



Occam Shmoccam



UCI 2019 Colloquium

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History



- 62 year-old H J, history of Diffuse Large B-Cell Lymphoma s/p R-CHOP in 2007 in remission.
- Symptoms started with numbness/tingling on the plantar surface of his L foot.
- 6 months later, developed pain in the same location of his L foot. Started on Gabapentin and Duloxetine by his PCP, which were ineffective.
- He was referred to see a neurologist and he had an MRI in late 2016 of his L-spine that reportedly showed a “disc bulge”.



History



- Early 2018, his pain gradually moved proximally to involve the L calf and behind the knee.
- In April 2018, he began using a cane because of difficulty walking.
- In May/June 2018 – He was referred to a neurosurgeon that evaluated him and felt that the pain was not originating from his spinal cord.
- 1 month prior to presentation to our clinic in 9/2018, he had two falls at work despite cane use feeling that his leg “gave out”.
- NO other weakness in any of his other extremities. NO diplopia, dysarthria, dysphagia, dyspnea, cramps. NO incontinence but mild urinary urgency.



History



Past Medical/ Surgical History:

- Diffuse Large B-cell Lymphoma – diagnosed in 2007 s/p R-CHOP in remission. No radiation.
- R Retinal Detachment s/p surgical repair
- Pheochromocytoma s/p adrenalectomy

Family History:

- Non-contributory

Social History:

- Married



We now invite Dr. Lewis to elicit further history and examine the patient



Neurological Exam



Physical Exam:

- **MS:** NL orientation and language.
- **CN:** Intact
- **Motor:**
 - Normal Neck Flex/Ext.
 - LLE: Flaccid and plegic below the knee. KF 2/5, KE 5/5, HF 4/5, ABduction 2/5, ADduction 4+/5.
- **Sensory:** Diminished anterolateral and plantar surface of L foot.
- **DTRs:** Absent L ankle jerk. 2+ in all remaining DTRs.
- **Gait:** Cautious, mildly Trendelenburg with steppage gait on the L. Requires cane.



Differential?

What would you do?



Evaluation



STUDIES:

MRI Lumbar Spine: Mild degenerative changes and neural foraminal narrowing.

MRI Brain: Bilateral white matter signal changes consistent with extensive microvascular angiopathy.

MRI L Foot: Negative.

MRI L Femur: Lesion involving the distal aspect of the left vastus lateralis muscle likely a benign lesion.



Evaluation



STUDIES:

PET/CT:

New increased FDG uptake in left sciatic nerve.

MRI Pelvis:

Edema in left gluteus maximus and slight asymmetry of the left sciatic nerve. No masses.



Evaluation



ELECTRODIAGNOSTIC STUDIES:

- Sensory: Left superficial peroneal and sural were absent. The right sural normal [Amp 8.5, Vel 44 m/s]
- Motor: Left peroneal (EDB + TA) and Tibial Absent. Right peroneal and Tibial normal DML and CMAPs and mildly low conduction velocities [velocity 36-37 m/s].



Evaluation



Needle EMG										
	Spontaneous				Volitional MUAPs					
Muscle	IA	Fibs/PSW	Fasc	Other	Dur.	Amp	Poly	Recruit	Rate	Comment
L. Tibialis anterior	Normal	3+	None	.	No MUPs	No MUPs		No MUPs		.
L. Gastrocnemius (Medial head)	Normal	3+	None	.	No MUPs	No MUPs		No MUPs	Normal	.
L. Vastus lateralis	Normal	None	None	.	Normal	Normal	None	Normal	Normal	.
L. Semitendinosus	Normal	3+	None	.	No MUPs	No MUPs		No MUPs		.
L. Gluteus medius	Normal	3+	None	.	No MUPs	No MUPs		No MUPs		.
L. Gluteus maximus	Normal	3+	None	.	Long	High		Single Unit		.
L. Lumbar paraspinals (mid)	Normal	None	None							
L. Lumbar paraspinals (low)	Normal	None	None							
L. Sacral paraspinals	Normal	1+	None							
L. Adductor longus	Normal	None	None	.	Normal	Normal	None	Normal	Normal	.



Evaluation



STUDIES:

Lab Studies:

Unremarkable CMP, CBC, LDH, CK, Immunofixation, EPG, ESR, CRP

Demyelinating Neuropathy Panel (WashU): Negative

Basic CSF Studies: normal

Cytology: NEGATIVE

Flow Cytometry: NEGATIVE

Bone Marrow Biopsy:

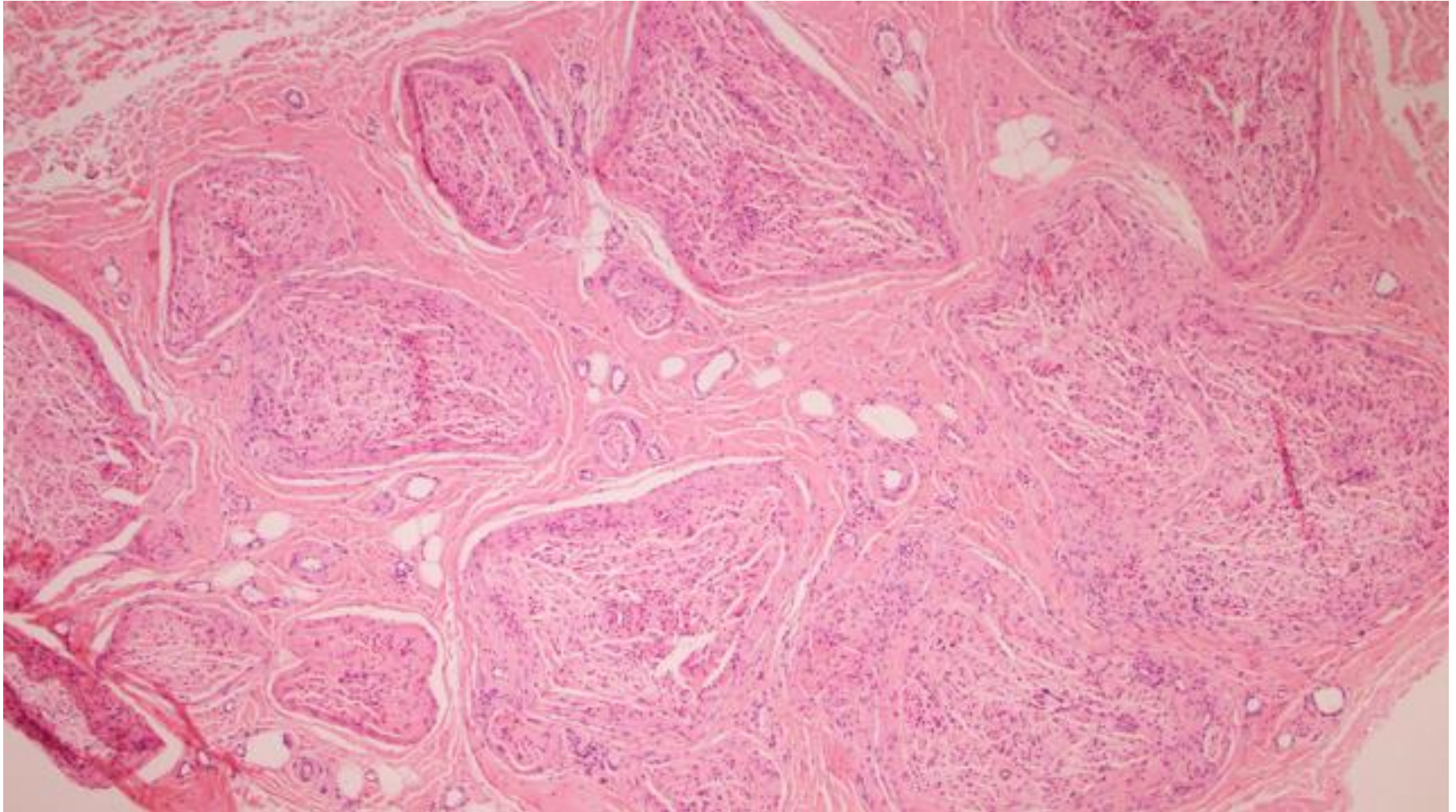
No definite morphologic evidence of involvement by lymphoma.



Evaluation



Left Sciatic Nerve Biopsy Pathology:

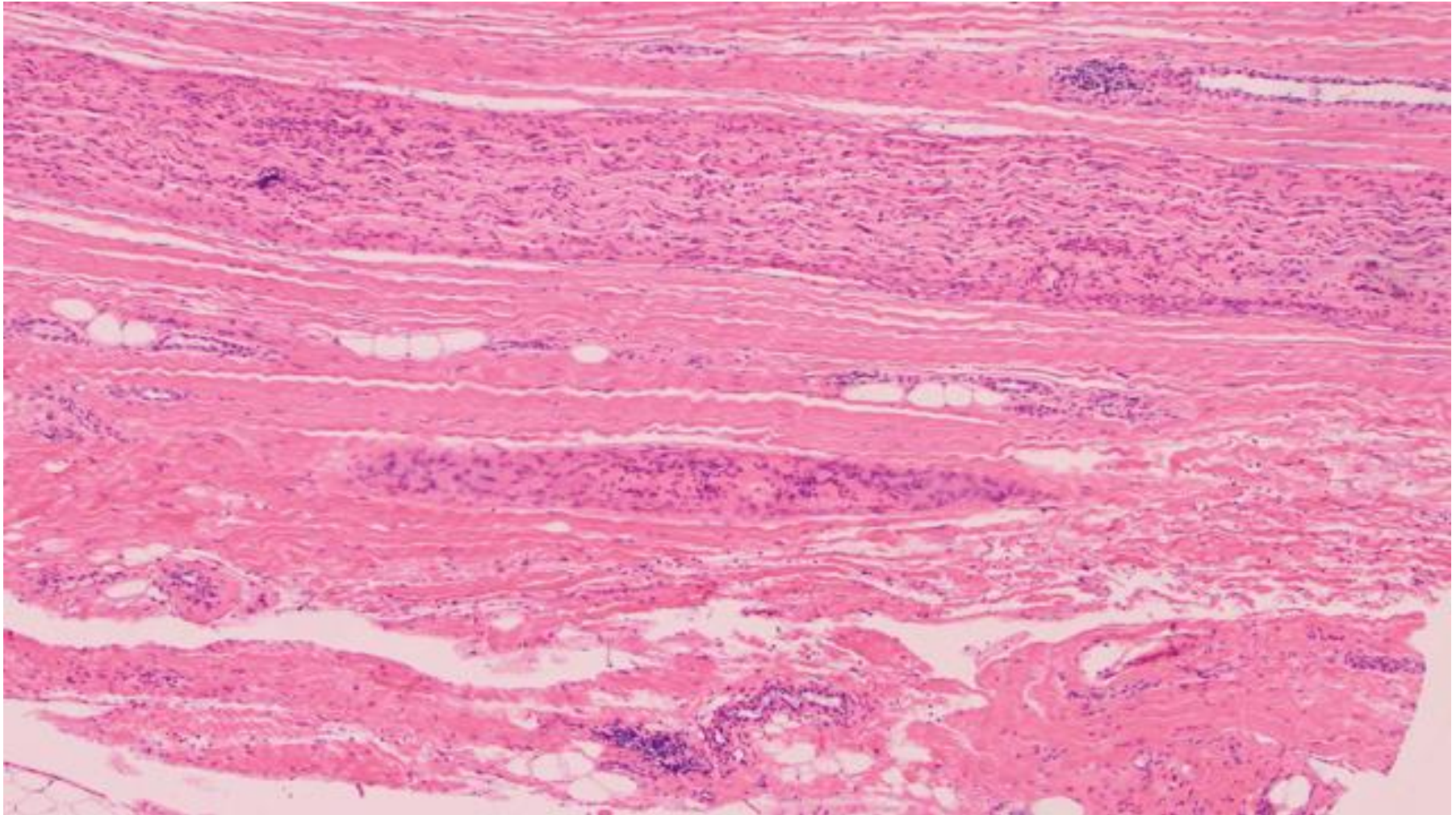




Evaluation



Left Sciatic Nerve Biopsy Pathology:

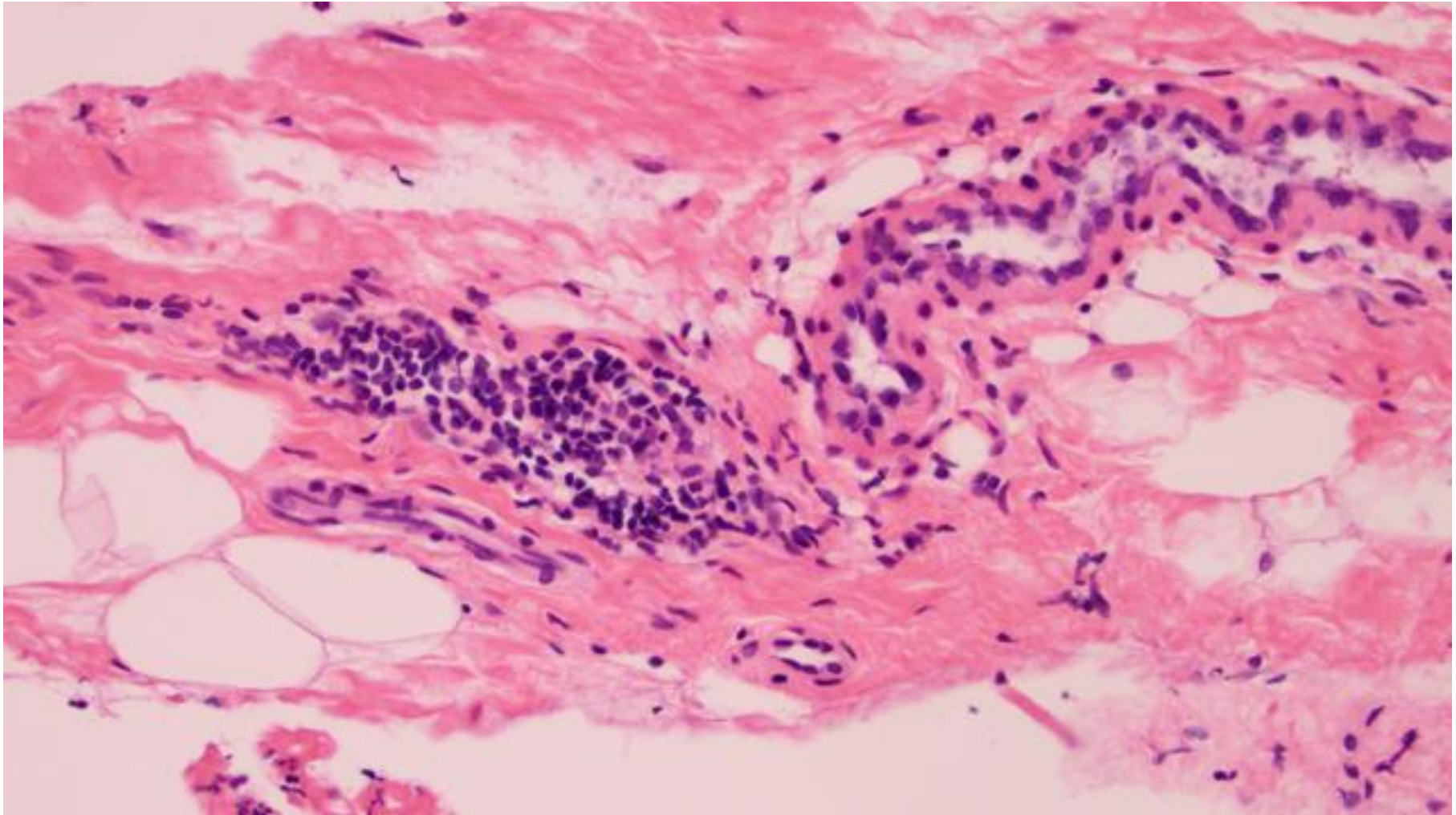




Evaluation



Left Sciatic Nerve Biopsy Pathology:

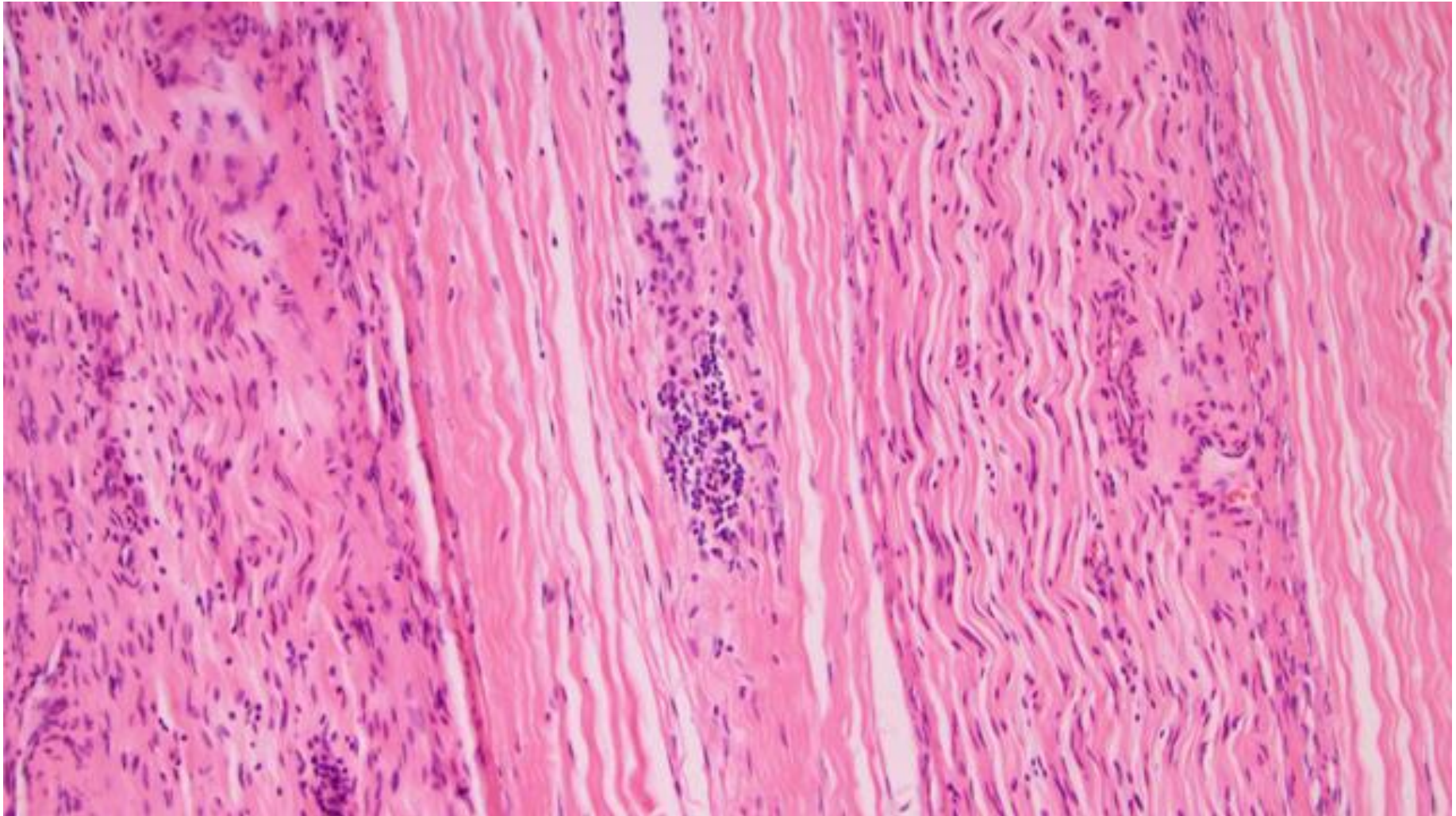




Evaluation



Left Sciatic Nerve Biopsy Pathology:



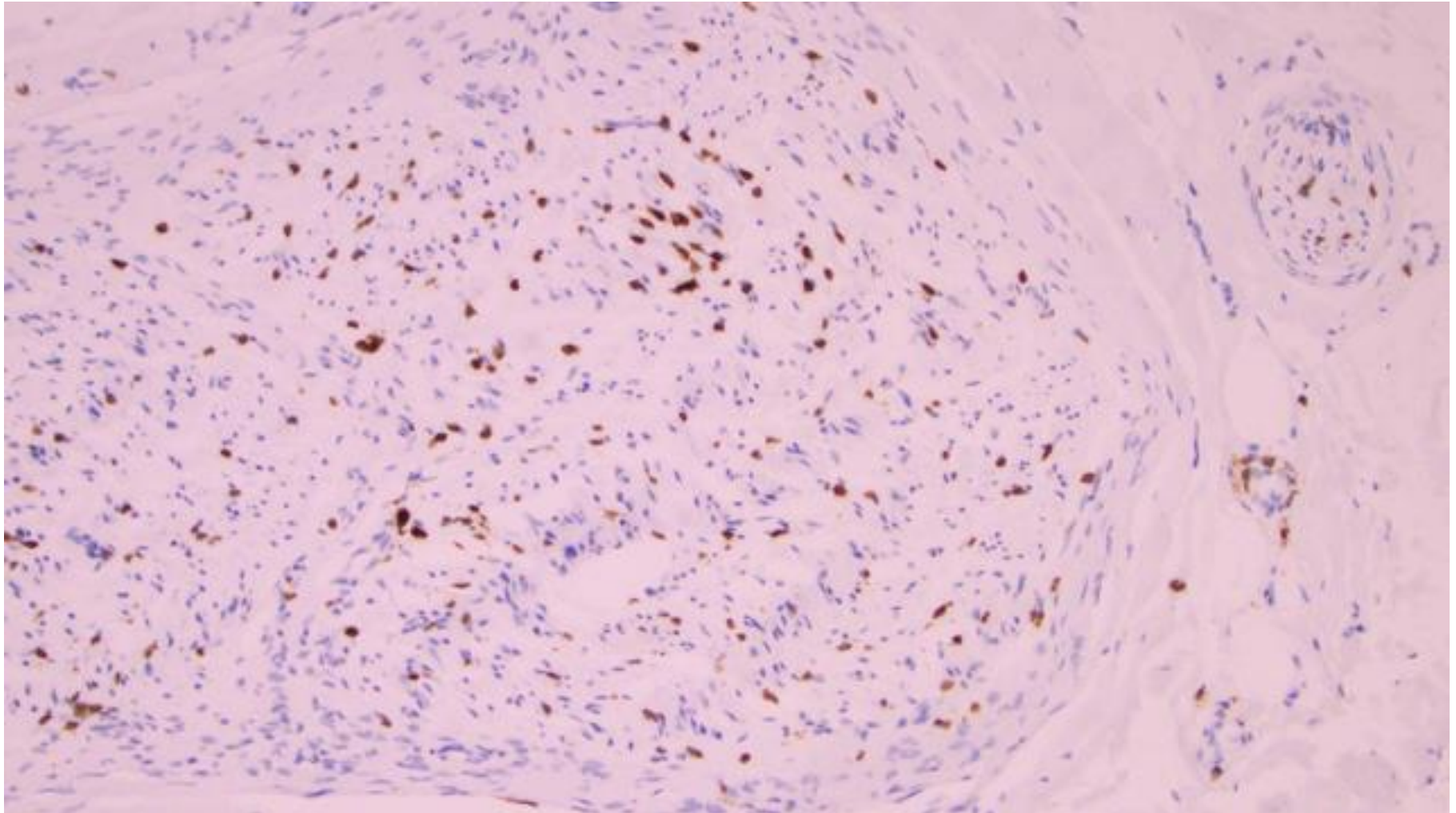


Evaluation



Left Sciatic Nerve Biopsy Pathology:

CD3



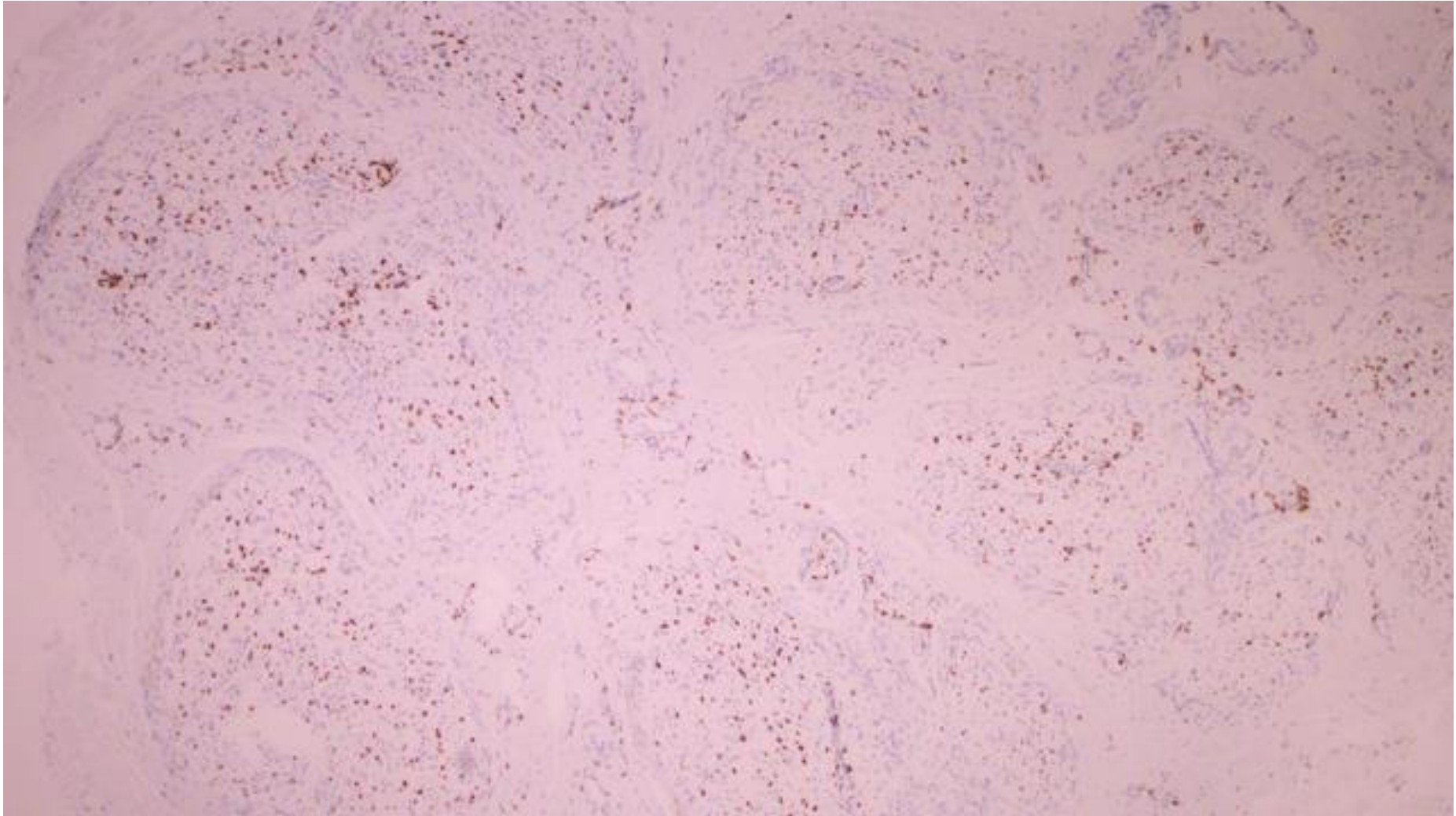


Evaluation



Left Sciatic Nerve Biopsy Pathology:

CD3



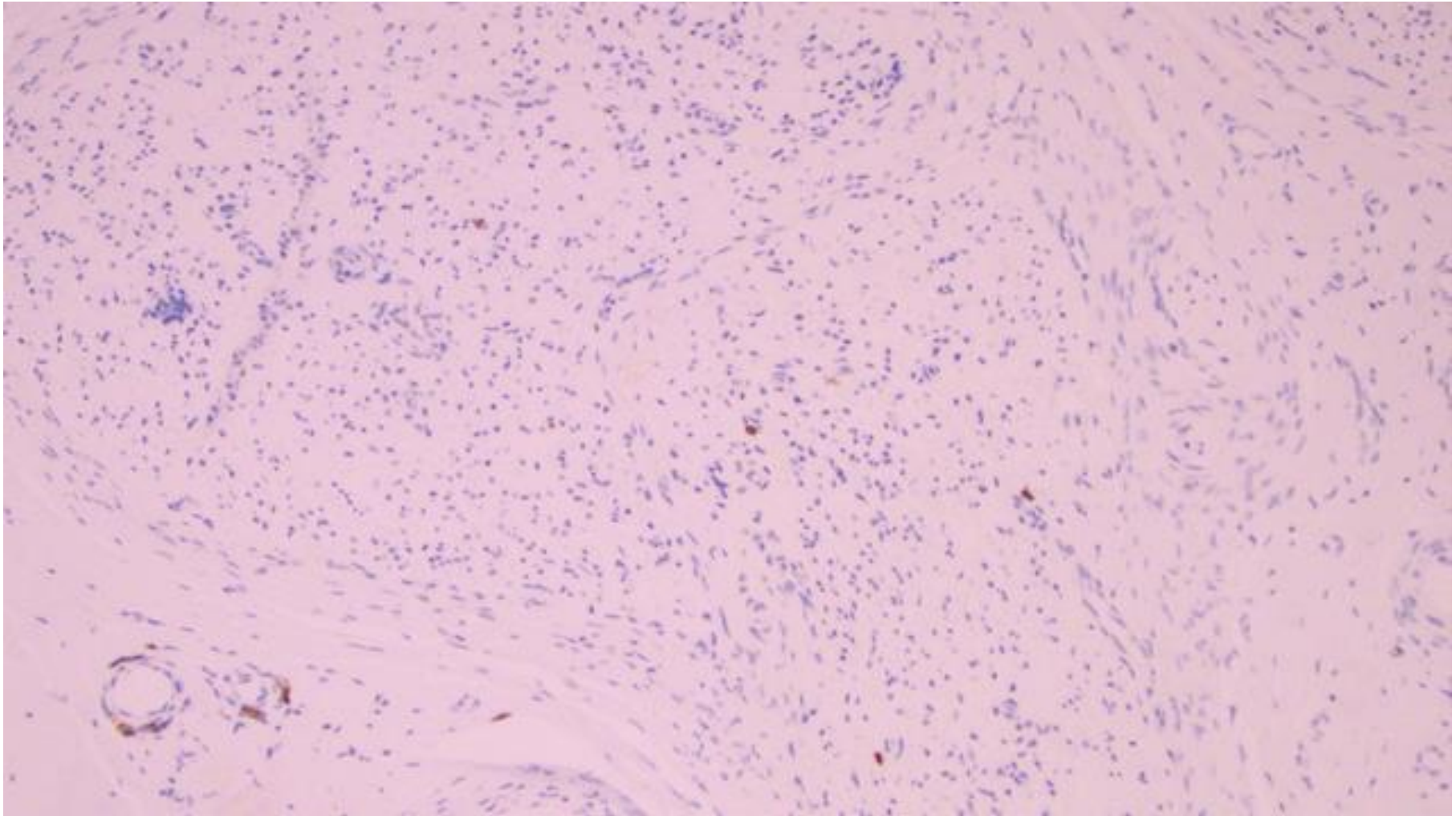


Evaluation



Left Sciatic Nerve Biopsy Pathology:

CD20



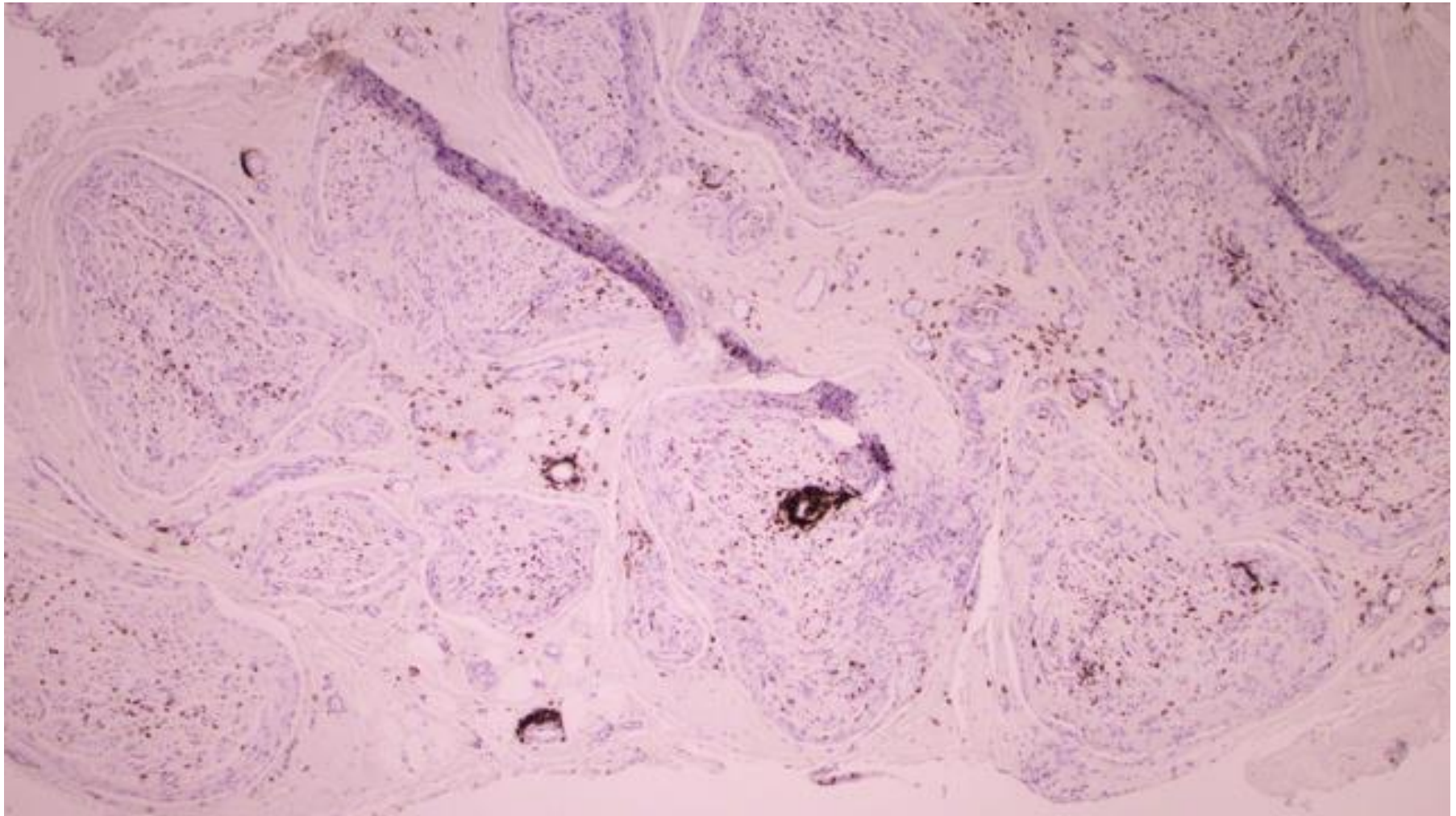


Evaluation



Left Sciatic Nerve Biopsy Pathology:

CD45



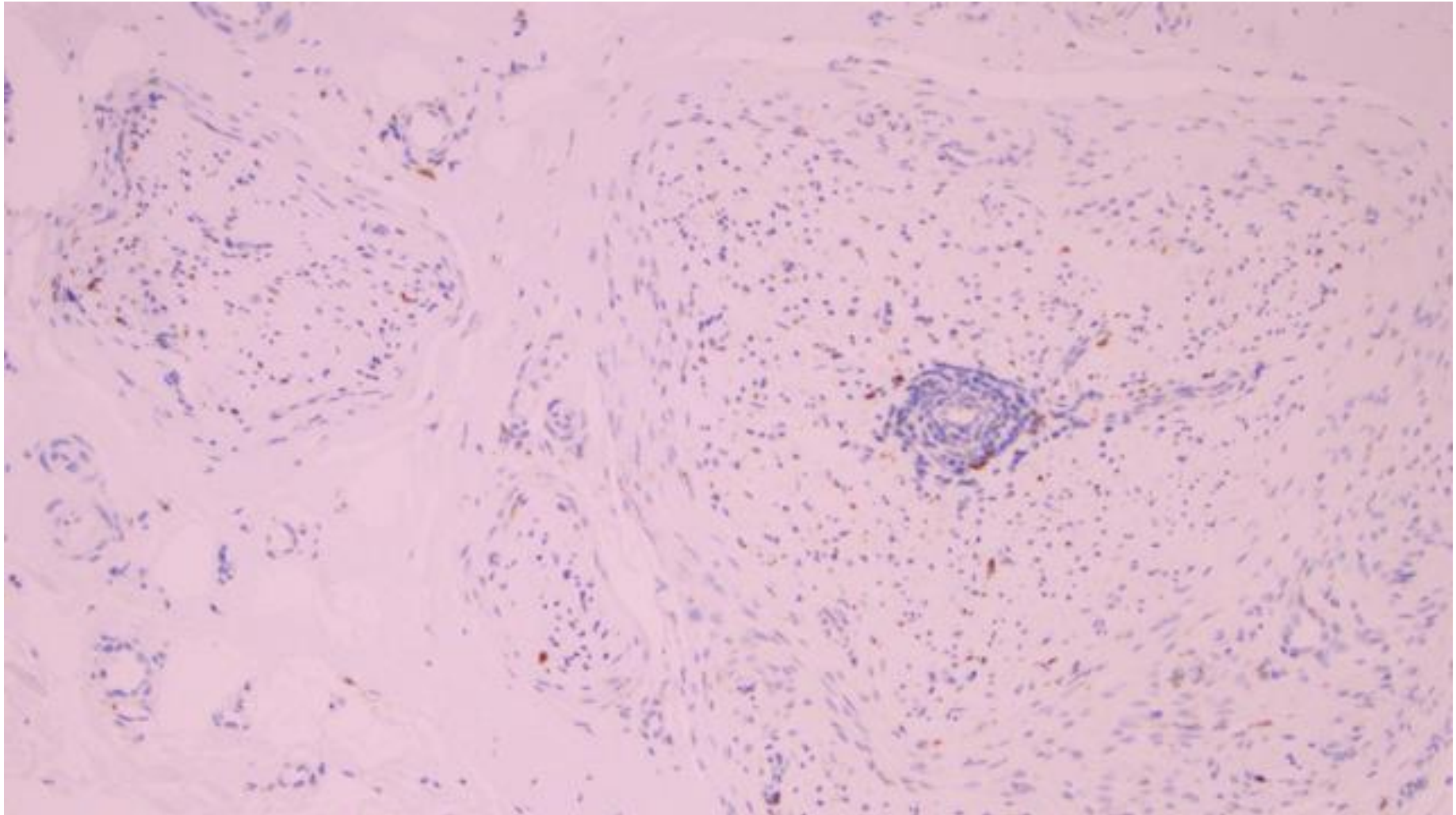


Evaluation



Left Sciatic Nerve Biopsy Pathology:

CD68

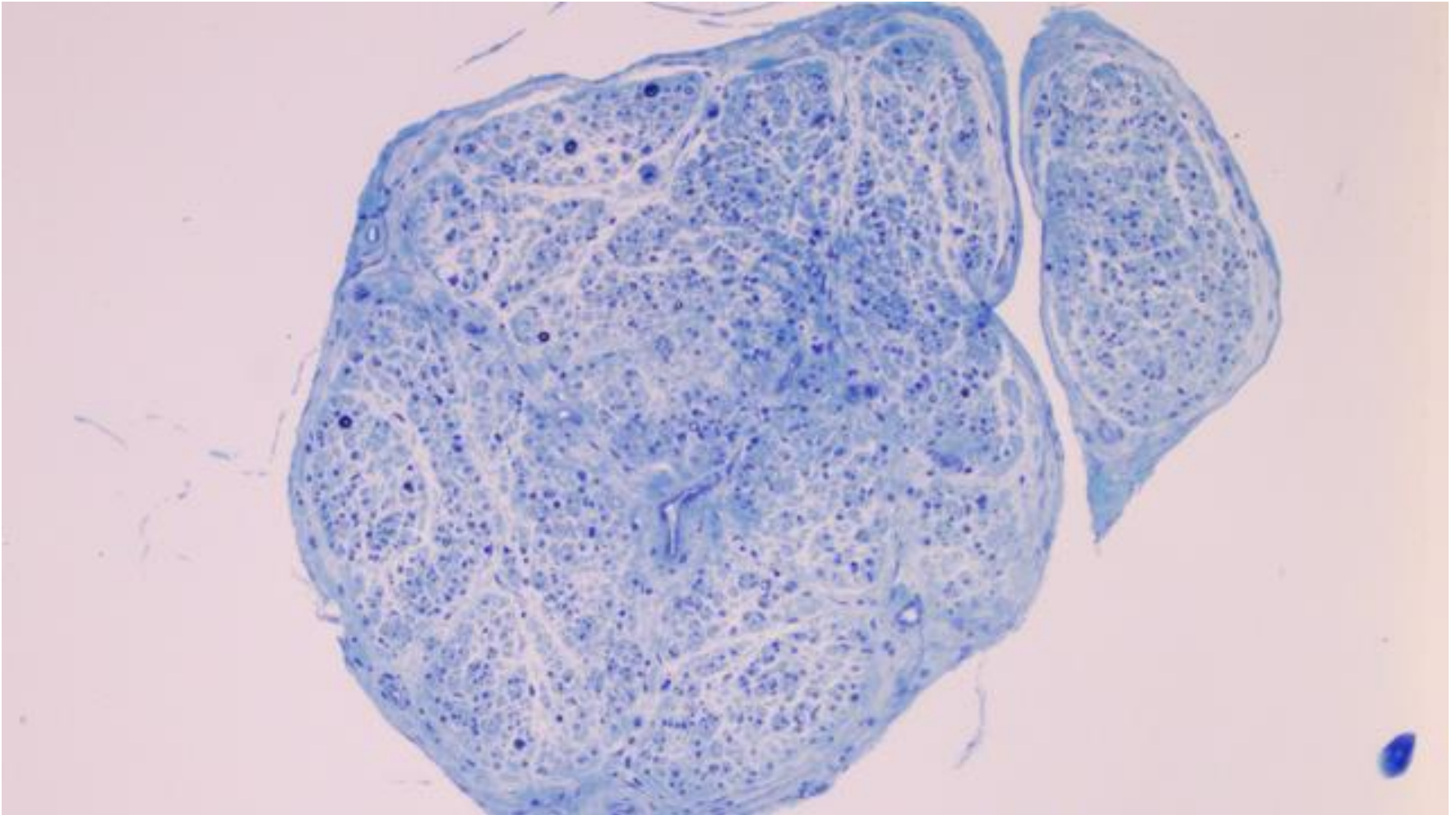




Evaluation



Left Sciatic Nerve Biopsy Pathology: Methylene Blue

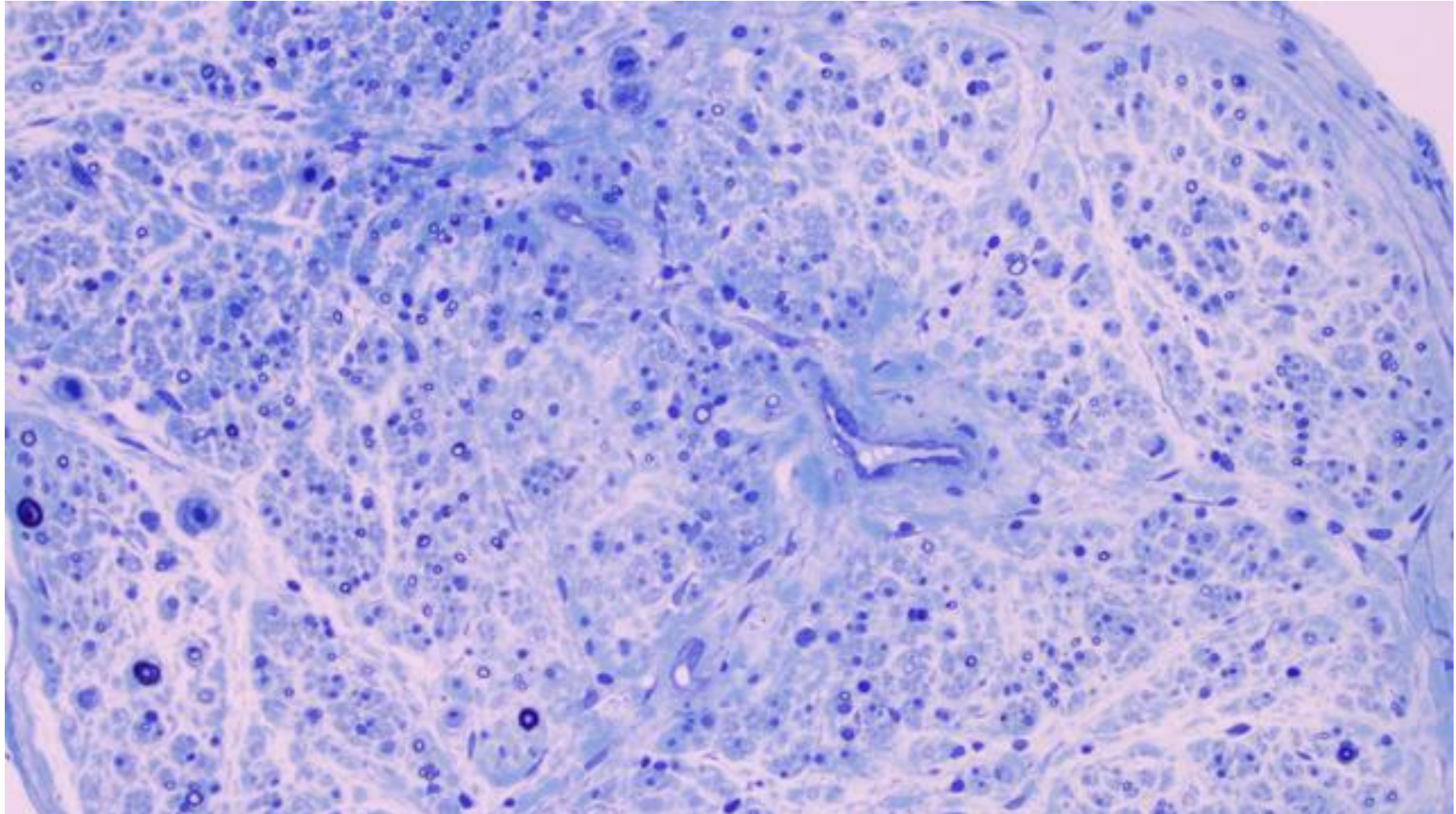




Evaluation



Left Sciatic Nerve Biopsy Pathology: Methylene Blue





Evaluation



Left Sciatic Nerve Biopsy Pathology Report:

Mononuclear inflammatory Cells (mainly T lymphocytes) within endoneurium corroborated by Mayo Clinic pathology laboratory.



Evaluation



STUDIES:

Follow up PET/CT scan 6 months later:

Resolution of increased metabolic activity in sciatic nerve.



The working diagnosis is...



FOCAL CIDP



Management



- Started on IVIG



Differential for Non-traumatic Lumbosacral Plexopathy



- **Tumor/Mass:**

Malignant Invasion, Metastasis, Benign Tumor, Amyloidosis

- **Infection:**

Local spread or generalized infection

- **Inflammatory:**

Diabetic lumbosacral radiculoplexus neuropathy, post-surgical inflammatory neuropathy, sarcoidosis

- **Radiation**

- **Hematoma**

Dyck PJB and Thaisetthawatkul P. 2014.



Differential for Non-traumatic Lumbosacral Plexopathy



Tumor/Mass:

- The majority of cases of lumbosacral plexopathy arise as a consequence of direct spread from pelvic tumors - most commonly rectal gynecological or sarcoma.
- Metastatic spread is another mechanism, typically breast, lung or prostate cancers.
- Typical presentations are pain in the buttock, perineum, posterolateral leg aspect, bladder dysfunction and leg edema.

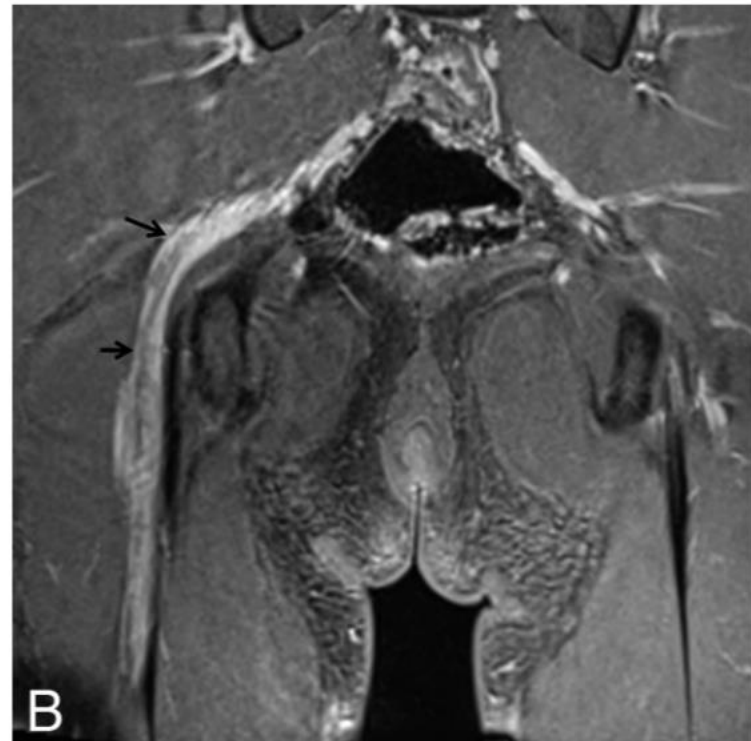
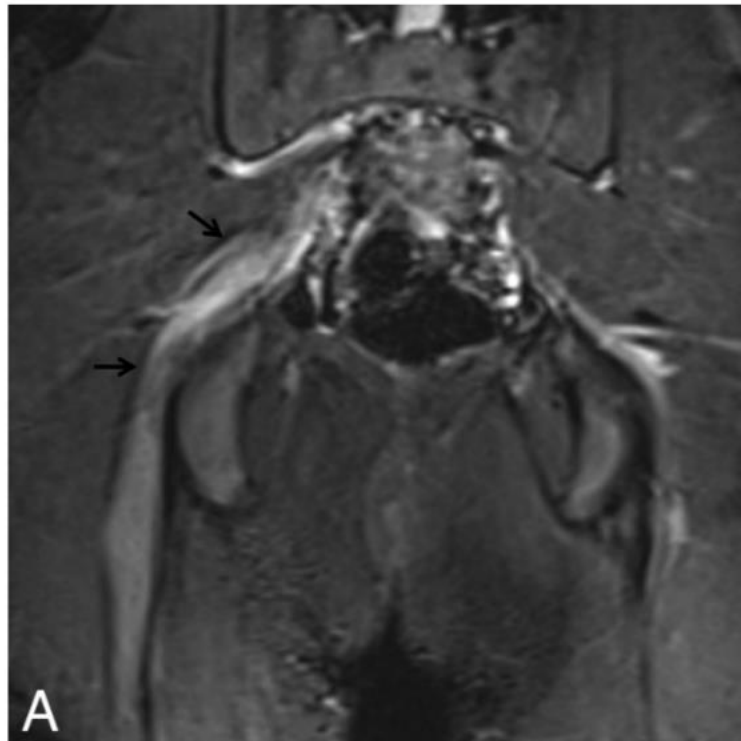


Differential for Non-traumatic Lumbosacral Plexopathy



Benign Tumors:

- Primary nerve sheath tumors, though uncommon, can occur particularly in neurofibromatosis and can present in younger patients.



Thawait et al. 2011.

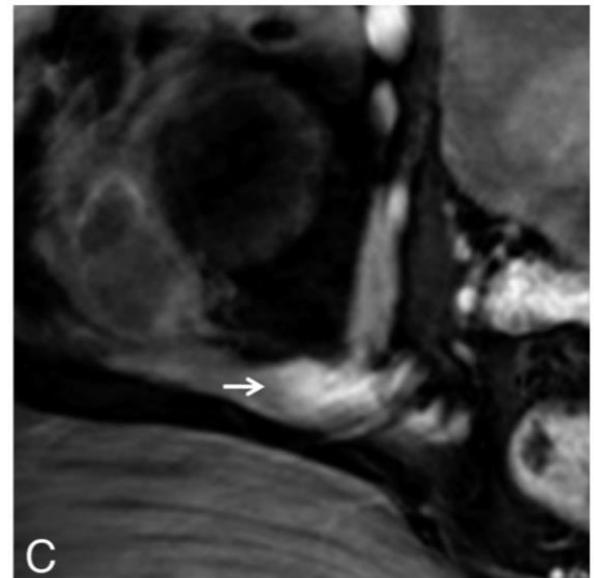
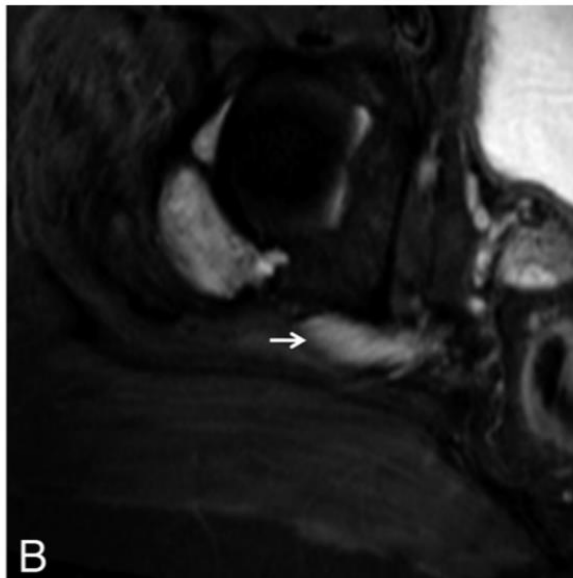
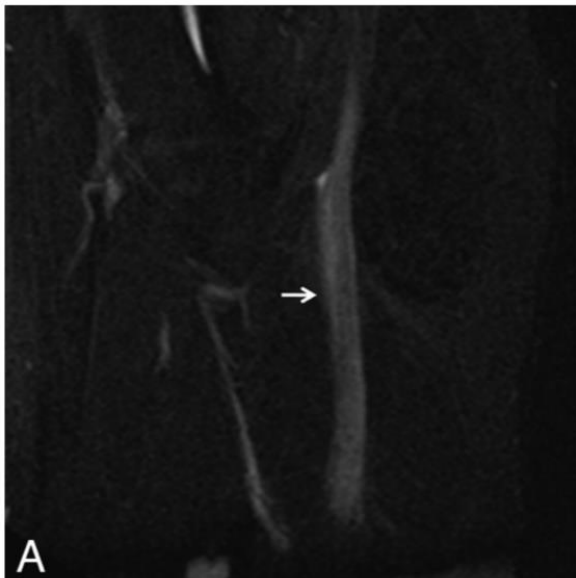


Differential for Non-traumatic Lumbosacral Plexopathy



Benign Tumors:

- Focal deposits of amyloid, are a rare but established cause referred to as an amyloidoma. This is typically secondary amyloid related to light chain deposition rather than inherited varieties.



1. Dyck PJB and Thaisetthawatkul P. 2014. 2. Thawait et al. 2011.

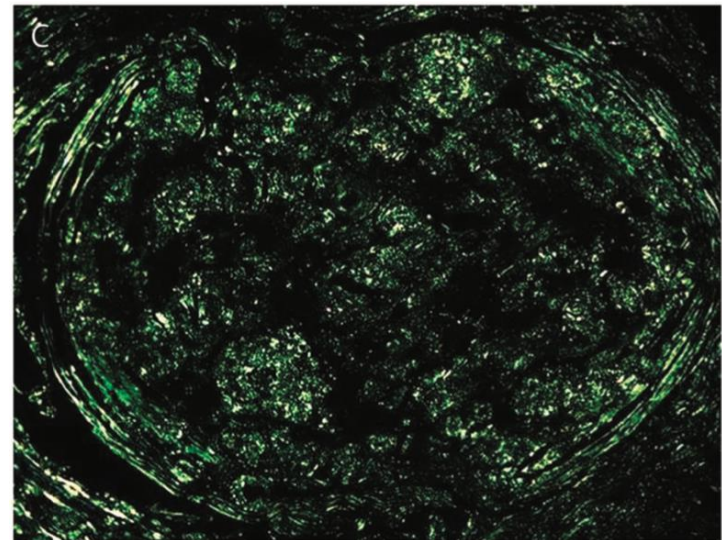
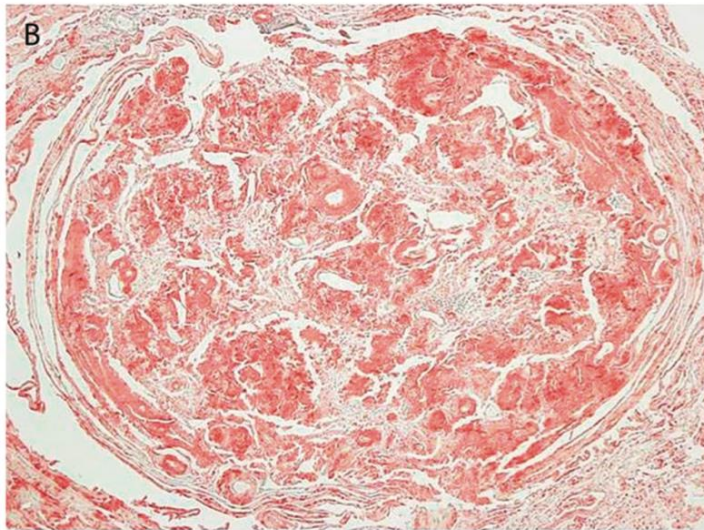


Differential for Non-traumatic Lumbosacral Plexopathy



Benign Tumors:

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1. Tracy, JA Et al. 2019. 2. Dyck PJB and Thaisetthawatkul P. 2014.



Differential for Non-traumatic Lumbosacral Plexopathy



Infections:

- Abscess or gas formation particularly in the psoas muscle can result in compression of the LS plexus.
- Diffuse Infiltrative Lymphocytosis Syndrome (DILS) is seen in HIV patients causing a painful, plegic lower extremity neuropathy with T-cell infiltrates on Biopsy.



1. Dyck PJB and Thaisetthawatkul P. 2014.

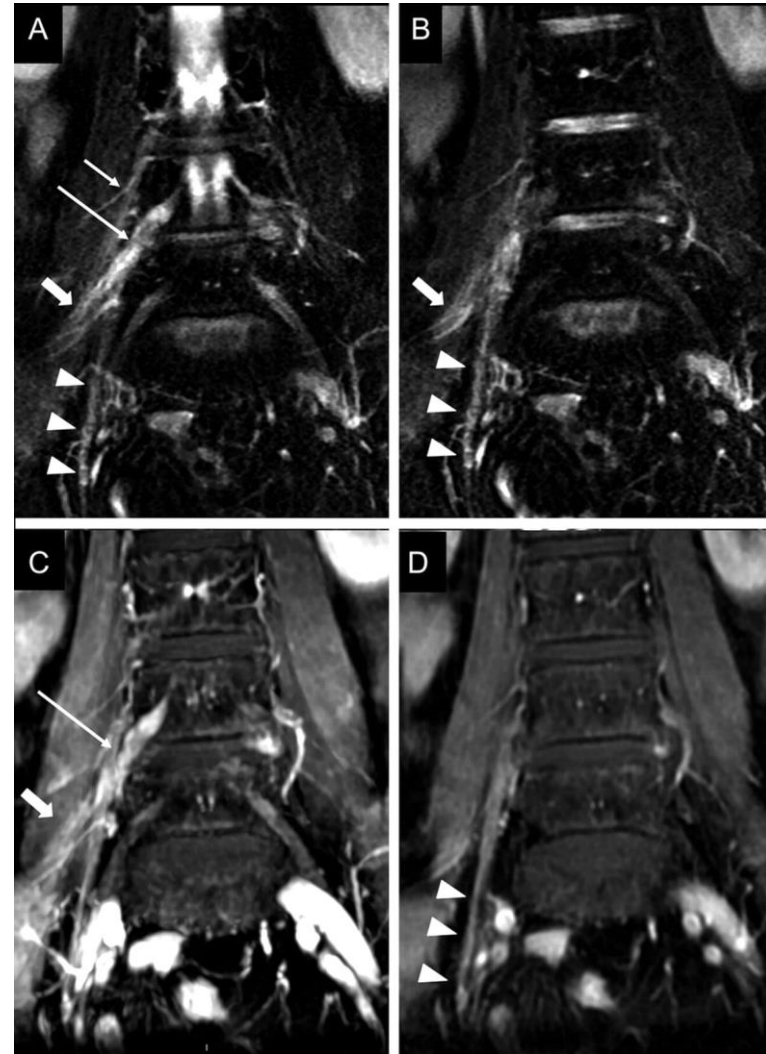


Differential for Non-traumatic Lumbosacral Plexopathy



Diabetic Amyotrophy:

- Asymmetrical proximal motor neuropathy often associated with back, hip and knee pain often accompanying significant weight loss. There is typically weakness and wasting in the hip flexors, adductors and quadriceps with subtle sensory features.





Differential for Non-traumatic Lumbosacral Plexopathy



Idiopathic Lumbosacral Plexopathy:

- Present with acute onset of spontaneous and painful lower limb weakness and sensory loss. It can also be bilateral. This presentation is very similar to the diabetic variety and there is pathological investigations suggesting that this may be the same process.



Differential for Non-traumatic Lumbosacral Plexopathy



Radiation Plexopathy:

- Lumbosacral plexus is frequently affected from radiotherapy to neoplasm in pelvic region. This can often includes lymph nodes in the abdominal aortic and ilioinguinal region.
- Electrodiagnostic studies may demonstrate Myokymia though absence of it does not infer that radiation is not the culprit.
- Effects can be seen usually after 6 months but can develop many years later. Some studies have demonstrated effects > a decade later.



Differential for Non-traumatic Lumbosacral Plexopathy



- Retroperitoneal Hematoma:



Thapa SS et al. 2016.



Focal CIDP resulting in Lumbosacral Plexopathy



References:

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- Thawait, SK et al. High resolution MR Neurography of diffuse peripheral nerve lesions. American Journal of Neuroradiology. 2011; **32**: 1365 – 1372.
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Thank You