KREP

2025

KARNATAKA RADIOLOGY EDUCATION PROGRAM

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CASE 2 :

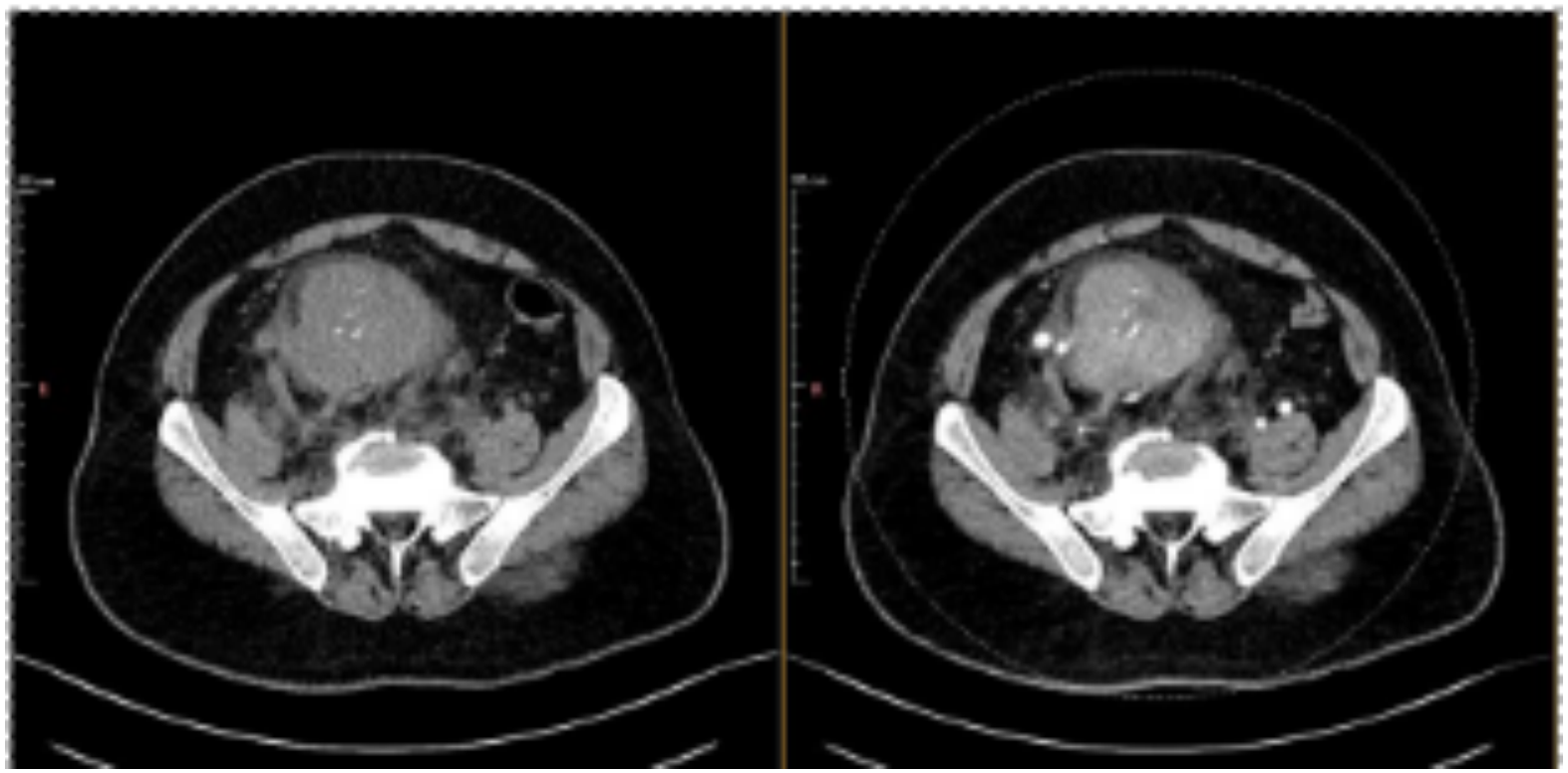
A 33yr old male patient presented with the following complaints :

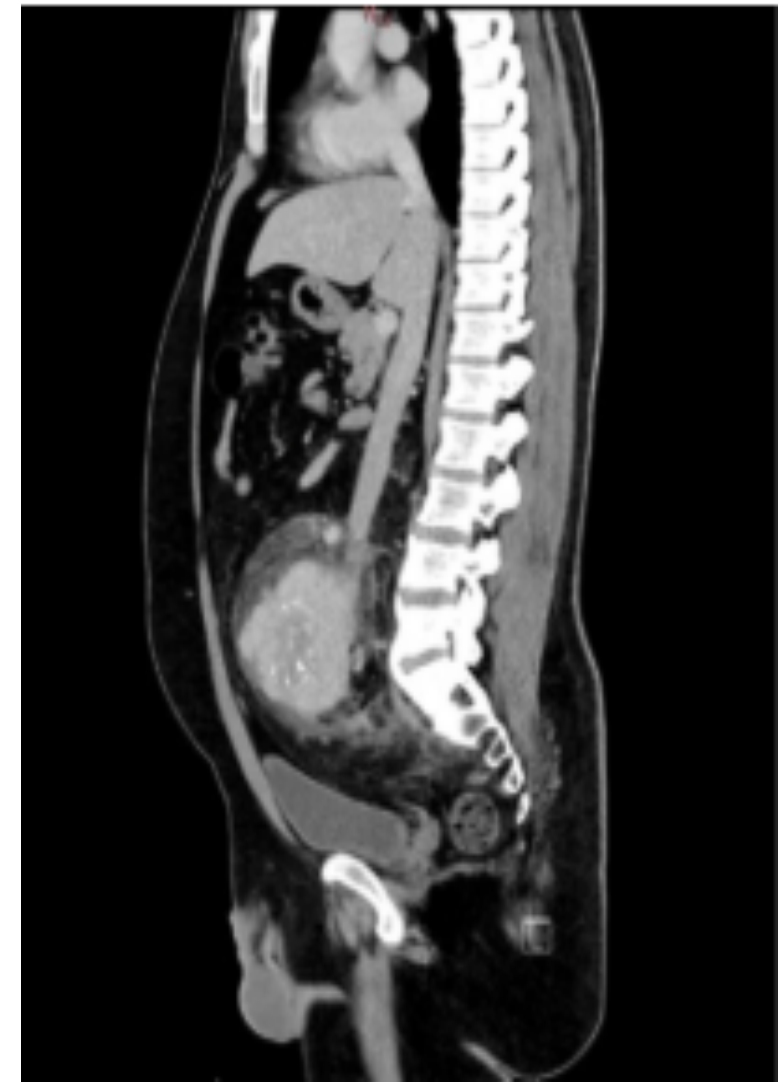
- Pain in lower abdomen radiating to back Since 1 Weeks,
- No nausea/ vomiting
- Bowel, bladder habits normal
- No fever

O/E :

- Abdominal Examination : Soft, nontender, firm mass felt in the right iliac fossa and hypogastric region and extending to left para midline.

- MRN : 10020001778304





DIFFERENTIAL DIAGNOSES :

1. **GIST -**
2. **FDC Sarcoma – x Progressive enhancement**
3. **Lymphoma - x Calcification**
4. **Metastases – x Primary**
5. **Soft-tissue sarcomas :– x Origin, Hypoattenuating, Destruction**
6. **Neurogenic and nerve sheath tumors, paraganglioma and pheochromocytoma - x Location, Salt and pepper/ Target appearance, Cystic changes**

MICROSCOPIC EXAMINATION	Bottle 1 labeled as Peritoneal Fat : Cores of fibrofatty tissue with prominent blood vessels. No evidence of malignancy is seen. Bottle 2 labelled as pelvic mass: The scores show a cellular lesion. It is composed of predominantly lymphoid infiltrate. Most of the lymphocytes appear small with hyperchromatic nucleus and scanty cytoplasm. Focally, these lymphoid cells are intense aggregates with central hyalinised blood vessels. In between these follicles/dense aggregates are spindle cells with oval elongated nucleus. Admixed with the spindle cells are many prominent thin walled blood vessels along with dense inflammatory infiltrate composed of lymphocytes, few plasma cells and eosinophils. There is mild nuclear atypia. No mitosis or necrosis are identified.

MICROSCOPIC EXAMINATION	Enlarged lymph node. It shows follicles with atretic germinal centers. It is traversed by sclerotic, hyalinised blood vessels. Mantle layer is thickened with focal onion skin like appearance. There is expansion of interfollicular zone with extensive vascular proliferation of high endothelial venules. Admixed are few small lymphocytes along with focal sheets of plasma cells. No no definite large atypical cells are seen.
IMPRESSION	Features are suggestive of Castleman's disease - hyaline vascular Castleman's disease. Advice: Immunohistochemistry (6-10 markers) for confirmation.

IHC REPORT	Immunohistochemistry performed and analyzed at Narayana Health. All cells are positive for LCA Follicles are highlighted by CD20. CD3 is positive in interfollicular cells. BCL6 and CD10 is positive in follicular center cells. CD21 and CD23 highlights follicular dendritic meshwork. Ki67 shows normal staining pattern. Controls are satisfactory.
IMPRESSION	Features are consistent with hyaline Vascular Castleman's disease. Kindly correlate clinically.

CASTLEMAN'S DISEASE :

Imaging of Castleman Disease

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- **Dysregulation or dysplasia of follicular dendritic cells and inflammatory mediators (interleukin-6 – IL-6) have been incriminated, mainly in the multicentric form of the disease.**
- **Increased serum IL-6 level is positively correlated with active disease, and removal of the CD mass may result in an abrupt drop in IL-6 levels and resolution of symptoms. Furthermore, treatment with IL-6 antagonists relieves symptoms and signs of the disease.**
- **Chronic low-grade inflammation and immunodeficiency including autoimmune diseases are considered to be possible causal factors of CD. Furthermore, viral infections (HHV-8, EBV) and others have been reported in association with CD.**
- **The distribution is as follows:**
  - **A. Thorax: ~70%**
  - **B. Abdomen/pelvis and retroperitoneum: 10-15%**
  - **C. Neck: 10-15%**

- **The unicentric or localized type of CD involves a single lymph node or localized lymph node station. It represents the most common form of the disease (> 90%) and is almost always asymptomatic and of hyaline-vascular type. The size of lesions varies greatly (1–12 cm).**
- **In contrast, multicentric CD occurs in about 10% of patients and is almost always symptomatic but with highly varying symptoms which may progress over several months or episodically over years**
- **Multicentric CD commonly results in a fatal outcome due to infectious complications, multi-organ failure and occasional development of malignancies. Neoplastic complications include Kaposi sarcoma and lymphoma (Hodgkin's and non-Hodgkin) as well as plasma cell dyscrasias.**
- **Associations :**

1. iMCD -TAFRO : MCD but with thrombocytopenia, anasarca, fever, bone marrow reticulin fibrosis (or renal insufficiency), and organomegaly (hepatosplenomegaly and lymphadenopathy).

2. POEMS-MCD : is a paraneoplastic syndrome caused by an underlying plasma cell neoplasm. Peripheral polyneuropathy as a defining feature, along with MCD symptoms.

Other features : Organomegaly, endocrinopathy, monoclonal plasma cells, hyperpigmentation, Raynaud phenomenon, hypertrichosis, and clubbing. Sclerotic bone lesions, papilledema, thrombocytosis, elevated vascular endothelial growth factor (VEGF) level, and abnormal pulmonary function can also be seen.

3. Haemophagocytic syndrome : Esp a/w HHV – 8.

Table 2: Consensus Diagnostic Criteria for iMCD

Type of Criteria	Description
Major criteria (need both)	Histopathologic lymph node features consistent with iMCD spectrum
	Enlarged lymph nodes (≥1 cm in short-axis diameter) at two or more lymph node stations
Minor criteria (need at least two of 11)	Laboratory criteria
	Elevated CRP level or ESR
	Anemia
	Thrombocytopenia
	Hypoalbuminemia
	Renal dysfunction
	Polyclonal hypergammaglobulinemia
	Clinical criteria
	Constitutional symptoms: night sweats, fever, weight loss, or fatigue
	Large spleen or liver
	Fluid accumulation: edema, anasarca, ascites, or pleural effusion
	Eruptive cherry hemangiomatosis or violaceous papules
	Lymphocytic interstitial pneumonitis
Exclusion criteria (must rule out diseases that can mimic iMCD)	Infection-related disorders (HHV8, clinical EBV-lymphoproliferative disorders such as infectious mononucleosis or chronic active EBV, other uncontrolled infections causing widespread inflammation and adenopathy such as CMV, toxoplasmosis, HIV, active tuberculosis)
	Autoimmune or autoinflammatory diseases (SLE, RA, adult-onset Still disease, JIA, autoimmune lymphoproliferative syndrome)
	Malignant or lymphoproliferative disorders (lymphoma, multiple myeloma, primary lymph node plasmacytoma, FDC sarcoma, POEMS syndrome)

Note.—Adapted and reprinted, with permission, from reference 23. CMV = cytomegalovirus, CRP = C-reactive protein, ESR = erythrocyte sedimentation rate, JIA = juvenile idiopathic arthritis, RA = rheumatoid arthritis, SLE = systemic lupus erythematosus.

IMAGING :

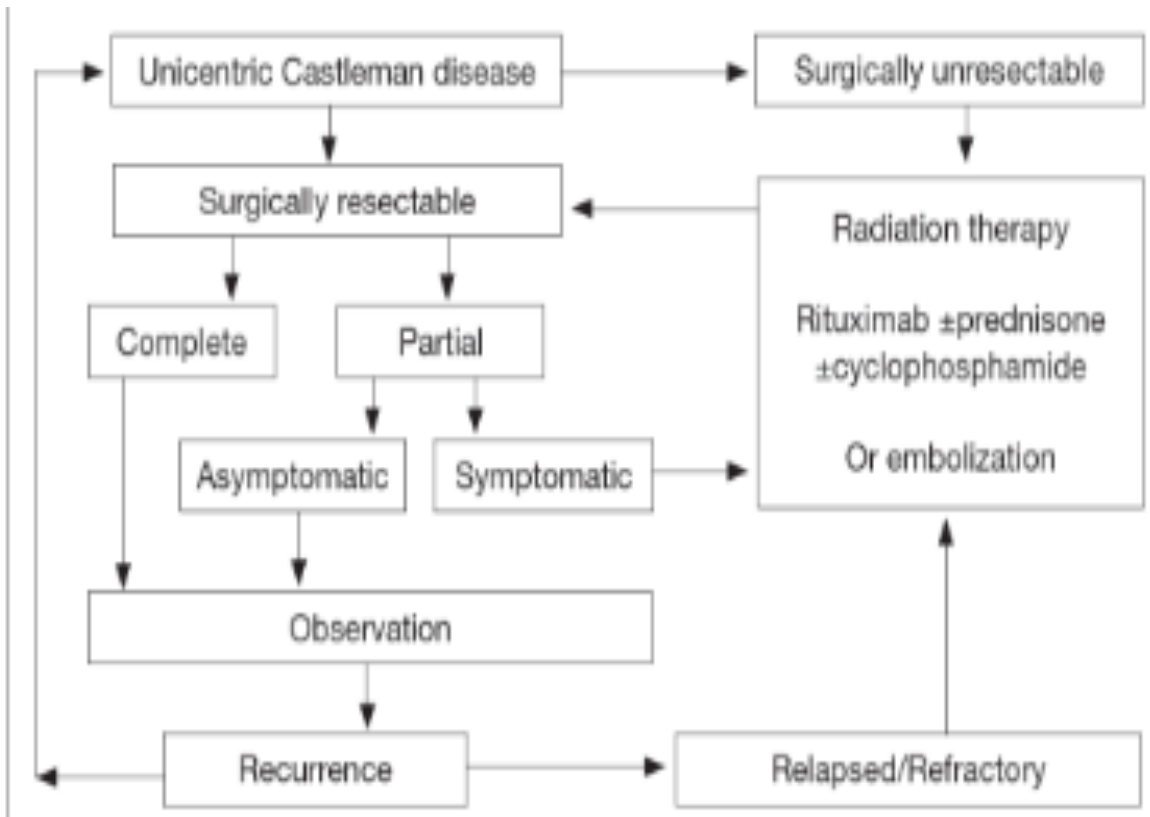
- **CT : Homogeneous mass of soft tissue density which is intensely enhanced following contrast administration and may rarely be accompanied by calcification(Arborising type in 15%). Heterogeneity is usually a reflection of intralesional sclerosis, necrosis and degeneration in larger lesions.**
- **MRI : Typically heterogeneous with an increased signal as compared to skeletal muscles on T1-weighted images and marked hyperintensity on T2-weighted images with areas of calcification showing as low signals on T2.**
- **PET : CD is usually positive on fluorodeoxyglucose (FDG) PET.**

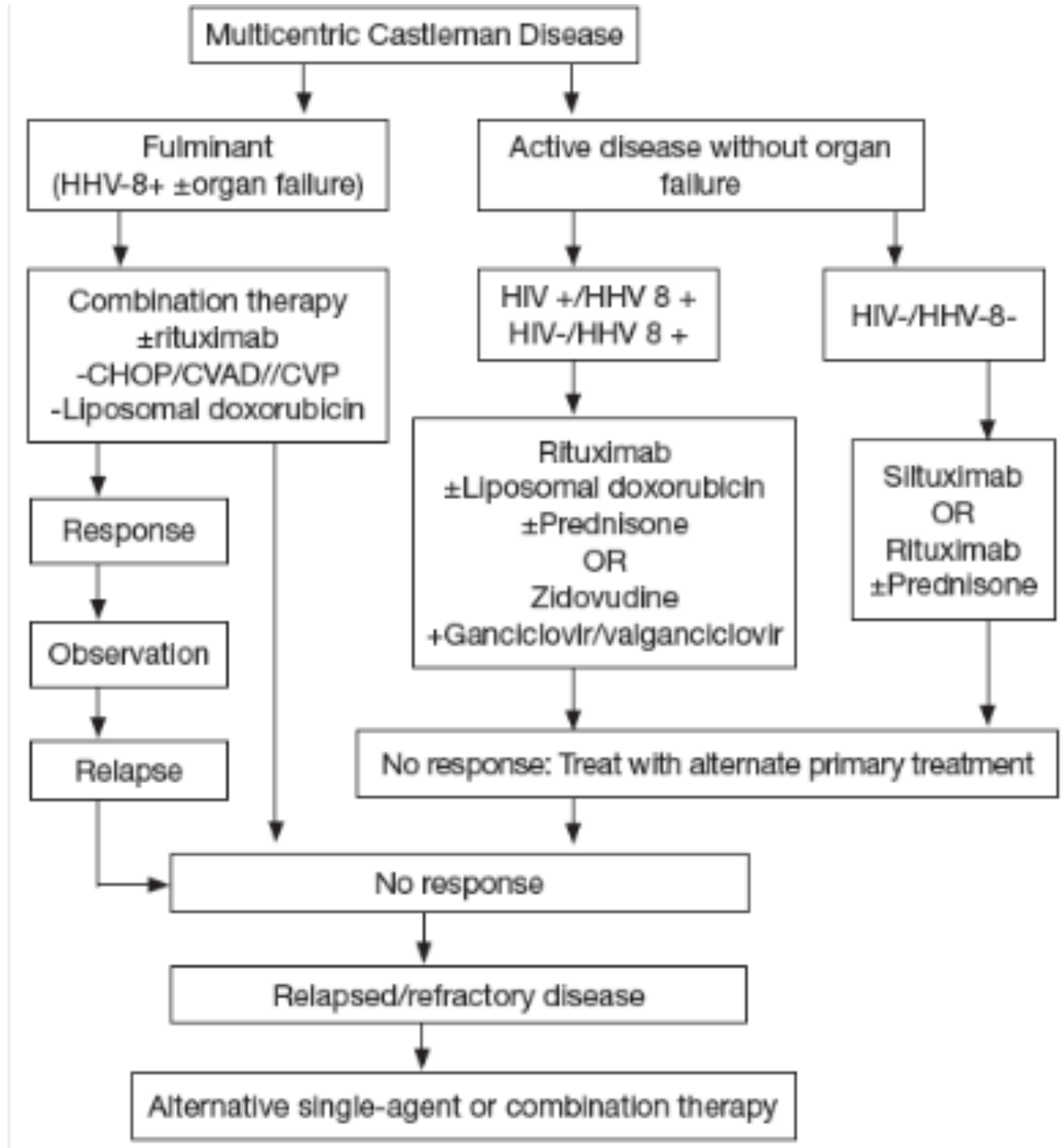
DIFFERENTIATING POINTS :

1. **Vs Neurofibroma, schwannoma, and paraganglioma.**
2. **Vs EBV-related disorders such as infectious mononucleosis or chronic active EBV**
3. **Vs lymphoma, multiple myeloma, plasmacytoma, and FDC sarcoma.**
4. **Vs metastases**
5. **Vs Sarcoidosis**

Table 3: Mimics of CD

Disease	Mimics What Subtype of CD	Differentiating or Suggestive Clinical Features	Differentiating or Suggestive Imaging Findings
Lymphoma	UCD, MCD	Development of B symptoms or abnormal results of laboratory tests (eg, anemia)	Increasing adenopathy, ascites, or new splenomegaly Higher SUV_{max} than in CD Lack of calcification
Systemic infections (CMV-, HIV-, or EBV-associated disorders)	MCD	History of exposure Positive results of serologic tests Abnormal results of laboratory tests (eg, elevated WBC count)	…
Autoimmune disease (RA, SLE, adult-onset Still disease, JIA)	MCD	Positive results of serologic tests for disease (eg, ANA, RF)	…
Multiple myeloma (MM)	MCD (POEMS-MCD)	Bone pain in MM High incidence of renal failure in MM Better survival in CD	More likely lytic lesions in MM vs sclerotic lesions in CD
Solitary plasmacytoma (EMP, SBP)	UCD (EMP), MCD (POEMS-MCD, SBP)	EMP most frequently involves head and neck region, gastrointestinal tract, and lungs	SBP typically manifests as lytic osseous lesion
FDC sarcoma	UCD, MCD	Most FDC sarcomas occur in head and neck region; lesions arising in CD occur in abdomen	FDC sarcoma shows progressive enhancement in delayed phases
Thymic epithelial tumors (eg, thymoma)	UCD	Possible clinical evidence of MG	Hypoenhancing anterior mediastinal mass that may have cystic spaces or calcifications Invasion of great vessels, fat, or pleural seeding suggests malignancy
Germ cell tumor (eg, teratoma)	UCD	Younger patients (2nd to 4th decades of life)	Fluid, fat, ± calcification at CT and MRI No hyperenhancement
Soft-tissue sarcoma (eg, undifferentiated pleomorphic sarcoma)	UCD	Sarcomas tend to occur in younger patients	Origin from bone or muscle More invasive and locally destructive Iso- or hypoattenuating to adjacent muscle at CT
Solitary fibrous tumor (SFT)	UCD	…	SFT shows intermediate T1-weighted and heterogeneous T2-weighted signal intensity at MRI SFT may show avid enhancement
Gastrointestinal stromal tumor (GIST)	…	May manifest with bowel symptoms	Submucosal location Lymphadenopathy is rare
Neural crest–derived neoplasms (paraganglioma, pheochromocytoma)	UCD	Symptoms of catecholamine excess Association with NF types 1 and 2 Avidity at gallium 68–DOTATATE imaging or iodine 123–MIBG scintigraphy	Characteristic locations within paraganglia Cystic changes, necrosis, hemorrhage, and internal calcifications Salt-and-pepper enhancement due to prominent flow voids
Neurofibroma and schwannoma	…	Associated with NF types 1 and 2 Typical nerve distribution location	Fusiform shape, peripheral T2 hyperintensity with central hypointensity (target sign), and scalloping of adjacent vertebrae
Pancreatic neuroendocrine tumor	UCD	Symptoms related to hormonal excess if secreting hormones	Located in pancreas Avidity at somatostatin receptor imaging
Metastasis	UCD, MCD	Evidence of primary neoplasm elsewhere	Metastatic lymph nodes are usually hypoenhancing Presence of metastases in liver and lung
Sarcoidosis	MCD	Symmetric hilar and mediastinal lymphadenopathy Usually no mass effect on adjacent structures	Hypoenhancing nodes ± nodal calcification FDG avid and high uptake at gallium 67 scintigraphy Perilymphatic micronodules Hepatosplenomegaly with hypoenhancing granulomas Pulmonary fibrosis in late stages


Unicentric Castleman disease
Surgically unresectable
Surgically resectable
Radiation therapy
Rituximab ±prednisone
±cyclophosphamide
Or embolization
Complete
Partial
Asymptomatic
Symptomatic
Observation
Recurrence
Relapsed/Refractory


Multicentric Castleman Disease
Fulminant
(HHV-8+ ±organ failure)
Active disease without organ failure
Combination therapy
±rituximab
-CHOP/CVAD//CVP
-Liposomal doxorubicin
HIV +/HHV 8 +
HIV-/HHV 8 +
HIV-/HHV-8-
Response
Rituximab
±Liposomal doxorubicin
±Prednisone
OR
Zidovudine
+Ganciclovir/valganciclovir
Siltuximab
OR
Rituximab
±Prednisone
Observation
Relapse
No response: Treat with alternate primary treatment
No response
Relapsed/refractory disease
Alternative single-agent or combination therapy