

# Interesting\*

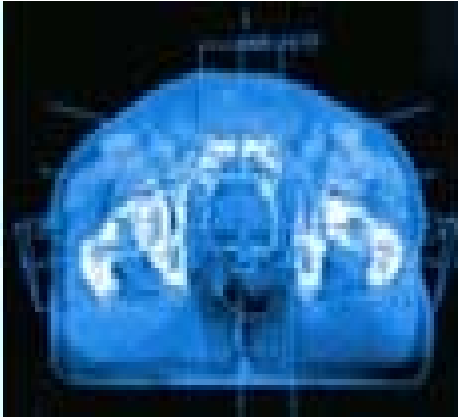
## Clinical Cases



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\* HOLDS THE ATTENTION



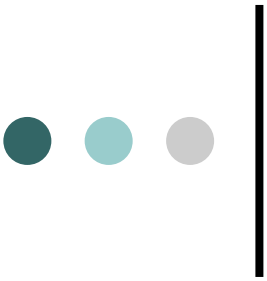
## ○ Thanks

- Patients (material of the talk)
- Prof.Dr. Saad Eissa (Surgical Pathology)
- Dr.Mohamed Maher (Radiodiagnosis)



# Case Report

## Case (I)



- Male patient 19 yrs. Old
- c/o: fever, Dyspnea, 3 months duration
- Cardiology advice:

9/03

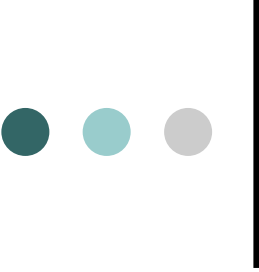
- CT chest : massive effusion

Aspiration of pericardial fluid and biopsy - +ve for malignancy (round cell tumor)

- Echo heart : Rt atrial mass 5x3 cm

- Marker study: +ve for **Embryonal RMS**

- Referred to NCI

- 
- Received IRS-V protocol in addition to RT
    - RT dose: 45Gy (conformal RT)
  - Developed lung metastases 3 months later
  - Subjected to second line CT and palliative RT

● ● ● | Cardiac tumors are rare (1-2 cases/year)

Benign 85%

Malignant 15%

○ 19 patients (1983 and 2003) China

○ 15 patients (1989 and 2002) Taiwan

○ 29 patients (1979 to 1999)

Eleven of the cases involved primary cardiac tumors [rhabdomyoma (n = 10) and fibroma (n = 1)].

Three out of 29 were malignant:

1 fibrosarcoma, 1 malignant mesenchymoma, and 1 rhabdomyosarcoma.

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○ Myxomas are the most common heart tumors seen in infancy and childhood, followed in frequency by rhabdomyomas, fibromas and lipomas.

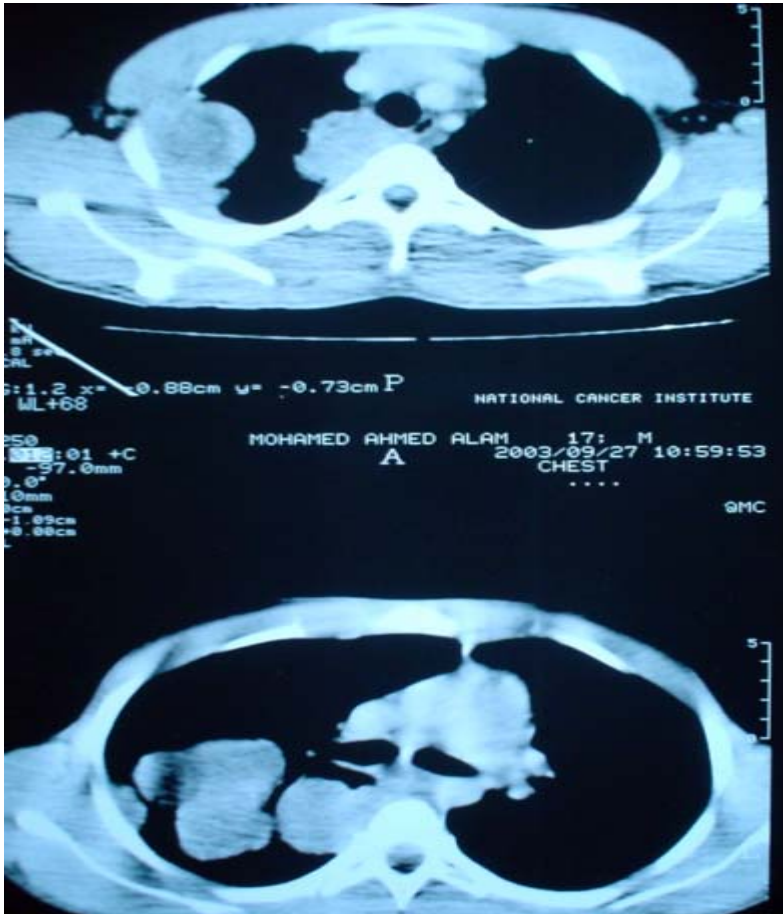
○ median age of patients was 7 +/- 5 years

○ The atrial mass was detected by transthoracic and ransesophageal echocardiography,

○ rhabdomyosarcomas of the heart are highly lethal, operation is indicated for emergency cases, in order to clarify the diagnosis, relieve symptoms, and improve short-term survival

# MRI chest

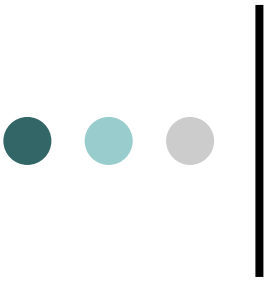
## CT chest





# **Case Report**

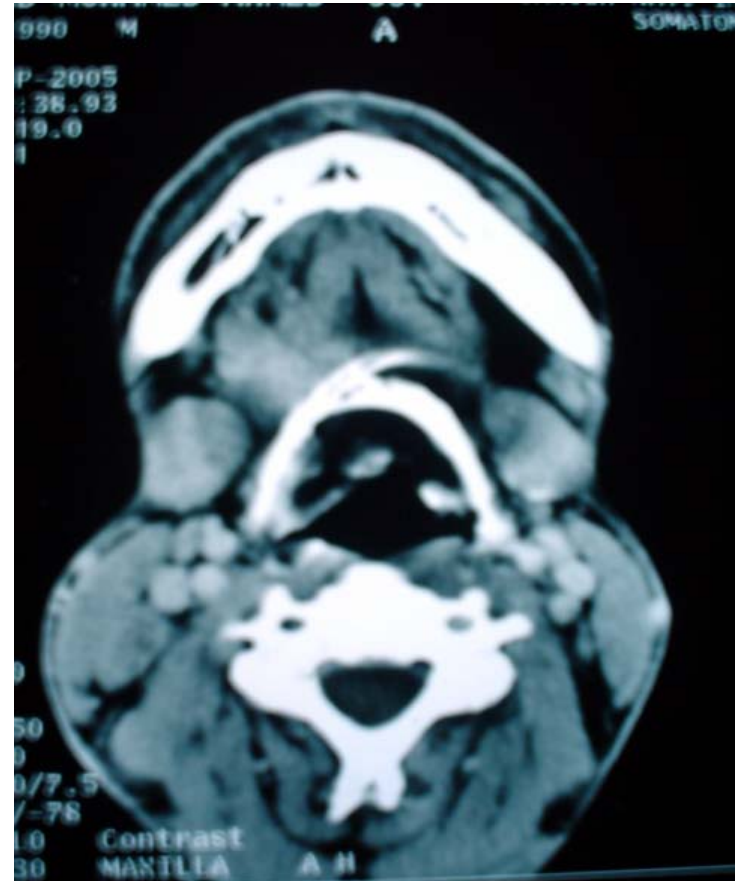
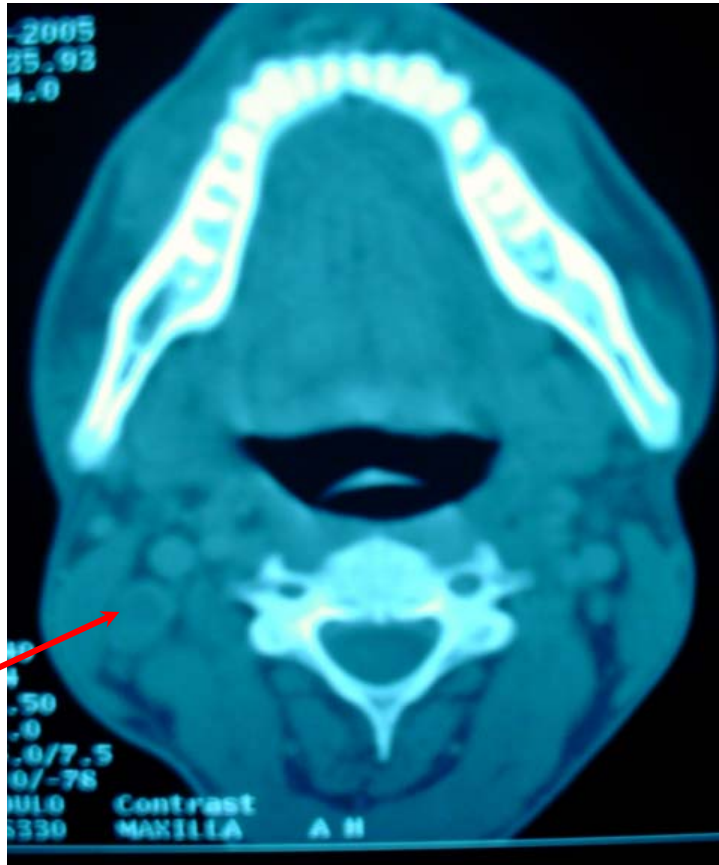
## **Case (II)**



- Male patient 28 yrs, having a **RIGHT TONSILLAR** mass (5x4cm)
- Excised in Fayoum, 8/04
- Referred to NCI 9/04

- 9/04 ● CT (PNS) :Multiple Bilateral Cervical L.Ns – Largest 2.3 cm in  $\Phi$  (Rt.)
- CT chest, abdomen & pelvis: **NAD**

# CT scan



1/05

Pathology: **Follicular Dendritic Cell Tumor (FDCT)**

+ve S100 & Vimentin

-ve CK, LCA, chromogranin, Synaptophysin, CD68, desmin, actin, CD45Ro and CD23.

○ BMA-(N) / LDH-(N)

○ Received 6 cycles of CHOP (Started 3/05 ended 8/05)

○ Re-evaluation:

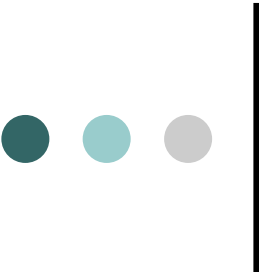
● CT (PNS): Multiple small Bilateral Cervical L.Ns – Largest 1.5 cm in  $\Phi$  (Rt.)

● CT chest, abdomen & pelvis: **NAD**

● Referred for RT

● ? After BND

9/05



# Follicular Dendritic Cell Tumor (FDCT) of the Tonsil

- Usually arises in the dendritic reticulum cells of the lymph nodes
- Low to intermediate malignancy
- Extra nodal cases are rare..
- Only 25 cases reported in the H&N region (most in the oropharynx)—**TEN IN TONSIL**
- **THE 11<sup>TH</sup> IS PRESENTED TODAY**

# Diagnostic Pitfalls

- Reticulum cells within the lymph nodes (Lennert since 1978)
- Histologically different from lymphomas Monda et al, 1986 (tumor/sarcoma)
- A disease of young/middle aged adults,
- Male to female ratio 1:1
- Most cases arise within the lymph node
- D.D.: dendritic cells, interdigitating, fibroblastic
- ? Misdiagnosed (4/10) as SCC

**Monoclonal FDCT markers**    ≡

Poorly differentiated neoplasia (epithelioid/spindled)



# Treatment and Prognosis

- Surgery is often followed by RT or adjuvant CT
- Five were managed only by surgery [tonsillectomy ± BND] (3pts.)
- Four were treated by PORT& one with PORT +CT
- One pt. received only adjuvant CT

# Adjuvant ttt.

In FDCT remains unproven

- Large tumors (>6cm)
- Intra-abdominal location
- Co-agulative necrosis
- High mitotic count (>5/10HPF)
- Cellular atypia
- Lack of adjuvant ttt

Unfavourable  
outcome

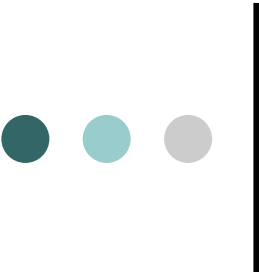
≡

**BUT**

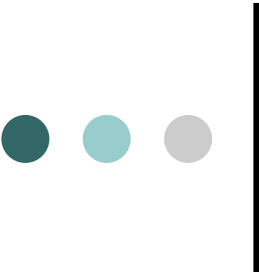
No survival benefit



# Case (III)

- 
- Male patient 63 yrs. Old
  - c/o: sore throat of 4 months duration
  - ENT consultation:
    - Oropharyngeal mass (2cm in  $\Phi$ )
    - CT scan: LT. polypoidal tonsillar mass  
3x2.5x2cm
    - Excision biopsy: spindle cell vascular tumor
    - Tonsillectomy: **kaposi sarcoma**
      - -ve immunostaining to CK
      - Serum HIV: -ve

1/05

- 
- PORT localized fields (4 weeks later)
  - Dose: 35 Gy in 4 weeks (6Mv photons)
  - 10 months later pt. developed lip nodule
    - Excision biopsy: Kaposi sarcoma
    - RT by EBT – 4 Mev (30 Gy in 3 weeks)

- **Between June 1986 and June 1998 (AIDS RELATED)**
- **Only 43 oral mucosa patients. [France]**

○ 1995 Jan-Feb;17(1):64-8

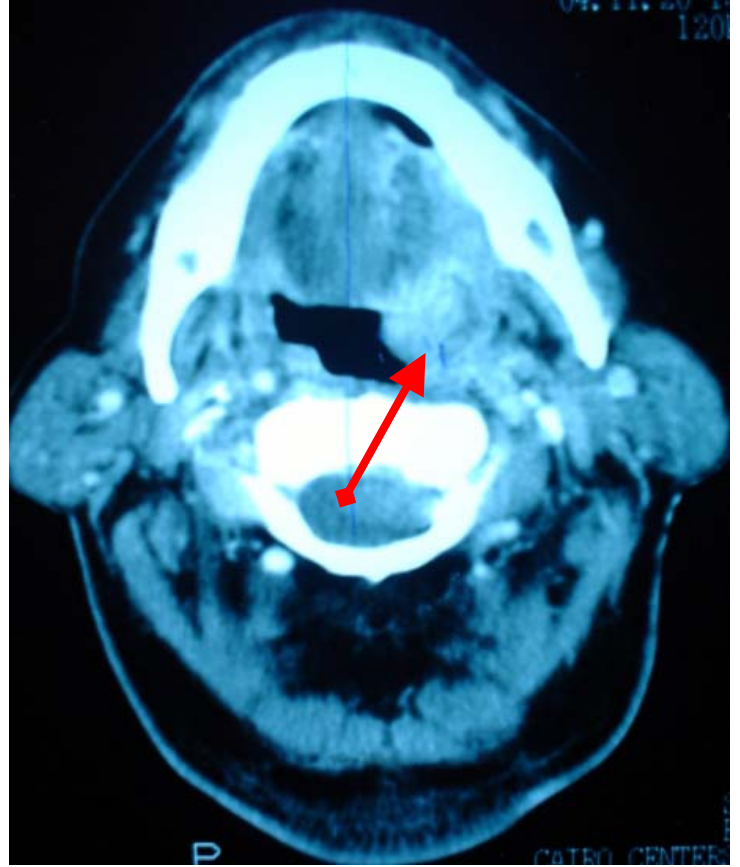
●  
**Kaposi's sarcoma of the oral cavity in a non-AIDS patient: case report and review of the literature.**

**Jindal JR, Campbell BH, Ward TO, Almagro US.**

Department of Otolaryngology, Medical College of Wisconsin, Milwaukee 53226.

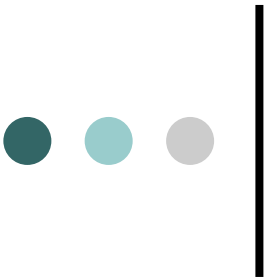
BACKGROUND. Not every patient with Kaposi's sarcoma (KS) has acquired immunodeficiency syndrome (AIDS). The "classic" form of KS is rare, and is associated with an indolent course. It is very distinct from AIDS-associated KS in which oral involvement is common and may be the initial presenting feature of this disease. Two other types of KS are recognized, the African and renal transplant-associated, which also are rarely associated with oral involvement.

- METHODS. We present the case of a 76-year-old man of Mediterranean ancestry who was found to have a biopsy-proven classical KS lesion of the hard palate. The patient was followed by the Radiation Therapy and Otolaryngology Services during and after his treatment. A review of the literature was also conducted.
- RESULTS. Radiotherapy was ineffective at a dose considered "standard" for KS in AIDS patients (1,500 cGy), but was effective when continued to 4,800 cGy. A 24-month follow-up showed no evidence of recurrence in the oral cavity.
- CONCLUSIONS. KS of the oral cavity, is almost always associated with AIDS in the United States, However, it can occur in any of the four types of KS. Although this neoplasm is typically highly radiosensitive, the treatment for each patient needs to be individualized.





# Case (IV)



- Female patient 20 yrs. Old
- c/o: Lt femur pain of 3 months duration
  - pathological fracture of lt. femur
  - Intramedullary nail + biopsy
- 2/05 ● Pathology : **Osteosarcoma**
  - 3months later Rt. Humerus pain
  - X-ray: osteolytic lesion
  - CT chest: (NAD)
  - Bone scan: 10/05
    - Wide spread hypervascular bony lesions



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**32257 Bünde**

Name d. P.:	<b>Rasema Abdulnasser</b>
Geb.-Datum:	11.11.1911
Untersuchungsgut u. klinische Angaben:	Unbekannt. Material ein gefärbter Ausstrich.
erhalten:	01.02.05

### **Pathologisch - anatomische Begutachtung**

Mikroskopisch finden sich unregelmäßig geformte Osteoid- u. Knochenbälkchen sowie Knorpelinseln umgeben von lockerer mehrschichtig oder netzförmig angeordneten spindeligen und rundlich-ovalen Zellen mit leicht polymorphen, teils bläschenförmigen, teils hyperchromatischen Kernen. Wiederholt sind Mitosefiguren nachweisbar. Die unregelmäßig geformten netzartigen Osteoid- u. Knochenbälkchen enthalten dichte Osteoblastensäume. Zur einen Seite hin wird quergestreifte Muskulatur infiltriert. Daneben sind reichlich Blut und Granulationsgewebe nachweisbar. Wiederholt finden sich auch sehr zellreiche Abschnitte mit nur kleinsten Einsprengungen von Osteoid. Immer wieder liegen im Tumor unterschiedlich große Knorpelinseln unregelmäßig verstreut.

**Gutachten:** Osteogenes Sarkom

**X-RAY AND BONE SCAN  
FINDINGS**

**?? METABOLIC DISEASE**



○ Lab. Results:

● LDH: 363 (N)

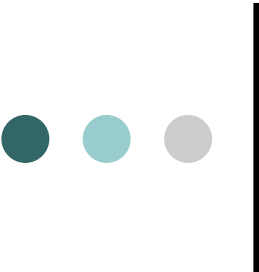
● ALKP: ↑ 405 (N: up to 92 u/l)

↑ ● S. Ca<sup>++</sup>: 12.1 mg/dl (N: up to 10.2)

● S. Phosphorus: 2 mg/dl (2.6-4.6) ↓



Parathyroid hormone: 433 (11-60)

- 
- Parathyroid scan:  $^{99m}\text{Tc}$  MIBI \*
  - Positive study for Lt. parathyroid adenoma
  - Hemithyroidectomy:
    - +ve for parathyroid adenoma
    - S.  $\text{Ca}^{++}$  & Phosphorus --- Normalized

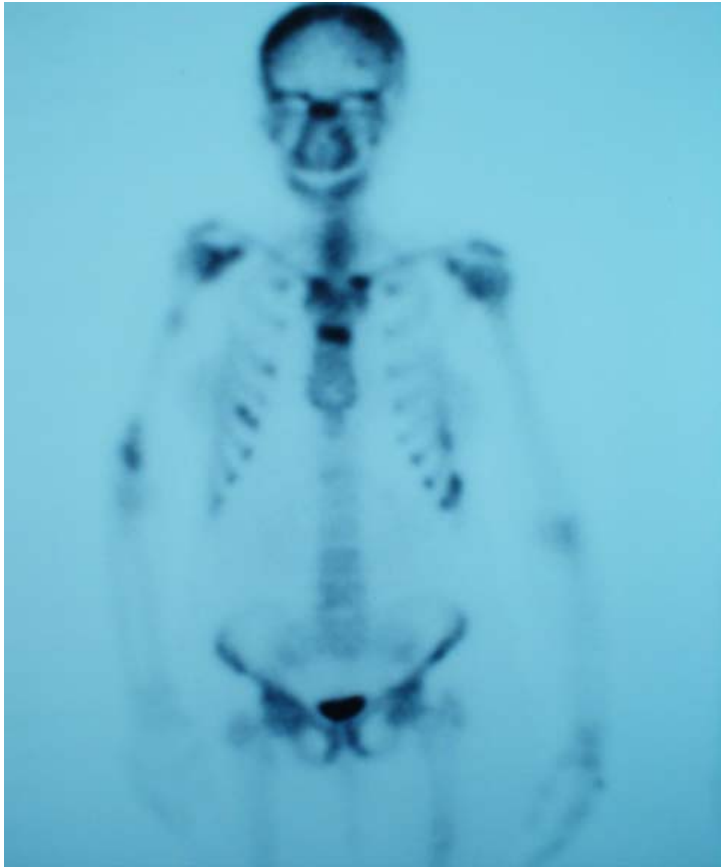
\* MIBI scan correctly detected and localized 19/21 adenomas (90%). parathyroid imaging using a single radionuclide with MIBI (procedure in the preoperative detection and localization of parathyroid adenomas in patients with primary hyperparathyroidism).



## Primary hyperparathyroidism (pHPT)

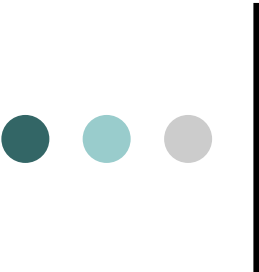
- (2-3/100,000 inhabitants/year)
- onset at a young age is one feature in multiple endocrine neoplasia (MEN) type 1 and MEN type 2A cancer syndromes
- Swedish registry data -- individuals operated on for parathyroid adenomas between 1958-99 (n = 12,468)
- Teaching hospital, Germany.
  - 322 patients who were operated on for pHPT from 1979-1993
  - 120 of the 322 patients had simultaneous thyroid resection for either nodular goitre or solitary adenoma; 9 patients had differentiated thyroid cancer
- The cure rate in the currently reported series of cases was 98% with no instances of recurrent laryngeal nerve injury or permanent hypoparathyroidism







# Case (V)

- 
- Male child 14 yrs. Old
  - c/o: headache & Lt. Nasal obstruction of 6 months duration & epistaxis
  - Lt. eye diminution of vision
  - ENT examination:
    - CT scan:
    - Carotid angiography: nasopharyngeal angio-
    - MRI:
    - Biopsy: **Angiofibroma**



# Review of Literature

- Juvenile nasopharyngeal angiofibroma (JNA) is a benign, highly vascular tumour
- JNA is uncommon and accounts for approximately 0.5% of all head and neck neoplasms.
- The median age at diagnosis is 15 years.
- Presenting symptoms commonly include:
  - nasal obstruction and epistaxis.
  - Approximately 20% of patients have evidence of skull base invasion at the time of diagnosis.
- Approximately 90% of patients with complete resection; local control rates varied from 61% to 87%.



# Staging

Radkowski et al,(1996)

- IA** Limited to nose and/or nasopharyngeal sinus
- IB** Extension into  $\geq 1$  sinus
- IIA** Minimal extension into PMF\*
- IIB** Full occupation of PMF with or without erosion of orbital bones
- IIC** Or posterior to pterygoid plates
- IIIA** Erosion of skull base – minimal ICE\*\*
- IIIB** Erosion of skull base – extensive intracranial extension with or without cavernous sinus invasion

\* pterygopalatine fossa

\*\* Intracranial extension

# Results of Surgery

The likelihood of local control after surgery was inversely related to tumour extent

Stage	Number of patients -	Disease free -	Symptom free	Recurrence
IA	3	3	0	0
IB	4	3	1	0
IIC	8	5	2	1
IIIA	21	12	3	6
IIIB	8	1	3	4
Total	44	24 (55%)	9 (20%)	11 (25%)



# Radiotherapy

- The local control rates after RT ranged from 73% to 100%.
- Tumour regression after successful RT is often slow.
- Clinical evidence of residual tumour in 50% of pts. - 1 year after RT, and in 10% of pts. 3 years after RT
- Median time to recurrence was 16 months (range, 12 to 101 months).
- All patients who experience a local recurrence after RT undergo successful surgical salvage /or a second course of RT, thus ultimate local control approaches 100%



# COMPLICATIONS

## ○ **Surgery (^30-40%)**

### Postoperative complications

- 17/44
- 5/18
- 11/30
  - Haemorrhage - mean blood loss of 350 cc (range, 50 cc to 750 cc).
  - Nerve palsies.
  - nasolacrimal duct stenosis
  - infection

## ○ **Radiotherapy (15%)**

- Hypopituitarism
- cataracts
- growth abnormalities,

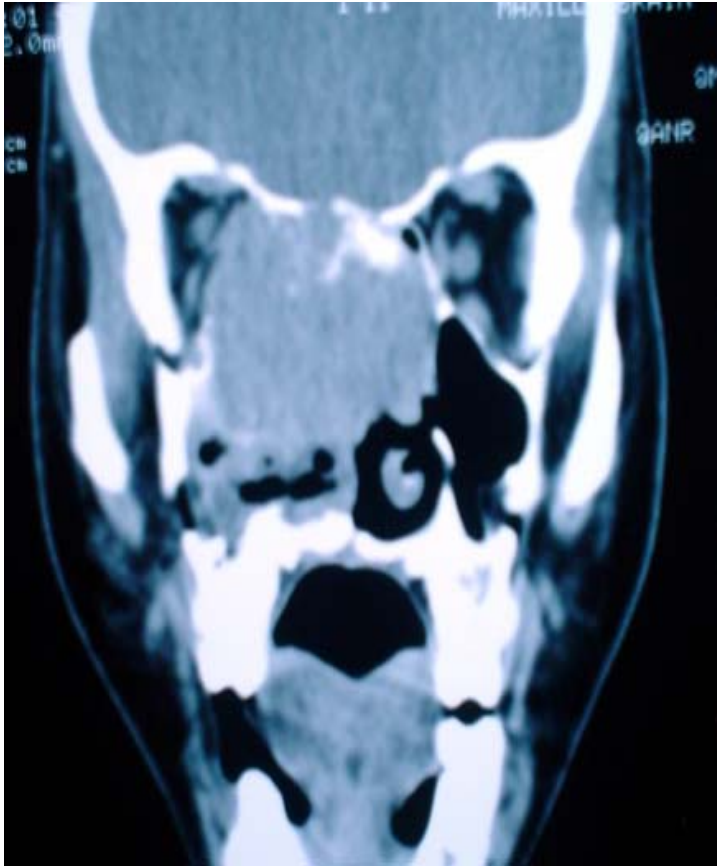


# CONCLUSIONS AND TREATMENT GUIDELINES

- Cure after surgery or RT is approximately 80% to 85%;
- Probability of ultimate local control approaches 100%.
- Completely resectable tumors with relatively limited morbidity should be treated surgically.
- Extensive tumours with significant intracranial invasion (likelihood of complete resection with acceptable morbidity is modest).  
Best treated with moderate-dose RT.
- Dose fractionation is 35-45 Gy in 19-25 fractions (once daily.) using 3-dimensional CT-based treatment planning.
- Tumours that exhibit significant anterior extension may be better treated using a anterior field and 1 or 2 lateral fields. The likelihood of significant complications with this approach is low.



## Juvenile nasopharyngeal angiofibroma (JNA)

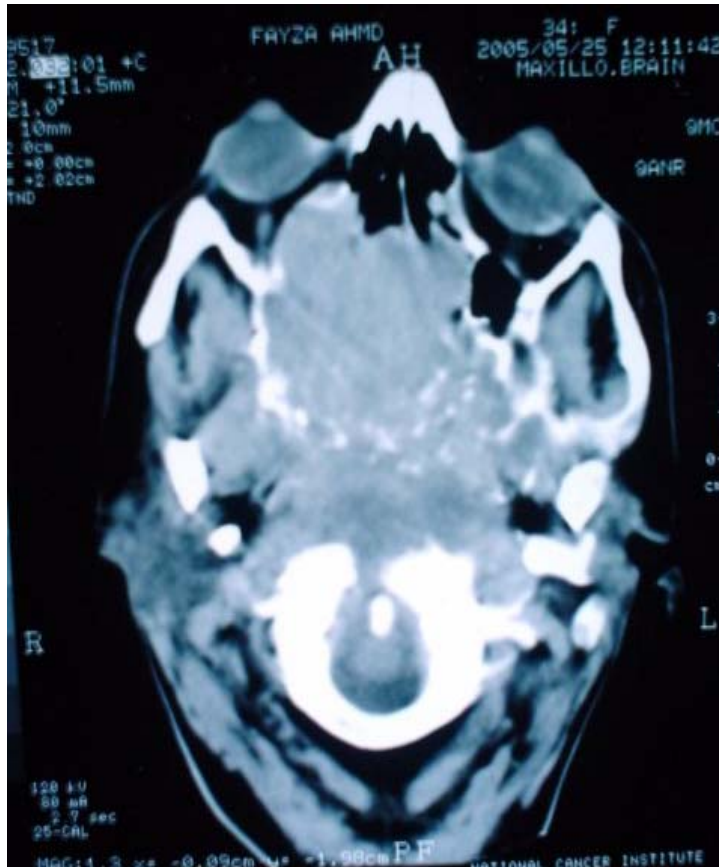


Skull base  
chordoma

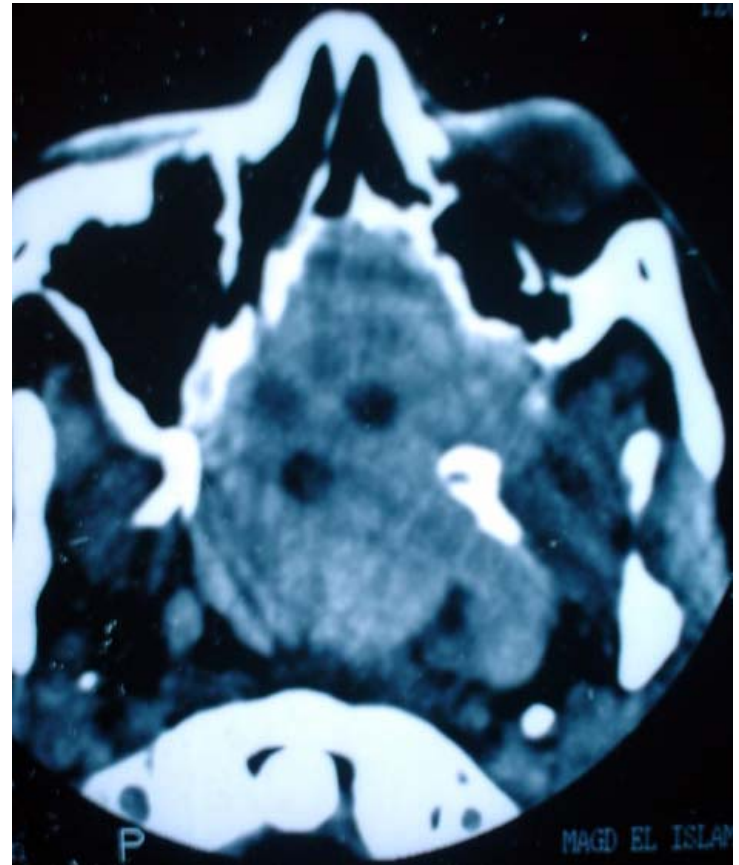




## Juvenile nasopharyngeal angiofibroma (JNA)



Skull base  
chordoma







THANK YOU

AND I HOPE IT WAS INTERESTING

**Ehab M. Khalil**