Mesenchymal chondrosarcoma

Mesenchymal chondrosarcoma is a rare, high-grade malignancy of bone or soft tissue with a unique, biphasic histology and poor prognosis. Because of its rarity and variable length of disease-free survival, the natural history of the disease remains poorly understood.

Intracranial extra-skeletal mesenchymal chondrosarcoma is a rare, malignant variant of chondrosarcoma which is characterized by undifferentiated mesenchymal cells interspersed with pockets of mature hyaline cartilage.

A 23-year-old female who underwent multiple craniotomies for tumor resection, as well as adjuvant radiotherapy and chemotherapy. They reviewed the literature for reported cases and discussed the histopathological features, radiological findings, therapeutic approaches and outcomes associated with this rare tumor [1].

To present clinical, radiographic, and histopathologic features of mesenchymal chondrosarcoma from one of the largest case series collected by a single, senior-level bone pathologist.

Twenty cases were reviewed in consultations spanning 45 years.

Eighteen tumors (90%) originated in bone, and 2 tumors (10%) were of extraskeletal origin. Of the skeletal tumors, locations included craniofacial bones (n = 9; 50%), ribs and chest wall (n = 4; 22%), sacrum and spinal elements (n = 3; 17%), and lower extremities (n = 2; 11%), whereas soft tissue tumors were located about the scapula (n = 1; 50%) and lower extremity (n = 1; 50%). Plain radiographs demonstrated calcified, osteolytic lesions with extraosseous extension. Typical histologic features were identified consisting of small, round or spindled cells, interspersed with hyaline cartilage islands. Seventeen patients (85%) were treated surgically, and 8 patients (40%) received adjuvant treatment. Seven patients (35%) were living at last follow-up, 1.8 to 12.5 years after diagnosis, and 8 patients (40%) died between 1.2 and 21.8 years after diagnosis.

Mesenchymal chondrosarcoma presents multiple challenges. Diagnostic pitfalls include inadequate biopsy samples, which may result in sample error. Sox9 has been proposed as a unique marker for mesenchymal chondrosarcoma which may improve diagnostic specificity. Treatment and prognosis vary considerably. Patients who receive surgery and chemotherapy seem to fare better. Multicenter studies with higher sample numbers may improve our understanding of this malignancy [2].

Case reports

Primary spinal intradural extramedullary mesenchymal chondrosarcoma: a case report [3].

2017

A case of intradural extramedullary MCS, located at the T11-T12 level, in a 14-year-old male. The tumor was documented by magnetic resonance imaging and treated with gross total resection (GTR) without adjuvant treatment. We further reviewed the relevant pediatric literature and discussed the management and outcome of intracranial and intraspinal MCS.

The patient's follow-up showed no evidence of disease 2 years from diagnosis. A total of 51 cases of intracranial and intraspinal MCS have been reported (24 intraspinal and 27 intracranial). Recurrence has been described in only 4 patients with intraspinal MSC, and among them 3 received adjuvant chemotherapy and radiotherapy. GTR seems to reduce the risk of recurrence and, due to a higher cancer-mortality rate for these patients, adjuvant chemotherapy and radiotherapy are recommended in case aggressive surgery is not possible.

According to our single experience, we would suggest that adjuvant therapy might be unnecessary in cases where a localized MCS undergoes GTR. Chemotherapy and radiotherapy should be recommended when GTR cannot be obtained. Further studies are needed to investigate a standard treatment approach for this rare tumor [4].

A 22-year-old woman with MCS of the arachnoid at the T12-L1 level with a 14-year-long observation. The tumor was totally resected using osteoplastic laminotomy with reconstruction of laminar roof. This small spindle cell tumor was initially microscopically suspected of synovial sarcoma, but correctly verified with widened immunophenotyping and molecular studies as MCS. At its first recurrence, the neoplasm showed microscopically a typical bimorphic pattern of small round cell component with foci of hyaline cartilage. The patient experienced three local recurrences: 4, 6, and 10 years after the initial resection, respectively. The techniques of laminotomy and relaminotomy were also used during three following operations. The repeated surgical removal, radiotherapy, and chemotherapy were the methods of complex oncological treatment. The patient remains now in complete remission, fully self-dependent with slight motor disturbance, and mild sensory deficits [5].

References

1)
Shabani S, Kaushal M, Kaufman B, Knipstein J, Lawlor MW, Lew S, Foy A. Intracranial extra-skeletal mesenchymal chondrosarcoma: Case report and review of literature of reported cases in adults and children. World Neurosurg. 2019 May 31. pii: S1878-8750(19)31498-6. doi: 10.1016/j.wneu.2019.05.221. [Epub ahead of print] PubMed PMID: 31158544.
2)
Shakked RJ, Geller DS, Gorlick R, Dorfman HD. Mesenchymal chondrosarcoma: clinicopathologic study of 20 cases. Arch Pathol Lab Med. 2012 Jan;136(1):61-75. doi: 10.5858/arpa.2010-0362-OA. PubMed PMID: 22208489.
3)
Saito R, Senbokuya N, Yagi T, Yoshioka H, Kanemaru K, Kinouchi H. Primary spinal intradural extramedullary mesenchymal chondrosarcoma: a case report. World Neurosurg. 2020 Oct 10:S1878-8750(20)32199-9. doi: 10.1016/j.wneu.2020.10.007. Epub ahead of print. PMID: 33049384.
4)
Di Giannatale A, Colletti M, Russo I, Ferruzzi V, Dell' Anna VA, Cozza R, Colafati GS, Messina R, Mastronuzzi A, De Vito R, Milano GM. Intraspinal mesenchymal chondrosarcoma: report of a pediatric case and literature review. Tumori. 2017 Nov 15;103(Suppl. 1):e66-e72. doi: 10.5301/tj.5000689. PubMed PMID: 29143958.
5)

Derenda M, Borof D, Kowalina I, Wesołowski W, Kloc W, Iżycka-Świeszewska E. Primary Spinal Intradural Mesenchymal Chondrosarcoma with Several Local Regrowths Treated with Osteoplastic Laminotomies: A Case Report. Surg J (N Y). 2017 Jul 20;3(3):e117-e123. doi: 10.1055/s-0037-1604159. eCollection 2017 Jul. PubMed PMID: 28825035; PubMed Central PMCID: PMC5553510.