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Images of Interest / Imagens de Interesse

Benign Notochordal Tumor: An Embryological Whisper

Tumor Benigno da Notocorda: Um Sussurro Embriológico

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Abstract

We present the case of a 31-year-old woman with a history of thoracolumbar scoliosis surgically corrected. In the context of lower back pain refractory to conservative treatment, imaging studies were performed, including MRI and CT of the lumbosacral spine and sacroiliac joints. An incidental finding revealed a nodule on the posterior aspect of the S1 vertebral body, with imaging characteristics suggestive of a Benign Notochordal Tumor. No other significant alterations were detected.
Benign notochordal cell tumors are rare lesions derived from Notochordal remnants, often discovered incidentally during imaging. These tumors are most frequently located in the clivus and sacrum, where they exhibit characteristic imaging findings, including mild sclerosis, preservation of trabecular bone, and the absence of aggressive features such as cortical destruction or soft tissue involvement. While Benign notochordal cell tumors are benign and typically indolent, their differentiation from chordomas, which are malignant and aggressive, is essential to prevent overtreatment and ensure proper management.

Keywords

Benign notochordal tumor; Axial skeleton lesions; Notochord remnants.

Resumo

Apresentamos o caso de uma mulher de 31 anos com antecedentes de escoliose toracolombar corrigida cirurgicamente. No contexto de lombalgia refractária ao tratamento conservador, foi realizado um estudo imagiológico com ressonância magnética e tomografia computorizada da coluna lombo-sagrada e articulações sacroilíacas. Como achado incidental, identificou-se um nódulo na vertente posterior do corpo vertebral de S1, com características imagiológicas sugestivas de tumor benigno da notocorda. Não foram detectadas outras alterações significativas.
Os Tumores Benignos da Notocorda são lesões raras derivadas de remanescentes da Notocorda, frequentemente descobertas incidentalmente durante estudos de imagem. Estes tumores localizam-se mais frequentemente no clivus e no sacro, onde apresentam características imagiológicas típicas, incluindo esclerose leve, preservação do padrão trabecular ósseo e ausência de características agressivas, como destruição cortical ou envolvimento dos tecidos moles. Embora os Tumores Benignos da Notocorda sejam benignos e, normalmente, indolentes, a sua diferenciação dos cordomas, que são malignos e agressivos, é essencial para evitar tratamentos excessivos e garantir uma correcta orientação.

Palavras-chave

Tumor benigno da notocorda; Lesões do esqueleto axial; Remanescentes da notocorda.

Case Description

We report the case of a 31-year-old female patient with a history of thoracolumbar scoliosis surgically corrected via thoracolumbar arthrodesis. Over the preceding six months, she experienced occasional low back pain refractory to conservative management with physiotherapy, prompting imaging evaluation of the lumbosacral spine and sacroiliac joints.

MRI (Fig. 1) revealed an incidental nodule on the posterior aspect of the S1 vertebral body. The lesion was well-defined, regular, and measured 30 mm, with heterogeneous T2 hyperintensity and T1 hypointensity. No evidence of intralesional fat, surrounding edema, diffusion restriction, or post-contrast enhancement were observed.
Subsequent CT of the lumbosacral spine (Fig. 2) demonstrated a mild osteosclerotic lesion with poorly defined margins with preservation of the trabecular bone pattern. No radiological

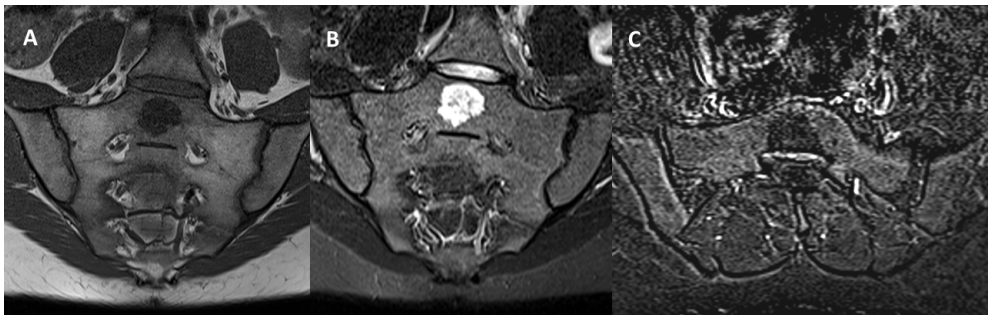


Figure 1 – Coronal oblique T1-weighted (A), STIR (B) and axial oblique T1 fat saturated post-contrast subtraction (C) MRI images demonstrate a lesion in the midline of the S1 vertebral body with heterogeneous T2 hyperintensity and T1 hypointensity. No evidence of post-contrast enhancement or surrounding edema was observed.

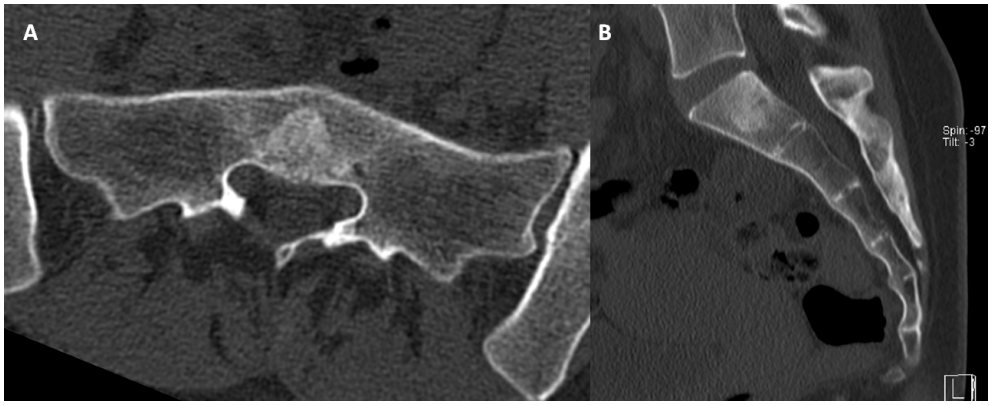


Figure 2 – Axial (A) and Sagittal (B) CT images of the lumbosacral spine show a mild osteosclerotic lesion in the midline of the S1 vertebral body, with poorly defined margins and preservation of the trabecular bone pattern. No evidence of bone destruction or soft tissue involvement is observed.

signs of aggressiveness, such as bone destruction or soft tissue involvement were observed.

The sacroiliac joints appeared normal on both MRI and CT, with no additional findings of inflammatory or degenerative changes or complications related to the surgical instrumentation observed.

Given the imaging characteristics, the lesion was categorized as Bone-RADS 3 (indeterminate lesion, requiring imaging follow-up). A decision was made to opt for imaging surveillance rather than an anatomopathological study, considering the absence of aggressive features and the need to assess its stability over time.

Discussion

Benign notochordal cell tumors (BNCTs) are rare lesions derived from remnants of the notochord and are typically discovered incidentally. Although their precise prevalence is unclear, autopsy studies suggest they may occur in up to 20% of the population, highlighting their relatively common presence as incidental findings in asymptomatic individuals.[1,2] They are distinct from chordomas, with which they share embryological origins, but differ in clinical behavior and prognosis[3,4] BNCTs are most commonly found in the clivus and sacrum, presenting as small, well-defined intraosseous lesions3. Radiographically, they exhibit mild sclerosis without cortical destruction or soft tissue extension.[1,4] MRI findings include low T1 and high T2 signal intensities, with little or no enhancement post-contrast.[2,4] In contrast, chordomas are aggressive, osteolytic, and often associated with significant soft tissue masses.(Table 1)[3,4]

Table 1 – Characteristics of Benign Notochordal Cell Tumors

Characteristic	Description
Location	Typically found in the clivus, sacrum, and vertebral bodies (especially in the midline)[3]
Size	Small[3]
Margins	Well-defined intraosseous lesions[3]
X-ray / CT Findings	Often demonstrates ill-defined mild sclerosis without cortical destruction. Trabecular pattern is usually preserved[2]
MRI Signal	Low T1 and high T2 signal intensities; may be heterogeneous[2,4]
Post-contrast Enhancement	Little to absent enhancement[2,4]
Soft tissue extension	Absent; confined within the bone[2]
Bone destruction	Absent; no cortical destruction or aggressive osteolysis[2]
Intralesional Fat	Typically absent, although some cases may show small foci[4]

BNCTs are considered benign lesions that do not require surgical resection.[4] However, sporadic cases associated with chordoma have been reported in the literature, and it remains controversial whether these are concomitant lesions or if BNCT may occasionally act as a precursor to chordoma.[3,4] That being said, most sources recommend imaging follow-up to confirm their radiological stability and rule out malignant progression, although they do not specify the optimal follow-up interval.[3,4,5]

Ethical Disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

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Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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