Solitary lymph node metastasis without local recurrence of primary chordoma

J. Sopta · G. Tulic · V. Mijucic · P. Mamontov ·
N. Mandic

Received: 9 July 2008 / Accepted: 17 September 2008 / Published online: 23 October 2008
© Springer-Verlag 2008

Abstract Chordoma is a malignant neoplasm believed to arise from notochord remnants. Its incidence is highest in the sixth decade and is generally regarded as a locally aggressive tumor with slow progression growth rate. Its metastatic incidence ranges from 5 to 40%, and it is generally believed that metastases without local recurrence of primary neoplasm are extremely rare. We report a case of a 38-year-old male patient with solitary inguinal lymph node metastasis without local recurrence of a previously surgically treated primary sacrococcygeal chordoma.

Keywords Chordoma · Sacrococcygeal · Surgically treated · Solitary lymph node metastasis · No recurrence of primary neoplasm

Introduction

Chordoma is a malignant neoplasm believed to arise from notochord remnants. It accounts for approximately 3–4%

of primary bone tumors and is localized along the axial skeleton [6, 22, 28]. This tumor develops predominantly in the sacrococcygeal (50%), spheno-occipital (35%) and cervical (15%) region and is generally regarded as a locally aggressive tumor with slow progression growth rate and metastatic incidence ranging from 5 to 40% [5, 20]. Its incidence is highest in the sixth decade, but it can also be found in younger population, even infancy (1%) [3, 4, 12, 26]. The male–female ratio among patients is 1.8–2.1.

Clinical, radiological (X-ray, CAT scan and MRI) and histological findings have been well discussed in the literature [7, 25]. Diagnostic certainty is based on histological examination, often suggestive of the diagnosis combined with ultrastructure and immunohistological study [2, 16, 23]. The clinical course involves local recurrences usually with realistic risk of metastatic development, notably in the sacrococcygeal chordomas case, with its incidence estimated at 17.5% of cases [5, 13, 24]. Sometimes developing in later stages of their course, histological findings are similar to those of the initial lesion. Mc Pherson stated that metastases without local recurrence of primary neoplasm are extremely rare. Organs which are frequently sites of metastases are: lung (48%), bones (26%) and sometimes liver. The sporadic cases of metastases in CNS [14], lymphatic organs [2, 5], breast [27] and skin and subcutaneous tissue [18] have been reported. Some cases of unusual clinical presentations of chordoma have also been emphasized [1, 19].

Macroscopically, this tumor mostly presents the lobular shape with fine fibro-vascular septas between the lobes. Microscopically, two distinct cellular patterns can be found: large, bright and vacuolated cells with excentric nucleus, called physaliphore-like cells and small, polygonal cells rich with eosinophilic cytoplasm and mild to moderate atypia of the nucleus [9, 17]. Both cell groups are

J. Sopta
Institute for Pathology, School of Medicine,
University of Belgrade, Belgrade, Serbia

G. Tulic (✉)
Institute for Orthopaedic Surgery and Traumatology,
Clinical Center of Serbia, 2 Pasterova Street,
11000 Belgrade, Serbia
e-mail: ditulio01@yahoo.com

V. Mijucic
Institute for Oncology of the Republic of Serbia,
Belgrade, Serbia

P. Mamontov · N. Mandic
Institute for Orthopaedic Surgery “Banjica”, Belgrade, Serbia

Fig.1 MRI and CAT scan presentation of a sacral tumor mass which infiltrates the surrounding muscular tissue and destroys the bone

arranged in band-like formations circumscribed by basophilic, mixoid stroma [22]. Mitoses are rare and not necessarily pathologic [28].

Immunophenotyping of the tissue samples enables the precise differentiation of chordoma from other neoplasms. This tumor has a strong positivity to vimentin (VIM), CK AE1/AE3 (pan-keratin), S-100 protein, epithelial membrane antigen (EMA) and CK (cyto-keratin) with low molecular weight [15, 21, 28].

Therapeutic management, ideally, consists of complete surgical excision of the initial tumor and adjuvant chemo or radio therapy [7, 8, 10].

Case report

This report is a case of a 38-year-old male patient with a history of an intermittent dull pain in the lumbosacral region of the spine without any neurological dysfunction, initially treated conservatively as a case of low-back pain syndrome. Due to slight paresthesias that occurred in both legs during treatment course, he underwent the X-ray and CAT scan diagnostic, which revealed expansive bone lesion of the sacrum. Further diagnostic with MRI delineated a $37 \times 35 \times 18$ mm solitary tumor mass of nodular shape, with bone destruction and local infiltration of the surrounding muscular tissue. The incisional biopsy was done. (Fig. 1)

Microscopically, tumor was lobulated. Individual lobes were separated by fibrous bands. The cells were arranged in syncytial nests and cords, lying in pink mucinous stroma. The tumor cells presented typical “physaliphorous appearance” with an abundant, pale, vacuolated bubbly

Fig. 2 Chordoma-physaliphorous cells with vacuolated cytoplasm and prominent vesicular nucleus (H&E, 100×)

cytoplasm. They showed mild to moderate nuclear atypia. Mitoses were infrequent, 1-2/HPF. Histological diagnosis was chordoma (Fig. 2).

For further characterisation of tumor, immunoperoxidase staining for cytokeratin (CK AE1/AE3), EMA, VIM, and S-100 protein were performed by streptavidinbiotin peroxidase technique using universal detection kit (Immunotech™). Tumor cells showed diffuse reactivity with antibodies against VIM and S100 protein, and focal to mild reactivity with pan-keratin (CK AE1/AE3) and EMA. (Fig. 3)

After definitive histopathologic diagnosis and in accordance with disease staging (local, extacompartmental lesion with infiltration of the surrounding muscles and subcutaneous tissue, absence of pulmonary dissemination)

Fig. 3 Immunoprofile of chordoma: immunoreactivity for vimentin (a), CK AE1/AE3(b), and S-100 protein (c)

Fig. 4 Lymph node involved by metastatic chordoma (a)- (H&E, 40×) Chordoma cells are positive for CK AE1/AE3 (b) and S-100 protein (c); LCA positive reaction in lymphoid tissue (d)

the radical surgery, meaning subtotal sacrectomy in the level of S2 segment, was performed through anterior and posterior approach simultaneously. Macro and micro analysis of the specimen obtaining at surgery confirmed the diagnosis. The width of the tumor's pseudocapsula was 1,7 mm, and from the oncological point of view this was interpreted as a marginal surgical procedure.

The patient was regularly followed at 3 months intervals, with no clinical, MRI or CAT scan signs of local recurrence or dissemination. After 13 months, the patient noticed the painless mass in his left inguinum. The control MRI revealed an enlarged solitary lymph node in this region, 25 × 30 mm in diameter, without local recurrence of a neoplasm. The lungs were free of metastases. This was initially treated by antibiotics which showed no result after two weeks of treatment, after which the excision biopsy

was done. The metastatic chordoma with identical histological and immunohistochemical characteristics of the primary tumor was confirmed. (Figs. 4, 5)

Twenty-eight months after index procedure, the patient is now disease-free and he is regularly followed-up in 3 months intervals. He had no functional impairment of urinary bladder and he has completely restored physiological walking pattern.

Discussion

According to the literature, almost 17–35% of the patients with chordoma present signs of local recurrence within various period of time after surgery [11, 13, 20, 24]. This can be followed easily or even preceded by distant

Fig. 5 The control MRI and CAT scan, 13 months after index procedure, that delineates the solitary inguinal lymph node metastasis without local recurrence of tumor

haemathogenous metastases, with lungs as a predominant site. That is why the regular MRI and CAT scan controls of all such patients are imperative, particularly in cases of a proven aggressive biological behaviour of a primary neoplasm. Very few authors described metastases in liver, skin or lymphatics. Solitary metastases without local recurrence of the primary tumor are supposed to be extremely rare [2, 5, 13, 20, 24].

To our best knowledge, there are just few cases in the literature that describe solitary lymph nodes metastases of chordoma, among whom we found not even one without verified local recurrence [2, 5, 26]. Furthermore, these patients, predominantly children or adolescents, were of younger age than usual for population of patients with chordoma. All of these cases ended lethally and were associated with already existing distant haemathogenous metastases in lungs or other organs.

The case here presented has almost all typical characteristics of chordoma: sacral localisation, non-specific clinical presentation in “low back pain” form and a long period of evolution. Radiological procedures that revealed an unclearly marginated large sacral mass that infiltrated and compromised the local muscles and other structures suggested the diagnosis of chordoma. The specific histological picture of a tumor samples obtained at surgery, showing mixoid tissue structure with well-defined physaliphorous cells, confirmed this presumption. Further immunophenotypisation of tumor cells with co-expression of VIM, cytokeratine, EMA and S-100 proteine excluded other types of neoplasms, such as chondrosarcoma, metastatic lucidocellular carcinoma and lyposarcoma.

Solitary lymph node metastasis that we clinically, radiologically and immunohistologically verified in this

patient 13 months after the index procedure, was not associated with local recurrence of primary neoplasm. We believe that this extremely rare clinical course and biological behaviour of primary chordoma represents rather unusual example in orthopaedic oncology.

Conflict of interest statement None of the authors has any potential conflict of interest.

References

1. Baboiu OE, Taylor WM (2006) Noninvasive sacral chordoma presenting as a benign soft tissue mass. *Ann Diagn Pathol* 10(2):95–99. doi:[10.1016/j.anndiagpath.2005.11.003](https://doi.org/10.1016/j.anndiagpath.2005.11.003)
2. Chambers PW, Schwinn CP (1979) Chordoma. A clinicopathologic study of metastasis. *Am J Clin Pathol* 72(5):765–776
3. Dahlin DC, Unni KK (1986) Bone tumors: general aspects and data on 8542 cases, 4th edn. Springfield, IL
4. de Noronha L, Werner B, Mendonça CM, Nomura L, Bleggi-Tores LF (1995) Saccrococcygeal chordoma in a 9-year-old boy. *Arq Neuropsiquiatr* 53(3-B):654–658
5. Fagundes MA, Hug EB, Liebsch NJ, Daly W, Efird J, Munzenrider JE (1995) Radiation therapy for chordomas of the base of skull and cervical spine: patterns of failure and outcome after relapse. *Int J Radiat Oncol Biol Phys* 33(3):579–584. doi:[10.1016/0360-3016\(95\)02014-3](https://doi.org/10.1016/0360-3016(95)02014-3)
6. Fletcher CD (2006) The evolving classification of soft tissue tumours: an update based on the new WHO classification. *Histopathology* 48(1):3–12. doi:[10.1111/j.1365-2559.2005.02284.x](https://doi.org/10.1111/j.1365-2559.2005.02284.x)
7. Fournier DR, Gokaslan ZL (2003) Current management of sacral chordoma. *Neurosurg Focus* 15(2):9E. doi:[10.3171/foc.2003.15.2.9](https://doi.org/10.3171/foc.2003.15.2.9)
8. Gallia GL, Haque R, Garonzik I, Witham TF, Khavkin YA, Wolinsky JP et al (2005) Spinal pelvic reconstruction after total sacrectomy for en bloc resection of a giant sacral chordoma. Technical note. *J Neurosurg Spine* 3(6):501–506

9. Hruban RH, Traganos F, Reuter VE, Huvos AG (1990) Chordomas with malignant spindle cell components: a DNA flow cytometric and immunohistochemical study with histogenetic implications. *Am J Pathol* 137(2):435–447
10. Hulen CA, Temple HT, Fox WP, Sama AA, Green BA, Eismont FJ (2006) Oncologic and functional outcome following sacrectomy for sacral chordoma. *J Bone Joint Surg Am* 88(7):1532–1539. doi:[10.2106/JBJS.D.02533](https://doi.org/10.2106/JBJS.D.02533)
11. Ishii K, Chiba K, Watanabe M, Yabe H, Fujimura Y, Toyama Y (2002) Local recurrence after S2–3 sacrectomy in sacral chordoma. Report of four cases. *J Neurosurg* 97(1 Suppl):98–101
12. Iwasa Y, Nakashima Y, Okajima H, Morishita S (1998) Sacral chordoma in early childhood: clinicopathological and immunohistochemical study. *Pediatr Dev Pathol* 1(5):420–426. doi:[10.1007/s100249900057](https://doi.org/10.1007/s100249900057)
13. Jeanrot C, Vinh TS, Anract P, De Pinieux G, Ouaknine M, Forest M et al (2000) Sacral chordoma: retrospective review of 11 surgically treated cases. *Rev Chir Orthop Reparat Appar Mot* 86(7):684–693
14. Kamel MH, Kelleher M, Aquilina K, Keohane C, Kaar G (2005) Intracranial metastasis from a sacrococcygeal chordoma. Case report. *J Neurosurg* 102(4):730–732
15. Kinoshita T, Okuyama T, Shimosegawa E, Yoshida Y, Yasui N, Ogawa T et al (2001) Chordoma with postoperative subcutaneous implantation and meningeal dissemination: MRI. *Neuroradiology* 43(9):763–766. doi:[10.1007/s002340100563](https://doi.org/10.1007/s002340100563)
16. Klingler L, Trammell R, Allan DG, Butler MG, Schwartz HS (2006) Clonality studies in sacral chordoma. *Cancer Genet Cytogenet* 171(1):68–71. doi:[10.1016/j.cancergencyto.2006.06.007](https://doi.org/10.1016/j.cancergencyto.2006.06.007)
17. Kyriakos M, Totty WG, Lenke LG (2003) Giant vertebral notochordal rest: a lesion distinct from chordoma: discussion of an evolving concept. *Am J Surg Pathol* 27(3):396–406. doi:[10.1097/00000478-200303000-00015](https://doi.org/10.1097/00000478-200303000-00015)
18. Lountzis NI, Hogarty MD, Kim HJ, Junkins-Hopkins JM (2006) Cutaneous metastatic chordoma with concomitant tuberous sclerosis. *J Am Acad Dermatol* 55(2 Suppl):S6–S10. doi:[10.1016/j.jaad.2005.08.061](https://doi.org/10.1016/j.jaad.2005.08.061)
19. McCormick M, Schroeder T, Benham S (2006) Sacral chordoma: a case report with radiographic and histologic correlation and a review of the literature. *WMJ* 105(5):53–56
20. McPherson CM, Suki D, McCutcheon IE, Gokaslan ZL, Rhines LD, Mendel E (2006) Metastatic disease from spinal chordoma: a 10-year experience. *J Neurosurg Spine* 5(4):277–280. doi:[10.3171/spi.2006.5.4.277](https://doi.org/10.3171/spi.2006.5.4.277)
21. Meis JM, Giraldo AA (1988) Chordoma. An immunohistochemical study of 20 cases. *Arch Pathol Lab Med* 112(5):553–556
22. Mirra JM, Brien EW (2001) Giant notochordal hamartoma of intraosseous origin: a newly reported benign entity to be distinguished from chordoma. Report of two cases. *Skeletal Radiol* 30(12):698–709. doi:[10.1007/s002560100422](https://doi.org/10.1007/s002560100422)
23. Naka T, Boltze C, Kuester D, Samii A, Herold C, Ostertag H et al (2005) Intralesional fibrous septum in chordoma: a clinicopathologic and immunohistochemical study of 122 lesions. *Am J Clin Pathol* 124(2):288–294. doi:[10.1309/5DLWL8EA7JUDWNVL](https://doi.org/10.1309/5DLWL8EA7JUDWNVL)
24. Osaka S, Kodoh O, Sugita H, Osaka E, Yoshida Y, Ryu J (2006) Clinical significance of a wide excision policy for sacrococcygeal chordoma. *J Cancer Res Clin Oncol* 132(4):213–218. doi:[10.1007/s00432-005-0067-3](https://doi.org/10.1007/s00432-005-0067-3)
25. Plathow C, Weber MA, Debus J, Kauczor HU (2005) Imaging of sacral chordoma: comparison between MRI and CT. *Radiologe* 45(1):63–68. doi:[10.1007/s00117-003-1002-8](https://doi.org/10.1007/s00117-003-1002-8)
26. Shinmura Y, Miura K, Yajima S, Tsutsui Y (2003) Sacrococcygeal chordoma in infancy showing an aggressive clinical course: an autopsy case report. *Pathol Int* 53:473–477. doi:[10.1046/j.1440-1827.2003.01496.x](https://doi.org/10.1046/j.1440-1827.2003.01496.x)
27. Tot T (2006) Metastatic chordoma of the breast: an extremely rare lesion mimicking mucinous cancer. *APMIS* 114(10):726–729. doi:[10.1111/j.1600-0463.2006.apm_437.x](https://doi.org/10.1111/j.1600-0463.2006.apm_437.x)
28. Yamaguchi T, Watanabe-Ishiiwa H, Suzuki S, Igarashi Y, Ueda Y (2005) Incipient chordoma: a report of two cases of early-stage chordoma arising from benign notochordal cell tumors. *Mod Pathol* 18(7):1005–1010. doi:[10.1038/modpathol.3800378](https://doi.org/10.1038/modpathol.3800378)