

Isolated Plexiform Neurofibroma Mass in Foot and Ankle with Vertical Talus Reconstruction

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INTRODUCTION / PURPOSE

The incidence of congenital vertical talus is known to be a rare pedal deformity found 1 in 10,000 with only 50% of these cases having an association with neuromuscular disorders¹. This unique case report utilizes the presentation of an isolated, large, and painful plexiform neurofibroma mass in the setting of a rigid, unilateral vertical talus. With the collaboration of the podiatric surgical team and orthopaedic surgical oncologist, a unique treatment protocol for the excision of a large neurofibroma in combination of flatfoot reconstruction is described.

LITERATURE REVIEW

Literature and previous case reports on the presentation of unilateral vertical talus in the presence of isolated, plexiform neurofibroma mass are limited. Llyod-Roberts and Spence described one case of severe flatfoot with talar-navicular subluxation in combination with neurofibromatosis, with no clinical significance of vertical talus in the disease². The original article mentioned above, highlights the rarity of this clinical presentation, as no recent research, known to the authors, has been described since.

PATIENT PRESENTATION



Fig 1a. Initial presentation

- **HPI:** 14 yo male presents with large soft tissue mass in right posterior medial ankle/foot impeding comfortable shoe wear and daily activities. Soft tissue mass has grown in size in combination with painful, progressive flatfoot deformity

- **PMHX:** Neurofibromatosis, Type I
- **PE:** Soft, 10-12cm mobile, non tender mass to palpation in right posterior medial ankle/foot. DP/PT palpable. Full ROM of right ankle with valgus and eversion rotational deformity of foot and ankle. No clinical signs of infection noted.
- **Imaging:** RLE MRI with severe flatfoot deformity in the presence of a large soft tissue mass that is complex with predominant T2 hyperintense signal, with areas of focal hypointensity.



Fig 1b. Initial presentation

DIAGNOSTIC RADIOLOGY

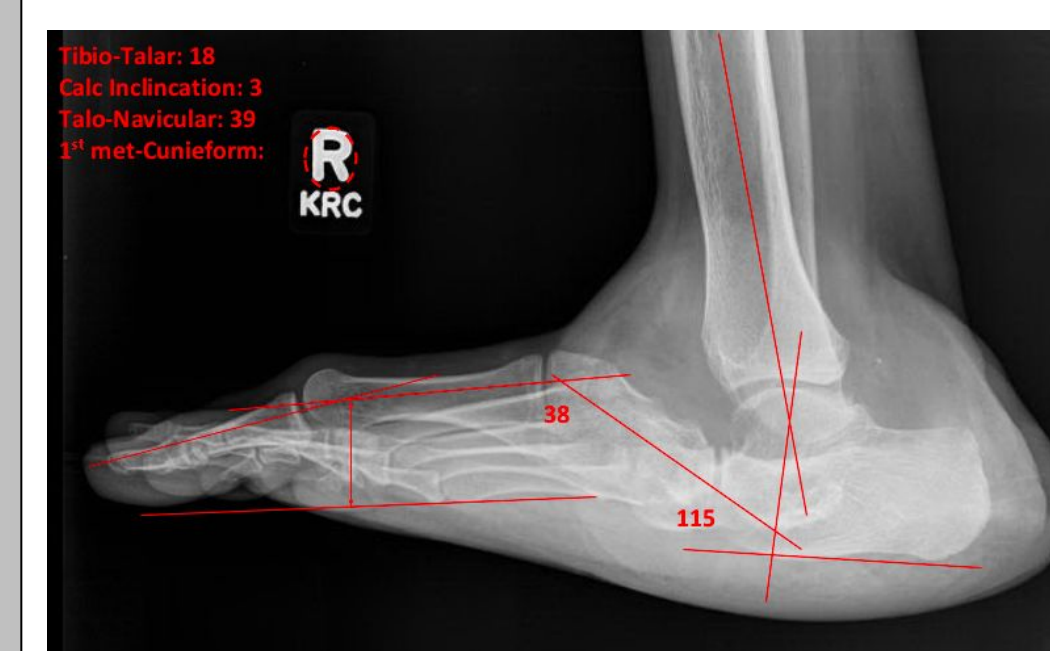


Fig 2a. Initial LAT X-Ray

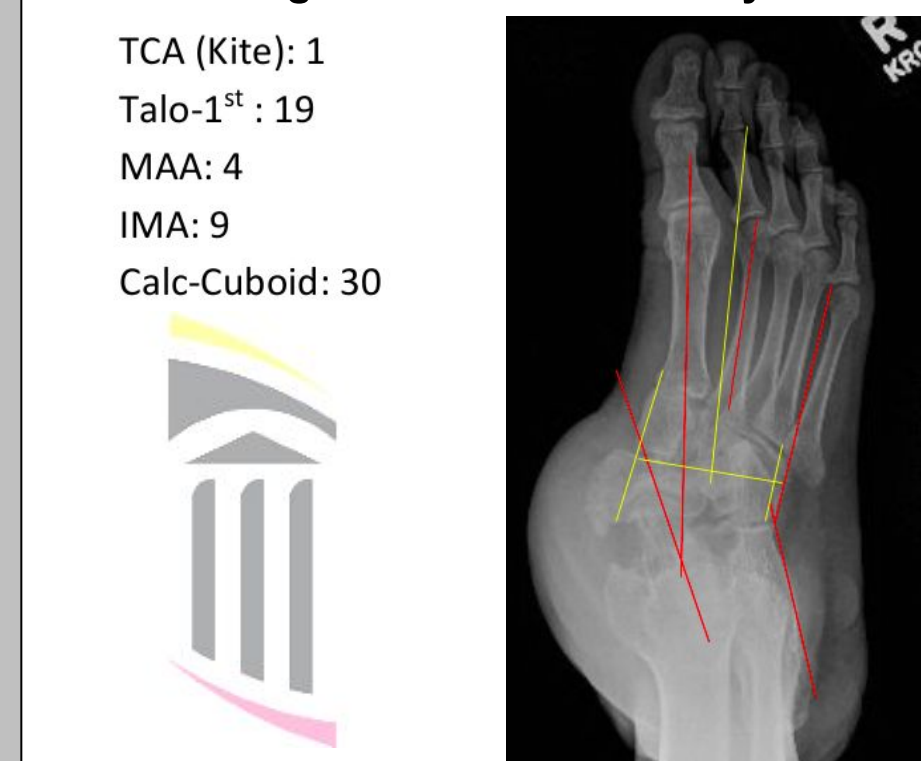


Fig 2b. Initial AP X-Ray



Fig 3. CT scan including sagittal view (top row) and coronal view (bottom row)

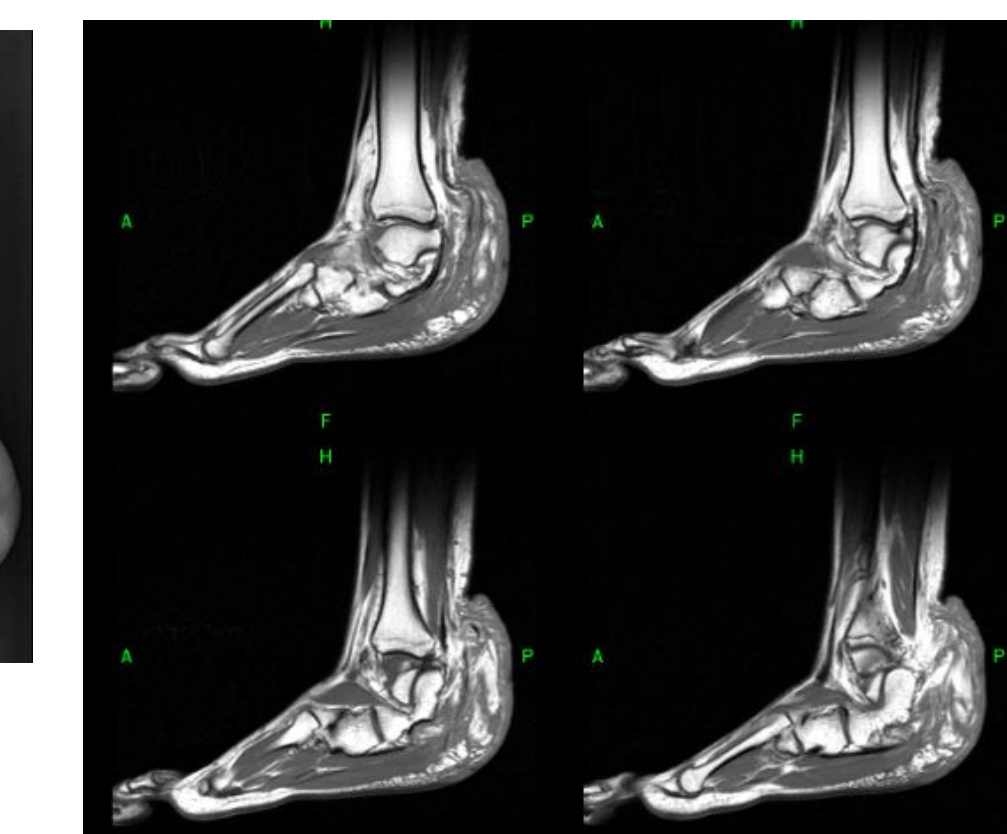


Fig 4a. MRI T1 Sagittal

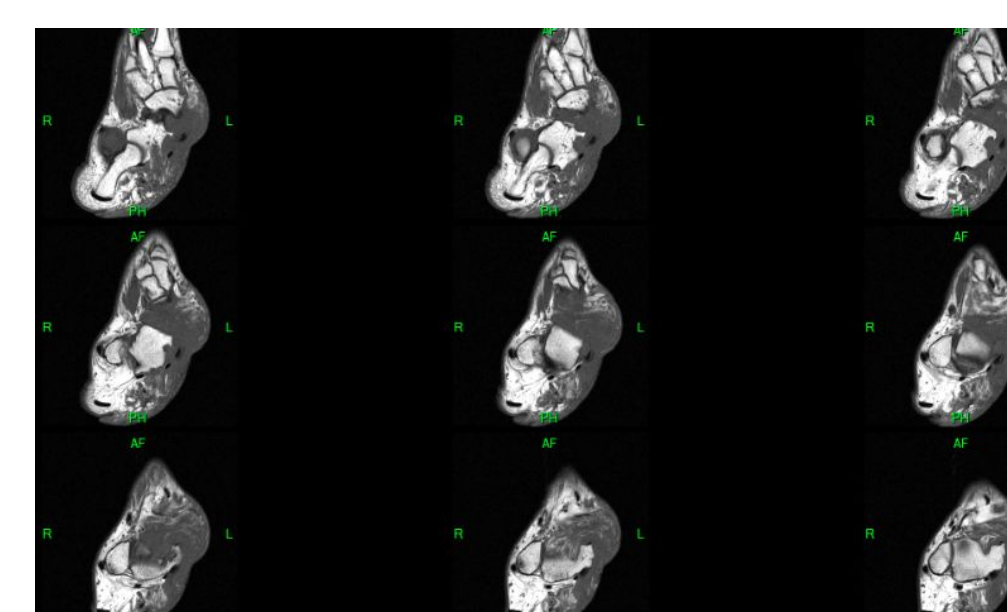


Fig 4b. MRI T1 Coronal

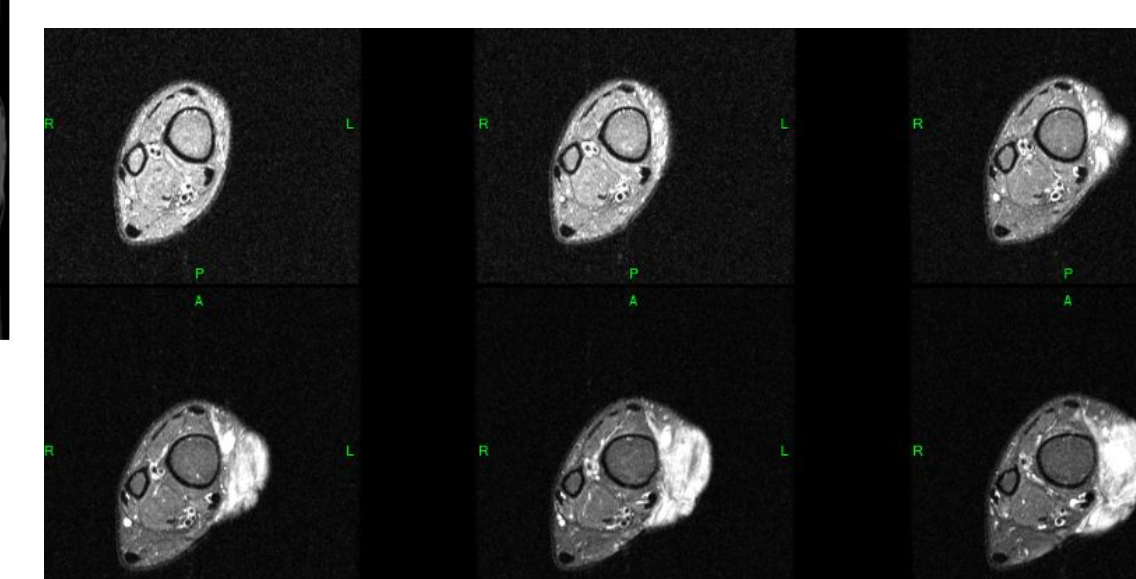


Fig 4c. MRI T2 Axial

PREOPERATIVE PLANNING

- Multi-specialty surgical team approach consisting of Foot and Ankle Reconstructive surgeon, Orthopaedic Surgical Oncologist, Vascular Surgeon and Plastic Surgeon (for potential free flap vs. STSG)
- Vascular angiogram performed preoperatively to evaluate arterial anatomy of the tumor

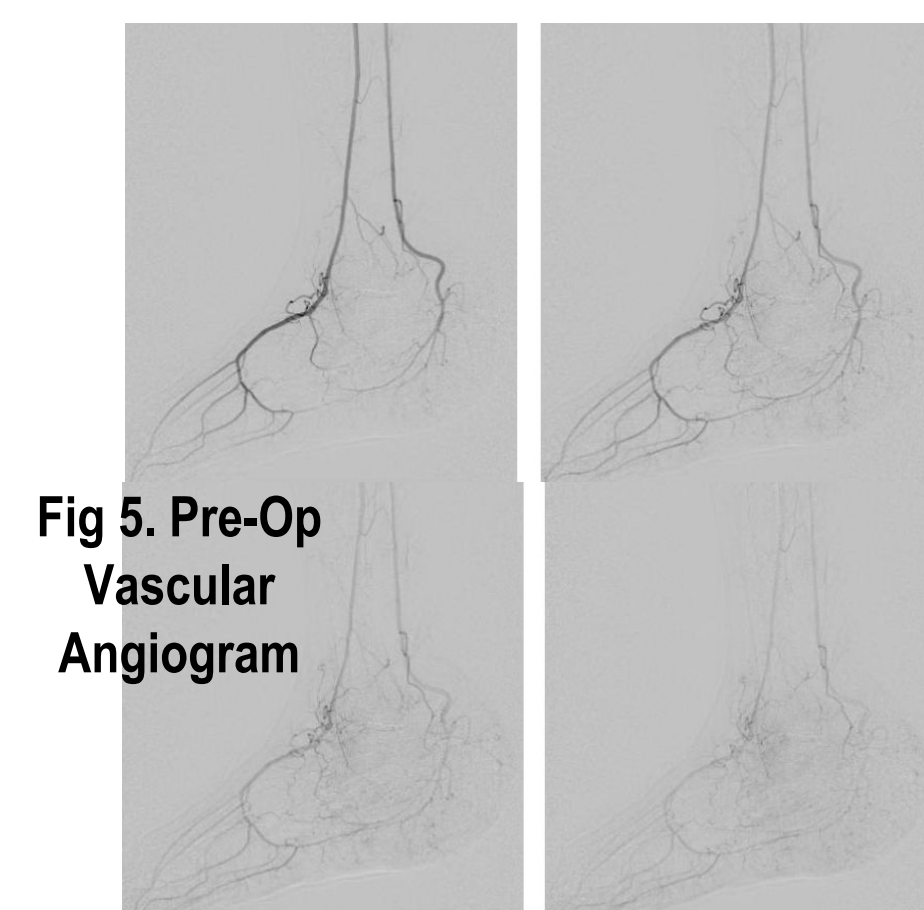


Fig 5. Pre-Op Vascular Angiogram

METHODS: FOOT / ANKLE RECONSTRUCTION

- **Step 1:** Open biopsy of right posterior medial ankle soft tissue mass. (Confirmed neurofibroma)
- **Step 2:** Radical resection of right lower extremity soft tissue mass approx. 15cm in diameter, with dissection of tumor off the medial plantar nerve, lateral plantar nerve, tibial nerve, posterior tibial artery and vein.
- **Step 3:** Correct TNJ/Dislocation via complete extensor tendon "Z" lengthening (Peroneus Tertius, EDL, EHL, TA)
- **Step 4:** Lateral tendon "Z" lengthening of Peroneus longus and Peroneus brevis
- **Step 5:** Correction of Equinus deformity via Achilles "Z" lengthening, posterior capsule release of both ankle and subtalar joint
- **Step 6:** Midfoot dorsal subluxation correction via TNJ and CCJ capsule release
- **Step 7:** Correction of forefoot abduction via Evans calcaneal osteotomy
- **Step 8:** Triple arthrodesis with PTT transection
- **Step 9:** Cotton osteotomy for correction of forefoot varus
- **Step 10:** Deltoid and flexor retinaculum reconstruction
- **Step 11:** Split thickness skin graft harvest and application
- **Step 12:** Wound vac application

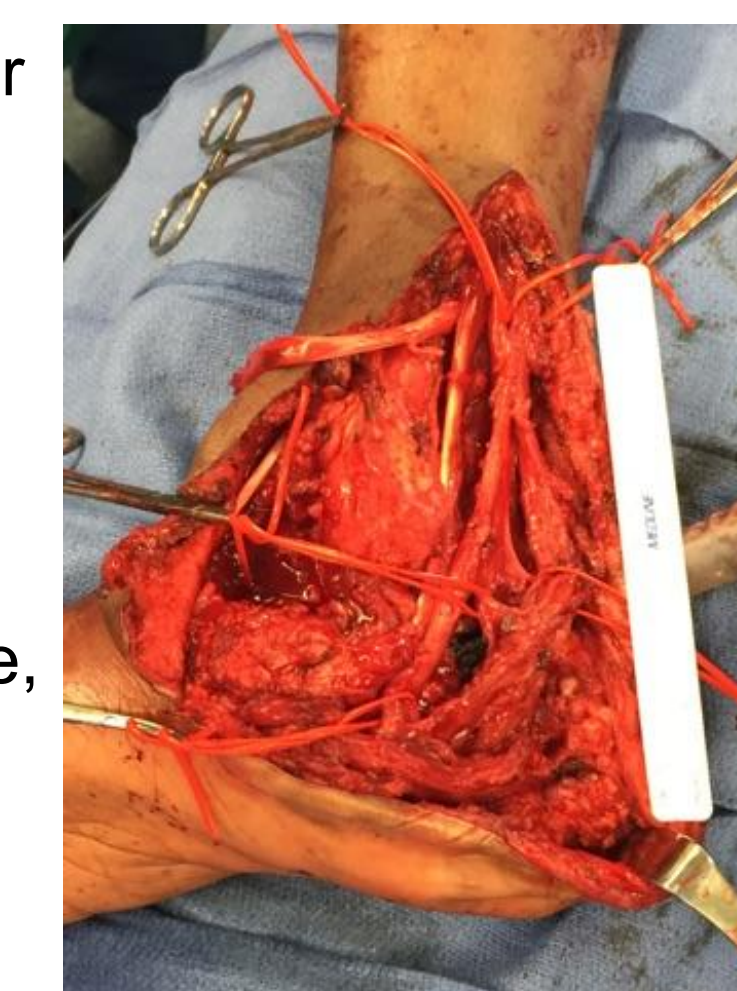


Fig 6. Intraoperative dissection of plexiform neurofibroma

POST OPERATIVE COURSE

- Wound vac application for adequate healing of large soft tissue defect to right foot
- Strict NWB to RLE for 6 weeks post operation
- Transition to 50% weight bearing in protected boot at 6 weeks, continue monitoring surgical incisions
- Advance to 100% WBAT in protected boot at 3 months
- At 5 months post op, transition from protected boot to ambulation in sneaker with custom inserts
- Aggressive physical therapy initiated at 6 months post operation for improvement of muscle strengthening

RESULTS: 3 YEAR FOLLOW UP

Upon presentation at the 3 year follow up appointment, the 17yo patient presents ambulating with non antalgic gait in men's athletic sneakers, fitted with custom molded orthotics. Patient indicates he is able to participate in intramural sports and run/walk without pain.

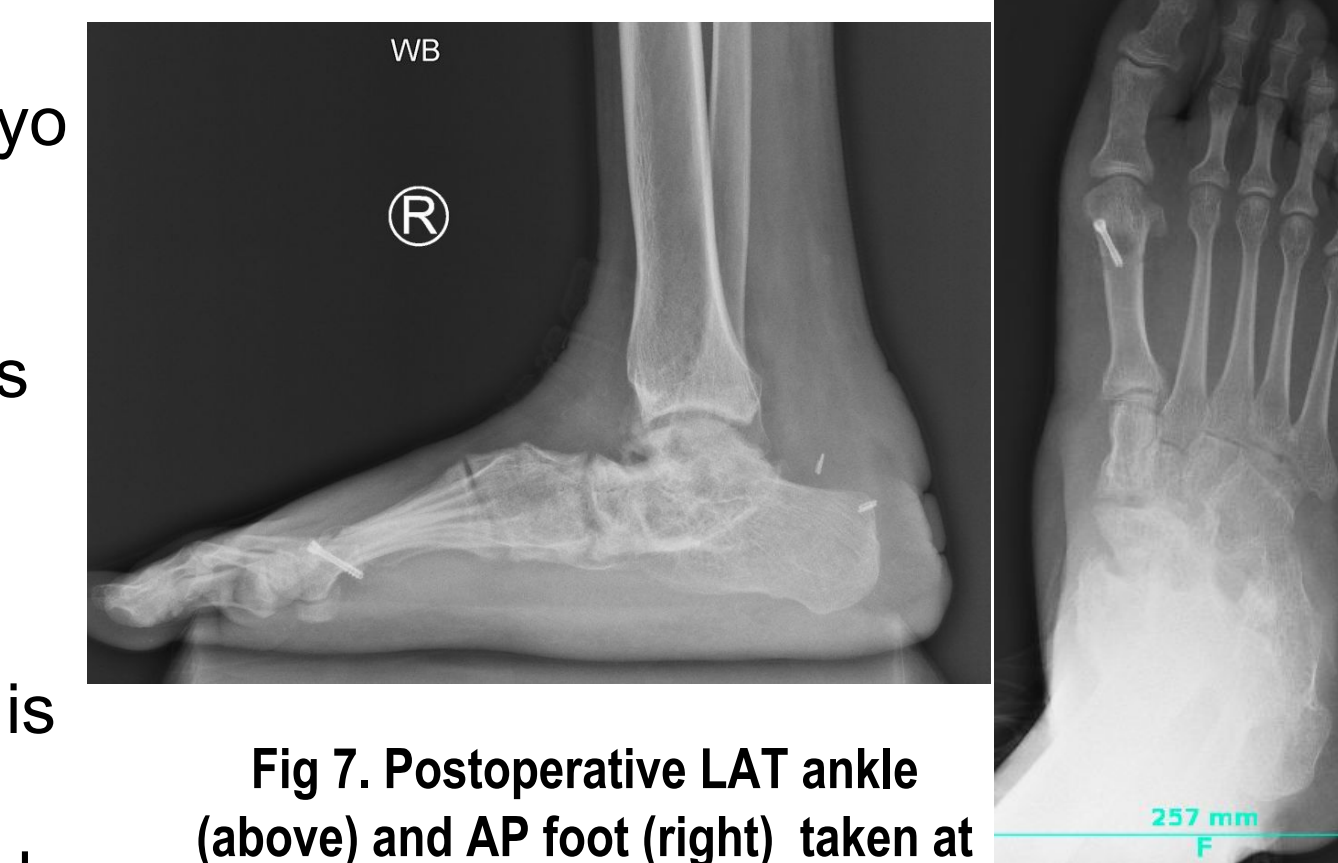


Fig 7. Postoperative LAT ankle (above) and AP foot (right) taken at 2.5 yr follow up visit

DISCUSSION

The foot and ankle reconstructive surgeon utilized a technique similar to Dobbs, et al. in the single stage surgical approach for correction of congenital vertical talus⁴. This patient's successful outcome indicates that the surgical intervention of posterior capsule release with tendon transfers and arthrodesis of the midfoot and subtalar joint are viable surgical options for delayed vertical talus correction in conjunction of excision of a large, plexiform neurofibroma. Today, the patient exhibits a neurovascularly intact plantigrade foot that is able to withstand the daily activity of a (now)17-year-old male. Following a literature search, the rarity of this case presentation was acknowledged. As clinicians, it was the responsibility of the surgical team to share the success of our operative intervention and treatment plan with the medical community.

REFERENCES

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- ⁴Dobbs, Matthew B., et al. "Early Results of a New Method of Treatment for Idiopathic Congenital Vertical Talus." *The Journal of Bone and Joint Surgery*, vol. 88, no. 6, June 2006, pp.1192-200.