Childhood onset polyarteritis nodosa is a rare vasculitis that affects multiple systems including the central and peripheral nervous system, kidneys, gastrointestinal system, and the skin. It is an autosomal recessive disorder that has a wide range of symptoms including necrotizing vasculitis, aneurysms in multiple locations, hypercoagulability, hepatosplenomegaly and infarcts in multiple areas of the brain. Multiple examples of nerve palsies affecting the face, as well as the upper but primarily the lower extremities. Genetic and genomic sequencing has shown that a homozygous or heterozygous mutation in the CECR1 gene is responsible for childhood onset polyarteritis nodosa. This mutation causes a decrease or non functioning of the enzyme adenosine deaminase 2 which leads to a propensity for inflammation in blood vessels, as well as an decrease in blood vessel integrity. There are 46 patients worldwide with this identified mutation. Treatment consists primarily of either immunosuppression or bone marrow transplant.

A 12 year old male with a history of well controlled childhood onset polyarteritis nodosa secondary to a DADA2 mutation. Additional comorbidities include ADHD, asthma and GERD. Referred to our pediatric pain clinic after he developed significant right sided foot drop and a sciatic mononeuropathy following a bone marrow biopsy. He was seen 3 months later in clinic for follow up. He had near complete resolution of foot drop. He reported minimal pain and no hyperalgesia of the right leg. His family reports that he has returned to full activity.

He was taken to interventional radiology where he underwent successful angiography and coilng of the lesion. He presented to pain clinic with a right foot drop and burning pain in his right lower leg. He initially had throbbing pain in his upper leg as well but that had resolved with initiation of gabapentin by his neurologist approximately six weeks previous to his visit. He was also undergoing a course of physical therapy which had significantly improved his foot drop. Since he was doing well with minimal sedating side effects from the gabapentin. The decision was made to continue titration of the gabapentin to maximize relief of his pain. He was thought to have a compressive mononeuropathy of the sciatic nerve. Over the next four days he developed worsening right leg pain, numbness, paresthesias and weakness. He also developed right lower quadrant abdominal pain that persisted and increased in intensity. He underwent a CT scan after presenting to the emergency department that showed a right iliac pseudoaneurysm adjacent to the biopsy tract.

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THE CASE

• Childhood onset polyarteritis nodosa is a rare vasculitis that affects multiple systems including the central and peripheral nervous system, kidneys, gastrointestinal system, and the skin.
• It is an autosomal recessive disorder that has a wide range of symptoms including necrotizing vasculitis, aneurysms in multiple locations, hypercoagulability, hepatosplenomegaly and infarcts in multiple areas of the brain.
• Multiple examples of nerve palsies affecting the face, as well as the upper but primarily the lower extremities.
• Genetic and genomic sequencing has shown that a homozygous or heterozygous mutation in the CECR1 gene is responsible for childhood onset polyarteritis nodosa.
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• There are 46 patients worldwide with this identified mutation
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THE CASE (contd.)

• A 12 year old male with a history of well controlled childhood onset polyarteritis nodosa secondary to a DADA2 mutation
• Additional comorbidities include ADHD, asthma and GERD
• Referred to our pediatric pain clinic after he developed significant right sided foot drop and a sciatic mononeuropathy following a bone marrow biopsy
• an initially uneventful bone marrow biopsy with sedation and was discharged home
• Over the next four days he developed worsening right leg pain, numbness, paresthesias and weakness.
• He also developed right lower quadrant abdominal pain that persisted and increased in intensity.
• He underwent a CT scan after presenting to the emergency department that showed a right iliac pseudoaneurysm adjacent to the biopsy tract.

THE CASE (contd.)

• He was taken to interventional radiology where he underwent successful angiography and coiling of the lesion.
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OUTCOME

• He was seen 3 months later in clinic for follow up
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REFERENCES
