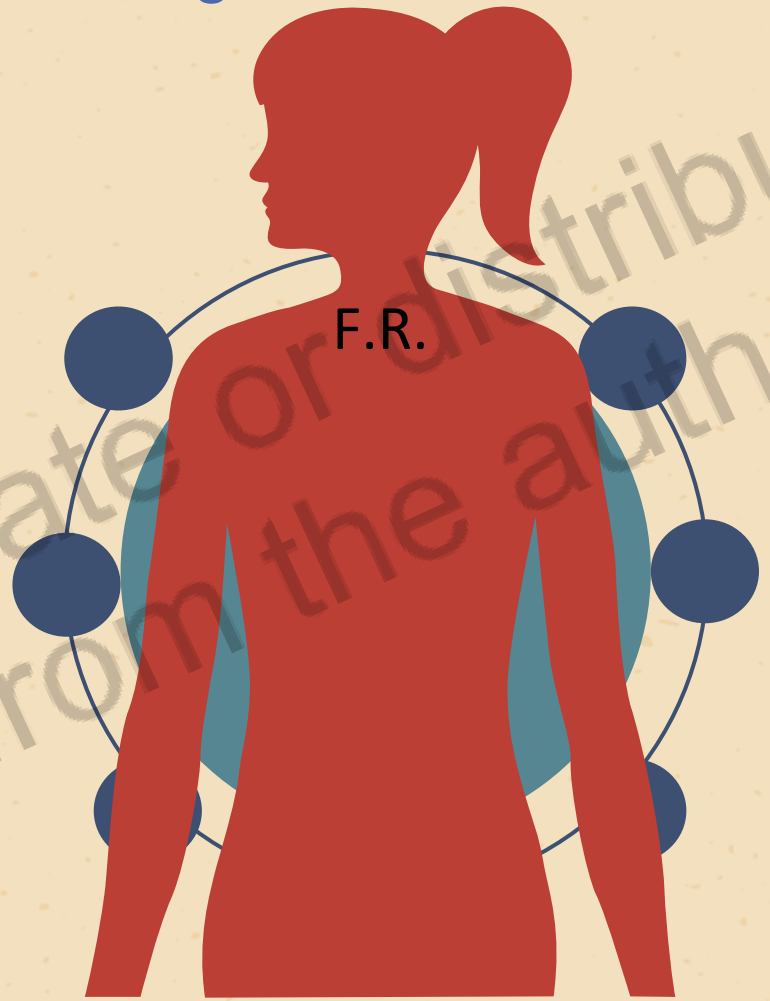
A case report on Gluteal Hidradenocarcinoma, initially diagnosed as Basal Cell Carcinoma,treated with Neoadjuvant RT followed by Wide Excision and Inguinal lymph node dissection

Stephen Lowell Ciocon and Ariz Rigor R. Reillo
Radiation Oncology Residents
Jose R. Reyes Memorial Medical Center

GENERAL DATA

CHIEF COMPLAINT:

Right Gluteal Mass



44 years old

- Female

- Filipino
- Married

History of present Illness

12 yrs PTC

- Noted small, brownish and pedunculated nodule in the right gluteal region which bled persistently after wearing tight clothing.
  - **Excision biopsy (07/24/2010):** Adenocarcinoma of eccrine origin with positive margins.
  - Advised immunohistochemical staining and further ancillaries but was lost to follow-up.

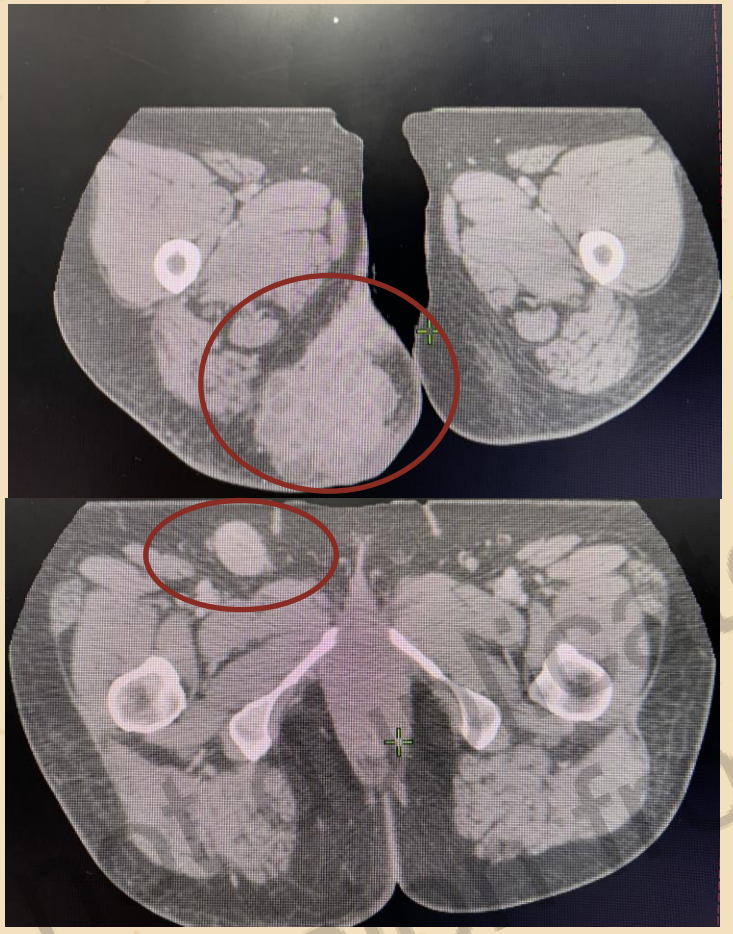
Interim

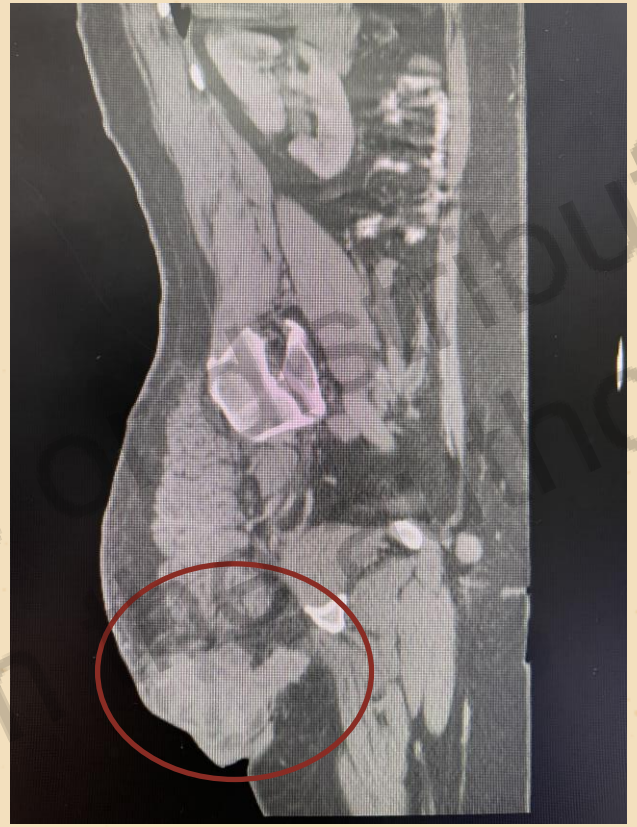
- Unremarkable with no gross evidence of lesion.

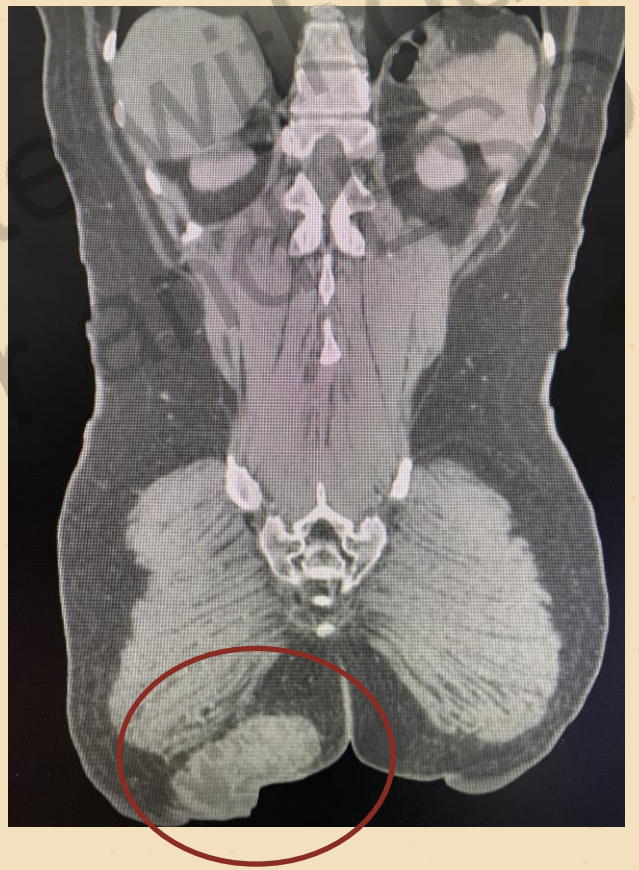
6 months PTC

- Patient noted **erythema in the previous excision site** which evolved into a plaque with a central ulceration associated with foul smelling discharge.
- Whole abdominal CT scan was requested

Whole Abdominal CT with IV contrast







- Whole abdominal CT scan (10/28/21): Lobulated minimally enhancing soft tissue density, measuring approximately 4.6 x 8.4 x 5.3 cm (CC x W x AP) in the cutaneous-subcutaneous right gluteal region with probable superimposed abscess formation . It appears to extend medially and superiorly into the perineal region with suspicious involvement of the right aspect of the anal verge.
- Enlarged and rounded lymph nodes identified in the right inguinal and external iliac regions.

6 months PTC

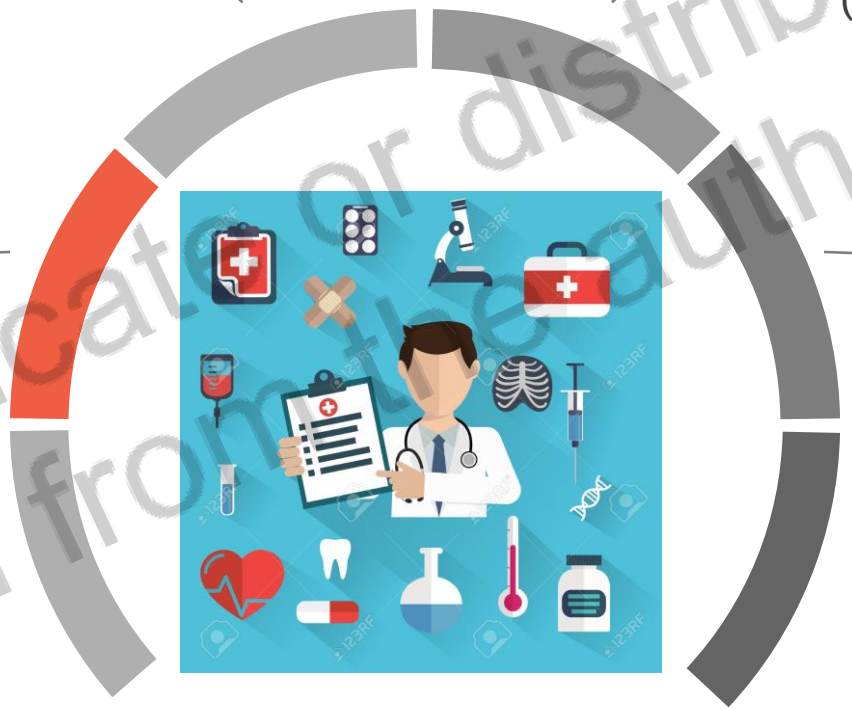
- **Repeat excision biopsy (11/09/21): Adnexal tumor t/c trichoblastoma or basal cell carcinoma.**
- **Immunohistochemical stains (01/22/22): Pancytokeratin (+), p63 (+), CK 20 (-), Ki67 (+) consistent with Basal cell carcinoma.**
- Metastatic work-up
  - **Chest CT scan (02/16/22):** No evidence of pulmonary nodules or mediastinal lymphadenopathy.
  - **Bone scan:** No gross evidence of osseous metastasis.
  - **Repeat CT scan of the abdomen (03/09/22)** showed slight interval increase in the size of the mass 4.9 x 9.3 x 7.3 cm with stable lymphadenopathies.
  - Patient was then referred to the Radiation Oncology department for neoadjuvant RT prior to surgical intervention.

Review of Systems

General
(-) fever, (-) headache, (-) dizziness.

Cardiovascular
(-) chest pain, (-) palpitations

HEENT
(-) blurring of vision, (-) hearing loss (-) sinonasal discharges



Gastrointestinal
(-) abdominal pain, (-) change in bowel habits, (-) diarrhea

Chest and Lungs
(-) chest pain, (-) cough, (-) flu

Musculoskeletal
(-) joint pain, (-) weakness

Past Medical History

Non Hypertensive, Non Diabetic, Non Asthmatic, No FAD

Personal and Social History

Non-smoker, Occasional alcoholic beverage drinker

Family History

No Hypertension, No DM, No Cancer

Physical Examination

Ht 163 m, Wt 70kgs, BSA 1.78 m^2

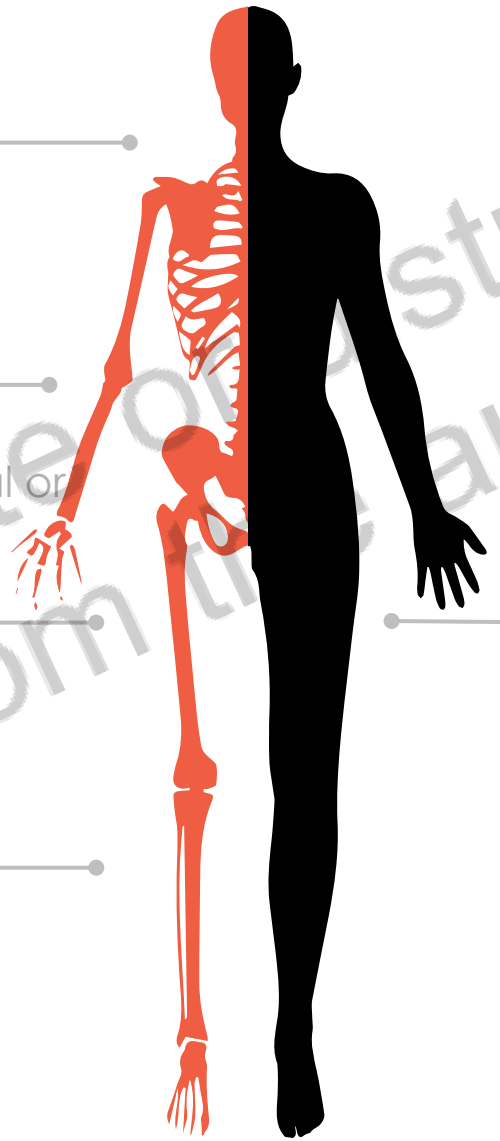
120/70mmHg, 65bpm, 15 cpm

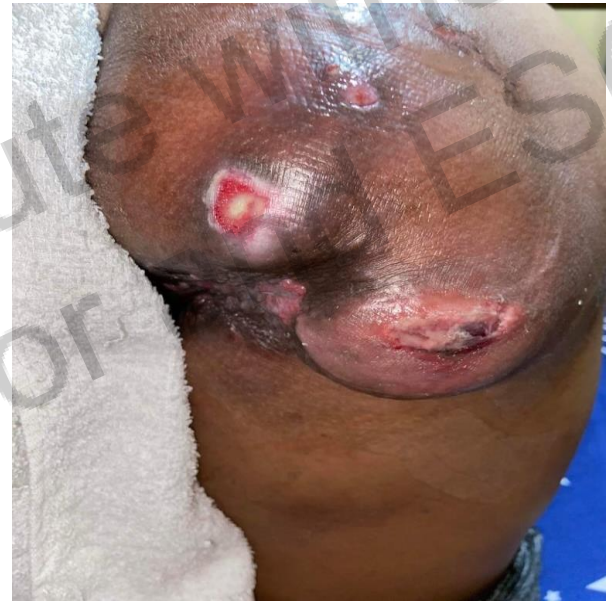
Flabby abdomen tympanitic, non tender, no organomegaly

HEENT: Pink palpebral conjunctivae, anicteric sclerae, No palpable cervical or supraclavicular

ECE, CBS, (-) rales, (-) wheeze

CBS, Normal rate and regular rhythm, no murmur





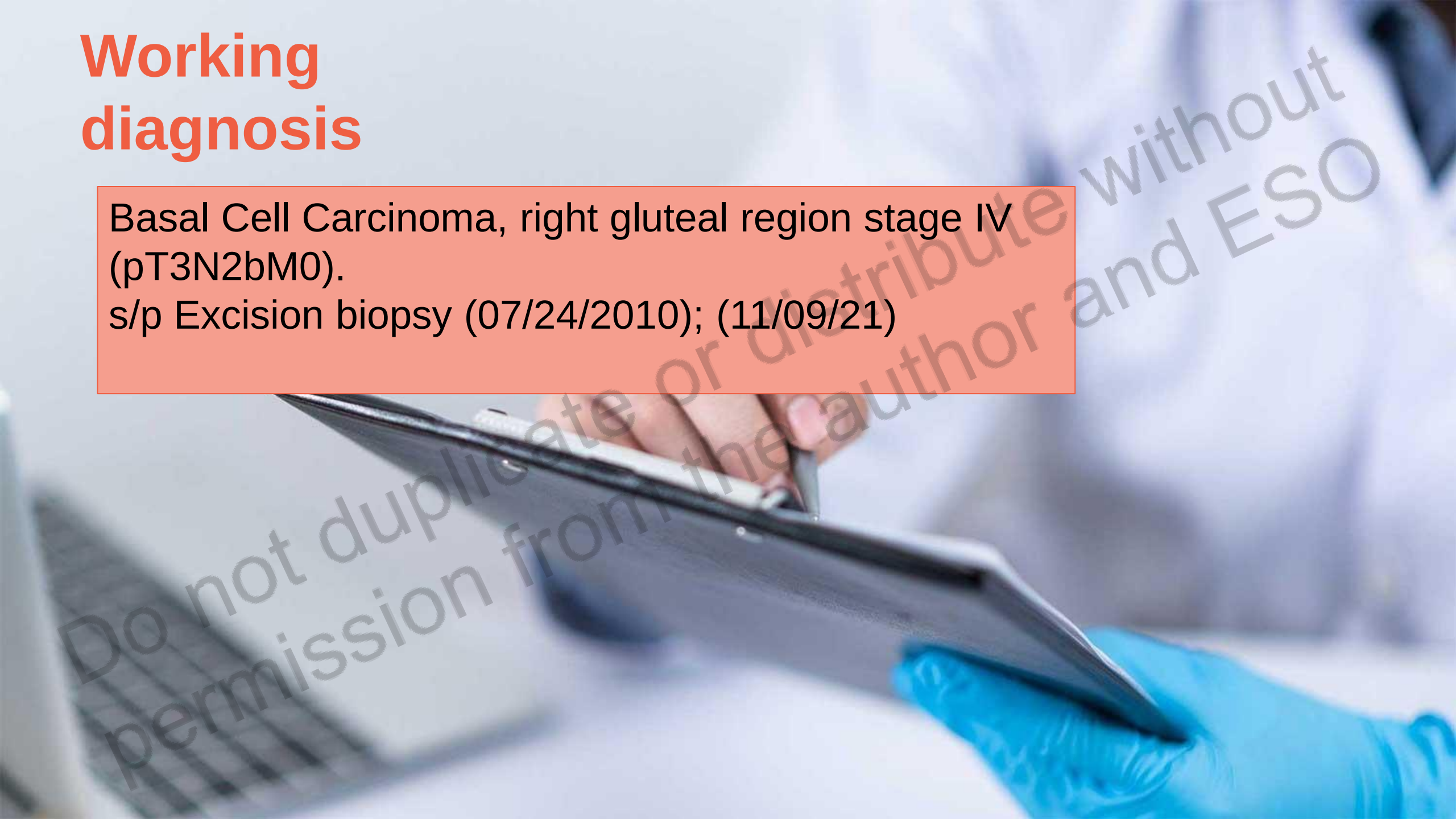
Large, lobulated mass with areas of ulceration and purulent discharge in the right gluteal region extending to the perianal region

Palpable right inguinal lymphadenopathy

Full and equal pulses, no cyanosis, no edema

Working diagnosis

Basal Cell Carcinoma, right gluteal region stage IV (pT3N2bM0).
s/p Excision biopsy (07/24/2010); (11/09/21)

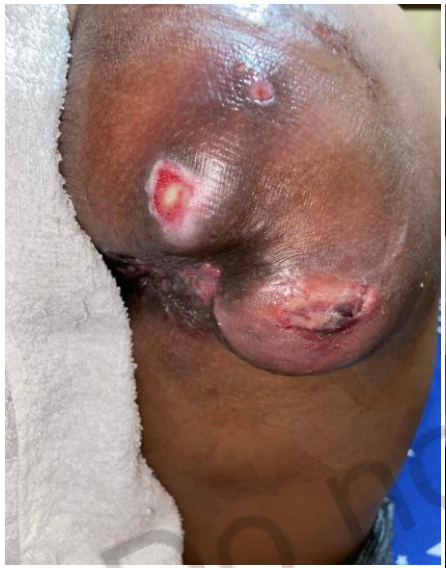


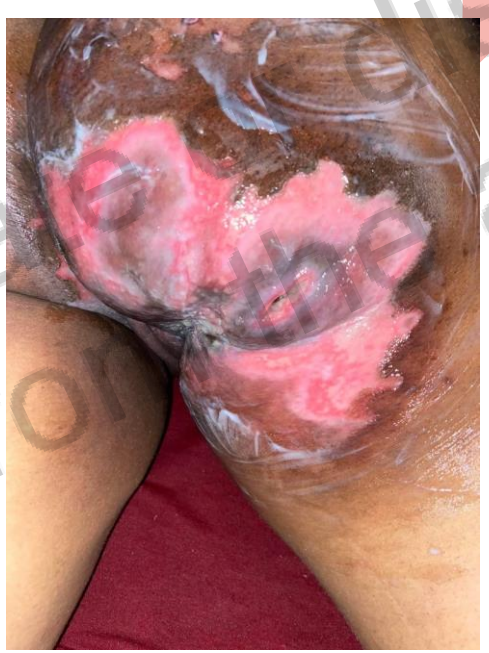
Treatment

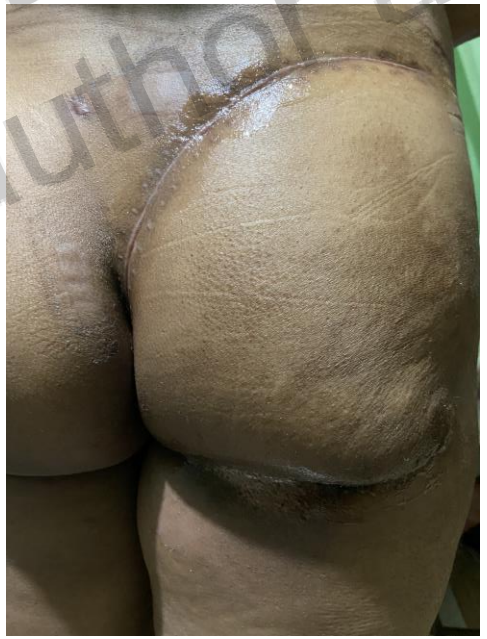
Radiotherapy
3D CRT, 2 Gy per fraction in 33 fractions
66 Gy gross tumor and pathologic lymph nodes
50 Gy regional lymph nodes

Post-RT MRI:
Partial interval decrease in the size of the gluteal mass was noted measuring 6.6 x 6.2 x 5 cm (previously 4.9 x 9.3 x 7.3 cm).
No significant change in the size of the pelvic lymphadenopathies

Wide Excision and Inguinal lymph node dissection



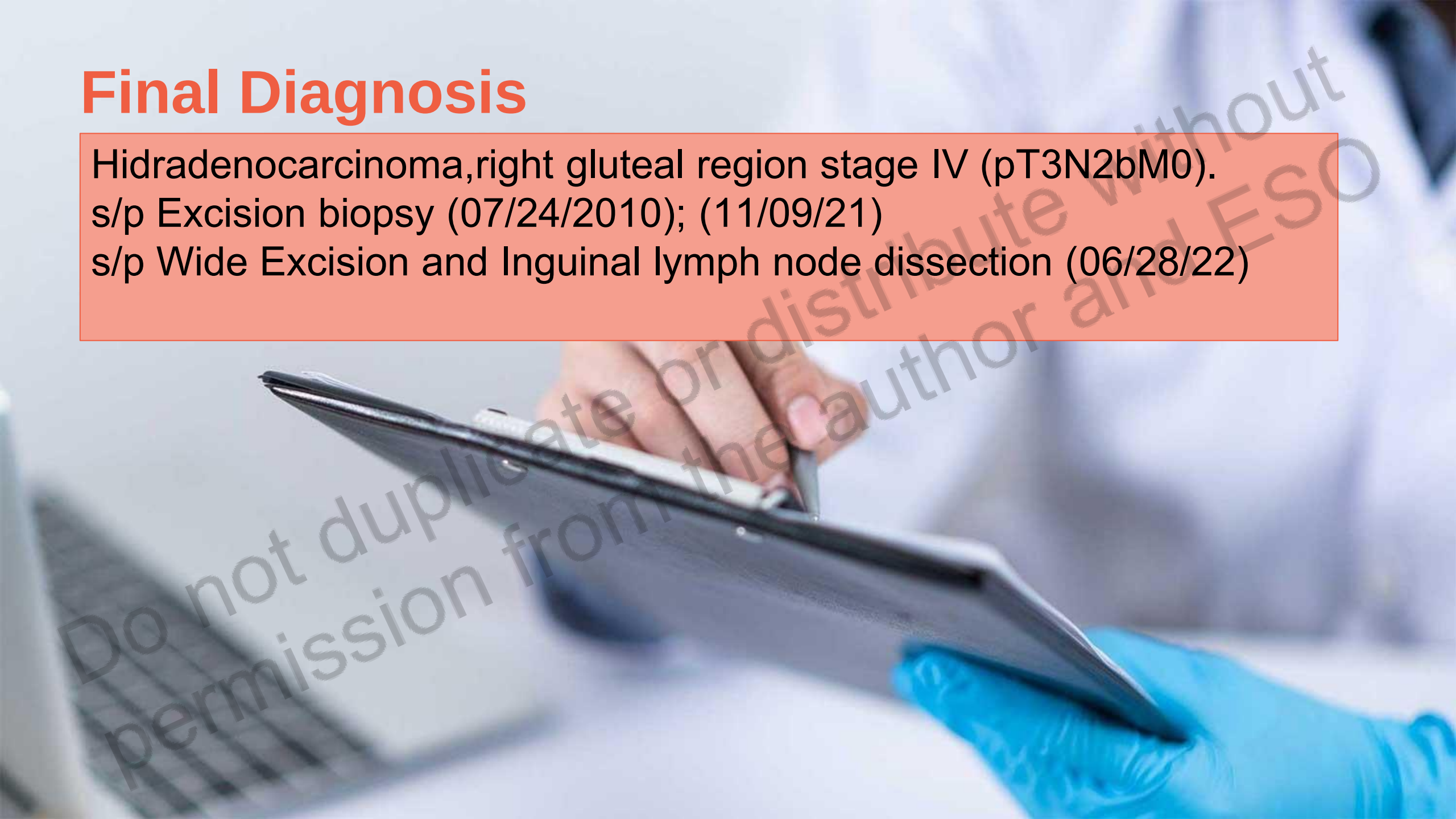




Final histopathology:
Hidradenocarcinoma
5.0 cm in widest dimension with negative surgical margins.
(+)LVSI Lymphovascular space invasion
(+)2 inguinal lymph nodes were noted.

Final Diagnosis

Hidradenocarcinoma,right gluteal region stage IV (pT3N2bM0).
s/p Excision biopsy (07/24/2010); (11/09/21)
s/p Wide Excision and Inguinal lymph node dissection (06/28/22)



Hidradenocarcinoma

Hidradenocarcinoma (HC) is an extremely rare primary eccrine carcinoma which accounts for less than 0.001% of all tumors[1]
Since the SEER database started on 1973 up to 2008, there has only been 226 cases of HC reported.[13]

MANAGEMENT AND OUTCOME

To date there is no consensus treatment for HC due to its aggressive and extremely rare nature

- **Surgery**
  - Remains the cornerstone of treatment, consisting of wide local excision with negative margins
  - Local recurrence rates following surgery range from 10–50% with 5-year post-surgical survival rate of less than 30%. [9]
- **Surgery +Adjuvant RT**
  - There are currently no prospective or randomized clinical trials to compare the clinical outcomes of neoadjuvant and adjuvant RT in HC.
  - The technique and dose of radiotherapy are not consensual. High doses ranging from 50 Gy–70 Gy are recommended
- **Surgery +Adjuvant chemotherapy**
  - Several chemotherapeutic agents have been reported which includes first line agents, 5- fluorouracil based regimen and capecitabine (oral 5-fluorouracil) while second line agents included doxorubicin, platinum-based agents, cyclophosphamide, vincristine, and bleomycin.[5]
  - Results were modest, varied and more often have failed to demonstrate a clear survival benefit with progression to metastatic disease.
  - Combination chemotherapy and radiation has also not shown a clear survival advantage or effect on local control

OUTCOME

Compared with the initial diagnosis of Basal cell carcinoma which carries an excellent 5-year overall survival of almost 100%, HC has a worse prognosis despite surgical intervention with less than 30% surviving after 5 years and warrants a more aggressive management.[9]

Dilemma

- How would you have managed this patient? What would be the treatment sequencing, RT dose and technique?
- What IHC would help determine the final histopathologic diagnosis of this patient?
- What further adjuvant treatment can we offer and how would we follow-up the patient?

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Non-melanoma skin cancer

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Complete response to cemiplimab in advanced squamous cell carcinoma

Carolina Capucho Pereira
Medical Oncology Department
Instituto Português de Oncologia de Lisboa Francisco Gentil

86 yo male, ECOG PS 1

PMH: ischemic heart disease, arterial hypertension, medicated for both conditions

No history of smoking or drinking alcohol

Family history: daughter and paternal aunt with breast cancer

Skin lesion in the chest that grew slowly over 2 years.

Excision in June 2018:

Malignant melanoma, superficial extension, Breslow 2.9 mm, without ulceration, free margins

pT3a

BRAF wild-type

Staging with CT chest-abdomen-pelvis: no distant metastasis

November 2018:

Wide local excision + sentinel node biopsy (left axilla)

Skin tissue without residual disease

1/2 nodes with metastasis (1 mm, without extra-capsular extension)

pN1a

2nd skin lesion removed in right lumbar area

Squamous cell carcinoma (SCC), 25 mm, without subcutaneous tissue or vascular invasion

Free margins > 5 mm

pT2

MM pT3a N1a (stage IIIB)

Cutaneous SCC pT2

Patient **refused right axillary LND**, and was closely monitored

April 2019:

FDG PET/TC and ultrasound: **right axillary lymphadenopathy**

Histology: **SCC metastasis**

Right axillary lymph node dissection

Quistic metastasis of SCC in 1/8 lymph nodes

Adjuvant radiotherapy to right axilla (3DRT, 50 Gy/25 fr)

Patient refused to participate in a clinical trial for adjuvant pembrolizumab (Keynote-630):

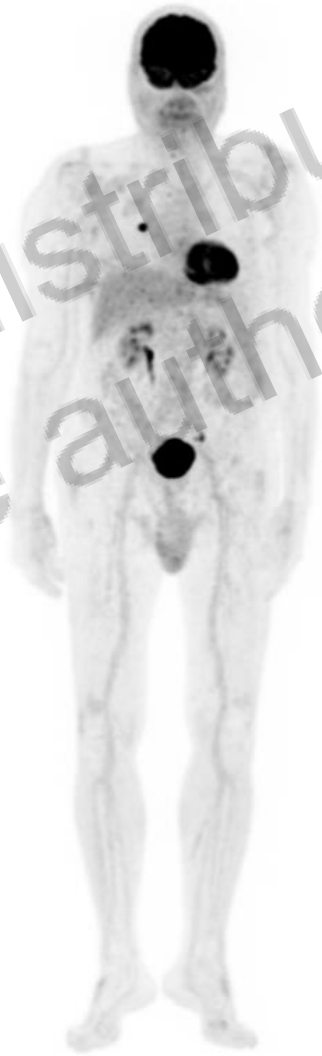
Monitoring

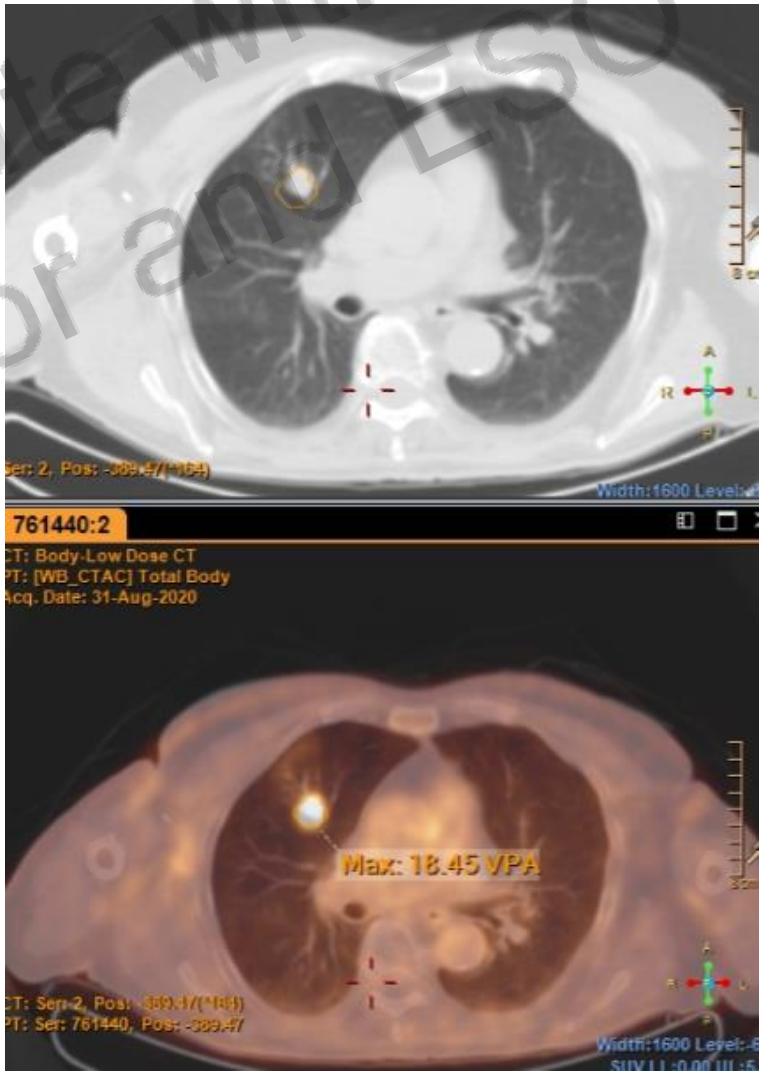
August 2020:

FDG PET/TC: single lung nodule (SUV 18.5)

CT chest: 25 mm nodule + satellite micrometastasis in upper right lobe

EBUS with biopsy: **carcinoma**, unable to further characterize sample

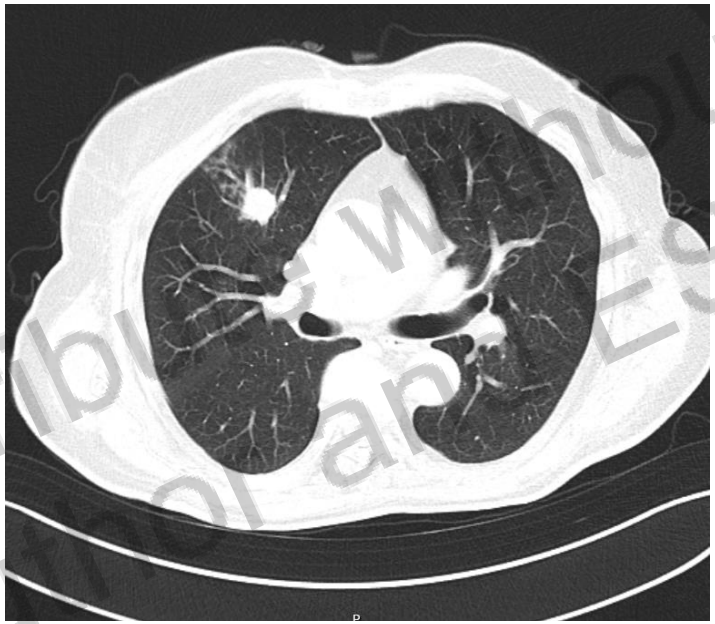


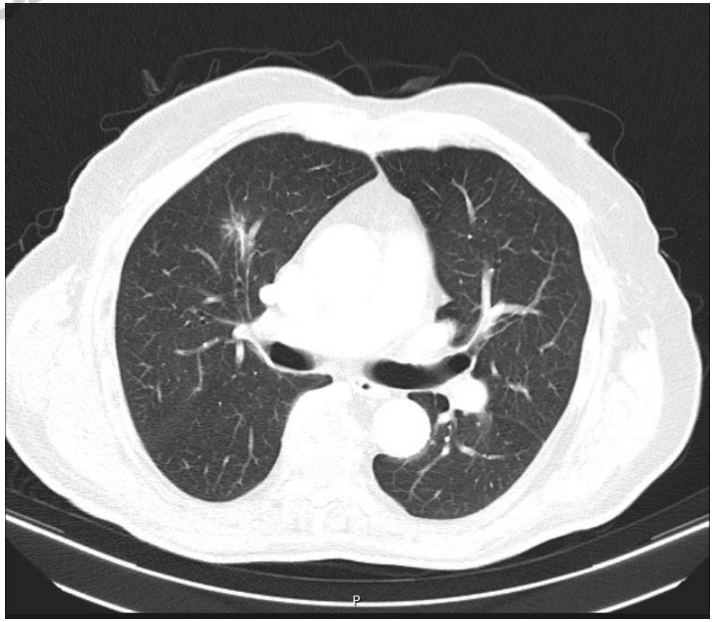


November 2020:

Started Cemiplimab 350mg IV q3w

CT chest after 6 cycles: **complete response**, residual changes in upper right lobe





Toxicities:

G1 fatigue

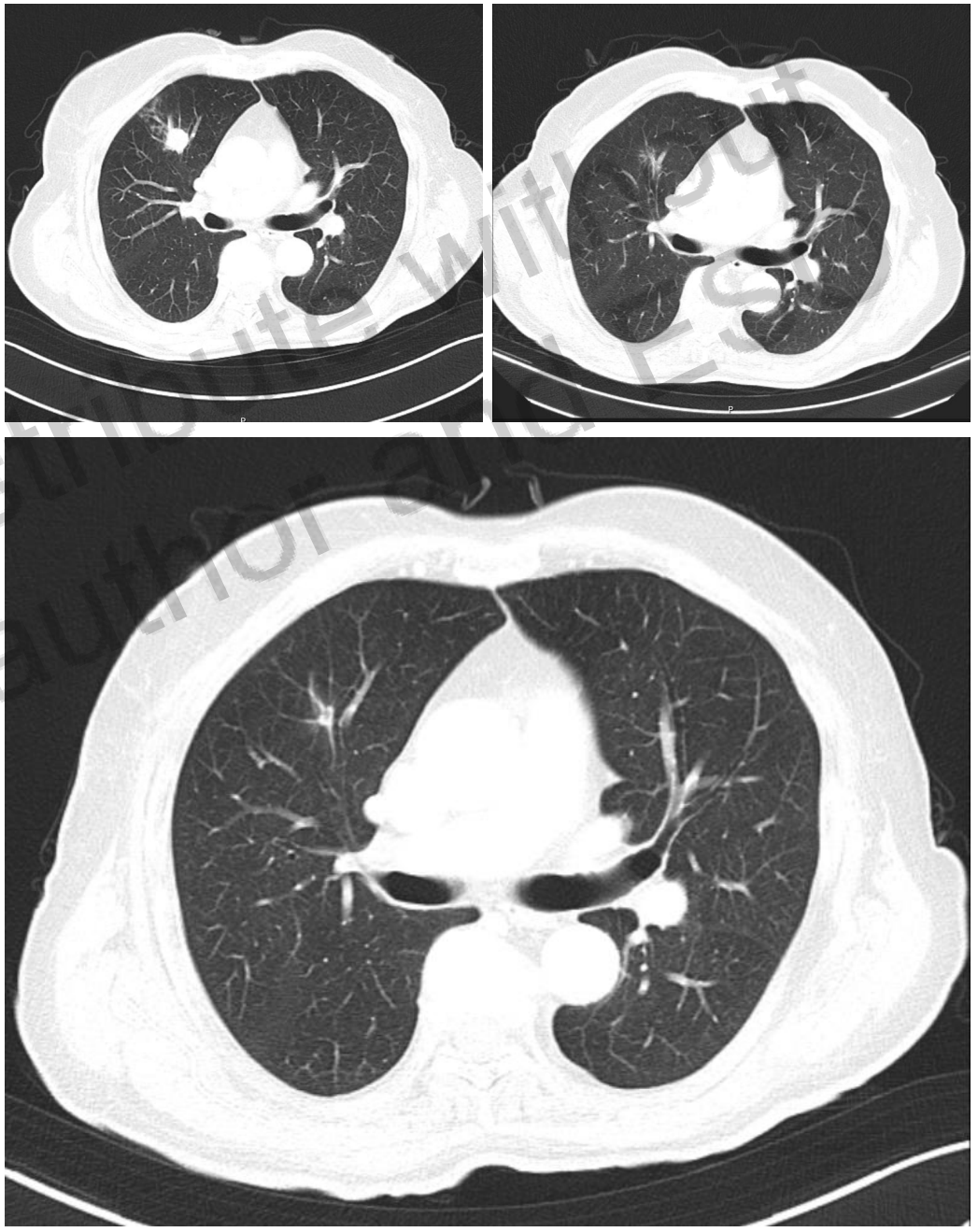
G1 incresed creatinine

Currently:

Continues with **Cemiplimab** (31 cycles)

Sustained complete response

No new toxicities



Discussion

1) Can we consider stopping treatment after 2 years if complete response continues to be sustained?

2) In case of disease progression, what options do we have in this elderly patient with history of ischemic heart disease?

3) In case of confirmed melanoma progression, is it reasonable to consider treatment with a different anti-PD-1?

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