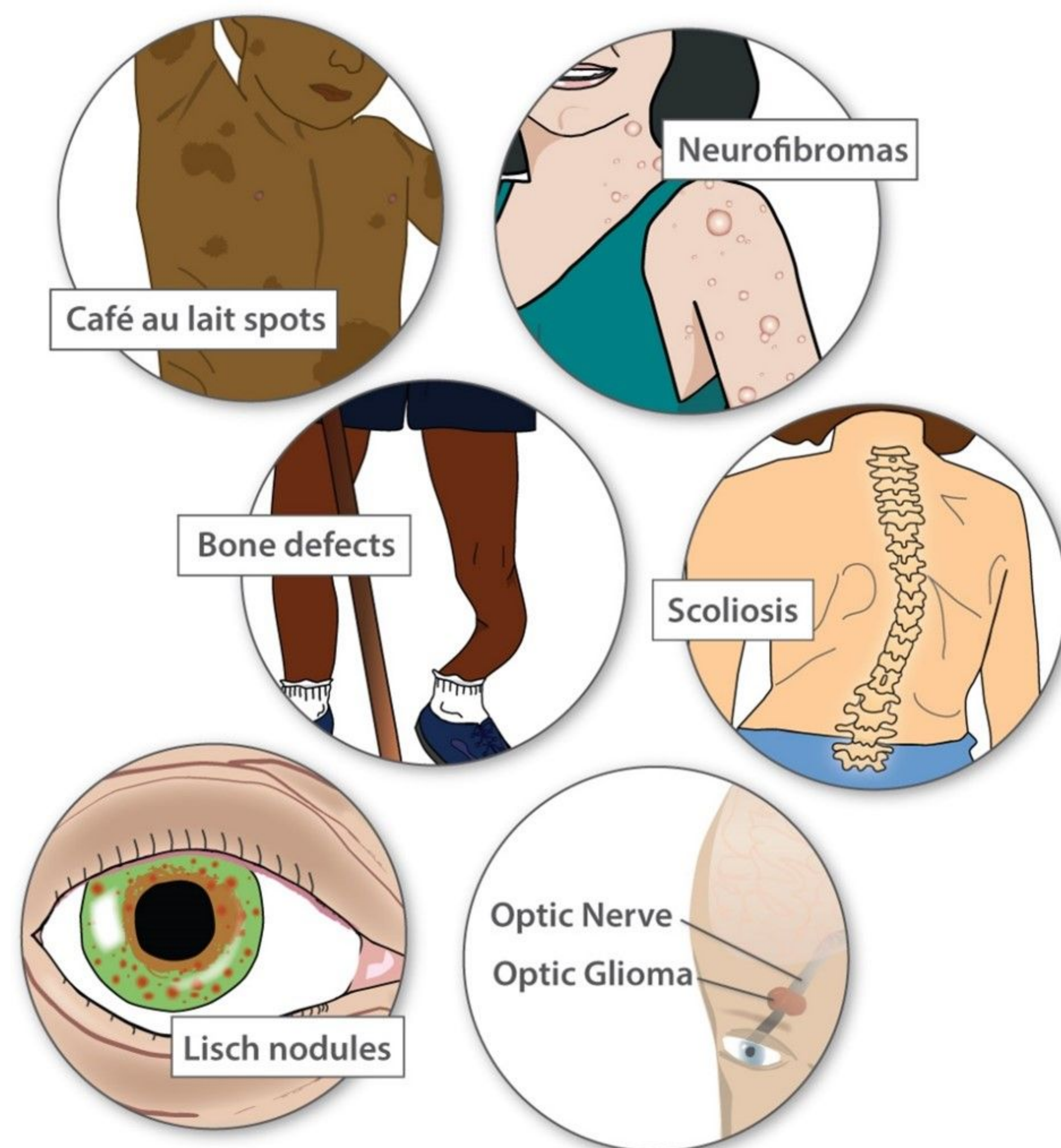


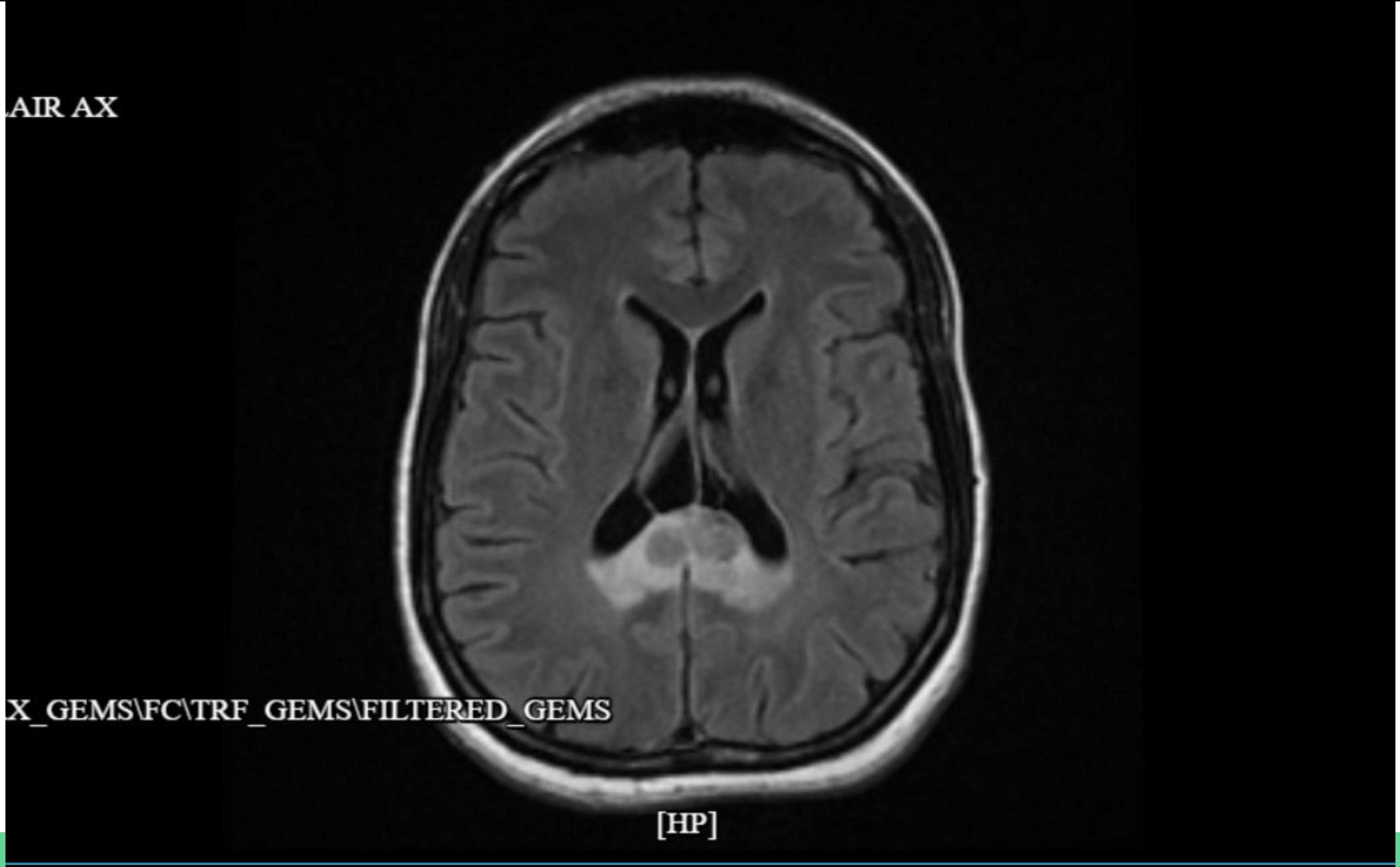
Epidural in Neurofibromatosis: proceed with caution!

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- Neurofibromatosis Type 1 is a Autosomal Dominant Disease
- Incidence of 1:3000 with 100% Penetrance
- Presentation: Cafe-au-lait spots, cutaneous neurofibromas and subcutaneous neurofibromas (in the peripheral NS), optic gliomas, scoliosis, Lisch Nodules, axillary freckling, Pheochromocytoma
- Current literature is equivocal about the need for radiologic testing prior to neuraxial anesthesia despite the possibility of intracranial and spinal tumors

- 35 yo G1P0 Female presented for induction of Labor at 39+1 weeks.
- PMhx of Hypothyroidism on levothyroxine, otherwise no other hx.
- Physical Exam: Multiple nodular lesions on her face, neck, chest, back, and arms as well as diffuse flat macular spots on her skin.
- Airway Exam: Mallampati 4 with multiple nodules in her airway.
- High suspicion for NF Type 1 with no formal diagnosis. Normal Neurologic Exam and no signs of Increased ICP.
- Proceeded with Epidural; initial attempt between L4-L5 which elicited R sided thigh discomfort, immediately aborted and placed successfully in L3-L4 (loss at 6cm and taped at 11cm at the skin)
- Birthing Course: Initially she felt relief however she started to feel a small window of pain around the left lower abdomen despite several boluses, other areas of were covered by the epidural. Attempt to pull out a couple of centimeters (from 11 cm to 9 cm) and bolused without resolve. Patient was given a dose of diphenhydramine and butorphanol, successfully delivered the baby vaginally a couple of hours later.
- Postpartum Neurology was consulted and Imaging was obtained; formally diagnosed with NF Type 1.
- Imaging of the spinal cord showed a large neurofibroma on the left between L5-S1.



Discussion

- The large neurofibroma between L5 and S1 was likely impinging and distorting the spine and the epidural space resulting in the patchy coverage of labor analgesia.
- We were fortunate that we avoided the need for general anesthesia and difficult airway management.
- There are several case reports that show uncomplicated neuraxial in labor while one was complicated by epidural hematoma after dural puncture. One radiologic study shows about 40% of NF patients have involvement of their CNS, mostly involving nerve roots.
- Currently there is lack of consensus among the anesthetic approach for these patients; the American College of Medical Genetics and Genomics did not feel there was a need for preop radiologic testing prior to spinal anesthesia
- We recommend that asymptomatic NF patients should have full radiologic work up before neuraxial anesthesia and when not possible a risk-benefit analysis should be conducted to determine safe practice.
- In facilities that have the infrastructure and policies in place, other forms of labor analgesia could be considered as well such as remifentanyl infusions or a fentanyl PCA pump.