Benign Cartilaginous Tumors of the Sacrum

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13.1 Introduction

Primary benign bone tumors of the sacrum are extremely rare, and cartilaginous lesions have been reported only as case reports or small series. The histotypes affecting the sacrum and reported in literature were osteochondroma, chondroblastoma, chondromyxoid fibroma, and periosteal chondroma. Since clinical, imaging, and histologic characteristics vary immensely, as well as type of treatment, each of these lesions will be discussed in separate paragraphs.

13.2 Osteochondroma

Osteochondroma is caused by a misplaced fragment of the growth plate and the consequent abnormal overgrowth of cartilage in unusual site. The result is a progressive endochondral ossification into a bony subperiosteal protuberance, covered by cartilaginous cap, that projects from the bone surface [1]. The lesion has a thin outer cortex and an internal cancellous structure in continuity with the medulla of the bone from which it arises [2, 3], rich of fatty or hematopoietic marrow [4]. Osteochondroma may be solitary or multiple (associated with an autosomal disorder called hereditary multiple exostoses) [3, 5].

Osteochondroma rarely involves the spine (approximately 3% of cases) [3, 6–8], with solitary lesions involving 1.3–4.1% and HME involving 3–9% of the cases [2, 8–13]. Sacrum is involved less commonly than mobile spine, with an occurrence of about 0.5% of all spinal osteochondromas (Table 13.1) [2, 6–9, 13–19]. Swelling is

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Table 13.1 Osteochondroma of the sacrum: review of the literature

Study	Year	Patients (*n*)	Age/ gender	Treatment	Approach and location	Outcome
Pugh et al. [14]	1946	1	–	–	–	–
Sung et al. [7]	1987	2	–	Surgery	PA, below S3	Good, no LR
Hanakita et al. [15]	1988	1	42/F	Surgery, hemilaminectomy L4-S1	PA, L5-S1 lamina	Good, no LR
Gille et al. [16]	2004	1	45/F	Surgery, lumbotomy	PA, S1	
Bess et al. [8]	2005	1	34/F	Surgery, MHE	PA, L5-S1 articular process	Good, no LR
Agrawal et al. [17]	2005	1	14/M	Surgery	PA, right ala sacrum	Good, no LR
Samartzis et al. [6]	2006	1	11/M	Surgery, en bloc excision with S1-S4 laminectomy	PA, S2 lamina	Good, no LR
Chin et al. [18]	2010	1	54/F	Surgery, en bloc excision	Abdominal-retroperitoneal approach, sacrum	Good, no LR
Kuraishi et a. [2]	2014	1	63/F	Surgery, hemilaminectomy right L5-S1	PA, S1 articular process	Good, no LR
Baruah et al. [19]	2015	1	21/M	Surgery, en bloc excision	PA, S3-S4 lamina	Good, no LR
Sciubba et al. [13]	2015	1	48/M	Surgery, en bloc excision	PA, S1	Good, no LR
		1	48/M	Surgery, en bloc excision	PA, S1	LR
		1	21/M	Surgery, en bloc excision	PA, S1	Good, no LR
		1	17/M	Surgery, en bloc excision	PA, S1	Good, no LR
		1	13/M	Surgery, en bloc excision	PA, L5-S3	Good, no LR

F Female, *M* Male, *MHE* multiple hereditary exostosis, *PA* posterior approach, *LR* Local recurrence

one of the main symptoms, slowly increasing during skeletal growth. Osteochondroma is usually painless considering that usually grows posteriorly into the soft tissue, out of the spinal canal, whereas rarely it grows anteriorly causing spinal cord or nerve root compression [8, 19–22]. Similar to other benign tumors, osteochondromas have predilection for young male patients younger than 20 years of age [9]. Due to the unusual site, half of the described cases were treated in adult age [2, 8, 13, 15, 16, 18].

X-rays and computed tomography (CT) scans show the pathognomonic features of bone components with pedunculated or sessile base. However, plain X-rays are usually insufficient because of overlapping of other osseous structures [6, 15, 17]. MRI is important to visualize the size of the cartilaginous cap and the eventual neural structures compression. Differential diagnosis from other neoplasms involving the sacrum is not difficult. Parosteal chondrosarcomas are suspected if increasing size is noticed after skeletal maturity [1, 3, 12]. Malignant transformations to chondrosarcoma range between 10 and 20% in hereditary multiple exostosis and 1–5% in solitary osteochondromas [3, 12, 16]. When the cartilaginous cap is greater than 1–3 cm, in presence of new onset of symptoms or when the tumor rapidly increases in size, malignant changes should be suspected [13].

The mainstay of treatment is observation because most lesions are asymptomatic. En bloc surgical excision of sacral osteochondroma with free margins or marginal margins usually constitutes adequate treatment for symptomatic tumors although it is important to consider the risk of injuring nearby pelvic organs and neurovascular structures [23]. Tumors of the sacrum can be removed through anterior, posterior, or combined approaches [24]. The choice of the appropriate approach is dictated by the location of the tumor and the anatomic peculiarity and hypervascularity of the sacrum. Complete excision of the cartilaginous cap and its overlying periosteum is recommended to reduce the risk of local recurrence [3, 6, 25, 26]. Some authors suggest a frozen section biopsy to confirm margin free of tumor [19].

The outcome and prognosis after surgery of sacral osteochondromas are excellent. The risk of recurrence after treatment of osteochondroma of the spine or the sacrum is not well known because of the rare occurrence. Based on literature review, Gille et al. estimated 4% risk of recurrence in spine, slightly higher than the estimated 2% recurrence in long bones [16]. A tumor recurrence may represent a suspicious of malignancy [3, 12].

13.3 Chondroblastoma

Chondroblastoma is a rare benign cartilage-producing tumor consisting of about 1% of all bone tumors [3]. Chondroblastoma is usually found in the epiphyseal or epimetaphyseal areas of long bones, in males (male:female ratio 2:1), and could be found in any ages, even if it occurs most frequently between 10 and 25 years old. The incidence of vertebral chondroblastoma is 1.4% of all chondroblastomas, and less than five cases of sacral involvement have been reported in literature (only one clearly described) [27]. The radiological findings of vertebral chondroblastomas are nonspecific and biopsy should be performed considering possible more frequent differential diagnosis: aneurysmal bone cyst, giant cell tumor, chondromyxoid fibroma, osteoid osteoma, osteoblastoma, chondrosarcoma, chordoma, and metastasis [3]. Chondroblastomas of the spine behave more aggressively than those of long bones with a higher rate of local recurrence (about one-third of patients) [27–30]. Some authors suggested that this may be related to the frequent extension to adjacent soft tissue and the spinal canal, which precludes complete tumor resection [31,

32]. Three cases of tumor-related death have been reported due to direct invasion to adjacent soft-tissue and neurological structures [27, 30, 33]. Therefore, complete excision and long-term follow-up are generally recommended as the treatment modality for vertebral or sacral chondroblastomas.

13.4 Chondromyxoid Fibroma

Chondromyxoid fibroma (CMF) is a rare benign cartilaginous tumor generally observed between 5 and 30 years of age. It accounts for about 0.5% of all primary bone tumors [3, 34]. CMF of the sacrum is exceedingly rare, with less than ten cases reported in the literature (Table 13.2) [35–43]. Many of them demonstrate expansile or erosive imaging pattern, with cortical destruction, sclerotic and lobulated borders, septation, and intralesional calcifications [34]. Because of these radiographic similarities with more aggressive sacral pathology, differentiating CMF from chondrosarcoma, chordoma, and giant cell tumor on the basis of imaging alone is not possible [40]. CT-guided biopsy with histologic evaluation is mandatory for diagnosis. Although follow-up and detailed documentation are rather lacking in the treatment of sacral CMF, surgery in terms of total en bloc resection or intralesional curettage represents the current accepted management options. Radiation therapy has been used for sacral CMF in only one patient who died of complications before 4 months of follow-up [37]. The risk of local recurrence for CMF ranges between 4 and 80% [41], but higher risk can be expected in younger age group with spine

Table 13.2 Chondromyxoid fibroma of the sacrum: review of the literature

Study	Year	Patients (*n*)	Age/gender	Treatment	Outcome
Markley et al. [35]	1982	1	–	–	–
Shulman et al. [36]	1985	1	15/M	Surgery, anterior-posterior resection	–
Zillmer et al. [37]	1989	1	58/F	Surgery and radiation therapy	Died of complications
Rodgers et al. [38]	1997	1	17/F	Surgery, curettage, and graft	Good, no LR
Wu et al. [39]	1998	1	59/F	Surgery, resection, and lumbopelvic fixation	–
Brat et al. [40]	1999	1	30/M	Surgery, partial sacrectomy	Good, no LR
Mehta et al. [41]	2006	1	26/F	Surgery, resection, and curettage	–
Ahuja et al. [42]	2011	1	59/F	Surgery, curettage, graft, and lumbopelvic fixation	Good, no LR
Minasian et al. [43]	2016	1	35/F	Surgery, partial sacrectomy	Good, no LR

lesions despite extensive curettage [36]. The longest reported follow-up for sacral CMF was a patient treated with curettage and bone grafting, who was found to be recurrencefree at 8-year follow-up [38]. Other authors reported no evidence of recurrence at about 1 year of follow-up after wide resection [40, 42].

13.5 Periosteal or Juxtacortical Chondroma

Periosteal chondroma in the spine is extremely rare and only two cases affecting the sacrum have been reported until now [44, 45]. Periosteal chondroma is a slow-growing benign cartilaginous tumor of bone surface with periosteal origin. Radiologically, it often demonstrates a periosteal shelf of bone and a superficial erosion of the cortex with endosteal sclerosis. Chondrosarcoma is the major differential diagnosis in cases of periosteal chondroma, followed by periosteal osteosarcoma (chondroblastic type), osteochondroma, osteoblastoma, and aneurysmal bone cyst [44, 46]. Akiyama et al. [45] reported a case with large periosteal chondroma that arose on the endopelvic surface of the sacrum, which was difficult to distinguish from chondroid chordoma. The treatment of choice for an asymptomatic tumor is observation as well as for osteochondromas, considering surgery only in presence of symptoms [17]. Singh et al. [44] performed an en bloc excision with good results in terms of pain relief and return to normal activities, without signs of local recurrence after 4 years of follow-up. Akiyama et al. [45] performed intralesional excision with curettage of the underlying cortical bone, and there has been no local recurrence at about 3 years of follow-up.

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References

1. Solomon L, Warwick D, Osteochondroma Nayagam S. Apley's system of orthopaedics and fractures. 9th ed. London: Hodder Arnold; 2010. p. 199–200.
2. Kuraishi K, Hanakita J, Takahashi T, Watanab M, Honda F. Symptomatic osteochondroma of lumbosacral spine: report of 5 cases. Neurol Med Chir. 2014;54(5):408–12.
3. Unni K, editor. Dahlin's bone tumors: general aspects and data on 11,087 cases. Philadelphia: Lippincott-Raven; 1996.
4. Rodallec M, Feydy A, Larousserie F, Anract P, Campagna R, Babinet A, et al. Diagnostic imaging of solitary tumors of the spine: what to do and say. Radiographics. 2008;28:1019–41.
5. Wise C, Clines G, Massa H, et al. Identification and localization of the gene for EXTL, a third member of the multiple exostoses gene family. Genome Res. 1997;7:10–6.
6. Samartzis D, Marco RA. Osteochondroma of the sacrum: a case report and review of the literature. Spine. 2006;31(13):E425–9.
7. Sung H, Shu W, Wang H, et al. Surgical treatment of primary tumors of the sacrum. Clin Orthop Relat Res. 1987;215:91–8.
8. Bess RS, Robbin MR, Bohlman HH, Thompson GH. Spinal exostoses analysis of twelve cases and review of the literature. Spine. 2005;7:774–80.

9. Albrecht S, Crutchfeld JS, SeGall GK. On spinal osteochondromas. J Neurosurg. 1992;77:247–52.
10. Robbins SE, Laitt RD, Lewis T. Hereditary spinal osteochondromas in diaphyseal aclasia. Neuroradiology. 1996;38:59–61.
11. Royster RM, Kujawa P, Dryer RF. Multilevel osteochondroma of the lumbar spine presenting as spinal stenosis. Spine. 1991;16:992–3.
12. Schajowicz F. Tumors and tumorlike lesions of bone and joints. New York: Springer; 1994. p. 141–256.
13. Sciubba DM, Macki M, Bydon M, Germscheid NM, Wolinsky JP, Boriani S, Bettegowda C, Chou D, Luzzati A, Reynolds JJ, Szövérfi Z, Zadnik P, Rhines LD, Gokaslan ZL, Fisher CG, Varga PP. Long-term outcomes in primary spinal osteochondroma: a multicenter study of 27 patients. J Neurosurg Spine. 2015;22(6):582–8.
14. Pugh H, Crile GJ, Robnett A. Exostosis of sacrum: report of a case. US Nav Med Bull. 1946;46:269–72.
15. Hanakita J, Suzuki T. Solitary sacral osteochondroma compressing the cauda equina—case report. Neurol Med Chir. 1988;28:1010–3.
16. Gille O, Pointillart V, Vital JM. Course of spinal solitary osteochon-droma. Spine. 2004;30:E13–9.
17. Agrawal A, Dwivedi S, Joshi R, Gangane N. Osteochondroma of the sacrum with a correlative radiographic and histological evaluation. Paediatr Neurosurg. 2005;41:46–8.
18. Chin K, Kim J. A rare anterior sacral osteochondroma presenting as sciatica in an adult: a case report and review of the literature. Spine J. 2010;10(5):e1–4.
19. Baruah RK, Das H, Haque R. Solitary sacral osteochondroma without neurological symptoms: a case report and review of the literature. Eur Spine J. 2015;24(Suppl 4):S628–32.
20. Ohtori S, Yamagata M, Hanaoka E, Suzuki H, Takahashi K, Sameda H, Moriya H. Osteochondroma in the lumbar spinal canal causing sciatic pain: report of two cases. J Orthop Sci. 2003;8:112–5.
21. Fiumara E, Scarabino T, Guglielmi G, Bisceglia M, D'angelo V. Osteochondroma of L-5 vertebra a rare cause of sciatic pain. Case report. J Neurosurg. 1999;91(2 suppl):219–22.
22. Govender S, Parbhoo AH. Osteochondroma with compression of the spinal cord. A report of two cases. J Bone Joint Surg Br. 1999;81-B:667–9.
23. Bottner F, Rodl R, Kordish I, Winklemann W, Gosheger G, Lindner N. Surgical treatment of symptomatic osteochondroma: a three- to eight-year follow-up study. J Bone Joint Surg Br. 2003;85:1161–5.
24. Ruggieri P, Angelini A, Ussia G, Montalti M, Mercuri M. Surgical margins and local control in resection of sacral chordomas. Clin Orthop Relat Res. 2010;468(11):2939–47.
25. Boriani S, Weinstein JN, Biagini R. Primary bone tumors of the spine. Spine. 1997;22:1036–44.
26. Weinstein JN, McLain RF. Primary tumors of the spine. Spine. 1987;12:843–51.
27. Akai M, Tateishi A, Machinami R, Iwano K, Asao T. Chondroblastoma of the sacrum: a case report. Acta Orthop Scand. 1986;57:378–81.
28. Ilaslan H, Sundaram M, Unni KK. Vertebral chondroblastoma. Skelet Radiol. 2003;32:66–71.
29. Bloem JL, Mulder JD. Chondroblastoma: a clinical and radiological study of 104 cases. Skelet Radiol. 1985;14:1–9.
30. Hoe el JC, Brasse F, Schmi M, et al. About one case of vertebral chondroblastoma. Pediatr Radiol. 1987;17:392–6.
31. Kim J, Kumar R, Raymond AK, Ayala AG. Non-epiphyseal chondroblastoma arising in the iliac bone, and complicated by an aneurysmal bone cyst: a case report and review of the literature. Skelet Radiol. 2010;39(6):583–7.
32. Leung LY, Shu SJ, Chan MK, Chan CH. Chondroblastoma of the lumbar vertebra. Skelet Radiol. 2001;30:710–3.
33. Chung OM, Yip SF, Ngan KC, Ng WF. Chondroblastoma of the lumbar spine with cauda equina syndrome. Spinal Cord. 2003;41:359–64.

34. Wilson AJ, Kyriakos M, Ackerman LV. Chondromyxoid fibroma: radiographic appearance in 38 cases and in a review of the literature. Radiology. 1991;179:513–8.
35. Makley JT, Cohen AM, Baada E. Sacral tumours: hidden problems. Orthopedics. 1982;5:996–1003.
36. Shulman L, Bale P, de Silva M. Sacral chondromyxoid fibroma. Pediatr Radiol. 1985;15: 138–40.
37. Zillmer DA, Dorfman HD. Chondromyxoid fibroma of bone: thirtysix cases with clinicopathologic correlation. Hum Pathol. 1989;20:952–64.
38. Rodgers WB, Kennedy JG, Zimbler S. Chondromyxoid fibroma of the ala of the sacrum presenting as a cause of lumbar pain in an adolescent. Eur Spine J. 1997;6:351–3.
39. Wu CT, Inwards CY, O'Laughlin S, Rock MG, Beabout JW, Unni KK. Chondromyxoid fibroma of bone: a clinicopathologic review of 278 cases. Hum Pathol. 1998;29:438–46.
40. Brat HG, Renton P, Sandison A, Cannon S. Chondromyxoid fibroma of the sacrum. Eur Radiol. 1999;9:1800–3.
41. Mehta S, Szklaruk J, Faria SC, Raymond AK, Whitman GJ. Radiologicpathologic conferences of the University of Texas M.D. Anderson Cancer Center: chondromyxoid fibroma of the sacrum and left iliac bone. AJR Am J Roentgenol. 2006;186:467–9.
42. Ahuja SK, McCanna SP, Horn EM. Treatment strategy for chondromyxoid fibroma of the sacrum. J Clin Neurosci. 2011;18:1550–2.
43. Minasian T, Claus C, Hariri OR, Piao Z, Quadri SA, Yuhan R, Leong D, Tashjian V. Chondromyxoid fibroma of the sacrum: a case report and literature review. Surg Neurol Int. 2016;7(Suppl 13):S370–4.
44. Singh AP, Singh AP, Mahajan S. Periosteal chondroma of the sacrum. Can J Surg. 2008; 51(5):E105–6.
45. Akiyama T, Yamamoto A, Kashima T, Ishida T, Shinoda Y, Goto T, Nakamura K, Kawano H. Juxtacortical chondroma of the sacrum. J Orthop Sci. 2008;13(5):476–80.
46. Boriani S, Bacchini P, Bertoni F, et al. Periosteal chondroma. A review of twenty cases. J Bone Joint Surg Am. 1983;65:205–12.