CASE REPORT

Lumbar vertebral chordoma arising from an intraosseous benign notochordal cell tumour: radiological findings and histopathological description with a good clinical outcome

^{1,2}T NISHIGUCHI, MD, ¹K MOCHIZUKI, MD, ³T TSUJIO, MD, PhD, ⁴T NISHITA, MD and ¹Y INOUE, MD, PhD

Department of ¹Radiology, ³Orthopedics and, ⁴Radiation Oncology 1-4-3 Asahimachi, Abeno, Osaka, 545-8585 Japan
²T & Co. Medical Imaging 3-4-8-2304, Awaji, Chuo, Osaka, 541-0047 Japan

ABSTRACT. Benign notochordal cell tumours have recently been described as intraosseous benign lesions of notochordal cell origin. The lesions are found in vertebral bodies in 20% of autopsy studies and are a potential precursor of chordoma. We report a rare case of lumbar vertebral chordoma that was thought to arise from a benign intraosseous notochordal cell tumour and which showed significant osteosclerotic change. Radiologically, the lumbar vertebral mass lesion showed hyperintensity on T_2 weighted images, with scanty enhancement on post-contrast T_1 weighted MR images. High uptake corresponding to the mass was noted on fluorine-18-fluorodeoxyglucose positron emission tomography. Bone biopsy revealed proliferation of the physaliphorous cells between thickened bone trabeculae; no nuclear mitosis was observed. Although the mass was diagnosed clinically as spinal chordoma, histopathology contained both benign notochordal cell tumour and conventional chordoma. After heavy particle (^{11}C)-charged radiation therapy was applied to the lesion with a sufficient radiation field margin, the tumour volume significantly decreased and there was improvement in the patient's symptoms. On follow-up radiological studies, the tumour had markedly regressed and there was no tumour regrowth or distant metastasis. In this case report, benign notochordal cell tumour and conventional chordoma are histopathologically identified in the L1 vertebral body, which contains osteosclerotic and osteolytic areas. It is suggested that the benign notochordal cell tumour coexists with a conventional chordoma and that this histopathological finding supports a hypothetical relationship between benign notochordal cell tumour and chordoma.

Received 26 July 2008
Revised 17 March 2009
Accepted 2 April 2009

DOI: 10.1259/bjr/63846600

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A 59-year-old woman presented with urinary incontinence and progressive perineal numbness of six months' duration. She also complained of having recently developed difficulty in walking. There was no history of major medical problems. No remarkable abnormality was observed in the laboratory tests. Physical examination showed paraparesis of the lower extremities and decreased sensation in the perineal region.

A plain radiograph (not shown) displayed faint sclerotic change in the L1 vertebral body; CT depicted osteosclerotic change with some small foci of osteolysis in the L1 vertebral body and posterior marginal erosion of the T12 and the L1 vertebral bodies (Figure 1a). On MRI, the mass exhibited homogeneous low intensity with small foci of high intensity on T_1 weighted images (Figure 1b) and heterogeneous low to high intensity in the intraosseous component and hyperintensity in the extraosseous component on T_2 weighted images

(Figure 1c), compared with the normal vertebral body. The mass extended into the spinal canal posteriorly and perivertebral space anterolaterally (Figure 1d). After intravenous administration of contrast material, the posterior aspects of the mass enhanced slightly (Figure 1e). Fluorine-18-fluorodeoxyglucose (FDG) positron emission tomography (PET) (not shown) revealed moderate FDG uptake (standardised uptake value=3.4) in the L1 vertebra. No abnormal FDG uptake was recognised in other areas. The clinical and radiological manifestations were indicative of conditions ranging from a benign bone disorder to a malignant spinal neoplasm, including Paget's disease, osteoblastic metastasis, Hodgkin's lymphoma, osteosarcoma or a multiple myeloma in polyneuritis-ostitis-endocrinopathy-myeloma-splenomegaly syndrome (POEMS) syndrome. Bone biopsy was performed by a right-sided transpedicle approach with a 13 G needle, and multiple histological samples were taken from the osteosclerotic part of the L1 vertebral body.

The photomicrograph of the specimen from the L1 vertebral body demonstrated different histology. One

Address correspondence to: Tomokazu Nishiguchi, Department of Radiology, 1-4-3, Asahimachi, Abeno, Osaka, 545-8585 Japan.
E-mail: tomokazu-n@med.osaka-cu.ac.jp

Figure 1. (a) A sagittal reformatted CT image (bone algorithm) demonstrates posterior marginal destruction at the T12 and L1 level (arrowheads). (b) Pre-contrast-enhanced T_1 weighted sagittal image (repetition time/echo time (TR/TE), 633/11.8 ms) shows a low-signal-intensity mass with small foci of high intensity (curved arrow). (c) T_2 weighted sagittal image (TR/TE, 4000/97.9 ms) demonstrates that the mass extends posteriorly, with narrowing of the spinal canal, and involves the posterior aspect of the T12 vertebral body (arrowhead). (d) T_2 weighted axial image (TR/TE, 4000/102.6 ms) depicts a significant extension of the mass into the perivertebral space (asterisks) and compression of the conus medullaris (arrow). (e) Post-contrast-enhanced T_1 weighted sagittal image (TR/TE, 633/11.8 ms) displays ill-defined slight enhancement on the posterior aspect of the mass (curved arrows).

showed proliferation of adipocyte-like vacuolated cells between thickened trabecular frameworks without intercellular myxoid matrix, nuclear atypia or intracellular matrix (Figure 2). The other specimen demonstrated proliferation of physaliphorous cells with a prominent intracellular eosinophilic matrix, increased intercellular myxoid matrix and nuclear atypia (Figure 2, inset image). Immunohistochemical stains were positive for vimentin, S-100, cytokeratin and CD-68 (not shown). Although results from haematoxylin and eosin staining were suggestive of metastatic renal cell carcinoma, the results from immunohistochemical stains revealed an intraosseous lesion of notochordal cell origin. The Ki67 index was mostly negative in the sampled specimen. Considering the locally aggressive features of the tumour and results from abdominal CT, which were negative for renal cell carcinoma, a spinal chordoma was the most likely diagnosis. However, histopathologically, the intraosseous tumour was predominantly a benign notochordal cell tumour.

The tumour extended significantly into the perivertebral area anteriorly and the intraspinal canal posteriorly, with compression of the conus medullaris and bilateral nerve roots at the L1 level (Figure 1d). The lesion was probably accessible by the retroperitoneal approach for tumour resection and concurrent spinal reconstruction, followed by adjuvant radiation therapy. In accordance with the patient's wishes, however, heavy particle ^{11}C -charged radiation therapy with a wide radiation field margin was initially applied to the lesion. The total dose of particle-charged radiation therapy was 64 Gy, fractionated in 16 applications and delivered by quadro portal over 3 weeks. On the patient's last visit to our department 24 months after radiation therapy, physical examination showed significant recovery of her motor weakness, and follow-up MRI displayed a significant reduction in tumour volume, especially in the spinal canal. No relapsing tumour in the L1 vertebral body was observed, although mild compression fractures developed in the T11 and L2 vertebrae (Figure 3).

Discussion

This paper describes the rare association between a benign notochordal cell tumour and the concurrent development of conventional chordoma. The tumour, histologically, was a benign notochordal cell tumour, with areas resembling conventional chordoma. Only the intraosseous component was biopsied, and, therefore, it was assumed that the extraosseous lytic component was a malignant chordoma owing to its aggressive features. The radiological findings in our case show mixed features of osteosclerotic and osteolytic change on CT scans. Significant osteosclerotic change in the anterior part of the L1 vertebral body, which suggests a slow-growing process on imaging, seemed to reflect the thickened bony trabeculae on histopathological analysis. On MRI, no contrast enhancement was observed in the anterior part of the lesion, and the finding histopathologically corresponded to a paucity of blood vessels. These imaging findings are atypical for conventional chordoma and are consistent with those of benign notochordal cell tumour [1]. In contrast, the appearance

Figure 2. Bone biopsy specimen of the L1 spine reveals proliferation of physaliphorous cells (asterisks) without abundant intercellular myxoid matrix and no mitotic figures between the thickened trabecular frameworks (arrows). In contrast, the inset image demonstrates atypical physaliphorous cells with nuclear atypia and increased intracellular matrix. (Haematoxylin and eosin stain; low-power field with original magnification of $\times 40$, and a smaller inset image of high-power field and original magnification of $\times 400$.)

of the posterior aspect of the L1 vertebra suggested conventional chordoma because it extended out of the vertebra into the extradural space of the bony spinal canal, was slightly enhanced and showed high uptake of FDG on PET imaging [2].

A benign notochordal cell tumour is a recently described intraosseous benign lesion of notochordal cell origin. Histologically, it is unencapsulated and is composed of adipocyte-like polymorphic vacuolated cells with mildly hyperchromatic nuclei. No mitotic figures, myxoid background or vascular network are present [3]. Therefore, a benign notochordal cell tumour is occasionally overlooked histopathologically and misdiagnosed as a normal fatty marrow. The lesion is positive for vimentin, S-100 protein, epithelial membrane antigen (EMA), low molecular weight cytokeratin (CAM) and CK 18, as is conventional chordoma. Thus, the differentiation between chordoma and benign notochordal cell tumours relies on imaging studies being performed in combination with biopsies, which are often small. Notably, the distinction between benign notochordal cell tumours and chordomas is of clinical importance, as their management and ultimate prognosis are different [1, 4]. A benign notochordal cell tumour is generally located within the vertebral bodies and does not extend extraosseously. Thus, no clinical symptoms or neurological deficits are clinically manifest, although a rare case with significant clinical symptoms has been reported [5]. On CT studies, the surrounding bony trabeculae are sclerotic, whereas, on MRI, the lesion is homogeneously hypointense on T_1 weighted images and mildly to markedly hyperintense on T_2 weighted images. After administration of intravenous contrast material, the lesion does not enhance. However, contrast-enhanced MRI has so far been performed on only a limited number

Figure 3. Follow-up T_2 weighted sagittal image displays a significant decrease in the size of the mass in the spinal canal, although the mass probably remains in the T12 and L1 vertebral bodies. The T11 and L2 vertebral bodies show a compression fracture in contrast to the sclerotic anterior part of L1, where vertebral morphology is preserved.

of patients with these lesions [1]. Conversely, the general imaging features of chordomas are osteolytic on CT scans, although the tumour occasionally contains calcifications or residual bony trabeculae. They are inhomogeneously hyperintense on T_2 weighted MRI, with variable contrast enhancement.

To date, chordoma has been hypothesised to arise from embryonic notochordal remnants in the vertebral bodies. The embryonic notochordal remnants are generally identified in the intervertebral disc of fetuses or neonates [3]. In adults, notochordal remnants in the vertebral bodies have been described in radiological images [5, 6]; however, to the best of our knowledge, there is no histopathological proof that these lesions, *e.g.* giant vertebral notochordal rest, echordosis physaliphora vertebralis or benign notochordal cell tumours, are identical to embryonic notochordal remnants [7–9]. These benign lesions are found in 20% of autopsy series. In contrast, the reported incidence of conventional chordoma occurring in the clivus or vertebral bodies is approximately 0.2 cases per 100 000 yearly [9]. This epidemiological discrepancy suggests a rare association between benign notochordal cell tumour and conventional chordoma. Previously, Yamaguchi et al [10]

reported the first histologically confirmed case of a classic chordoma arising in a precursor benign notochordal lesion in the coccyx. Subsequently, Deshpande et al [11] gave support to the histopathological relationship, described by Yamaguchi et al [10], between intraosseous benign notochordal cell tumours and chordoma. In conjunction with these two compelling reports in the sacrococcygeal region, this is the first case that appears to illustrate, radiologically, and confirm histologically, benign notochordal cell tumour and classic chordoma arising within the same vertebra. Although the radiologically aggressive extraosseous component was not biopsied, it is possible that our case also supports the current hypothesis that benign notochordal cell tumour can exist as a precursor and transform into a classic chordoma [10]. Further collection of such cases is desired to verify this hypothesis.

Conclusions

In this case report, imaging studies and histopathology suggest that conventional chordoma has arisen from a benign notochordal cell tumour at the L1 level.

Acknowledgments

The authors are grateful to Dr Kenichi Wakasa and Dr Takehiko Yamaguchi for generous comments on the histopathology. We thank Dr Tadashi Kamada for invaluable performance and follow-up on radiation therapy and Dr Craig Kolodziej for helpful comments on our manuscript.

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