Skeletal Radiology (2022) 51:1899–1900
https://doi.org/10.1007/s00256-022-03998-7

TEST YOURSELF: ANSWER

Test Yourself Answer: A 38-year-old male presenting with a 1-year history of medial right knee pain

Susan Hesni[1] · Daniel Lindsay[2] · Asif Saifuddin[1]

Received: 22 December 2021 / Revised: 22 December 2021 / Accepted: 17 January 2022 / Published online: 20 January 2022

Answer

Intraosseous schwannoma of the right femur.

Discussion

MRI (Fig. 1) demonstrated a lobular intermediate T1W/PDW and hyperintense fat-suppressed PDW FSE lesion with no reactive change, and uniform enhancement following contrast. Minor extraosseous extension was noted. CT (Fig. 2) demonstrated a lytic lesion with a thin sclerotic margin. CT-guided biopsy was performed with histology revealing a tumour composed of spindle cells with possible vague nuclear palisading embedded in a collagenous stroma. Immunohistochemistry revealed the tumour was positive for S100 (Fig. 3).

Schwannomas are benign nerve sheath tumours originating from Schwann cells that typically arise in soft tissues. Intraosseous location is extremely rare, accounting for 0.1–0.2% of bone tumours [1, 2]. Three mechanisms for intraosseous location have been suggested: primary intraosseous development, development within a nutrient canal, and extraosseous schwannoma eroding into bone [3]. When developing within bone, the origin of these tumours is believed to relate to intraosseous vasomotor nerve fibres that accompany nutrient arteries [2, 4]. The mandible is the commonest site followed by the sacrum, although involvement of other bones, including the spine and extremities, has been reported [1, 2, 5]. When occurring in long bones, the commonest location is in the diametaphysis [6].

The condition typically occurs in adults in the 4th–6th decades of life [1] and there is a slight female predominance [2]. Although usually sporadic, cases may arise on a background of schwannomatosis [2, 4]. Intraosseous schwannomas are slow growing and may be asymptomatic in a quarter of patients. When symptomatic, patients may present with swelling, pain, or pathological fracture [1, 2, 5, 7, 8], while malignant transformation has not been reported [4, 9].

Histologically, intraosseous schwannomas are indistinguishable from their soft tissue counterparts, characterised by bland spindle cells with elongated nuclei embedded in a collagenous stroma. Alternating areas of hyper- and hypocellularity, known as Antoni A and Antoni B areas respectively, may be seen [1, 9]. Foci of nuclear palisading (Verocay bodies) are also characteristic, but are not required for the diagnosis [1]. Strong immunoreactivity for S100 can confirm the morphological diagnosis, as seen in this case [7, 9].

Radiographs and CT typically show a well-defined lytic lesion, with or without a sclerotic rim. There may be adjacent cortical thinning or destruction, although extraosseous extension is generally not seen [1, 2, 4, 7]. The lesions do not usually demonstrate any intralesional mineralisation [1, 8]. On MRI, schwannomas return low-to-intermediate signal on T1W images and heterogeneous signal on T2W images with heterogeneous or diffuse contrast enhancement [1, 5, 8, 9]. If performed, FDG-PET may reveal increased uptake [10]. Imaging features are non-specific, and therefore, histological review is required for definitive diagnosis.

The main differential diagnosis includes other benign bone lesions such as enchondroma, giant cell tumour, fibrous dysplasia, and non-ossifying fibroma as well as primary and secondary malignant bone lesions, particularly when seen in non-typical sites [4, 9, 10]. Treatment of intraosseous schwannoma is usually curettage and bone grafting or surgical resection, with a low risk of local recurrence [4, 5, 9].

The case presentation can be found at https://doi.org/10.1007/s00256-022-03997-8.

✉ Susan Hesni
susan.hesni@nhs.net

[1] Department of Radiology, Royal National Orthopaedic Hospital (RNOH), Brockley Hill, Stanmore HA7 4LP, UK

[2] Department of Histopathology, Royal National Orthopaedic Hospital (RNOH), Brockley Hill, Stanmore HA7 4LP, UK

In conclusion, the diagnosis of intraosseous schwannoma cannot be made on imaging alone and histological review is required to exclude other neoplastic lesions. CT-guided biopsy provides an effective and relatively non-invasive way of obtaining tissue, and can allow for appropriate operative planning [5].

Author contributions Dr Susan Hesni – responsible for the main body of text, corresponding author

Dr Daniel Lindsay – provided histological slides and information regarding the histological features

Dr Asif Saifuddin – reviewed the paper and provided the radiological images and annotations

Declarations

Conflict of interest The authors declare no competing interests.

References

1. Drumond GC, Nakagawa SA, Costa FD, de Souza MYT, Comunello J, Chung WT. Intraosseous schwannoma: case report and review of the literature. Rev Bras Ortop [Internet]. 202055(2):258–62. Available from: http://www.ncbi.nlm.nih.gov/pubmed/32346205. Accessed 9 Dec 2021
2. Kashima TG, Gibbons MRJP, Whitwell D, Gibbons CLMH, Bradley KM, Ostlere SJ, et al. Intraosseous schwannoma in schwannomatosis. Skeletal Radiol [Internet]. 2013 31;42(12):1665–71. Available from: http://link.springer.com/10.1007/s00256-013-1712-6. Accessed 9 Dec 2021
3. McAleese T, Clesham K, Moloney D, Hughes A, Faheem N, Merghani K. Intraosseous schwannoma of the femur in a patient with monoclonal gammopathy of undetermined significance. Int J Surg Case Rep [Internet]. 2020;72:494–8. Available from: http://www.ncbi.nlm.nih.gov/pubmed/32698273. Accessed 9 Dec 2021
4. Ida CM, Scheithauer BW, Yapicier Ö, Carney JA, Wenger DE, Inwards CY, et al. Primary schwannoma of the bone. Am J Surg Pathol [Internet]. 201135(7):989–97. Available from: https://journals.lww.com/00000478-201107000-00006. Accessed 9 Dec 2021
5. Wang YQ, Hu JX, Yang SM, Jiang L, Liu XG, Yuan HS, et al. Intraosseous schwannoma of the mobile spine: a report of twenty cases. Eur Spine J [Internet]. 2018 18;27(12):3092–104. Available from: http://link.springer.com/10.1007/s00586-018-5766-y. Accessed 9 Dec 2021
6. Kato H, Kanematsu M, Ohno T, Oshima K, Nagano A, Hatano Y, et al. Intraosseous schwannoma of the ilium. Clin Imaging [Internet]. 201539(1):161–4. Available from: https://linkinghub.elsevier.com/retrieve/pii/S0899707114002320. Accessed 9 Dec 2021
7. Huajun J, Wei Q, Yuxuan W, Jingjing Y. Intraosseous schwannoma of the proximal humerus with pathologic fracture. Eur J Med Res [Internet]. 2021 9;26(1):72. Available from: https://eurjmedres.biomedcentral.com/articles/10.1186/s40001-021-00541-7. Accessed 9 Dec 2021
8. Wang G, Wen X, Qu L, Qi X, Yang C. Intraosseous schwannoma involving multiple bones of the foot: a case report. J Foot Ankle Surg [Internet]. 201655(1):201–6. Available from: https://linkinghub.elsevier.com/retrieve/pii/S106725161500304X. Accessed 9 Dec 2021
9. Reyniers P, Wafa H, Sinnaeve F, Debeer P, Sciot R. Intraosseous schwannoma of the glenoid: case report and literature review. SICOT-J [Internet]. 2021;7:2. Available from: http://www.ncbi.nlm.nih.gov/pubmed/33416488. Accessed 9 Dec 2021
10. Igai H, Kamiyoshihara M, Kawatani N, Ibe T, Shimizu K. Sternal intraosseous schwannoma mimicking breast cancer metastasis. J Cardiothorac Surg [Internet]. 2014 Jun 27;9:116. Available from: http://www.ncbi.nlm.nih.gov/pubmed/24969640. Accessed 9 Dec 2021