Case 15643

Eurorad

Primary intraosseous psammomatous melanotic schwannoma of the sacrum

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Section: Musculoskeletal system
Area of Interest: Musculoskeletal bone Education
Procedure: Education
Imaging Technique: CT
Imaging Technique: MR
Special Focus: Neoplasia Case Type: Clinical Cases
Authors: Gavin A. McKenzieBenjamin M. HoweCaterina GianniniPeter S. RoseDoris E. Wenger
Patient: 60 years, female

Clinical History:

60-year-old man who presented with acute necrotising gallstone pancreatitis. During his hospitalisation, he underwent unenhanced CT examination of the abdomen and pelvis that revealed an indeterminate heterogeneous, predominantly osteolytic lesion involving the mid to upper sacrum that contained foci of intralesional matrix and a significant component of intralesional fat.

Imaging Findings:

The epicenter of the lesion was on the midline involving predominantly the 1st 3 sacral segments and it insinuated between the adjacent neural foramina. It extended laterally to, but did not involve or traverse the sacroiliac joints. There was a small focus of cortical irregularity along the anterior aspect of the sacrum at the level of S2 where there was a tiny associated soft tissue mass. MRI demonstrated a markedly heterogeneous destructive mass that did not invade or originate from the S1-S3 nerve roots, but rather insinuated in between, suggesting intra-osseous rather than neural origin. The mass demonstrated complex heterogeneous signal with areas of hypointense and hyperintense signal on T1 and T2-weighted imaging as well as a significant component of internal fat.

Discussion:

Melanotic schwannoma (MS) is a rare pigmented tumour considered an intermediate between melanoma and schwannoma [1]. Less than 200 cases of MS and 8 cases of primary intra-osseous have been reported [1- 6]. The two described MS subtypes are non-psammomatous and psammomatous, with the latter more often associated with Carney complex, a familial autosomal dominant multiple endocrine neoplasia associated with psammomatous melanotic schwannoma (PMS) approximately 50% of the time, and lends to a poorer clinical prognosis secondary to its pathologic associations [7]. Immunohistochemical stains are the mainstay for MS diagnosis, with PMS characteristically demonstrating psammoma bodies [1].

MRI demonstrates variable degrees of enhancement and heterogeneous T1/T2 signal, with T1 hyperintensity and

T2 hypointensity being ascribed to haemorrhage and/or melanin. MS and PMS usually demonstrate indolent imaging features on radiographs and CT with well-circumscribed margins and narrow zone of transition, but can be locally aggressive with malignant features. CT typically demonstrates a hyperdense mass with calcifications, however, calcifications may be absent and were originally described as being present in only 10% [1, 8, 9-15]. Primary intra-osseous MS and PMS are usually lytic lesions involving the axial skeleton [3-5].

The imaging features of MS and PMS are nonspecific with the differential diagnosis including primary sarcoma, malignant melanoma, nerve sheath tumors or bone metastases (such as malignant melanoma or haemorrhagic metastases). The presence of calcifications could have been secondary to matrix production (osteosarcoma or chondrosarcoma), or could have been secondary to remnants of residual bone matrix. The component of fat was an interesting feature of the lesion that could have been secondary to intra-lesional fat or a prominent focus of preserved yellow marrow. Regardless, the presence of fat strongly mitigated against a diagnosis of malignancy and raised the possibility of haemangioma, intra-osseous lipoma or teratoma. Treatment options for melanotic schwannoma vary from serial follow ups to surgical excision, with the benefit of adjuvant or post-excisional radiotherapy or chemotherapy unknown [8, 16].

Based on the rarity of MS and PMS with nonspecific imaging features, biopsy is required for the histopathological diagnosis. PMS can be intra-osseous and contain calcifications, melanin and blood products. Since 50% of the time PMS is associated with Carney complex, clinical evaluation for the associated manifestations is recommended. The biological behaviour of PMS and MS remains largely benign, but unpredictable. Given the reports of local recurrence, metastasis, and malignant transformation [2], close clinical and imaging follow-up are recommended.

Differential Diagnosis List: Primary intraosseous psammomatous melanotic schwannoma, Sarcoma, Malignant melanoma, Matrix-producing tumours

Final Diagnosis: Primary intraosseous psammomatous melanotic schwannoma

References:

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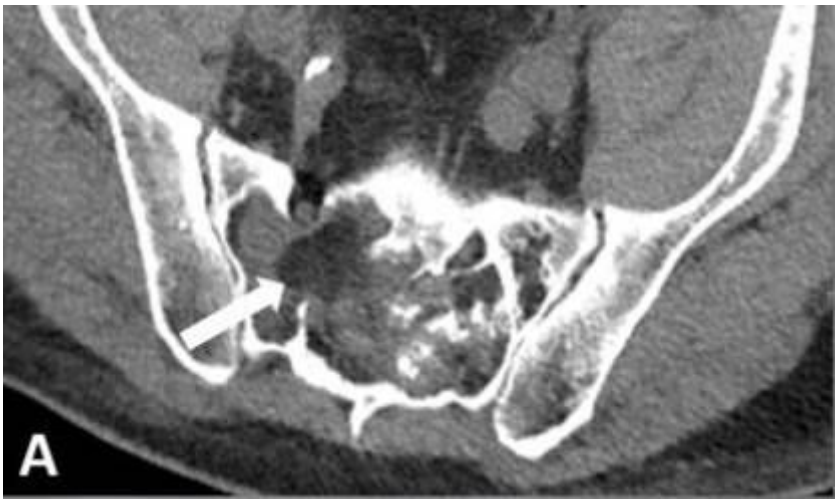
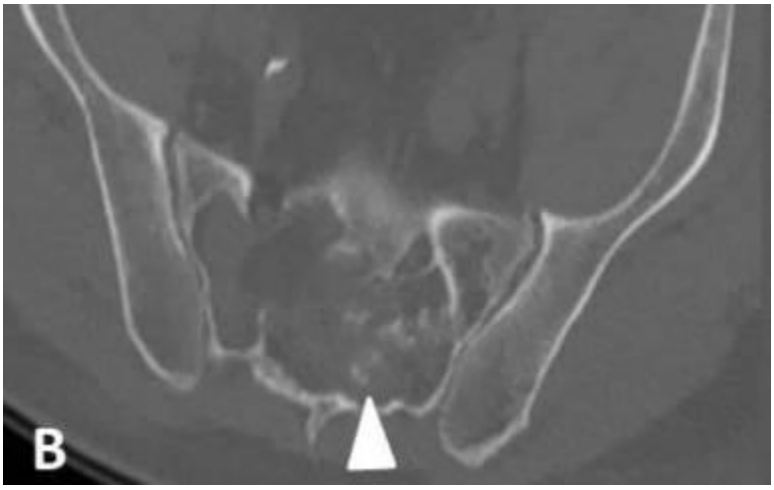
Figure 1

a



Description: Non-contrast CT demonstrates a lytic heterogeneous sacral mass with internal calcifications (arrowhead, B) and macroscopic fat (arrow, A) involving the first three sacral segments with a small focus of soft tissue breakthrough anteriorly (arrow, C). **Origin:** McKenzie G, Department of Musculoskeletal Radiology, Mayo Clinic, Rochester, MN

b



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c

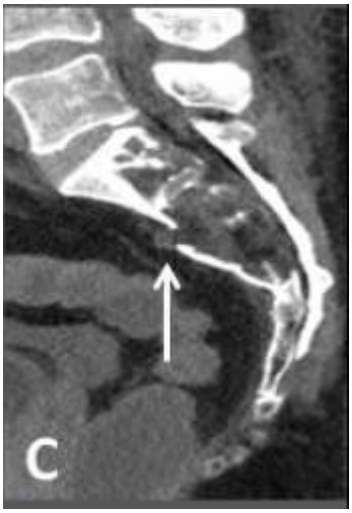

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Figure 2

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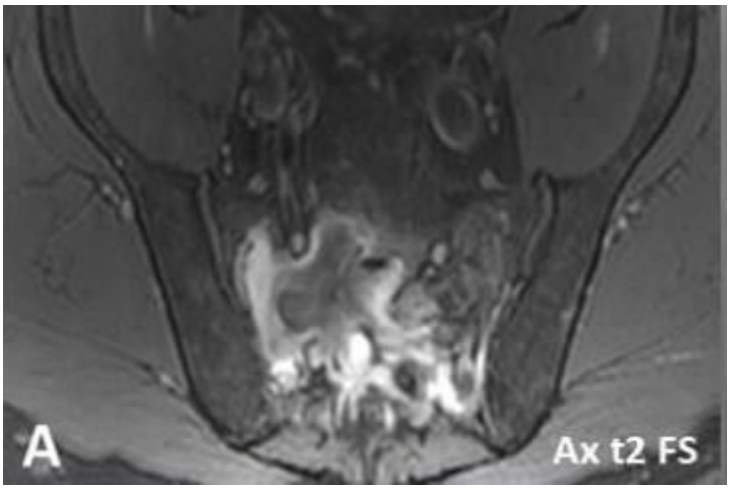
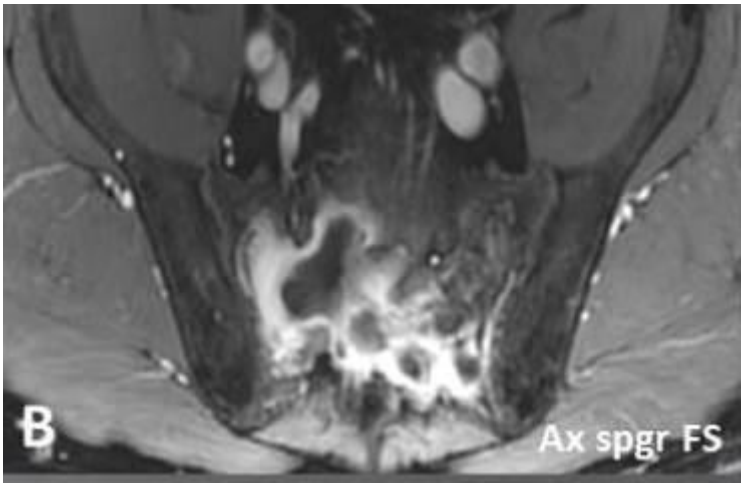


Description: Axial T2-weighted fat saturated (A) and SPGR post-gadolinium (B) sequences demonstrate a T2 heterogeneous peripherally enhancing lesion. Sagittal SPGR post-gadolinium sequence (C) demonstrates the small soft tissue focus with cortical breakthrough. **Origin:** McKenzie G, Department of Musculoskeletal Radiology, Mayo Clinic, Rochester, MN

b



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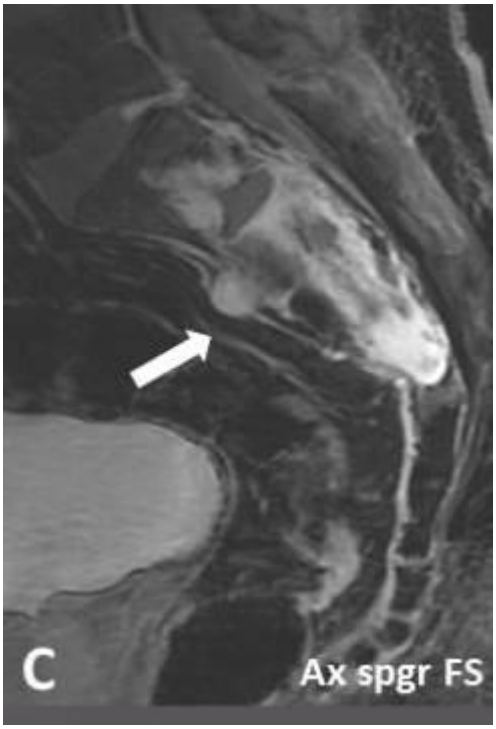

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Figure 3

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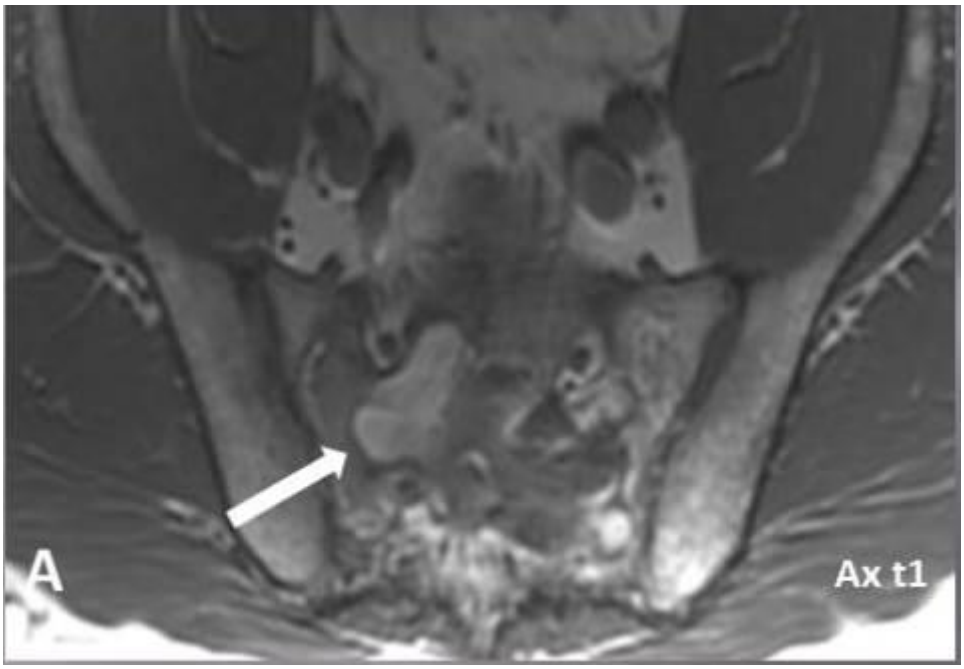
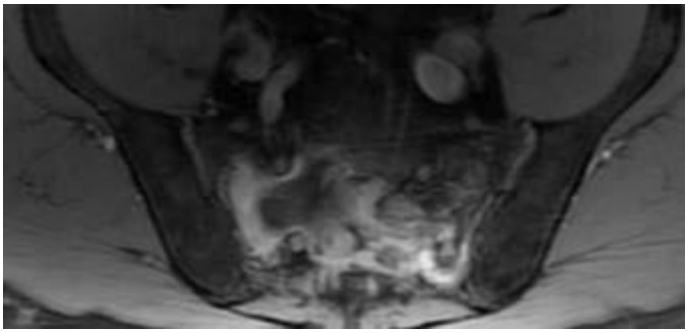


Description: Axial T1 (A) MRI sequence demonstrates inhomogenous T1 hyperintensity with superimposed dominant T1 hyperintense focus (arrows) which can be seen with fat, melanotic, haemorrhagic, proteinaceous or maybe mineralised lesions. **Origin:** McKenzie G, Department of Musculoskeletal Radiology, Mayo Clinic, Rochester, MN

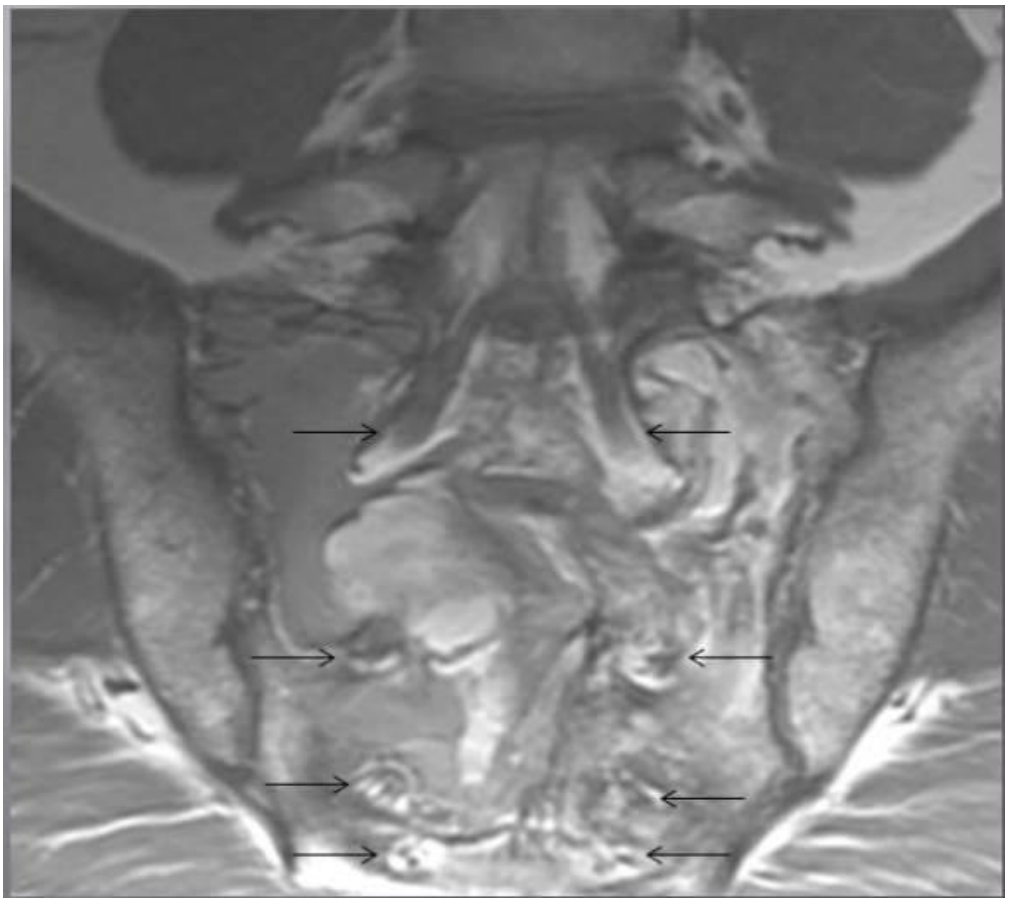
b



Description: Axial T1 fat sat non-contrast MRI sequence demonstrates inhomogeneous T1 hyperintensity which can be seen with melanotic, haemorrhagic, proteinaceous or maybe mineralised lesions. **Origin:** McKenzie G, Department of Musculoskeletal Radiology, Mayo Clinic, Rochester, MN

Figure 4

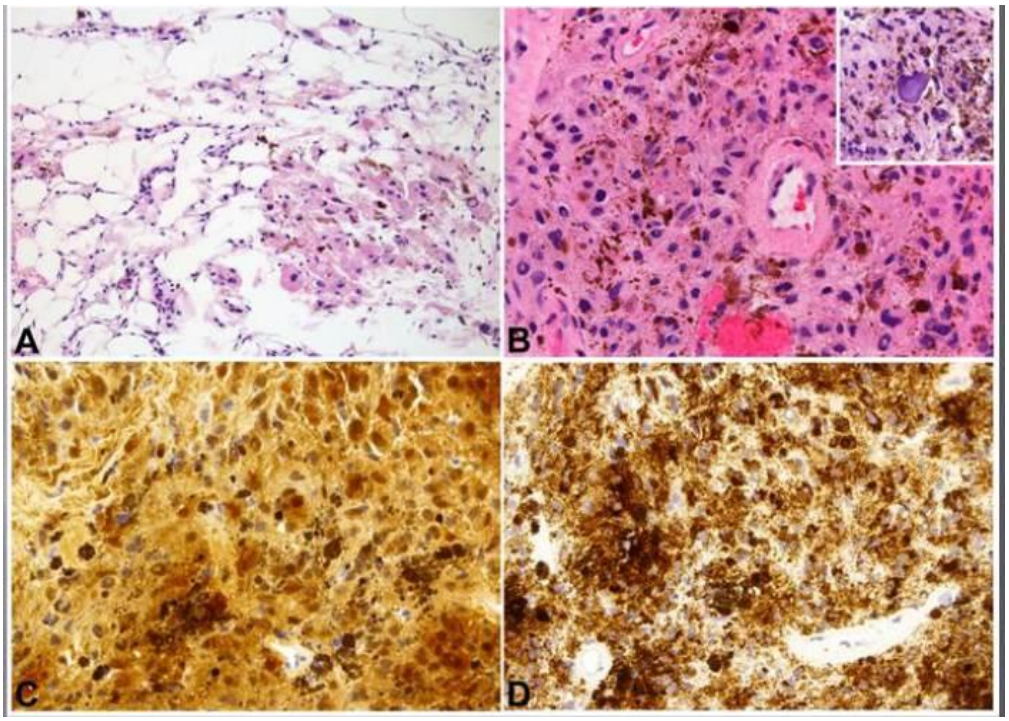
a



Description: Coronal oblique T1-weighted sequence in plane with the neuroforamen demonstrate the nerve-sparing nature of the mass with overall preservation of the S1-S4 nerves (arrows), most consistent with primary intra-osseous origin. **Origin:** McKenzie G, Department of Musculoskeletal Radiology, Mayo Clinic, Rochester, MN

Figure 5

a



Description: H&E sections (A, B) demonstrate tumour cells immersed in adipose tissue (A), and psammomatous calcification (B). Immunohistochemical stains show S100 (schwanninan marker positivity (C)) and HMB45 (immature melanosomes (D)), supporting a psammomatous melanotic schwannoma. **Origin:** McKenzie G, Department of Musculoskeletal Radiology, Mayo Clinic, Rochester, MN