KREP

2025

KARNATAKA RADIOLOGY EDUCATION PROGRAM

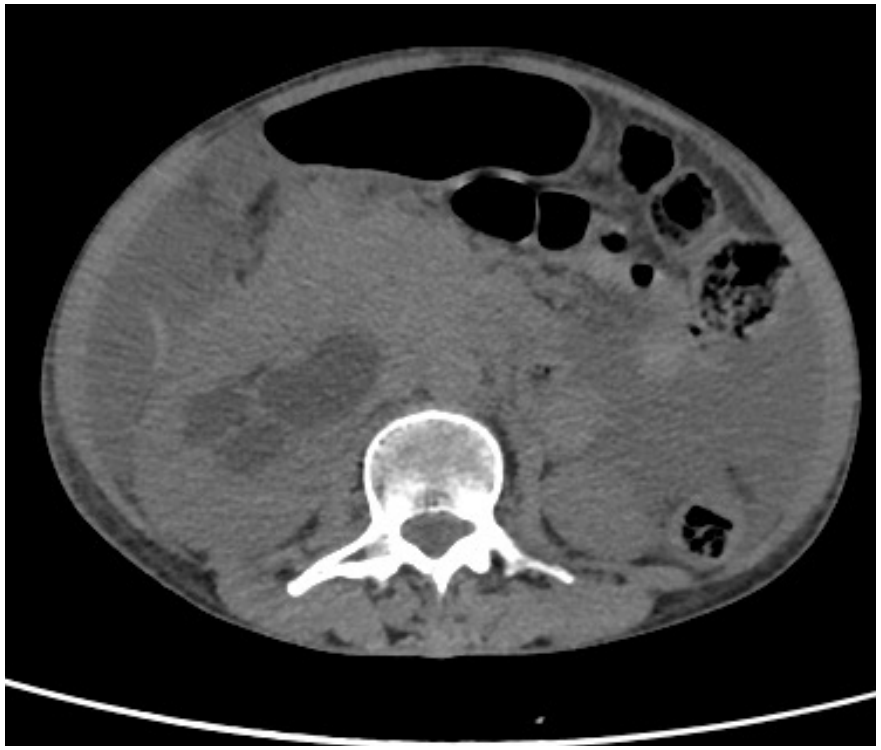
CASE PRESENTATION

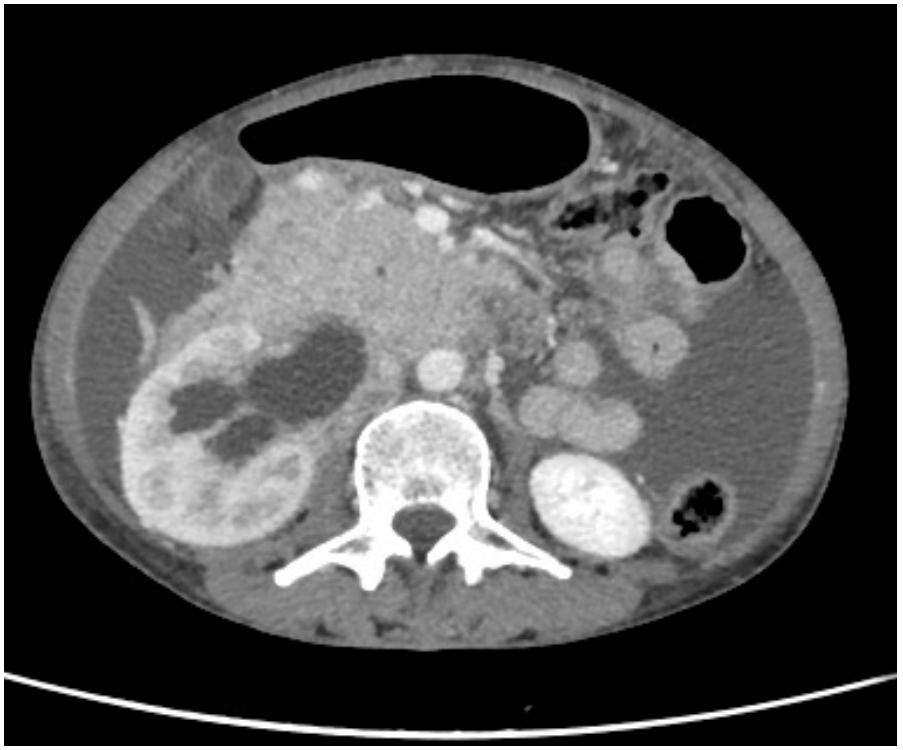
MENTOR: DR G C PATIL
KMCRI,HUBBALLI.

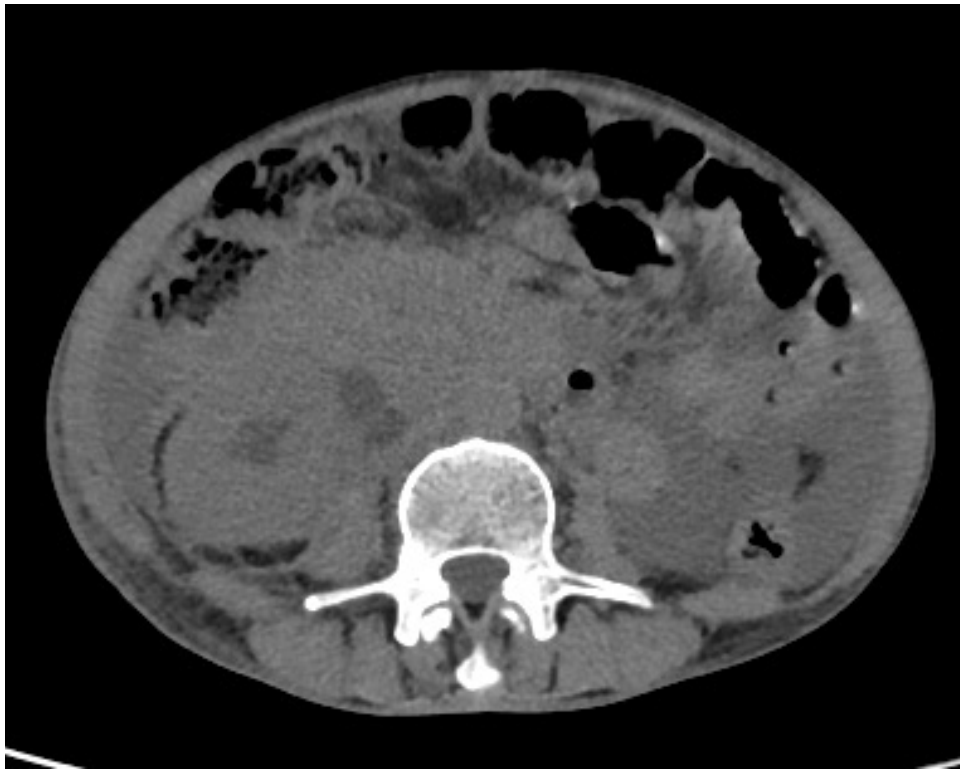
CASE 1

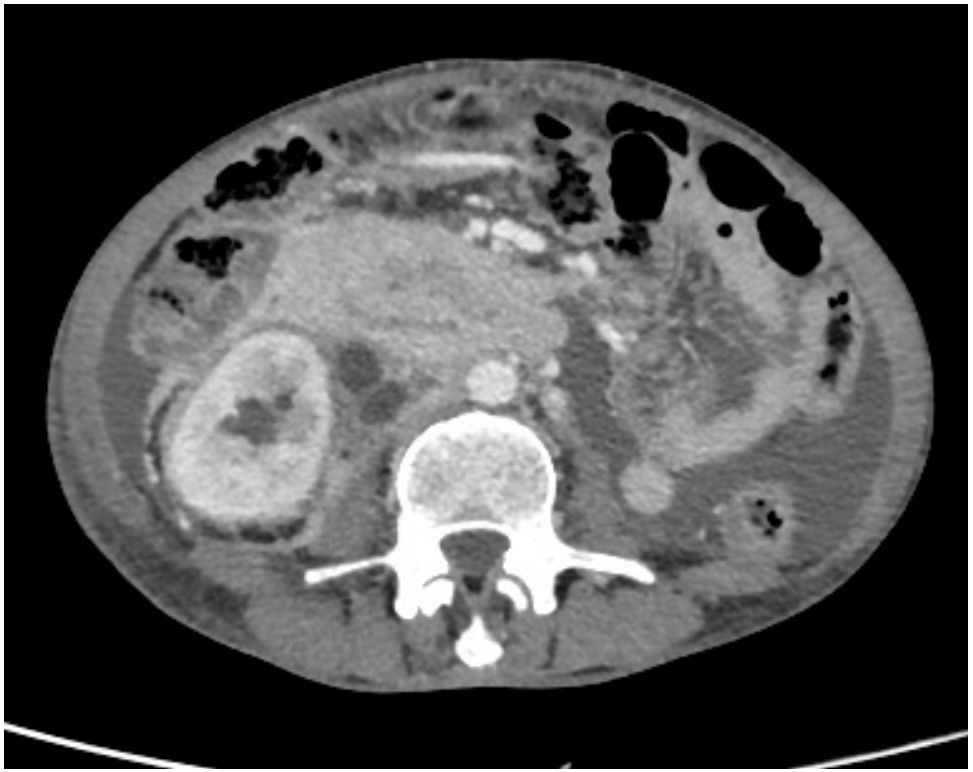
61-Year-old female patient, with RVD positive status, presented with abdominal pain and distension since 2 months

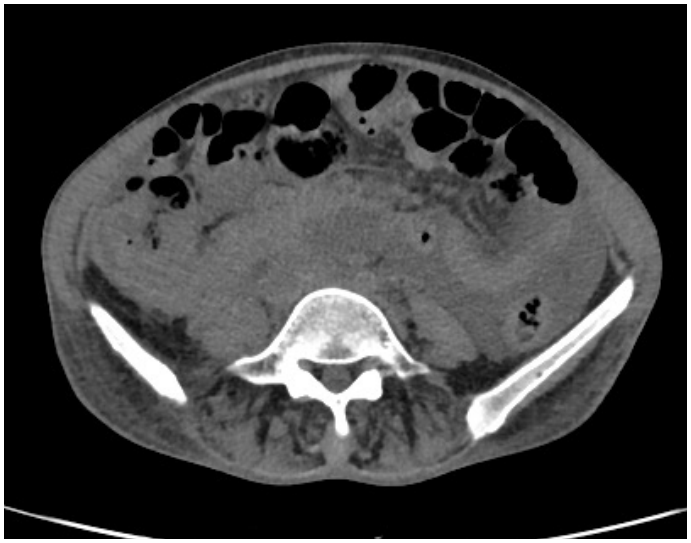
IMAGING FINDINGS

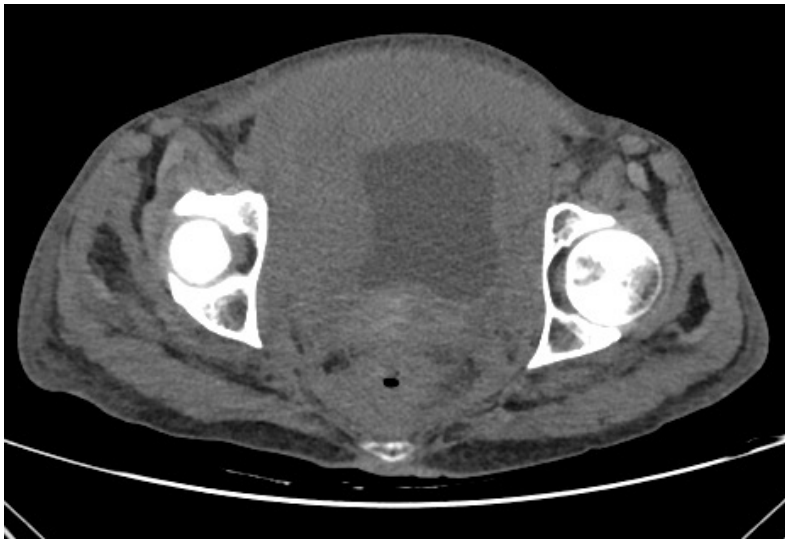


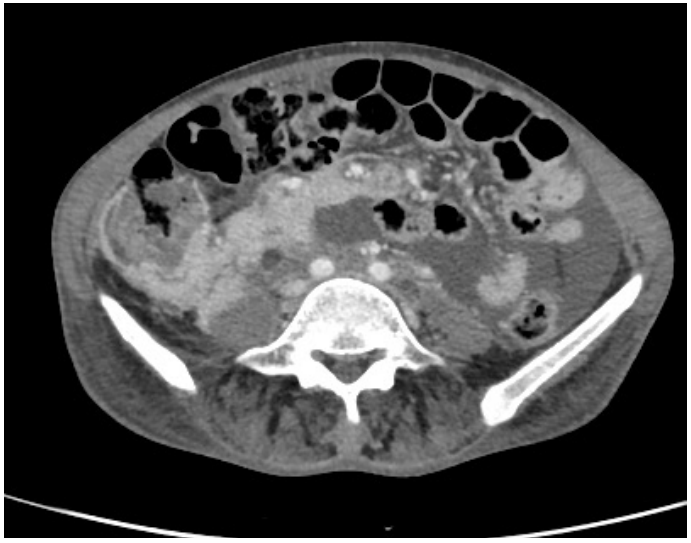


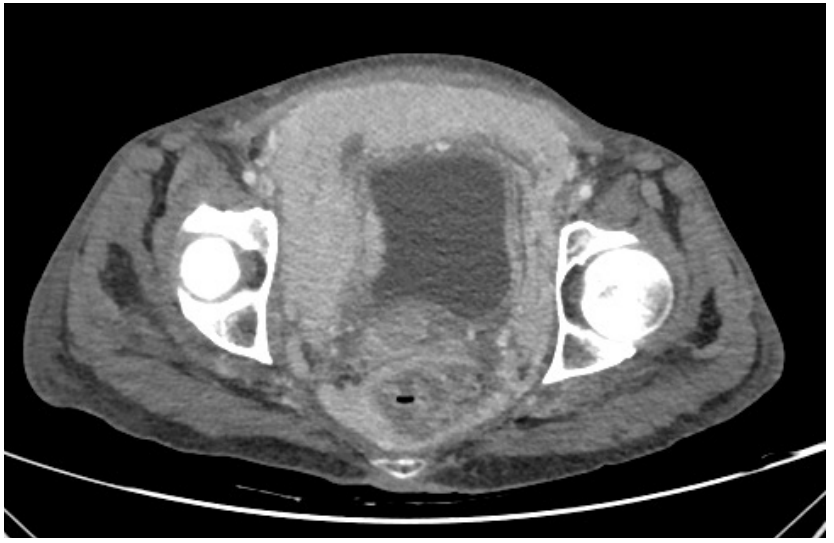


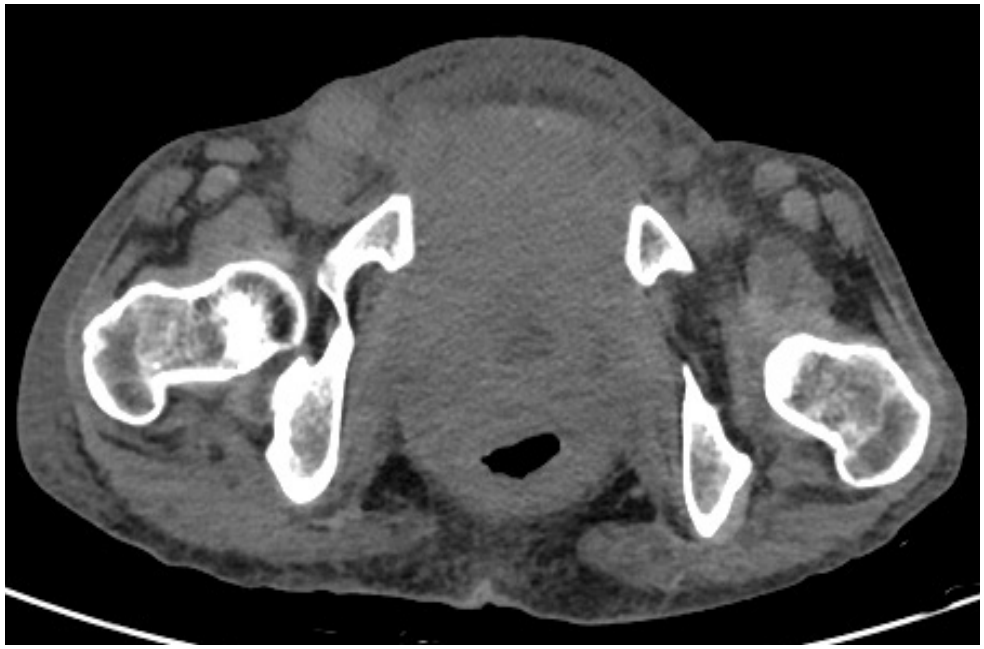


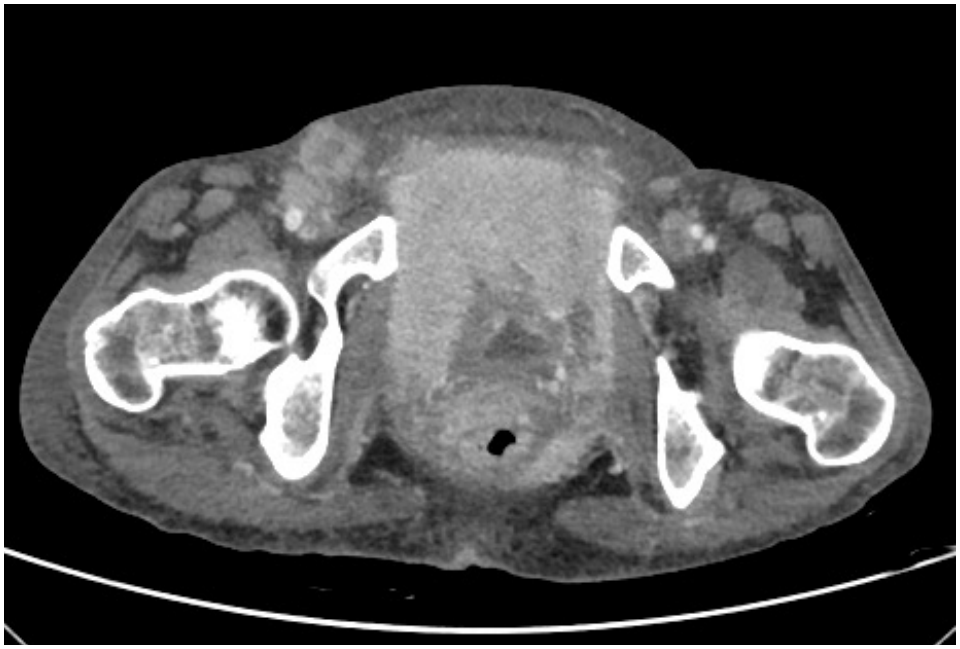


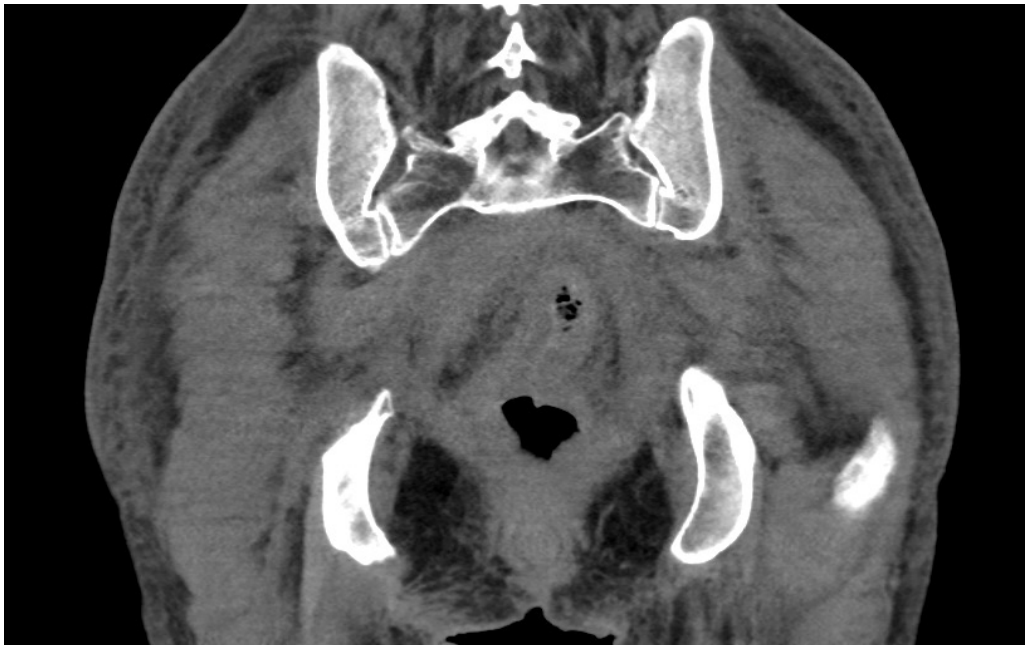


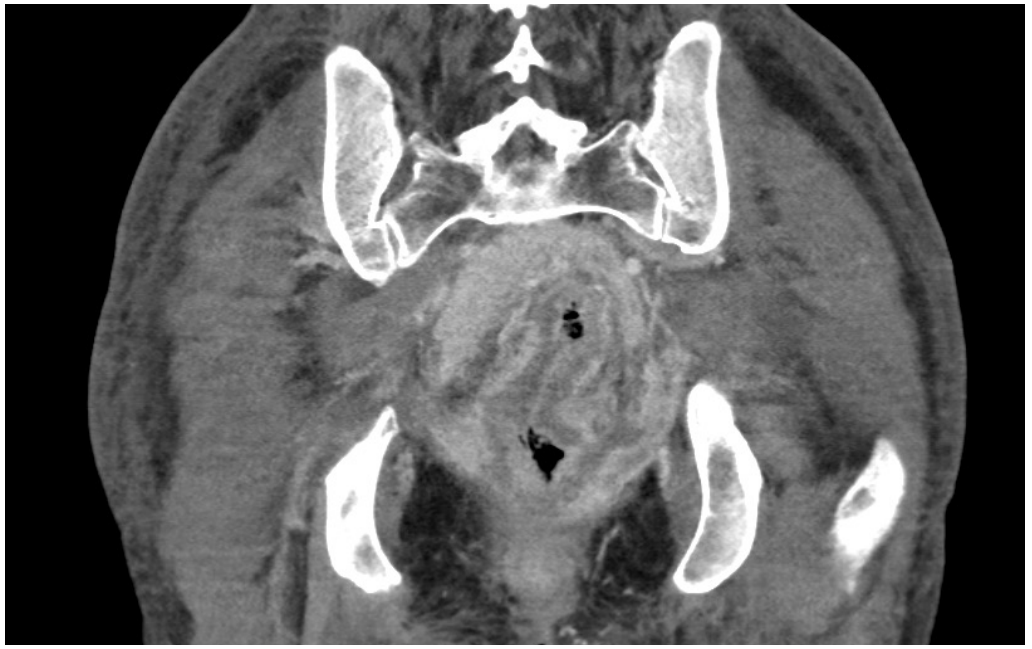


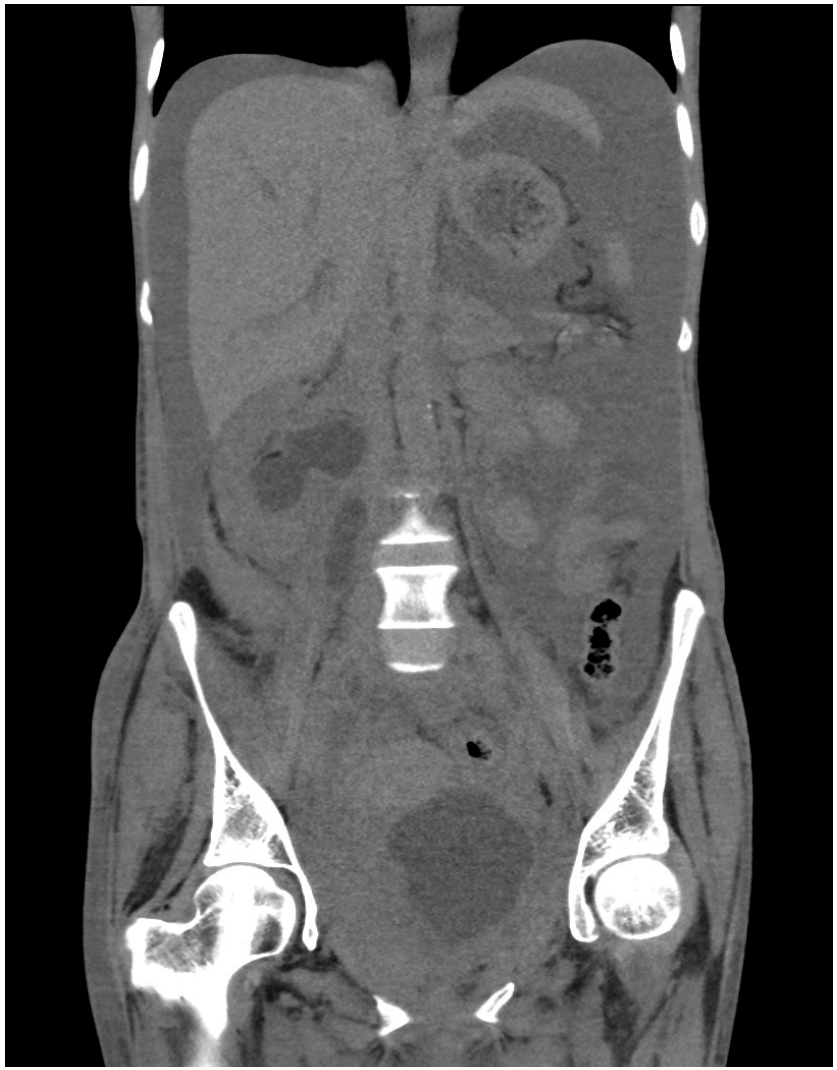


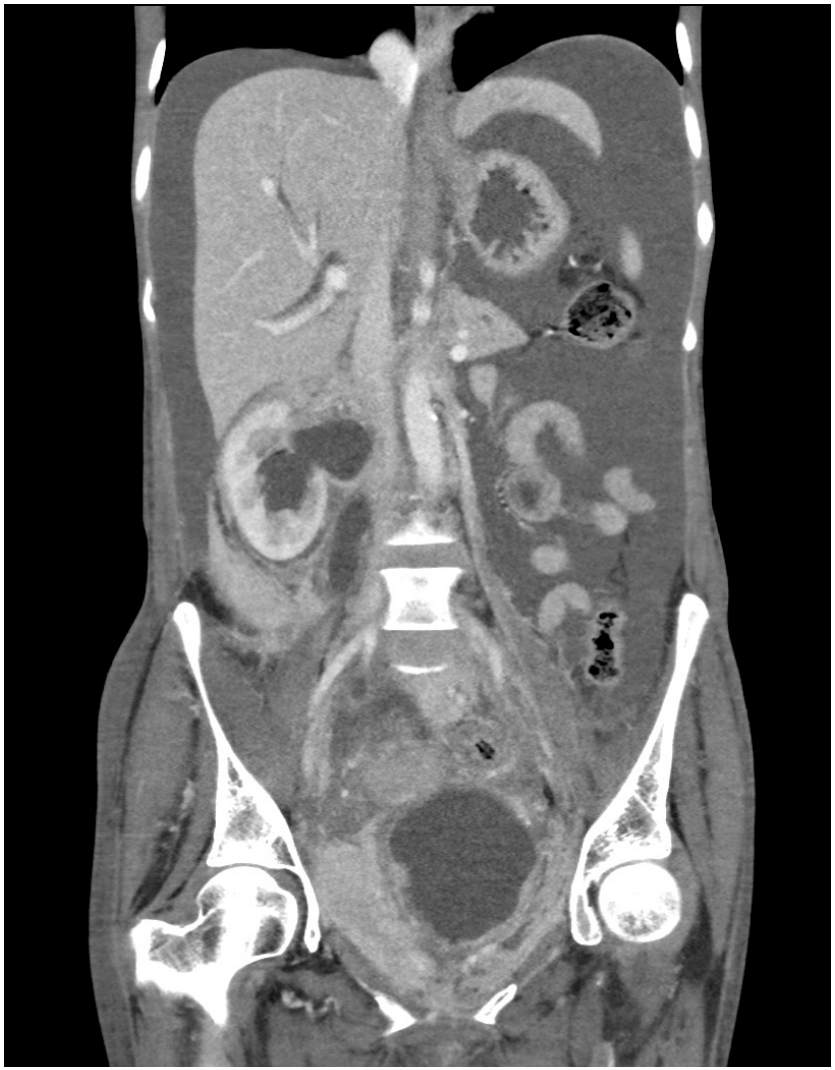


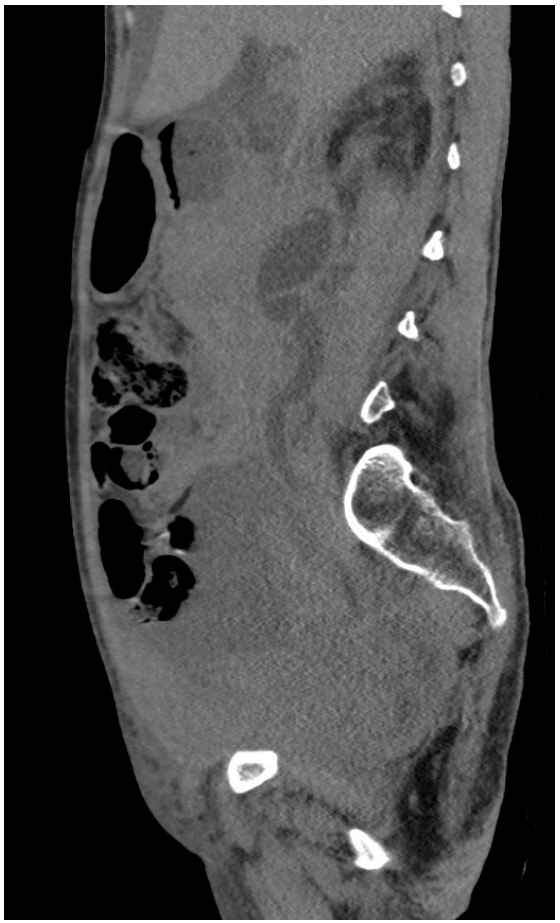


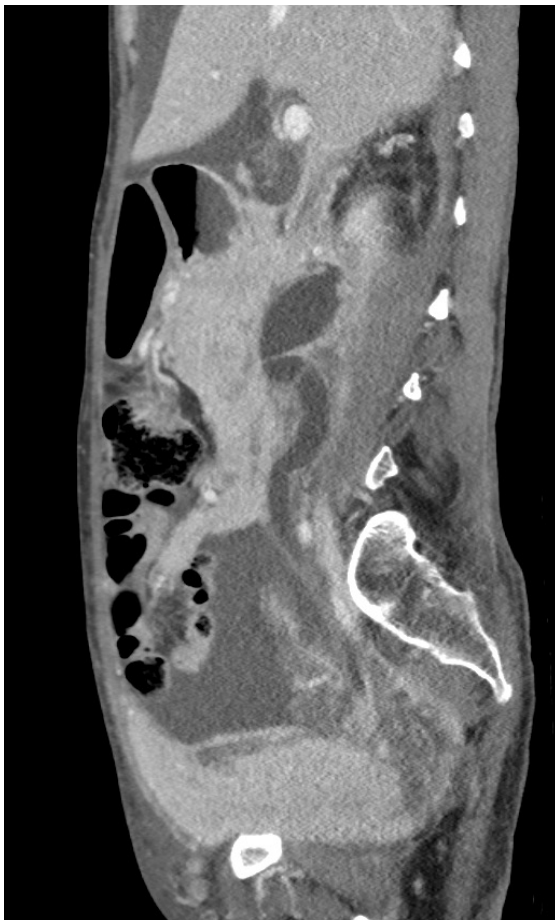


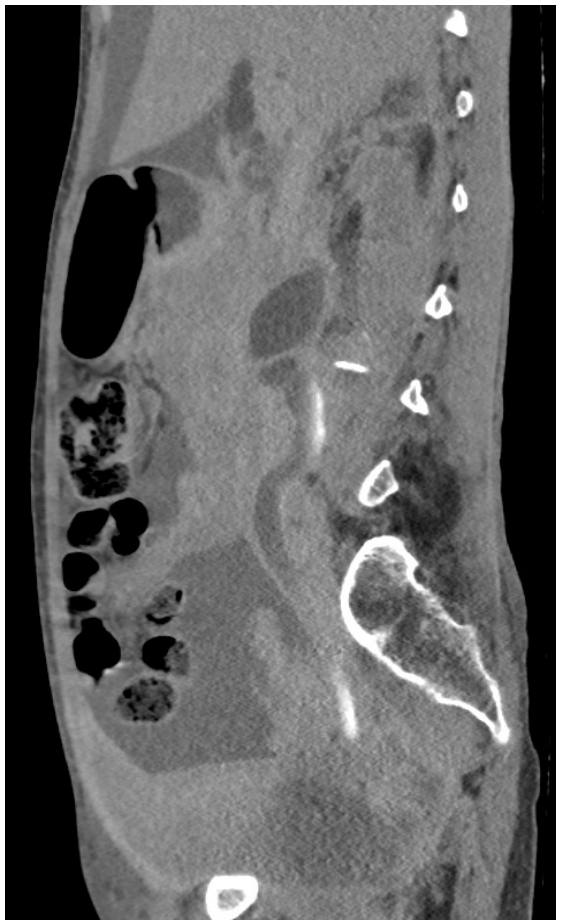


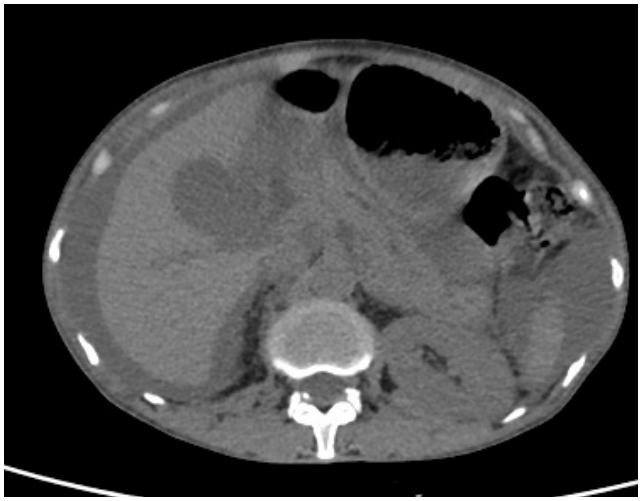


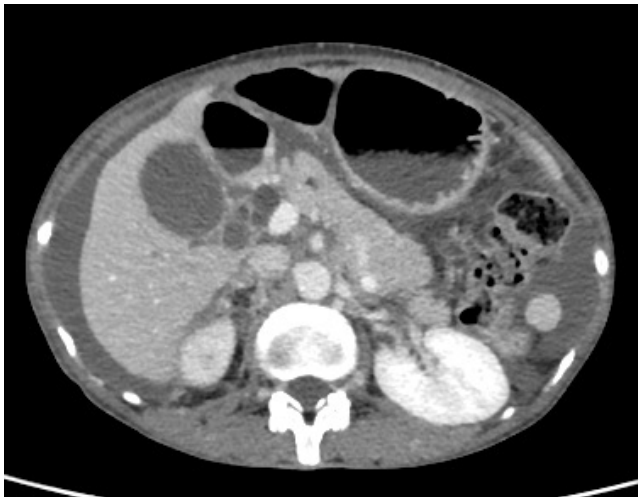


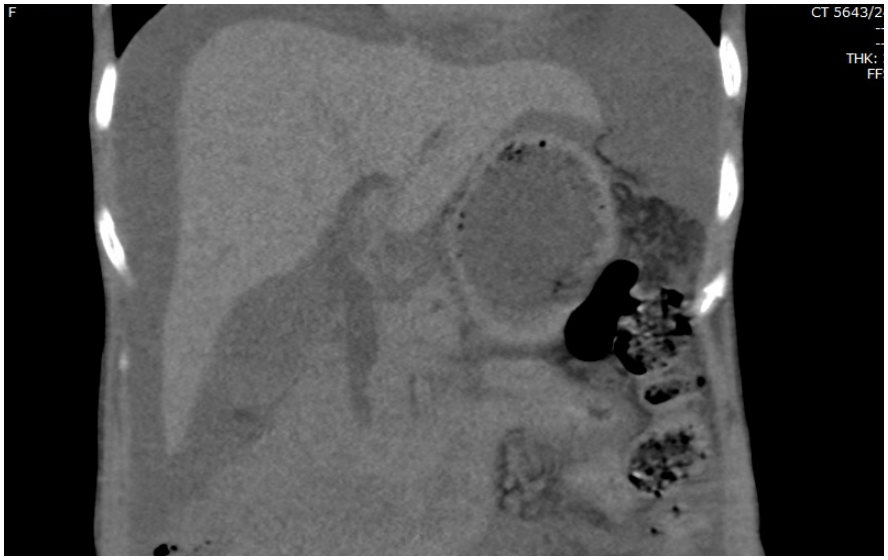








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To summarize

- Gross collection in peritoneal cavity with omental thickening and nodularity along the superior mesenteric vessels, in peripancreatic, para-duodenal and in the pelvic cavity adjacent to the bladder, presacral and perirectal region showing enhancement.
- Bilateral inguinal lymphadenopathy-likely necrotic.
- Symmetrical thickening of rectum, sigmoid colon and caecum and proximal part of ascending colon.
- Right moderate hydronephrosis with dilatation of proximal ureter with hypodense enhancing content in the distal ureter - secondary to the involvement of the ureter.
- Pyelitis with ureteritis and cystitis.
- Dilated intrahepatic biliary radicles and common bile duct with prominent main pancreatic duct.

DIFFERENTIAL DIAGNOSES

1. Peritoneal lymphoma
2. Kaposi sarcoma

ANATOMICAL SITE : Right inguinal lymph node

BIOPSY NO : 7429/24

GROSS: Received specimen consists of 2 grey white to grey yellow cores of tissue, largest measuring 1.5cm in length and smallest measuring 1cm in length and one tiny bit of tissue measuring 0.1x0.1x0.1cm. Entire tissue processed.

Grossed by : Dr.Prasanna and Dr.Rishi

MICROSCOPY : Sections studied shows pleomorphic cells arranged in nests, sheets. Individual cells shows high N:C ratio, coarse chromatin, nuclear irregularities. Surrounded by fibroadipose tissue showing infiltration by mononuclear inflammatory cells.

Result category- Neoplastic-malignant

IMPRESSION: Histologic features suggest possibility of POSITIVE FOR MALIGNANCY. Possibilities are: 1. HIGH GRADE LYMPHOMA, 2. MELANOMA, 3. POORLY DIFFERENTIATED CARCINOMA.

Kindly discuss and furnish clinical details.

Note: All malignant specimens will be preserved for 6 months and non malignant specimens will be preserved for one month only.

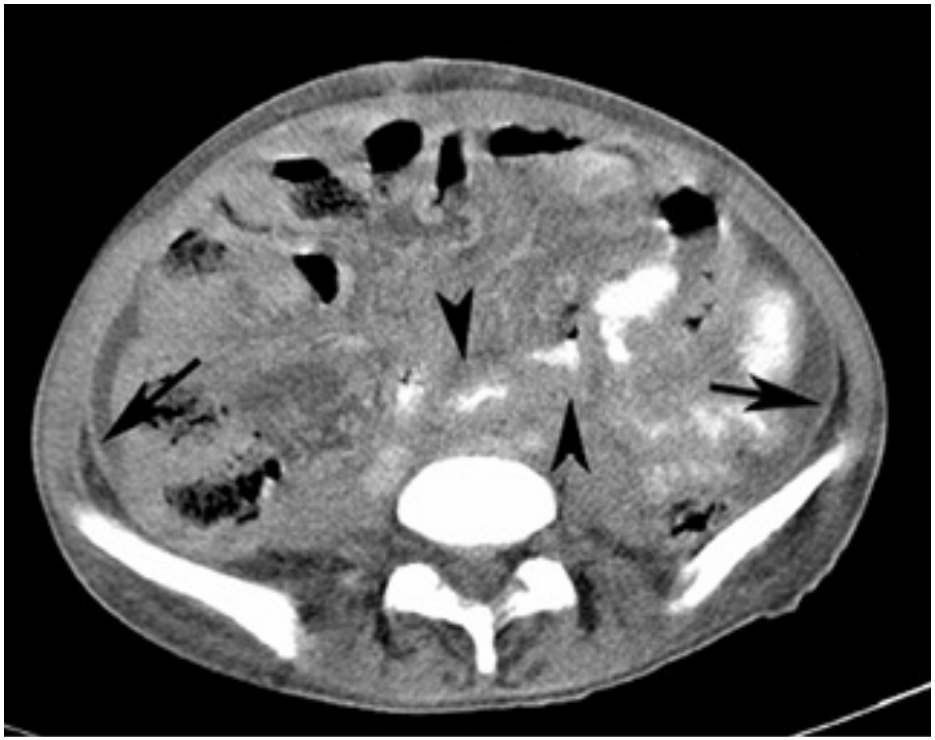
LYMPHOMA

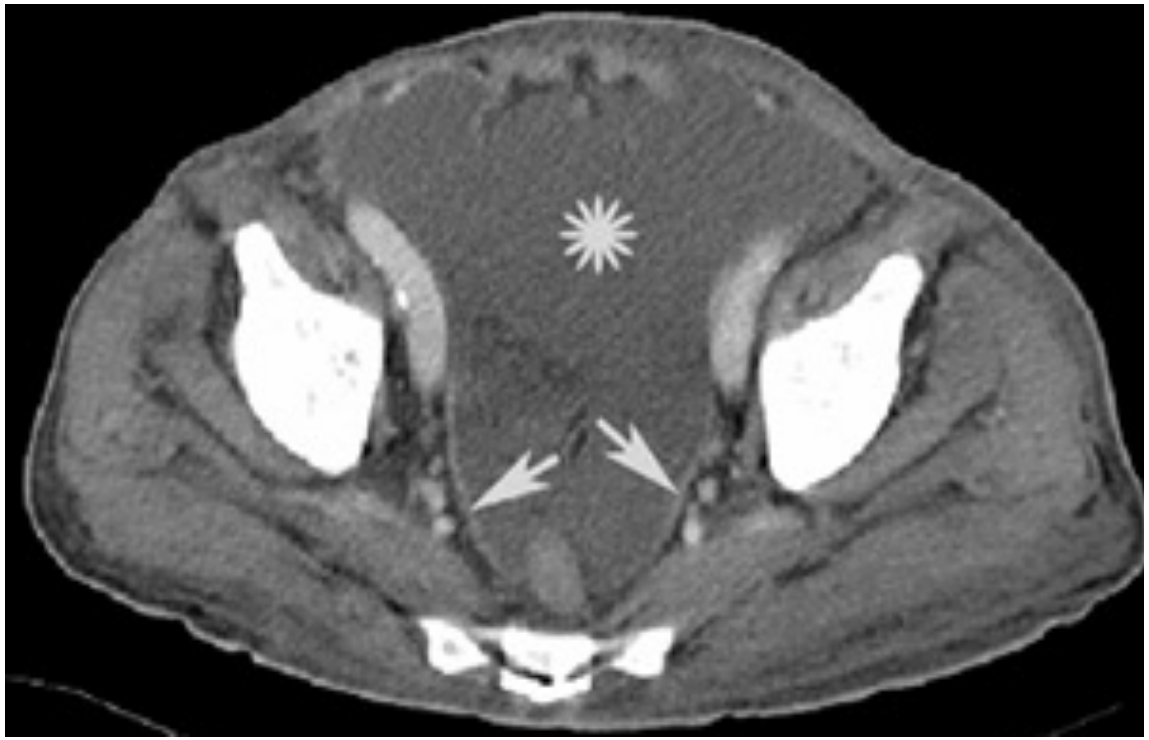
- Lymphoma is a complex group of neoplasms derived from lymphoid cell lines.
- It comprises of two groups – Hodgkin's lymphoma (HL) and Non-Hodgkin's lymphoma (NHL)
- HL – incidence peaks at 20-30 years of age
- NHL – Incidence increases with age after 20 years – associated with pre-existing immunosuppression
- FDG PET/CT –imaging technique of choice for staging of the disease

IMAGING IN LYMPHOMA

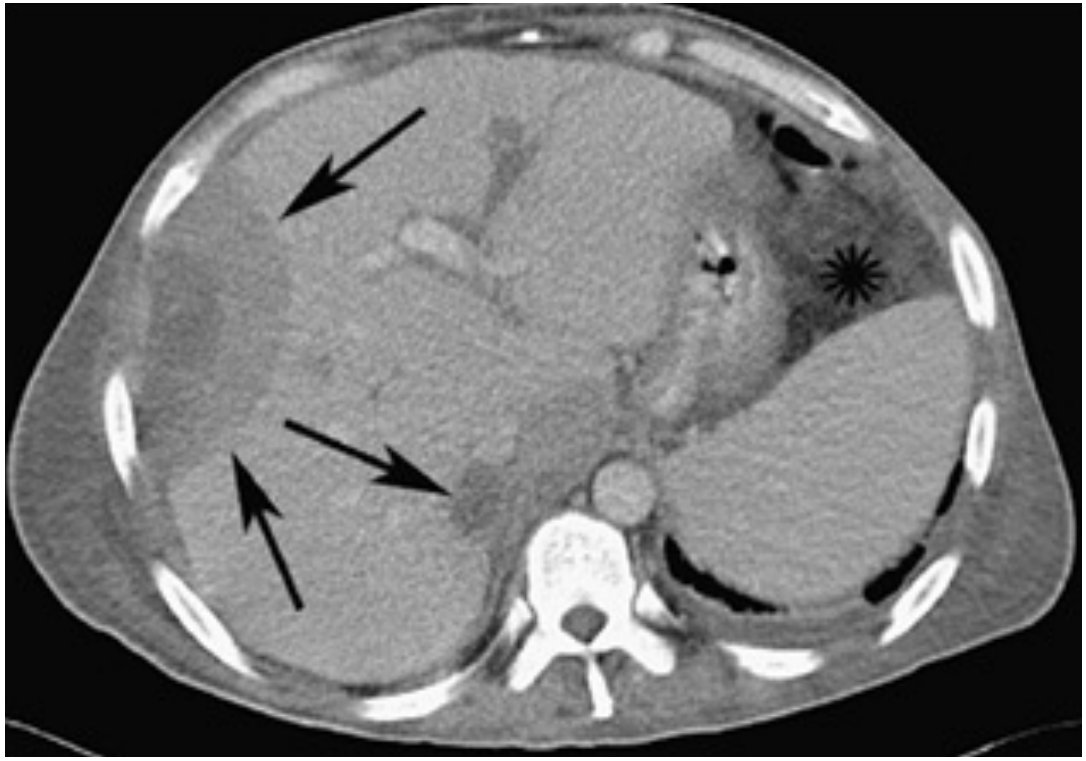
- USG:
  - Nonspecific
  - Pattern of vascular perfusion of lymph nodes (Central and peripheral perfusion) on doppler study may suggest diagnosis.
  - Also assess major viscera
  - No value in whole-body staging
- CT:
  - Demonstrate full extent of disease
  - Localize most appropriate lesion for percutaneous image-guided biopsy
  - Highly sensitive in the detection of peritoneal thickening and subcentimetric nodules, specially in the presence of ascites.
- MRI:
  - Whole body –DWI – for staging and response assessment – under research

- Peritoneal thickening - indicative of abdominal malignant seeding - smooth diffuse and irregular-nodular thickening
- Omental involvement:
  - Small areas of nodularity and fat stranding
  - Thick soft tissue mass replacing the entire omentum anterior to transverse colon
  - Can extend to pelvic cavity
- Associated secondary sign - non loculated, non septated ascites.





Karaosmanoglu D, Karcaaltincaba M, Oguz B, Akata D, Özmen M, Akhan O. CT findings of lymphoma with peritoneal, omental and mesenteric involvement: Peritoneal lymphomatosis. European Journal of Radiology. 2009 Aug;71(2):313–7.



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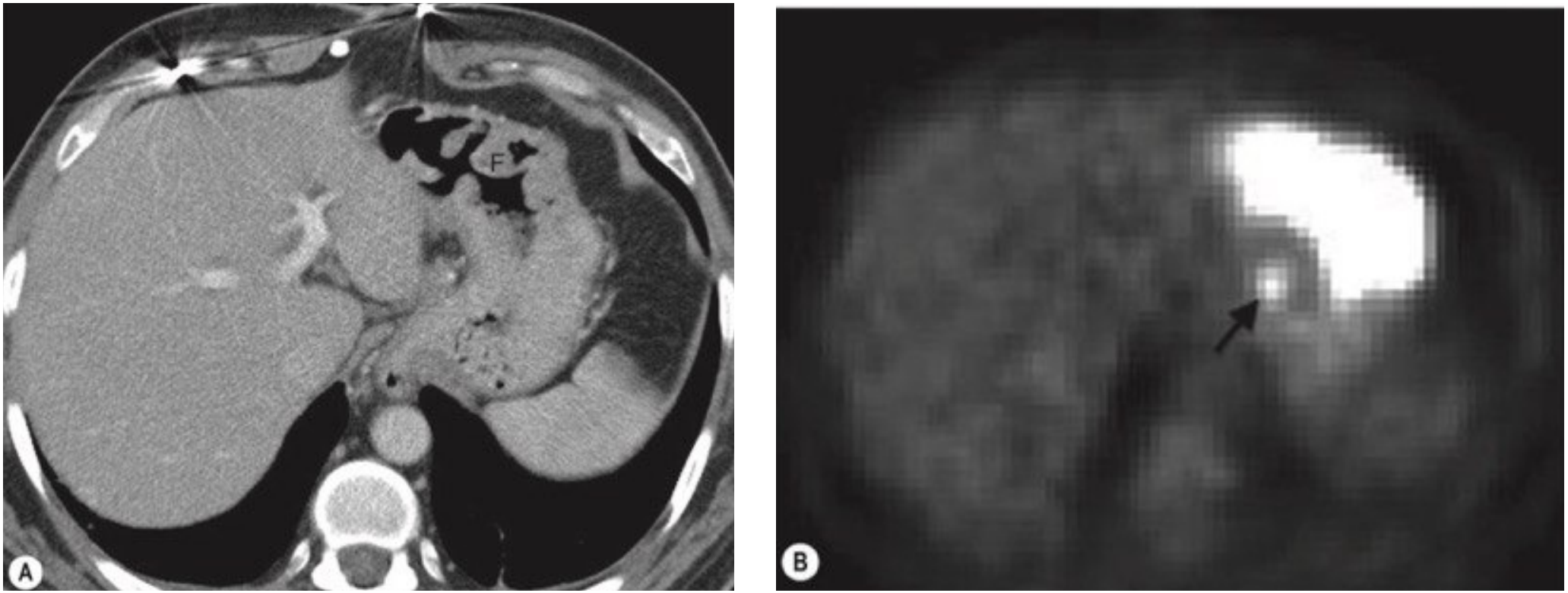
- Differentials for omental involvement:
  - Metastases from carcinoma- ovary, colon, stomach, pancreas, breast, endometrium; adenocarcinoma of unknown primary
  - Lymphomas
  - Sarcomas
  - Melanomas

- Lymphoma can occur at any part of the body
- Isolated or conglomerated lymphadenopathies are the most common imaging findings.
- Extensive infiltration of the peritoneum secondary to lymphomas is rare and many primary and secondary peritoneal neoplasms have similar imaging findings.
- Omental and intestinal lymphomas - dissemination secondary to gastrointestinal carcinomas
  - via the pathways: gastrocolic ligament, transverse mesocolon and visceral peritoneal surfaces

Staging of primary GI lymphoma

- As per Consensus Conference in Luguano in 1993
- Stage I: Tumor confined to gastrointestinal tract- single primary site and multiple noncontiguous lesions
- **Stage II**: Tumor extends into the abdominal cavity from the primary gastrointestinal site – **Most common presentation**
  - II 1: local nodal involvement
  - II 2: distant nodal involvement
- Stage III: Penetration through serosa to involve adjacent organs or tissues
- Stage IV: Disseminated extra-nodal involvement or a gastrointestinal tract lesion with supradiaphragmatic nodal involvement.



Source: Gore RM, Levine MS. Textbook of Gastrointestinal Radiology : Expert Consult. London: Elsevier Health Sciences; 2015.

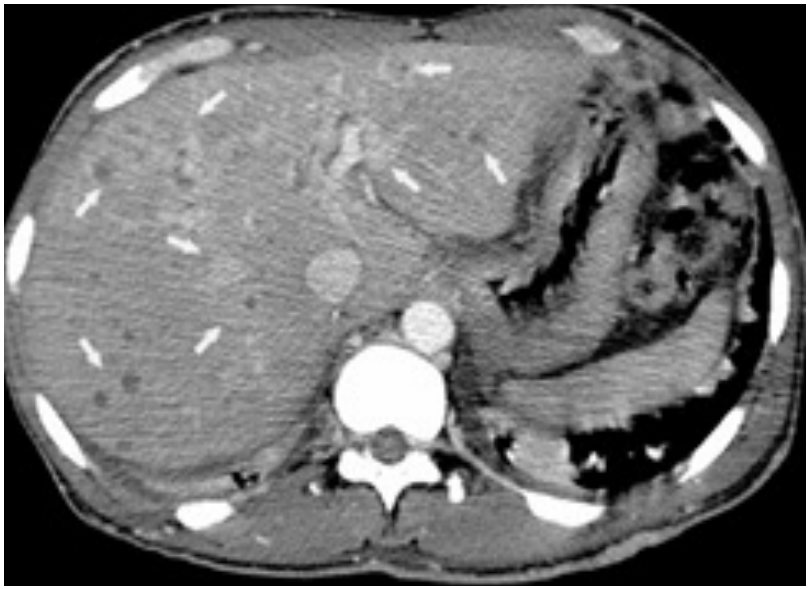
KAPOSIS SARCOMA

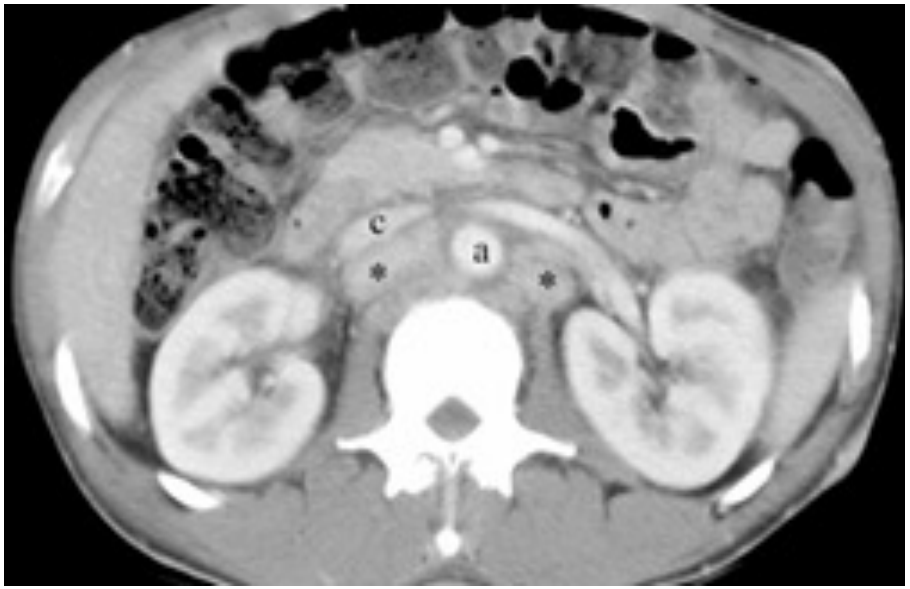
- Low-grade vascular tumour
- Four variants:
  - Classic KS
  - Endemic (African) KS
  - Iatrogenic (organ transplant–related) KS
  - **Acquired immunodeficiency syndrome (AIDS)–related KS**

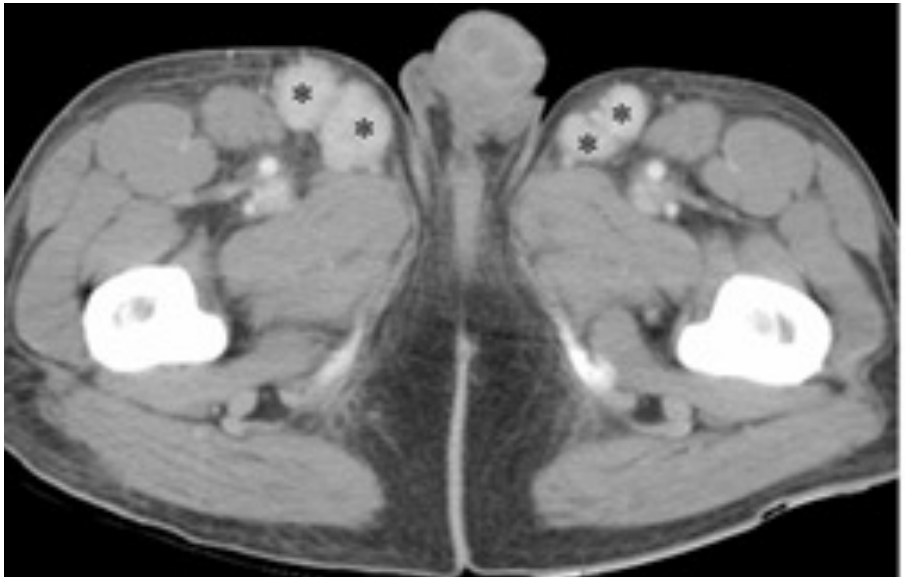
  Low CD4 lymphocyte count (150-200 cells/mm3)

GASTROINTESTINAL DISEASE IN AIDS RELATED KAPOSIS SARCOMA

- Most common visceral involvement in disseminated disease
- Concomitant cutaneous lesions is more common
- Most frequently affected site - duodenum.
- Differential diagnosis:
  - Lymphoma
  - Opportunistic infections
  - Hematogenous metastasis
- Hyperattenuating lymphadenopathies of the porta hepatis or peripancreatic, retroperitoneal, mesenteric, inguinal or pelvic groups (80% of cases)





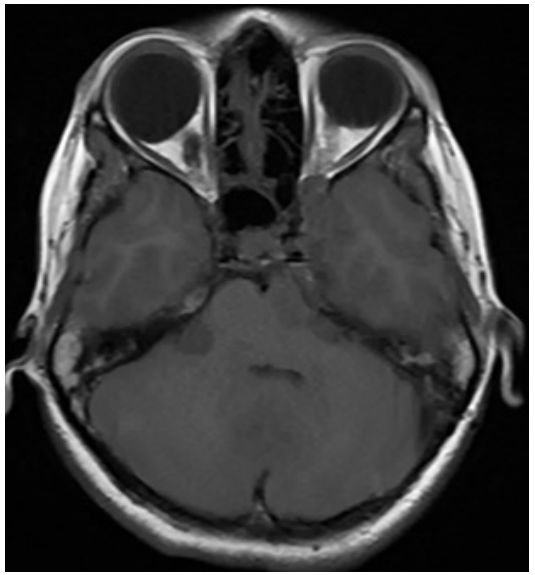
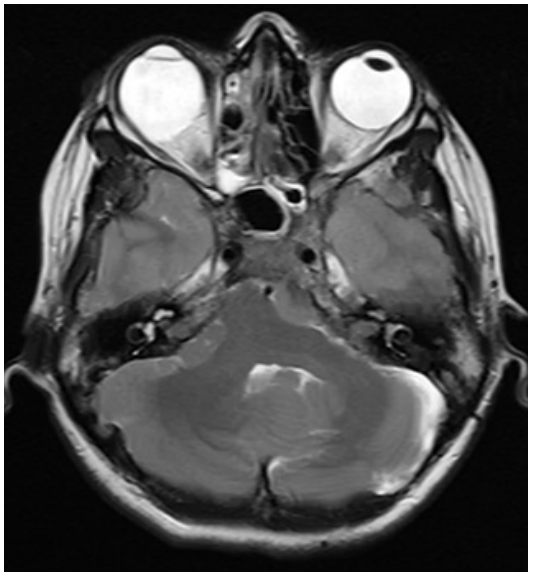
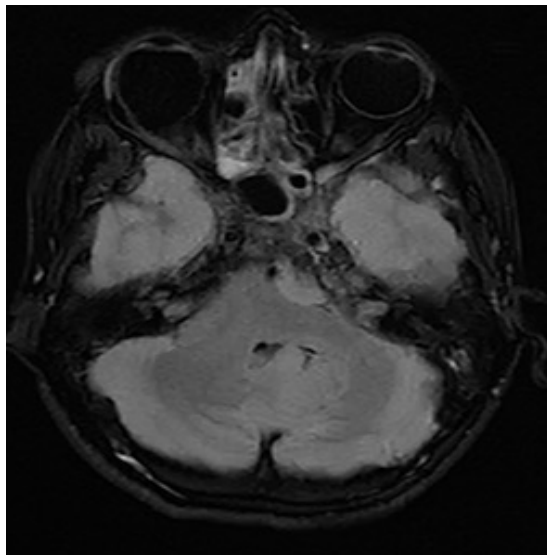
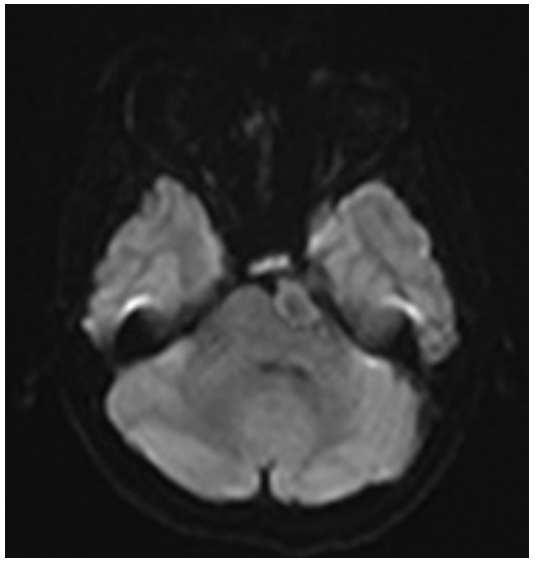
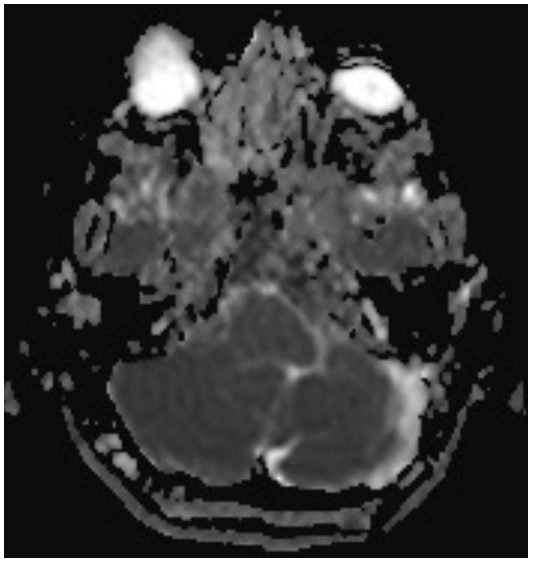
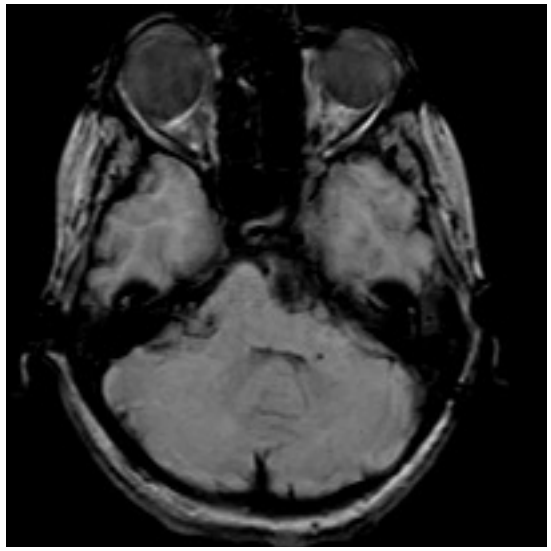


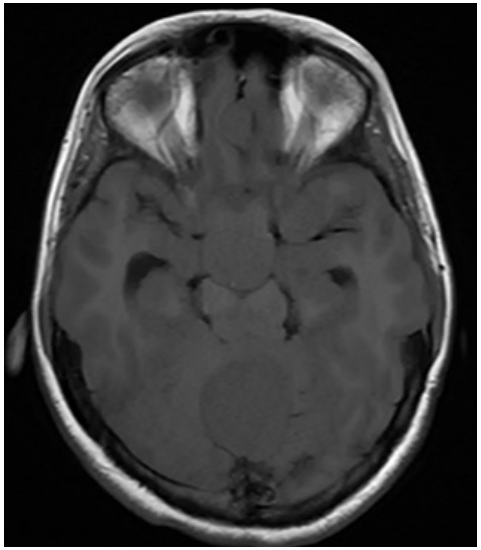
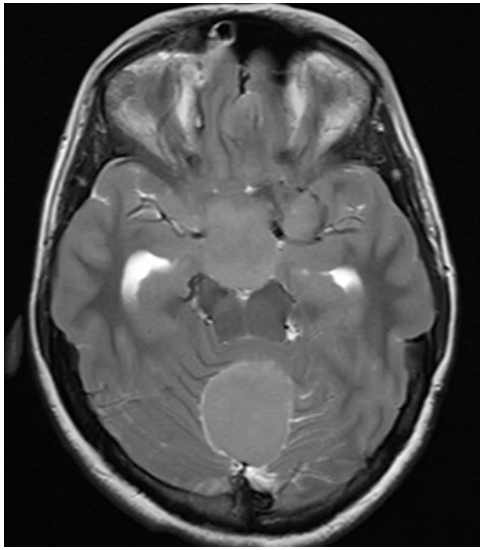
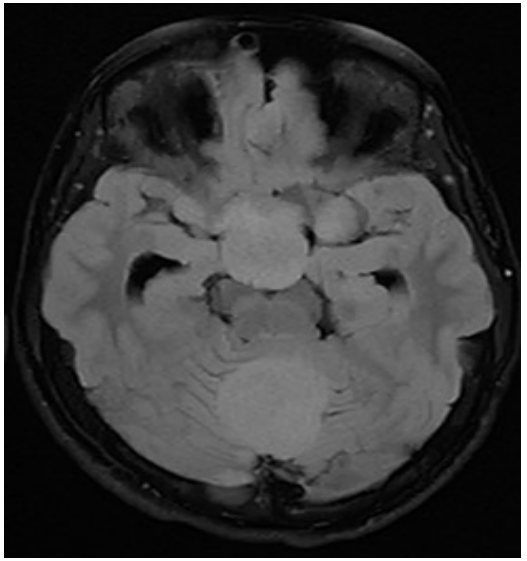
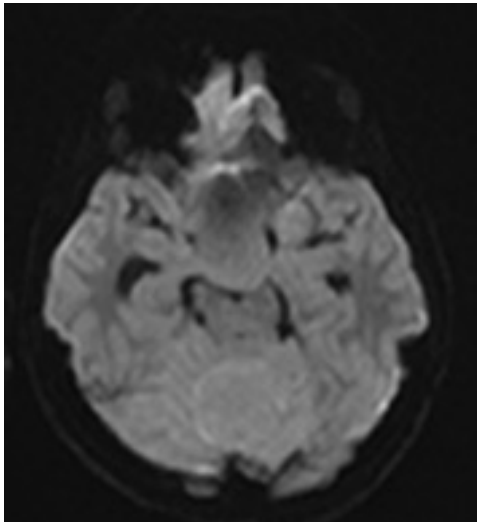
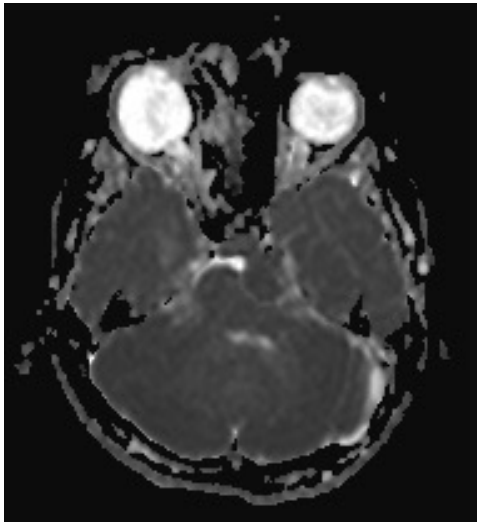
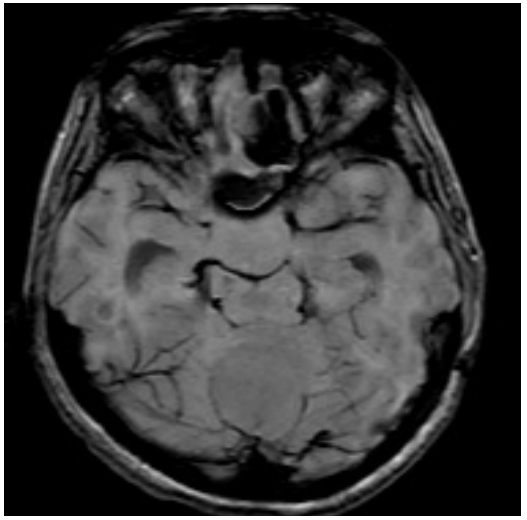
Restrepo CS, Martínez S, Lemos JA, Carrillo JA, Lemos DF, Ojeda P, et al. Imaging Manifestations of Kaposi Sarcoma. RadioGraphics. 2006 Jul;26(4):1169–85.

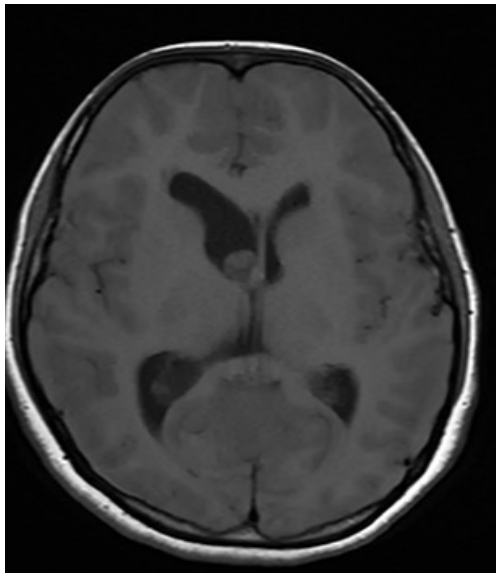
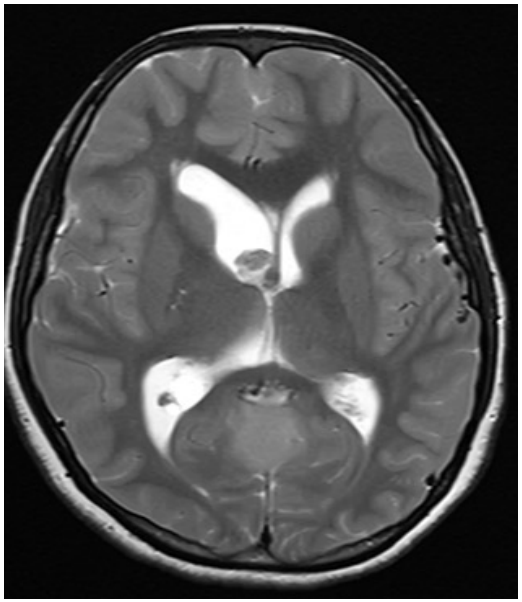
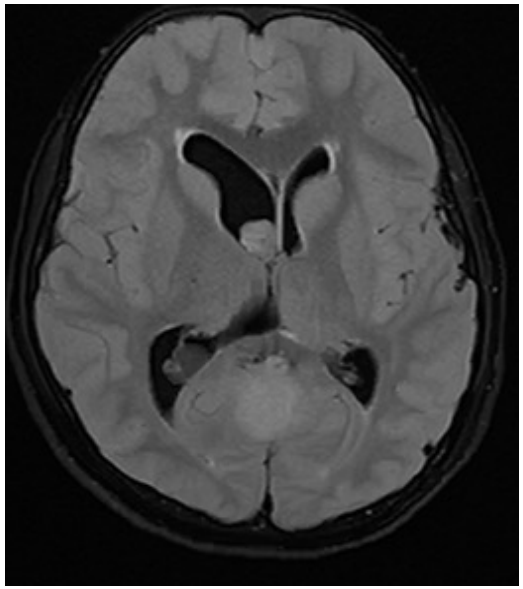
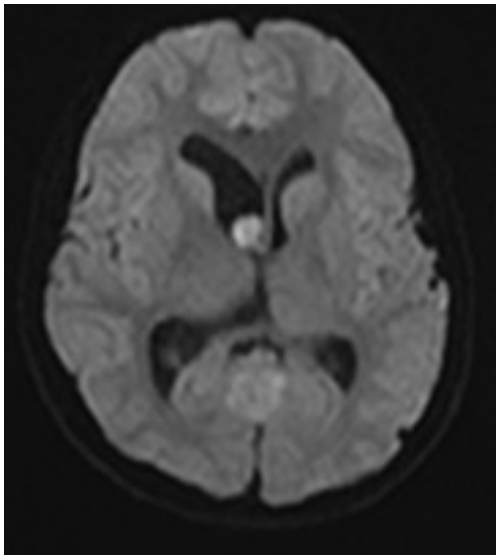
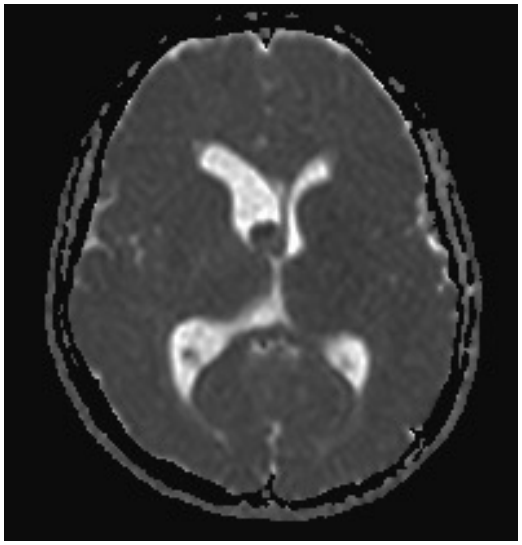
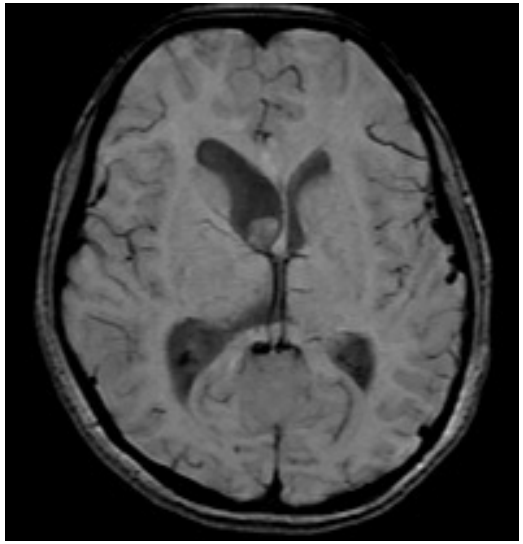
CASE 2

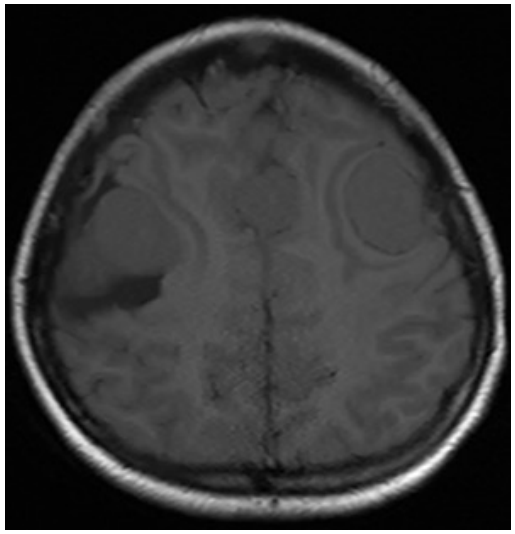
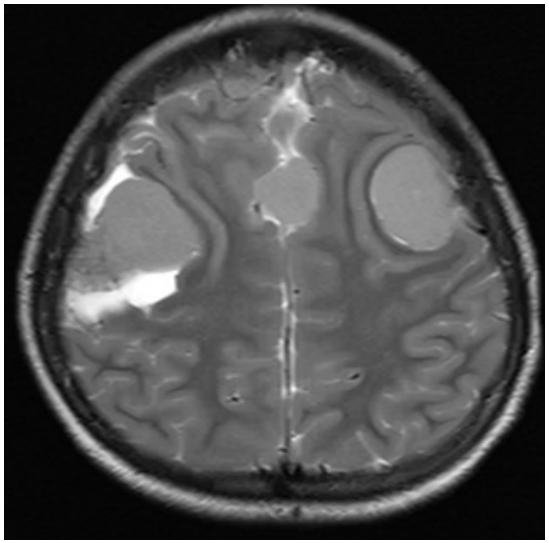
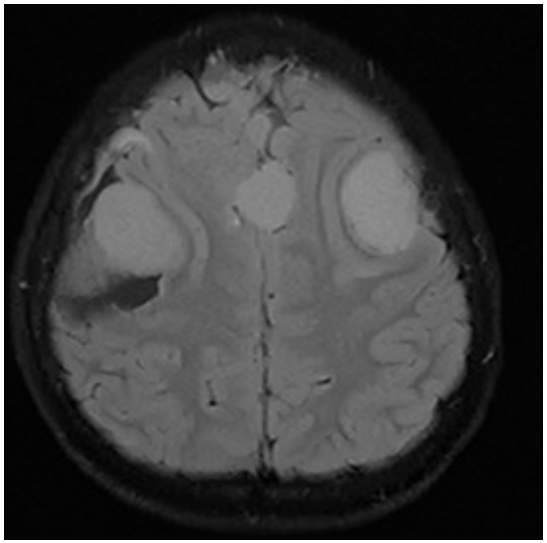
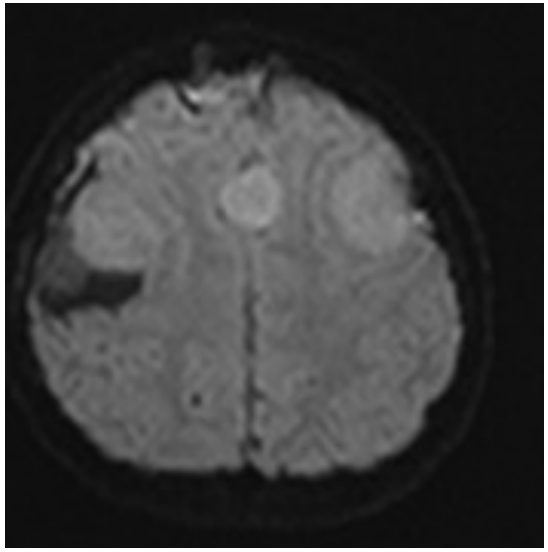
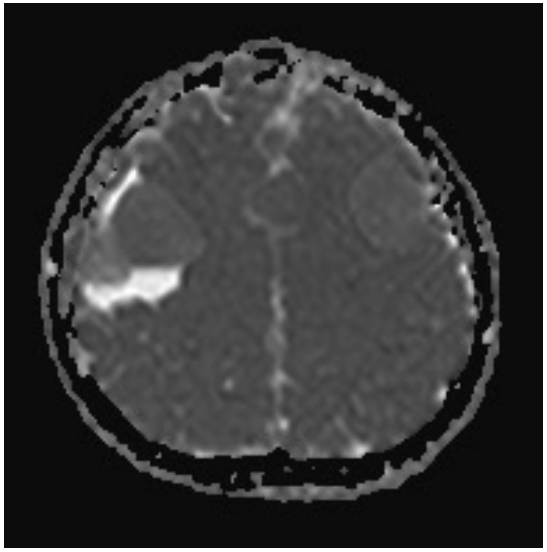
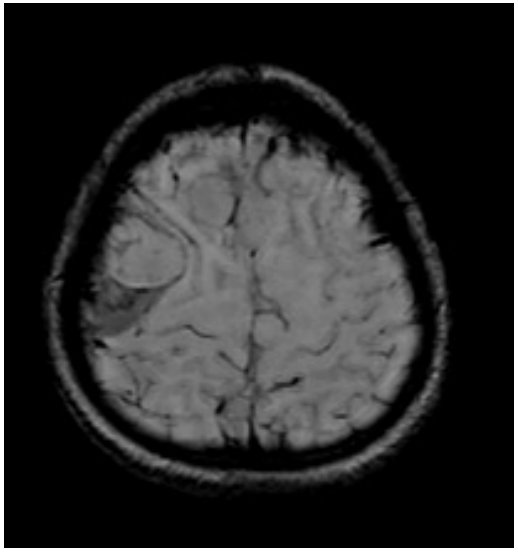
29-year-old female presented with headache

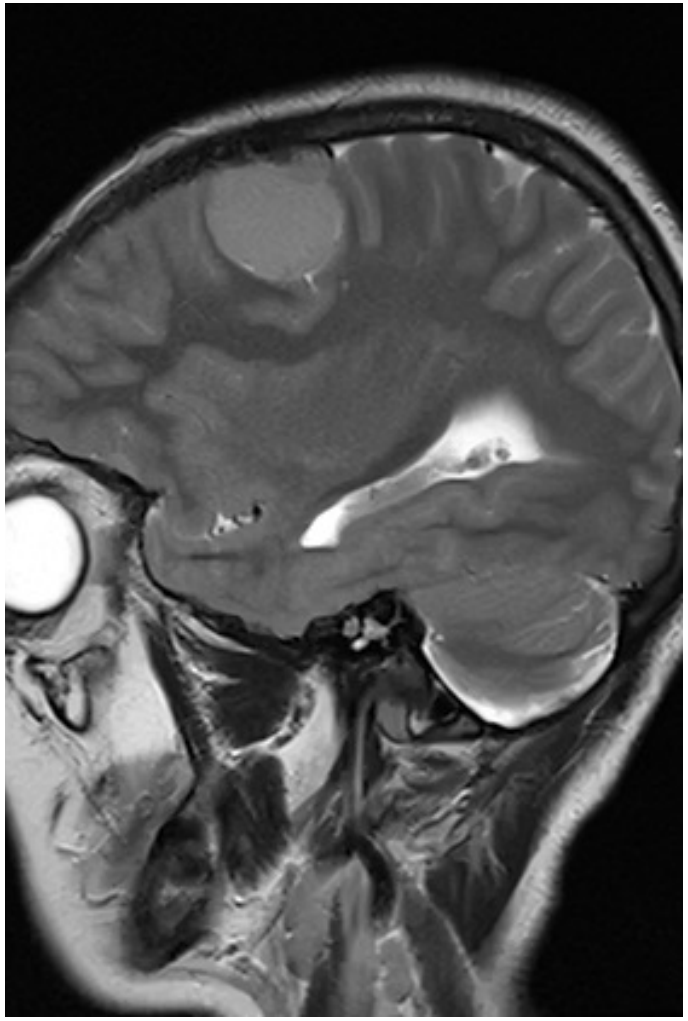
MRI of brain with MRI screening of whole spine was performed

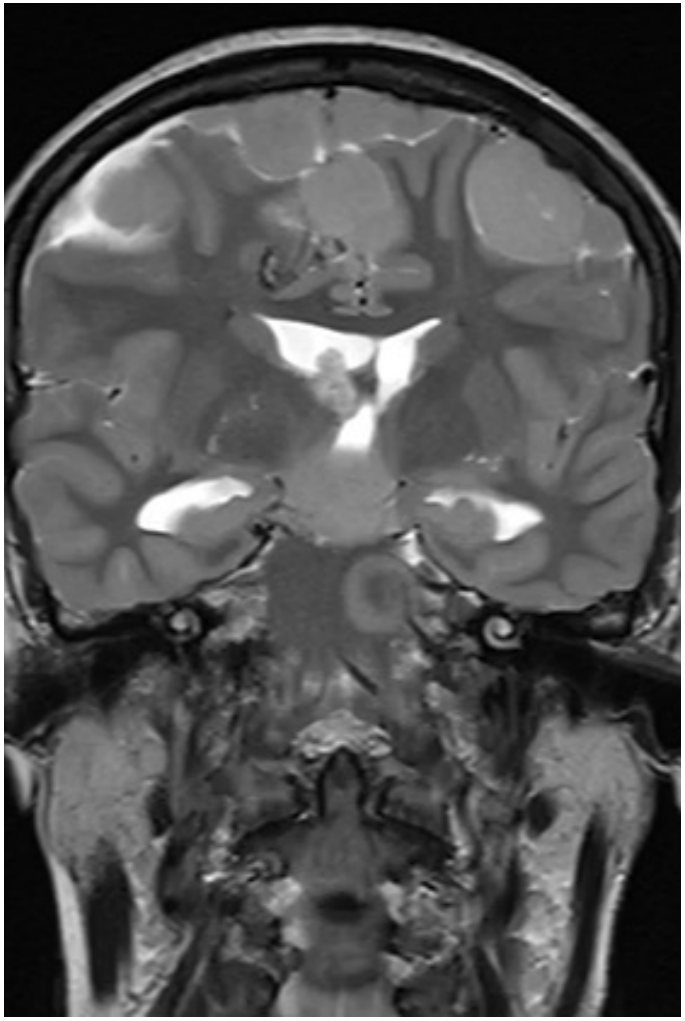







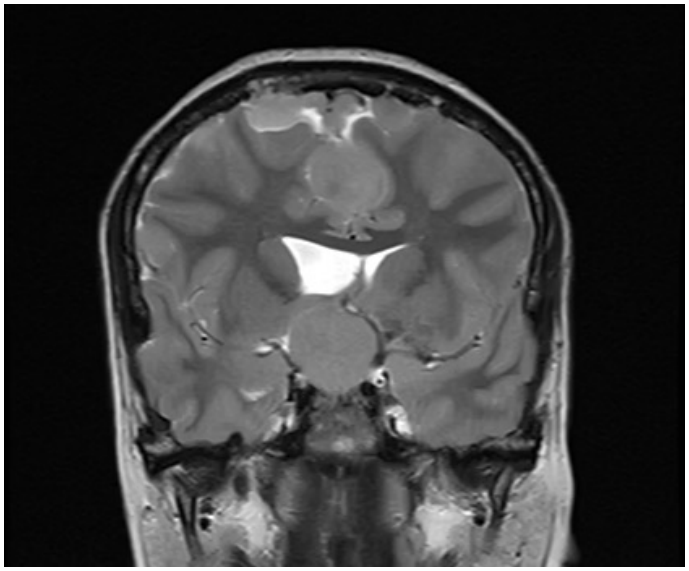







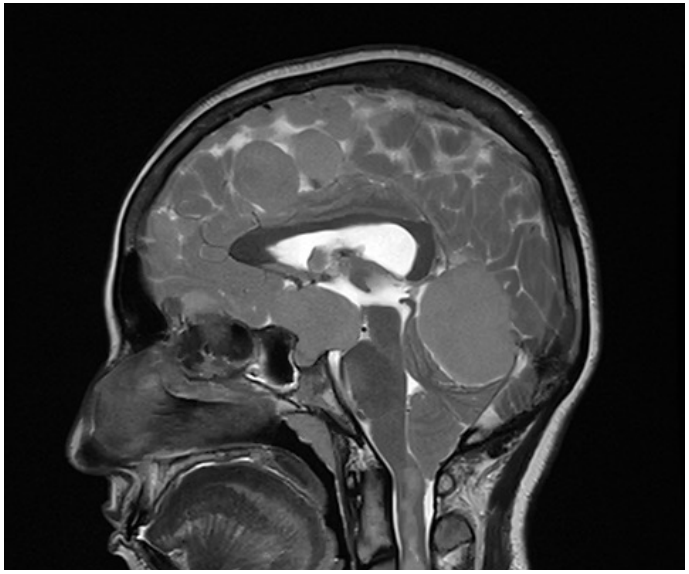







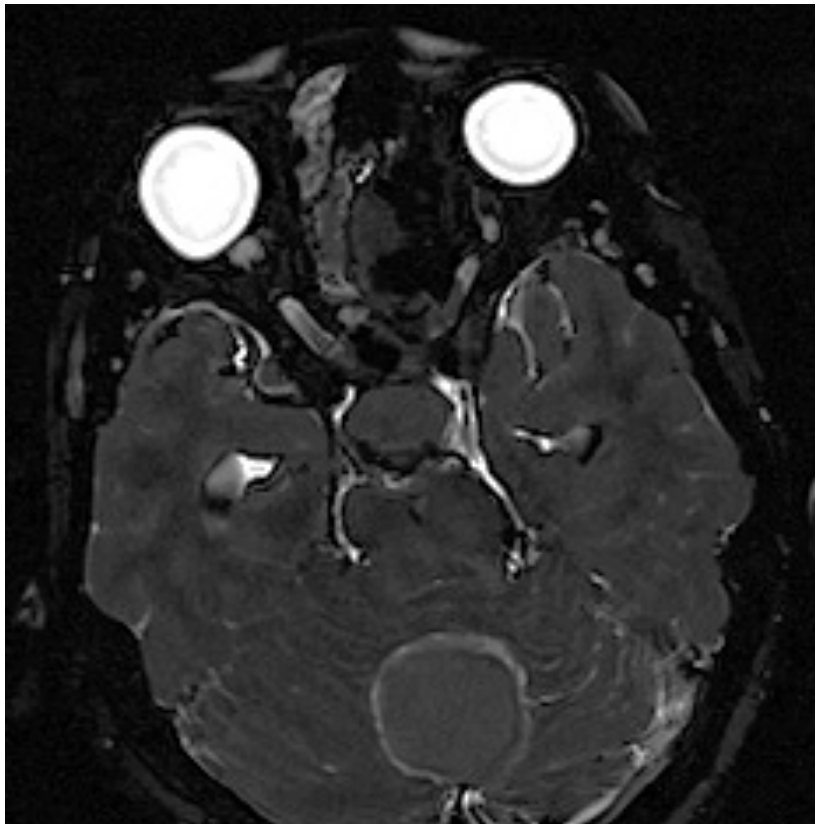







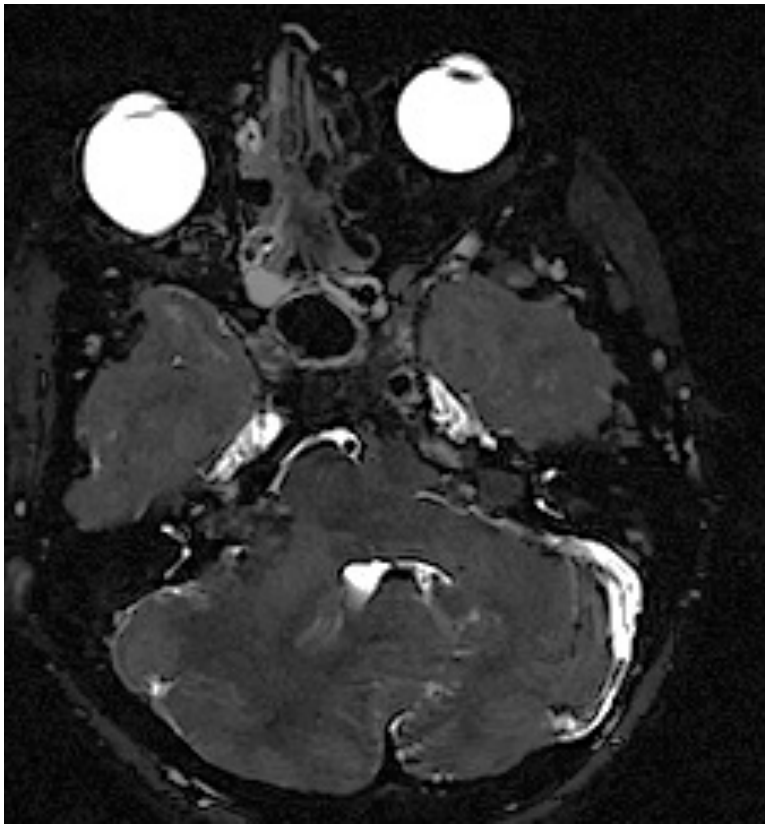


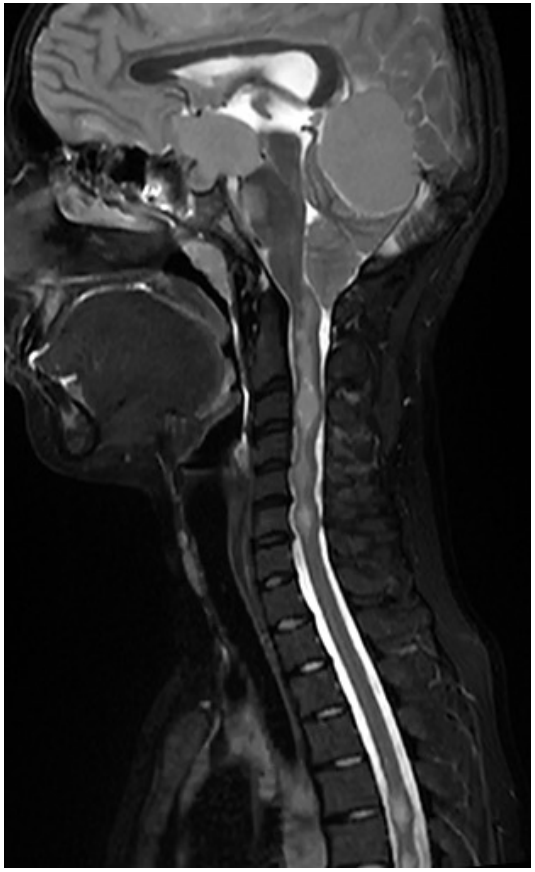
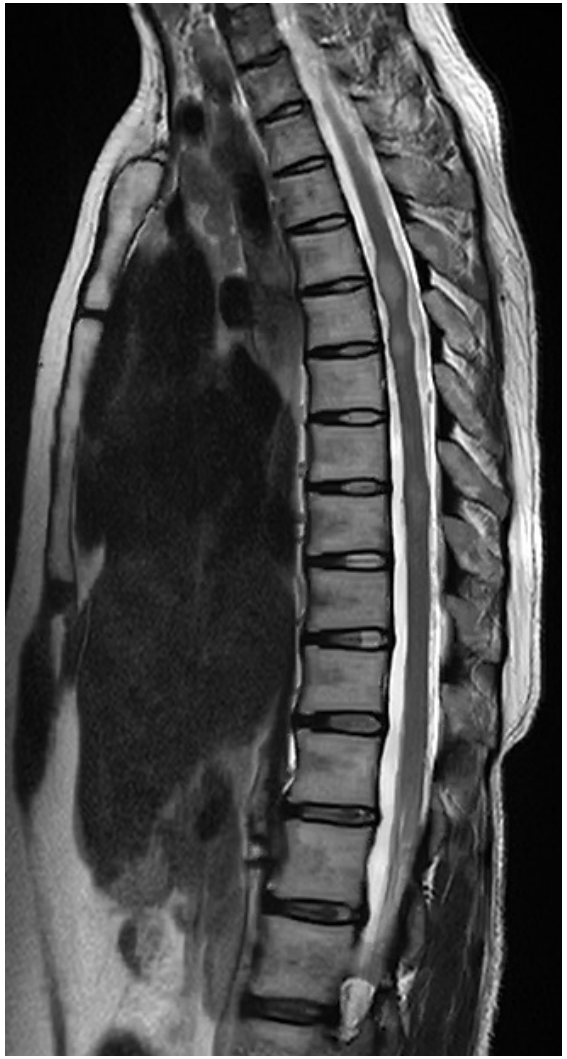
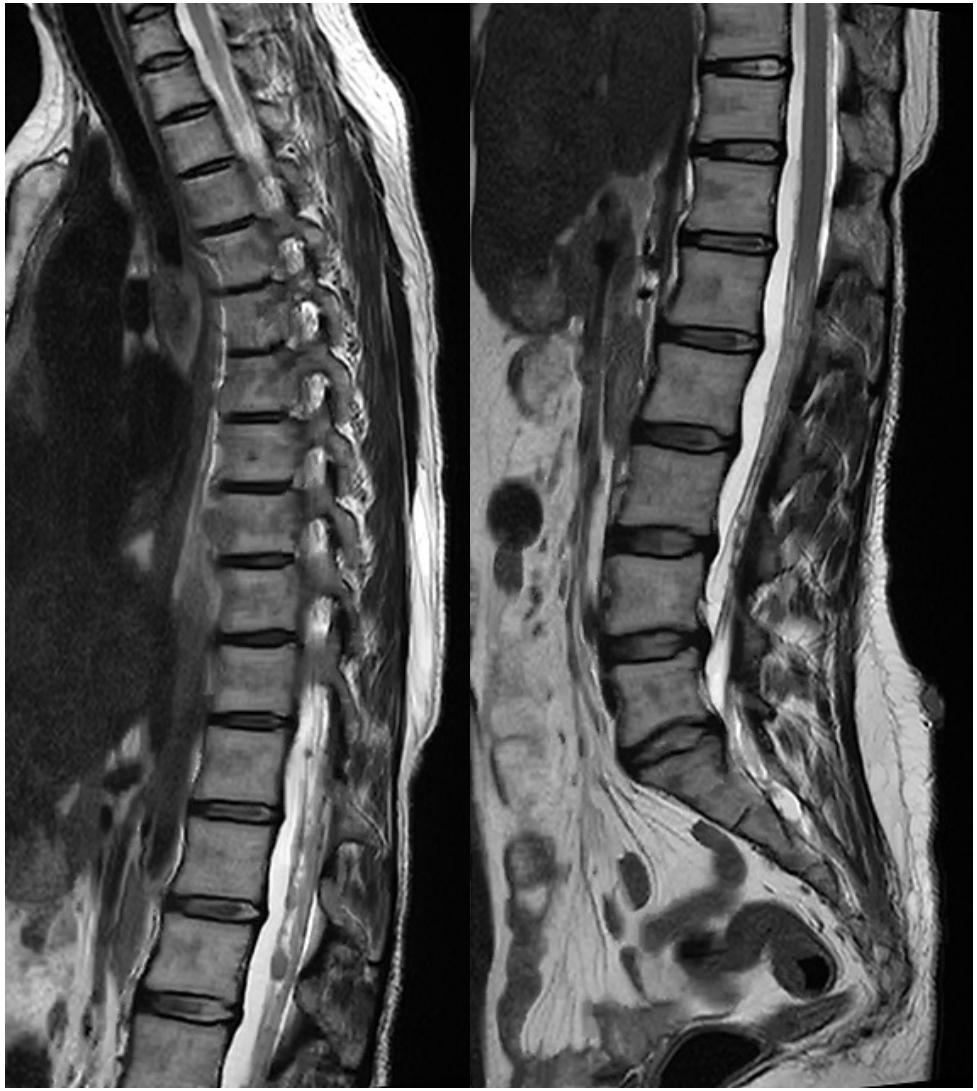










SUMMARY

- Well defined T1 hypo to isointense, T2/ FLAIR hyperintense lesion showing mild diffusion restriction on DWI with low ADC values and no blooming foci on SWI noted in bilateral cerebellopontine angle with broad base towards dura and extending into internal acoustic meati – Bilateral vestibular schwannomas
- Well defined T1 hypo to isointense, T2/ FLAIR hyperintense lesion showing mild diffusion restriction on DWI with low ADC values and no blooming foci on SWI is noted in suprasellar region with broad base towards dura along planum sphenoidale - ?Meningioma.
- Well defined T1/T2 isointense not inverting on FLAIR, not showing diffusion restriction on DWI and no blooming foci on SWI is noted in frontal horn of right lateral ventricle.
- Multiple well defined T1 hypo to isointense, T2/ FLAIR hyperintense lesions, showing mild diffusion restriction on DWI with low ADC values and no blooming foci on SWI are noted along high frontal convexities and falx cerebelli, with broad bases toward dura.- Meningiomas

- Multiple T2 hyperintense lesions are noted in intramedullary location are noted in cervical and upper thoracic spinal cord causing focal areas of bulge in the cord - ?Spinal ependymomas.
- Few well defined T2 iso to hypo intense lesions are noted in intradural extramedullary location in upper lumbar region with broad base toward anterior dura- ?Spinal meningiomas.

To conclude, it's a case of bilateral vestibular schwannomas, multiple meningiomas in brain and spine with spinal cord ependymomas.

Possible diagnosis to be considered - Neurofibromatosis 2

NEUROFIBROMATOSIS -2

- Inherited neurocutaneous syndrome with familial cancer predisposition.
- NF2 gene on chromosome 22q12 – encodes for protein Merlin (aka Schwannomin)
- Autosomal dominant disorder – inherited in 50 % individuals
  - 50% - acquired germline mutations
  - 30% - Mosaic genetic alterations
- *"MISME"* – **M**ultiple **i**nherited **S**chwannomas, **M**eningiomas and **E**pendymomas *(Predominantly benign neoplasms)*

CLINICAL PRESENTATION

- Age at presentation : Second to fourth decade (Average age17 to 24 years)
- First visible manifestation – Cutaneous schwannoma and/or juvenile cataract
- Café-au-lait spots in 25% cases
- 8^{th} cranial nerve dysfunction – progressive sensory neural hearing loss, tinnitus and difficulties in balance
- Meningiomas – asymptomatic > seizures, focal neurological deficits
- Spinal cord ependymomas – asymptomatic in 75% cases

CNS lesions in NF 2

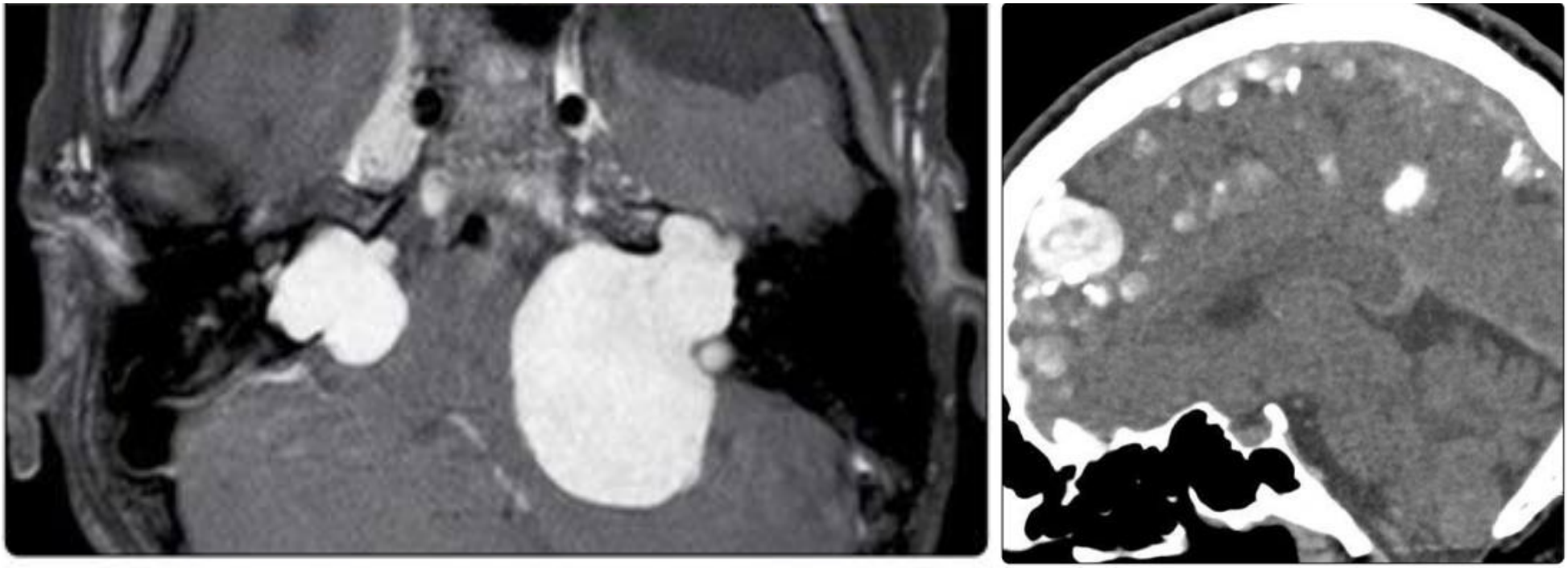
- Bilateral vestibular schwannomas
- Non vestibular schwannomas (50%)
- Meningiomas (50%)
- Cord ependymomas (multiple)
- Schwannomas of nerve roots

DIAGNOSTIC CRITERIA of NF 2

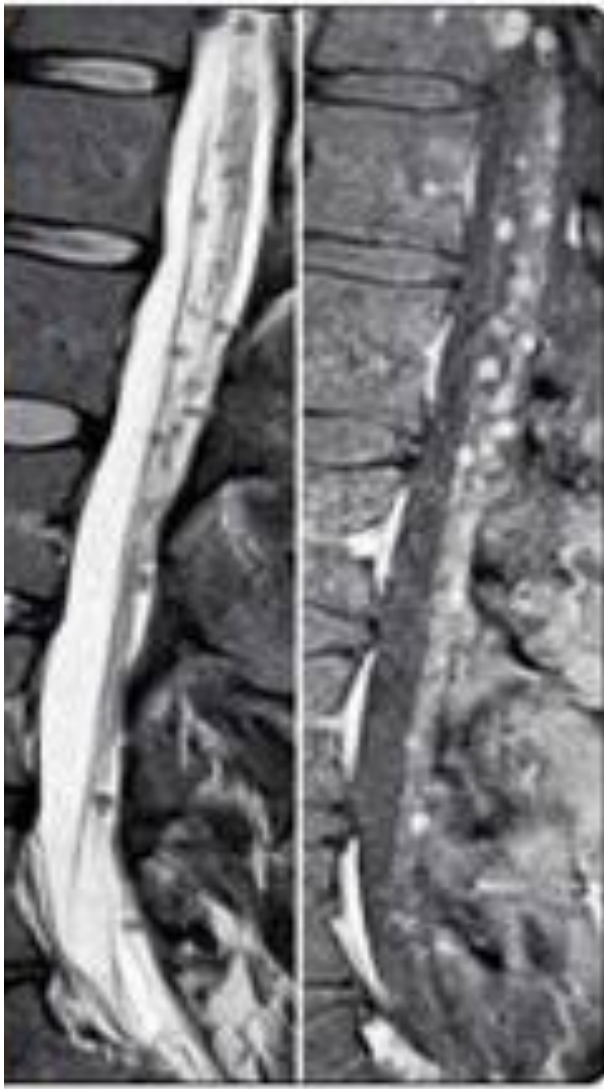
- Definite NF2
  1. Bilateral vestibular schwannomas (VS)
  2. First degree relative with NF2 and unilateral VS diagnosed before 30 years age
  3. Or First degree relative with NF2 and two of the following-
     - Meningioma
     - Glioma
     - Schwannoma
     - Juvenile posterior subcapsular lenticular opacities or cataract

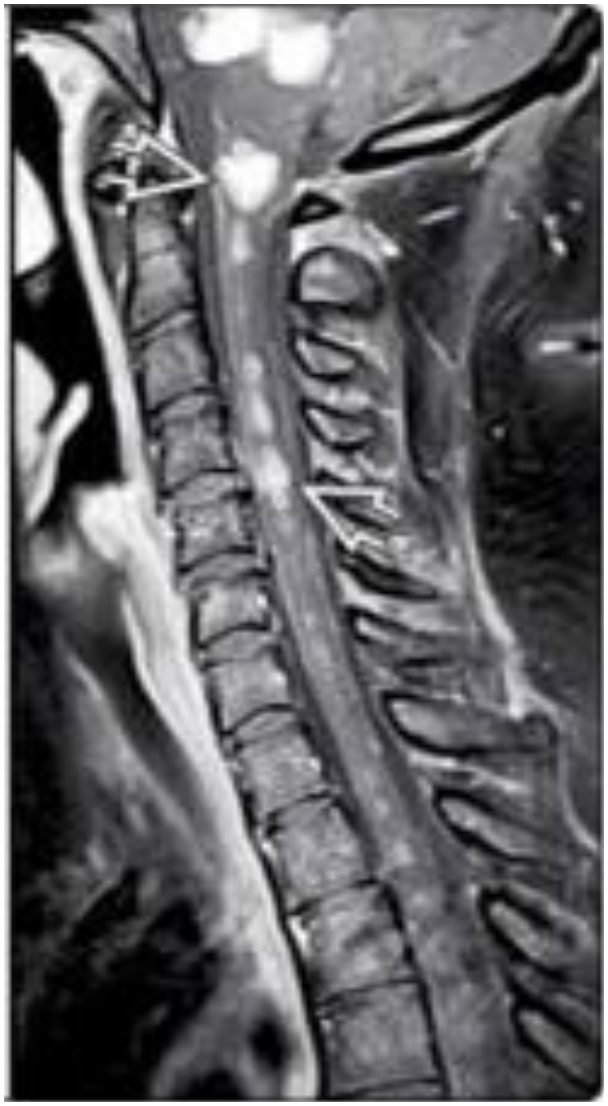
- Probable NF2

1. Unilateral VS diagnosed before 30 years age and one of the following –
   - Meningioma
   - Glioma
   - Schwannoma
   - Juvenile posterior subcapsular lenticular opacities or cataracts
2. Two or more meningiomas and one of the following –
   - One VS diagnosed before 30 years age
   - One glioma, schwannoma, or lens opacity



Source: Osborn AG, Hedlund GL, Salzman KL. Osborn’s brain : imaging, pathology, and anatomy. Philadelphia, Pa: Elsevier; 2018.





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NF- 1

- Peripheral neurofibromatosis
- Major feature: Neurofibromas

(Schwann cells and fibroblasts on histopathology)

- NF-1 gene – chromosome 17q11.2 – Neurofibromin – TSG with negative regulation of RAS oncogene
- Non CNS and cutaneous manifestations: more than that seen in NF2
- Present in childhood
- High mortality – related to malignant neoplasms

NF - 2

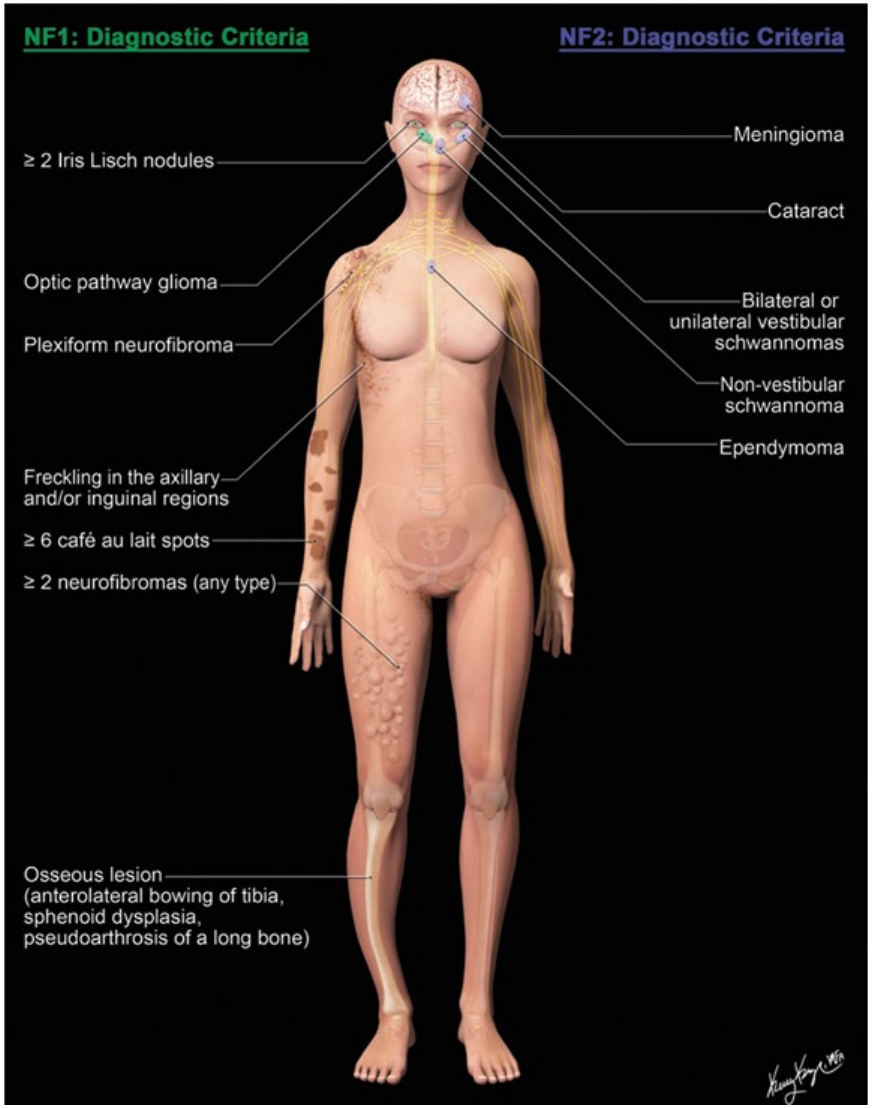
- Central neurofibromatosis
- Major feature: Schwannomas

(Only Schwann cells on histopathology)

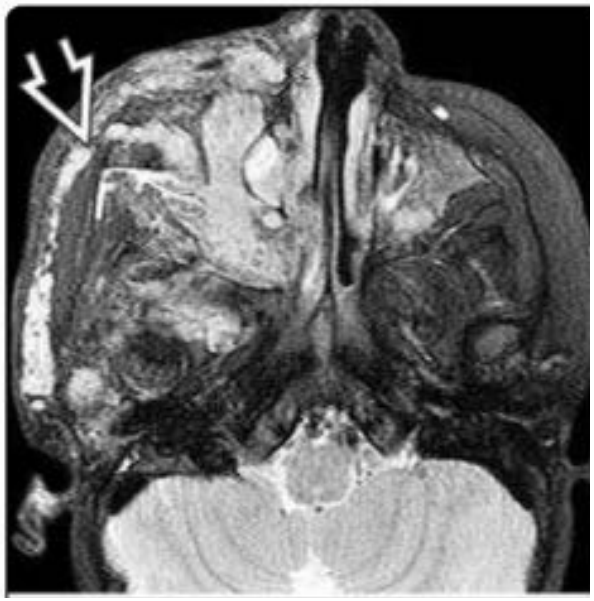
DIAGNOSTIC CRITERIA of NF 1

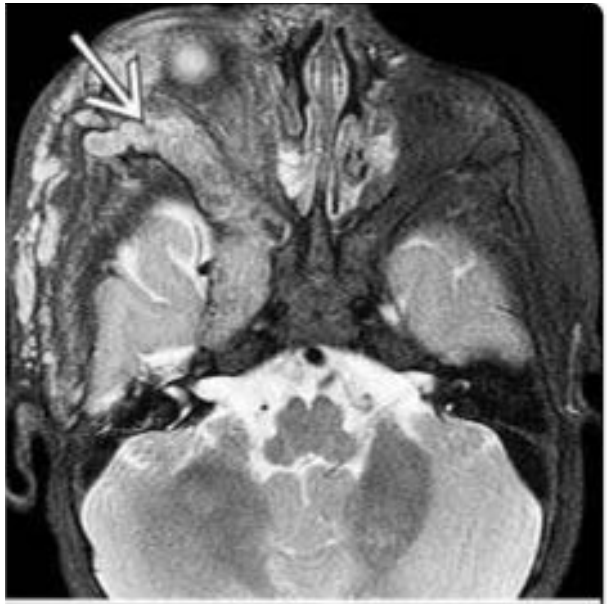
At least two of the following –

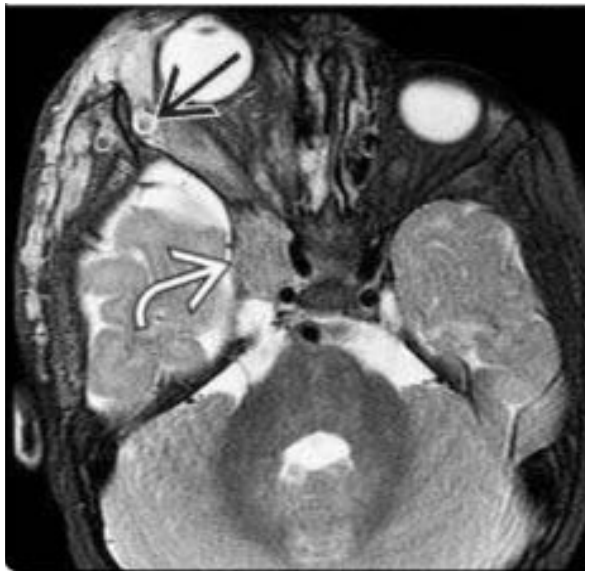
- Cutaneous lesions:
  - 6 or more café-au-lait spots (earliest manifestation)
    - Pre pubertal >0.5 cm and post pubertal >1.5cm
  - Freckling of armpits or groin
  - 2 or more neurofibromas (any type)
  - 1 plexiform neurofibroma
- Eye abnormalities:
  - 2 or more Lisch nodules
  - Optic pathway pilocytic astrocytoma
- Distinctive bone lesion
  - Sphenoid dysplasia/ absence
  - Long bone cortex dysplasia/thinning
- Family history : First degree relative with NF1

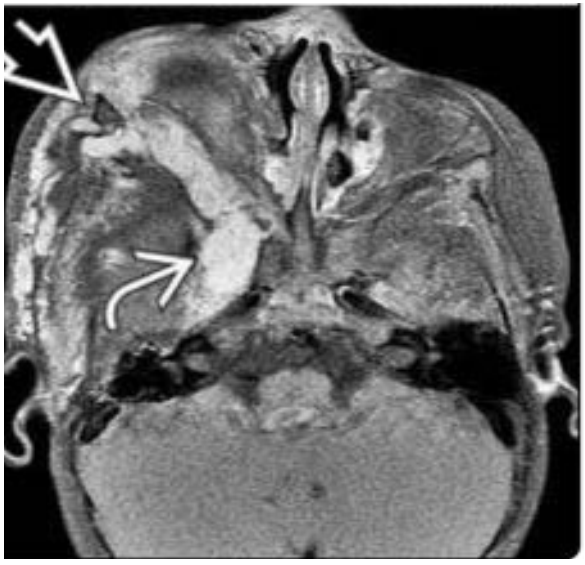


Wang MX, Dillman JR, Guccione J, Habiba A, Maher M, Kamel S, et al. Neurofibromatosis from Head to Toe: What the Radiologist Needs to Know. RadioGraphics. 2022 Jul;42(4):1123–44.









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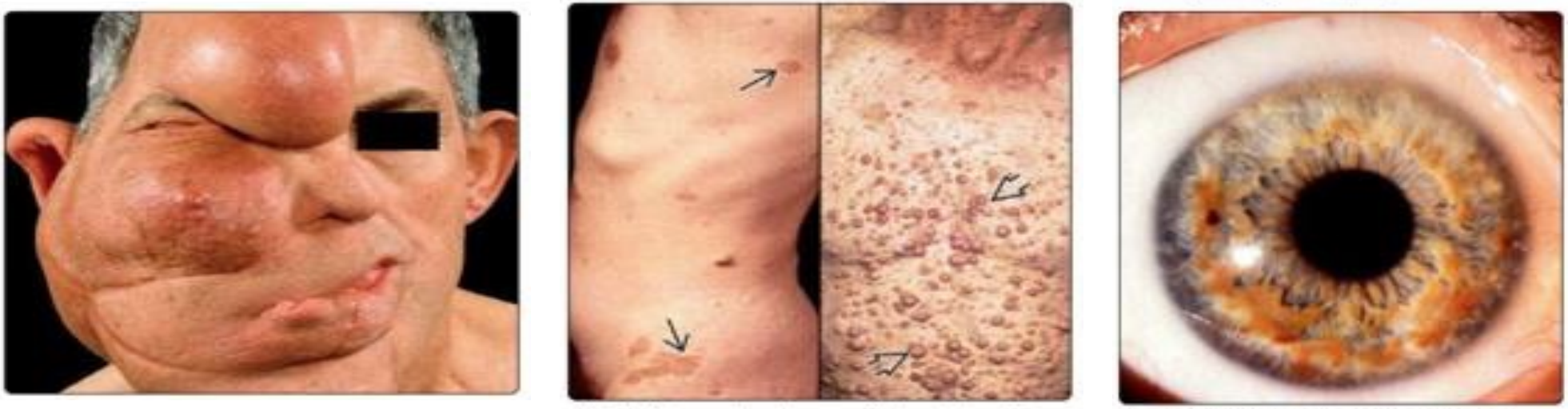

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SCHWANNOMATOSIS

- Germline mutations of SMARCB1 or LZTR1 genes
  - Familial in 85% cases and sporadic in 40%
- Seen in 1 in 40,000 births (Rarer than NF 1 and 2)
- Multiple schwannomas of the spine, subcutaneous tissues and non vestibular cranial nerves
- Meningiomas occur (lesser than NF2)
- Bilateral VS – characteristically absent.