Sacral chordoma

- Characterization of Soft Tissue Reconstruction Following Chordoma Resection
- Robotic Radiosurgery for Sacral Chordoma: Preserving Function in a Clinical Conundrum Scenario
- Layered Repair in Extensive Sacral Defects with Bilaminar Gluteal Flaps: Demirdover Flaps
- Notochord-derived lesion presenting with cerebrospinal fluid rhinorrhea: illustrative case
- Pelvic and Perineal Reconstruction After Bowel, Gynecological or Sacral Tumor Resection: A Case Series
- Hybrid therapy and use of carbon-fiber-reinforced polyetheretherketone instrumentation for management of mobile spine chordomas: A case series and review of the literature
- Management for chordoma of the spine and sacrum with stereotactic radiosurgery
- A population based survival analysis of skull base and sacral-coccygeal chordomas in an elderly population: 2000-2021

Epidemiology

This spinal chordoma, constitute over 50% of primary bone tumors of the sacrum, and show a male predominance [1].

Molecular basis

The molecular basis for the clinical behavior remains unknown.

The expression of phosphatase and tensin homolog deleted on chromosome 10 (PTEN) and mammalian target of rapamycin (mTOR) were detected immunohistochemically in 40 sacral chordoma tissues and 10 adjacent normal tissues. Correlations between PTEN and mTOR expression and clinicopathological factors were analyzed. Kaplan-Meier survival curves and log-rank test were used to analyze the continuous disease-free survival time (CDFS). The expression of PTEN in sacral chordoma was significantly lower than that in adjacent normal tissues, while the levels of mTOR expression in sacral chordoma were significantly higher than that in adjacent normal tissues ($P = 0.000$, $P = 0.030$). The positive expression of mTOR appears to correlate with the negative expression of PTEN in sacral chordoma ($P = 0.021$). PTEN-negative expression and mTOR-positive expression were associated with tumor invasion into the surrounding muscles ($P = 0.038$, $P = 0.014$). Log-rank test showed that PTEN-negative and mTOR-positive expressions had an important impact on the patients' CDFS ($P = 0.011$, $P = 0.015$).

Results suggest that PTEN and mTOR might play an important role in the local invasiveness of sacral chordoma. PTEN and mTOR might be recognized as important prognostic predictors of recurrence and could be used as potential therapeutic targets for the treatment for sacral chordoma [2].

Clinical features

May produce pain, sphincter disturbance or nerve root symptoms from local nerve root compression. It may occasionally extend cephalad into the lumbar spinal canal. It is usually confined anteriorly by

the presacral fascia, and only rarely invades the wall of the rectum [3].

Diagnosis

A firm fixed mass may be palpable between the rectum and the sacrum on rectal exam.

Characteristic radiographic findings: centrally located destruction of several sacral segments, with an anterior soft-tissue mass that occasionally has small calcifications. CT and MRI show the bony destruction. This is usually difficult to see on plain x-rays. MRI also shows the soft-tissue mass.

Open or CT guided percutaneous posterior biopsy can confirm the diagnosis. Transrectal biopsy should be avoided because of the potential of rectal spread of tumor [4].

Chest CT and bone scan: to R/O mets for staging purposes.

Treatment

Sacral chordoma treatment.

Outcome

see Sacral chordoma outcome.

Case series

Sacral chordoma case series.

Case reports

A 64 years old female patient with a locally recurrent sacral chordoma came to our attention after ten interventions performed at another institute. A surgical treatment was performed using argon cryosurgery.

In this case, it was not possible to perform a marginal or wide excision, but it was intralesional with the removal of three major blocks. The fragments sent for the histological analysis measured total 35 × 30 × 8 cm with a weight of 4.260 g.

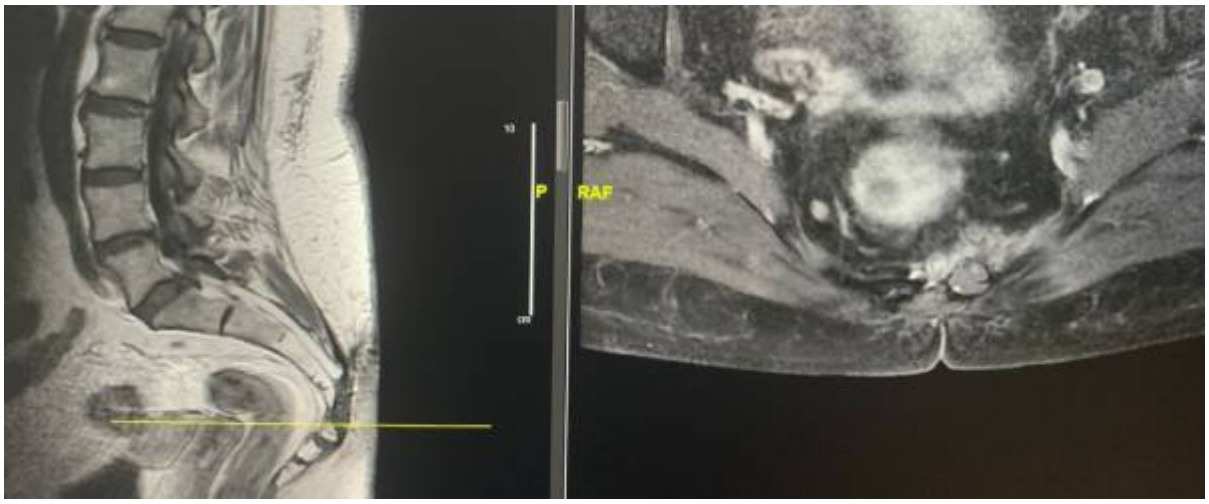
Given the gelatinous structure of the tumor and the possibility of contamination of operatory field, cryosurgery may be indicated when previous surgeries and the dimensions of tumor mass do not allow a resection with negative margins improving radiotherapy efficacy in the local control of tumor [5].

Case report from the HGUA

q10907

61-year-old woman, presenting a lytic tumor on the coccyx, diagnosed as Sacral chordoma by ultrasound-guided puncture, with a negative extension study -assessed by scintigraphy

CT of the lumbosacral-coccygeal spine



It is compared with a previous MRI study.

Report: Post-surgical changes at the level of the sacrococcyx after excision of chordoma at said level.

Ill-defined soft tissue increase in the left lateral region of the sacrococcygeal junction, in the surgical bed, which extends both anteriorly and posteriorly to the coccyx region of about 2.8 x 1.4 x 2.3 cm (TR x CC x AP), in relation to probable tumor recurrence at that level.

Prone . Reopening of the previous scar and enlargement with a crescentic incision in the transverse plane. Subperiosteal muscle dissection of the sacrum and coccyx. Under radiological control, the limit of sacral resection is identified in S3. With the help of a piezoelectric motor, a sacrectomy is performed maintaining a wide tumor margin at all times. Dissection of the anterior face of the Sacrum from the mesorectum with the help of a monopolar. Distal segments of the sacrum and coccyx are removed en bloc. Hemostasis with fibrillarsurgicel and bone wax. Blake-type drainage is left. Subcutaneous plane closure. Leather with monofilament with mattress stitches + clips.

References

1)

O'Neill P, Bell BA, Miller JD, Jacobson I, Guthrie W. Fifty Years of Experience with Chordomas in South-east Scotland. Neurosurgery. 1985; 16:166–170

2)
Chen K, Mo J, Zhou M, Wang G, Wu G, Chen H, Zhang K, Yang H. Expression of PTEN and mTOR in sacral chordoma and association with poor prognosis. Med Oncol. 2014 Apr;31(4):886. doi: 10.1007/s12032-014-0886-7. Epub 2014 Feb 18. PubMed PMID: 24535608.
3)
Azzarelli A, Quagliuolo V, Cerasoli S, et al. Chordoma: Natural History and Treatment Results in 33 Cases.J Surg Oncol.1988;37:185–191
4)
Mindell ER. Current Concepts Review. Chordoma. J Bone Joint Surg. 1981; 63A:501–505
5)
Pipola V, Girolami M, Ghermandi R, Tedesco G, Evangelisti G, Gasbarrini A. Cryosurgery in the excision of a giant local recurrent sacral chordoma: a case report and literature review. Eur Spine J. 2018 Dec;27(12):3059-3063. doi: 10.1007/s00586-017-5438-3. Epub 2018 Jan 25. PubMed PMID: 29372428.