

Case Reports

Pseudo-ataxia due to Osteoid Osteoma

Juanette McKenzie¹, Curtis Oettel-Flaherty², Douglas Noel³, Ruth H. Walker^{4,5} & Andrew K. Sobering^{2*}

¹ Department of Physiology, Neuroscience and Behavioral Sciences, St. George's University, St. George's, Grenada, WI, ² Department of Biochemistry, St. George's University, St. George's, Grenada, WI, ³ Clinical Teaching Unit, St. George's University, St. George's, Grenada, WI, ⁴ Department of Neurology, James J. Peters Veterans Affairs Medical Center, New York, NY, US, ⁵ Department of Neurology, Mount Sinai School of Medicine, New York, NY, US

Abstract

Background: Ataxia is diagnosed by typical features on examination suggestive of a cerebellar etiology and can invoke extensive diagnostic testing. Osteoid osteomas (OOs) are benign bone tumors of the lower limbs that occasionally present with focal neurological signs.

Case Report: A 3-year-old male presented with apparent progressive gait ataxia and non-specific leg pain. Initial imaging was unremarkable. However, 12 months later, a lesion was identified in the distal right femur, which was found to be an OO. The gait disorder and pain resolved after surgery.

Discussion: This case highlights the challenges of diagnosing a gait disorder in young children.

Keywords: Osteoid osteoma, ataxia, gait disturbance, socioeconomic status, Caribbean, West Indies

Citation: McKenzie J, Oettel-Flaherty C, Noel D, Walker RH, Sobering AK. Pseudo-ataxia due to osteoid osteoma. Tremor Other Hyperkinet Mov. 2019; 9. doi: 10.7916/vt1n-ga19

*To whom correspondence should be addressed. E-mail: asobering@sgu.edu

Editor: Elan D. Louis, Yale University, USA

Received: December 14, 2018 **Accepted:** December 19, 2018 **Published:** February 6, 2019

Copyright: © 2019 McKenzie et al. This is an open-access article distributed under the terms of the Creative Commons Attribution–Noncommercial–No Derivatives License, which permits the user to copy, distribute, and transmit the work provided that the original authors and source are credited; that no commercial use is made of the work; and that the work is not altered or transformed.

Funding: None.

Financial Disclosures: None.

Conflict of Interest: The authors report no conflict of interest.

Ethics Statement: This study was performed in accordance with the ethical standards detailed in the Declaration of Helsinki. The authors' institutional ethics committee has approved this study and all patients have provided written informed consent. All patients that appear on video have provided written informed consent; authorization for the videotaping and for publication of the videotape was provided.

Introduction

Ataxia is typically diagnosed by the presence of a wide-based gait, incoordination of limb movements, and typical speech and eye movement abnormalities; investigations are directed at identifying a cerebellar etiology. Other causes of a wide-based gait, such as proprioceptive sensory deficit from peripheral neuropathy, psychogenic etiology (astasia abasia), or cervical myelopathy, can typically be diagnosed by the presence of other features upon neurologic evaluation. Ataxia is not uncommon in pediatric neurology and the differential diagnosis is very broad.¹ Neurologic disorders are often challenging to diagnose, especially in the pediatric population because of the limited ability of children to identify and verbally articulate specific symptoms. Diagnostic challenges are amplified in settings of limited resources because extensive imaging studies are often required.

Osteoid osteoma (OO) accounts for approximately 10% of benign skeletal neoplasms^{2,3} and usually occurs in boys in the long bones of the

lower limb such as the femur,⁴ but may manifest at other locations.⁵ Diagnosis is based upon imaging. OO can present with a variety of neurologic symptoms, including muscle weakness and atrophy, diminished tendon reflexes, or gait disturbance,^{6,7} and it is recognized that these presentations are often associated with a delayed diagnosis.^{8,9} Surgery is usually curative, and patients are expected to make a full recovery after excision of the tumor;^{10–13} however, a high degree of clinical suspicion is needed to ensure early diagnosis and treatment.^{6,14}

Case report

A 3-year-old male with no significant medical history or trauma was referred to our pro bono movement disorders clinic by an orthopedic surgeon because of a 6-month history of progressive gait problems and pain. As best as he could communicate, the child described pain in his lower body and hips. The pain was sufficient to disrupt his sleep; however, acetaminophen brought relief. Fever often accompanied

the pain. Physical examination showed no tenderness, erythema, or swelling of the lower limbs.

Neurological examination revealed a markedly wide-based gait and inability to perform tandem gait or walk on the toes (Video 1).



Video 1. Gait Disturbance Due to Osteoid Osteoma. Six months after symptom onset, the patient had a significantly wide-based gait and was unable to perform tandem gait or walk on his toes. Our retrospective analysis reveals a subtle gait asymmetry favoring the right leg that is evident while running; however, no other signs or symptoms localizing to the right leg are evident. Two months following surgery, the patient is walking normally.

Deep tendon reflexes were normal, and no abnormality of muscle tone, strength, or bulk was observed. Neurological examination was otherwise normal, and no other cerebellar signs involving speech, eye movements, or upper limb coordination were evident. Radiology of the hips and lumbar spine were not remarkable (not shown). X-ray of the right femur was also normal (Figure 1A). Brain and cervical spinal magnetic resonance imaging (MRI) were normal. He tested negative for sickle cell disease, human T-cell lymphotropic virus, and rheumatoid arthritis serology. The alpha-fetoprotein level was normal, and common laboratory investigations were not informative.

Six months later, the lower body pain, sleep disruption, fever, and ataxia worsened significantly. A subsequent set of X-rays showed a circumscribed opacity on the distal end of the right femur (Figure 1B). The patient and his mother travelled by air to a neighboring island for surgical management and follow-up care. Exploratory surgery under fluoroscopic guidance revealed a mass, which was excised. Histopathology of the excised mass showed features consistent with OO. The patient's pain diminished after surgery and subsequently disappeared after 2 weeks. Postoperative radiology shows the bone scar (Figure 1C). The ability to walk normally was documented 2 months post surgery (Video 1).

Written informed consent was obtained from the parents of the patient to publish his clinical history and gait video. Because of the small population size of the nation where the patient resides, our Institutional Review Board has requested that we do not reveal the precise location of this patient and his family to protect their privacy.

Discussion

This child underwent an extensive, and expensive, neurologic evaluation of his gait disorder following referral from an orthopedic surgeon; however, no neurologic basis for the gait disturbance was found. The term pseudo-ataxia has been used to describe ataxia as

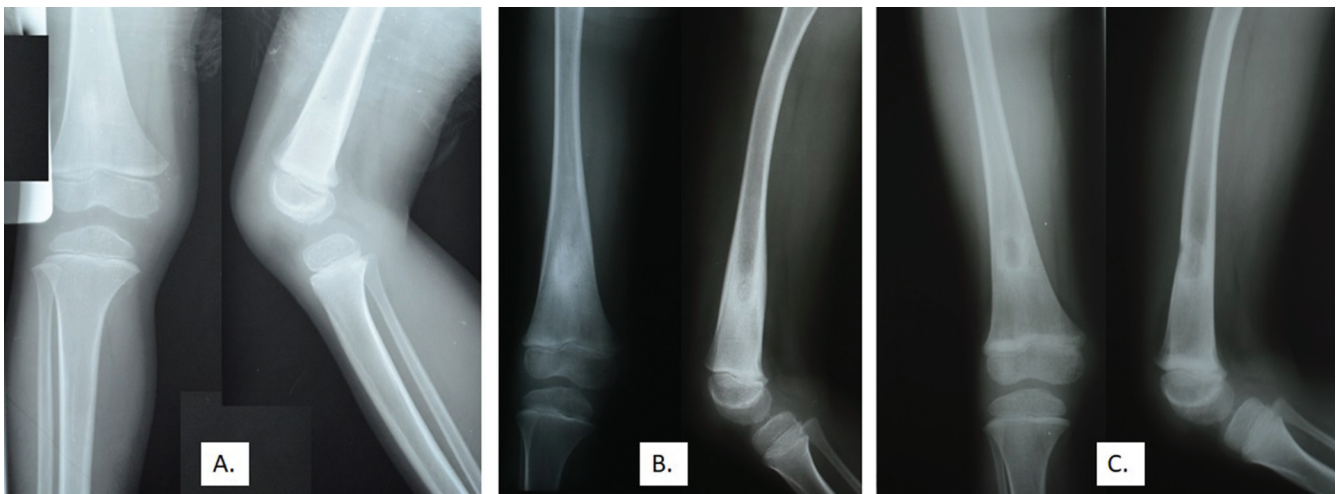


Figure 1. X-rays Showing Osteoid Osteoma of Right Femur (Anterior and Lateral Views). (A) at presentation 6 months after symptom onset, (B) 1 year after symptom onset, and (C) 2 weeks post surgery.

a manifestation of an epileptic seizure,^{15,16} i.e. in the absence of cerebellar disease. Here, we interpret the manifestation of the gait disorder in our patient as pseudo-ataxia caused by an OO and lower limb pain, which was not otherwise apparent on initial clinical examination. On retrospective review of the videos, a subtle gait asymmetry, favoring the right leg, is evident while running and performing tandem gait. However, no other signs or symptoms, specifically no focal tenderness, localizing to the right lower limb were identified. Cerebellar dysfunction resulting in gait ataxia typically also causes other cerebellar signs, such as nystagmus, dysarthria, and limb ataxia; however, involvement of primarily the cerebellar vermis, as can be seen in alcoholic cerebellar degeneration, may only affect gait. Despite the absence of other localizing signs, and normal eye movements, speech, and upper limb coordination, the urgency of identifying a cause such as a posterior fossa tumor merited neuroimaging. The cost of the brain and spine MRIs was covered by a local charitable organization as part of a charitable outreach program.

To our knowledge, OO presenting with pseudo-ataxia has not been previously reported. The reports of OO with accompanying neurological signs have focused on spinal localization of the tumor.¹⁷ Gait disturbance due to OO has been described in a case report,¹⁸ and limping is known to be caused by this type of tumor.¹⁹ Pain is thought to be due to the abundance of peripheral nerve endings and prostaglandins at the tumor site.^{14,20,21}

A delay in diagnosis is not unusual when OOs present with neurological manifestations.⁸ Instances where there is a suggestive clinical history but no changes in the plain radiographs occurs in a small percentage of cases.² Computed tomography (CT) scanning offers the best chance for identification of the nidus;^{22,23} however, given the low index of suspicion for a bone lesion and the high cost of CT, further bone imaging was not considered.

Diagnosis of OO is challenging, especially in the setting of an economically disadvantaged area in which patients often have to pay out of pocket for diagnostic examinations such as CT and MRI.²⁴ Other barriers faced in resource-limited communities stem from limited access to radiological services.²⁵ Although there have been improvements since 1990, the Caribbean country where this patient resides is below the fifth decile in terms of healthcare access according to a quality index study performed in 2017.²⁶

This case highlights the challenges of diagnosing gait disorders in children, in addition to the insidious presentation of OO. Additionally, we note the added challenges of diagnosing and managing rare diseases in communities of low socioeconomic status.^{27–29} It is clear that early diagnosis and timely treatment require a high index of suspicion, and an awareness of the possible presentation with neurological signs in the absence of early radiological findings on a plain radiograph.³⁰

Acknowledgments

We thank the patient and family for participation in this study. We also thank the Urban Humanitarian Projects of the Iota Epsilon Alpha

medical honor society of St. George's University for donations to pay for brain and cervical spine MRI.

References

1. Pavone P, Praticò AD, Pavone V, Lubrano R, Falsaperla R, Rizzo R, et al. Ataxia in children: early recognition and clinical evaluation. *Ital J Pediatr* 2017;43:1–9. doi: 10.1186/s13052-016-0325-9
2. Hashemi J, Gharahdaghi M, Ansari-pour E, Jedi F, Hashemi S. Radiological features of osteoid osteoma: pictorial review. *Iran J Radiol* 2011;8: 182–189. doi: 10.5812/kmp.iranjradiol.17351065.3392
3. Hakim DN, Pelly T, Kulendran M, Caris JA. Benign tumours of the bone: a review. *J Bone Oncol* 2015;4:37–41. doi: 10.1016/j.jbo.2015.02.001
4. Jaffe HL. "Osteoid-osteoma" a benign osteoblastic tumor composed of osteoid and atypical bone. *Arch Surg* 1935;31:709–728. doi: 10.1001/archsurg.1935.01180170034003
5. Daniele S, Merendi G, Rocchi L. Osteoid osteoma of the distal fibula: a case report of a very rare location treatment. *J Bone Res* 2017;5:1–4. doi: 10.4172/2572-4916.1000181
6. Hsich GE, Davis RG, Darras BT. Osteoid osteoma presenting with focal neurologic signs. *Pediatr Neurol* 2002;26:148–152. doi: 10.1016/S0887-8994(01)00359-9
7. Gille P, Nanchin P, Aubert D, de la Salle R, Giordan H, Bidet R, et al. Intraoperative radioactive localization of osteoid osteoma, four case reports. *J Pediatr Orthop* 1986;6:596–599. doi: 10.1097/01241398-198609000-00013
8. Kiers L, Shield LK, Cole WG. Neurological manifestations of osteoid osteoma. *Arch Dis Child* 1990;65:851–855. doi: 10.1136/adc.65.8.851
9. Lacorte D, Bonifacci V, Bertelli S, Gallo P, Sprocati M. Late diagnosis of osteoid osteoma in adolescence: a case report. *Acta Med Mediterr* 2012;28:29–31.
10. Moberg E. Further observations on "corticalisosteoid" or "osteoid osteoma." *Acta Radiol* 1952;38:279–293. doi: 10.3109/00016925209177018
11. Pfaff J, April MD. Osteoid osteoma. *J Emerg Med* 2016;50:780–781. doi: 10.1016/j.jemermed.2016.01.005
12. Yoshioka K, Matsuda E, Murakami H, Tsuchiya H. Microendoscopic excision of osteoid osteoma in the pedicle of the third lumbar vertebra. *Asian Spine J* 2015;9:958–961. doi: 10.4184/asj.2015.9.6.958
13. Swee RG, McLeod RA, Beabout JW. Osteoid osteoma. *Radiology* 1979; 130:117–123. doi: 10.1148/130.1.117
14. Motamed K, Seeger LL. Benign bone tumors. *Radiol Clin North Am* 2014; 49:1115–1134. doi: 10.1016/j.rcl.2011.07.002
15. Javadzadeh M, Hassanvand-Amouzadeh M, Esmail-Nejad SS, Abasi E, Alipour A, Mollamohammadi M. Ataxia in childhood: epidemiological, clinical and neuroradiologic features, and the risk of recurrence. *Iran J Child Neurol* 2017; 11:1–6.
16. Winchester S, Singh PK, Mikati MA. Ataxia. In: Dulac O, Lassonde M, Sarnat HB, editors. *Handbook of clinical neurology*. Amsterdam: Elsevier B.V.; 2013. p 1213–1217. doi: 10.1016/B978-0-444-52910-7.00043-X
17. Rivinus TM, Jamison DL, Graham PJ. Childhood organic neurological disease presenting as psychiatric disorder. *Arch Dis Child* 1975;50:115–119. doi: 10.1136/adc.50.2.115
18. Cabasson S, Yvert M, Pillet P, Pédespan JM. Présentation neurologique d'un ostéome ostéοide du col fémoral. *Arch Pediatr* 2012;19:1196–1199. doi: 10.1016/j.arcped.2012.08.022

19. Santili C, Júnior WL, Goiano E de O, Lins RAB, Waisberg G, Braga S dos R, et al. Limping in Children. *Rev Bras Ortop* 2015;44:290–298. doi: 10.1590/S0102-36162009000400002
20. Esquerdo J, Fernandez CF, Gomar F. Pain in osteoid osteoma: histological facts. *Acta Orthop* 1976;47:520–524. doi: 10.3109/17453677608988731
21. Rosenthal DI, Springfield DS, Gebhardt MC, Rosenberg AE, Mankin HJ. Osteoid osteoma: percutaneous radio-frequency ablation. *Radiology* 1995; 197:451–454. doi: 10.1148/radiology.197.2.7480692
22. Chai JW, Hong SH, Choi J-Y, Koh YH, Lee JW, Choi J-A, et al. Radiologic diagnosis of osteoid osteoma: from simple to challenging findings. *RadioGraphics* 2010;30:737–749. doi: 10.1148/rg.303095120
23. Errani C, Kreshak J, Ruggieri P, Alberghini M, Picci P, Vanel D. Imaging of bone tumors for the musculoskeletal oncologic surgeon. *Eur J Radiol* 2013;82:2083–2091. doi: 10.1016/j.ejrad.2011.11.034
24. Ramdass MJ, Mooteeram J, Beharry A, Mencia M, Barrow S. An 8-YEAR analysis of bone tumours in a Caribbean island. *Ann Med Surg* 2015;4: 414–416. doi: 10.1016/j.amsu.2015.10.009
25. Mollura DJ, Azene EM, Starikovskiy A, Thelwell A, Iosifescu S, Kimble C, et al. White paper report of the rad-aid conference on international radiology for developing countries: identifying challenges, opportunities, and strategies for imaging services in the developing world. *J Am Coll Radiol* 2010;7: 495–500. doi: 10.1016/j.jacr.2010.01.018
26. Barber RM, Fullman N, Sorensen RJD, Bollyky T, McKee M, Nolte E, et al. Healthcare access and quality index based on mortality from causes amenable to personal health care in 195 countries and territories, 1990–2015: a novel analysis from the Global Burden of Disease Study 2015. *Lancet* 2017; 390:231–266. doi: 10.1016/S0140-6736(17)30818-8
27. Demeter S, Reed M, Lix L, Macwilliam L, Leslie WD. Socioeconomic status and the utilization of diagnostic imaging in an urban setting. *Can Med Assoc J* 2005;173:1173–1177. doi: 10.1503/cmaj.050609
28. Stinson J, Connelly M, Kamper SJ, Herlin T, April KT. Models of Care for addressing chronic musculoskeletal pain and health in children and adolescents. *Best Pract Res Clin Rheumatol* 2016;30:468–482. doi: 10.1016/j.berh. 2016.08.005
29. Schieppati A, Henter J-I, Daina E, Aperia A. Why rare diseases are an important medical and social issue. *Lancet* 2008;371:2039–2041. doi: 10.1016/S0140-6736(08)60872-7
30. Ghosh PS, Mitra S, Moodley M. Pearls & oysters: osteoid osteoma of the scapula masquerading as neuralgic amyotrophy. *Neurology* 2012;79:7–9. doi: 10.1212/WNL.0b013e31825f0534