Images in Cytopathology

Low-Grade Chondrosarcoma of Petrous Apex Diagnosed by Intraoperative Crush Smear Cytology

Chondrosarcomas account for 6% of skull base neoplasms and 0.15% of all intracranial tumors. They are thought to arise from chondrocytes within the rest of enchondral cartilage present within the skull base. Although skull base chondrosarcomas are generally low to intermediate grade with indolent growth and low metastatic potential, they are neurologically devastating. Discussed here, a case of low-grade chondrosarcoma radiologically arising from the petrous apex, which could be diagnosed on intraoperative crush cytology.

A 35-year-old woman was admitted, who presented with left-sided tongue atrophy, tinnitus, and headache for a period of 13 years. She had no complaints of visual impairment, seizures, or loss of consciousness. Axial T2-weighted magnetic resonance imaging (MRI) of the brain showed a mass with heterogeneous but predominantly high signal intensity involving the left petrous apex. A computed tomography (CT) of her brain showed that the lesion had specks of calcification and the tumor was eroding the skull base. The mass was seen extending to the cerebellopontine angle cistern and cavernous sinus. The radiological impression was in favour of a malignant chondroid tumor.

The patient was planned for surgery and intraoperative consultation was sent to the pathology department. We received multiple grey-white tissue bits amounting to 0.2 g. The tissue was easily crushable and mucoid in consistency. On microscopic examination, the smears revealed good cellularity displaying sheets and singly scattered round to oval cells in an abundant myxoid background. The cells are large with abundant pale blue to foamy cytoplasm and eccentric to subcentric nuclei. Correlating with the radiological findings and based on the cytomorphological features of the crush smears, a diagnosis of low-grade chondrosarcoma was favored.

Subsequently, excision of the tumor was done and histopathological examination revealed a lobular architecture with abundant cartilaginous matrix separated by narrow fibrovascular septae. Each lobule was comprised of round to slightly elongated cells separated by abundant myxoid stroma. These cells lie within lacunae and occasional binucleated forms were also noted. However, atypia was mild with low to absent mitotic figures. On immunohistochemistry (IHC), tumor cells were strongly positive for S-100 and Vimentin antigens. Pancytokeratin and Epithelial membrane antigens (EMA) were negative in the tumor cells ruling out chordoma. Thus, the histopathological diagnosis of low-grade chondrosarcoma of the left petrous apex corroborated with the intraoperative diagnosis on crush smears.

Figure 1: (a) MRI brain showing a mass with heterogeneous but predominantly high signal intensity involving the left petrous apex. (b) Crush smears showing good cellularity consisting of sheets of round to oval cells in a myxoid background (H & E, ×100). (c) High power showing dispersed round to ovoid cells with eccentric to subcentric nuclei and abundant pale blue to foamy cytoplasm (H & E, ×400). (d) Histopathology showing cords and rows of chondrocytes present within lacunae in a myxoid background. (H & E, ×400) (e) Tumor cells positive for S-100 (DAB, ×400) (f) Tumor cells positive for Vimentin (DAB, ×400)
Post-surgery MRI of the patient did not show any residual tumor. At 6 months’ follow-up, the patient did not show any local recurrence or locoregional metastasis.

The grading system for chondrosarcomas consists of grade I (well differentiated or low-grade), grade II (moderately differentiated or intermediate), and grade III (poorly differentiated or high-grade).[1] Based on cellularity and nuclear atypia, cytological grading of chondrosarcoma is possible.[2] Grade I chondrosarcomas show abundant chondroid matrix with variable cellularity.[2] The chondrocytes are in clusters with normal to slightly enlarged nuclei, inconspicuous small nucleoli, and absent mitosis. Occasional binucleated cells may be seen. Grade 2 chondrosarcomas show less chondromyxoid stroma and more cellularity comprising of chondrocytes in loose clusters.[2] The chondrocytes demonstrate nuclear enlargement, hyperchromasia, and frequent multinucleation. Grade 3 chondrosarcomas demonstrate high cellularity and marked atypia of chondrocytes with the presence of frequent mitosis.[2] Diagnosing low-grade chondrosarcomas on intraoperative crush smears poses difficulty since these tumors share similar cytomorphological features with other chondromyxoid rich lesions of the central nervous system such as cystic schwannoma with myxoid degeneration, chordoma, and chondromyxoid fibroma.[3‑5] Although the histological features of these tumors are distinct, there is a considerable overlap of cytological features. Cytologically cystic schwannoma with myxoid degeneration shows clusters of spindle cells along with cyst macrophages in a myxoid background. In chordoma, the tumor cells are sparser, the chordoid matrix more abundant, cells are larger than normal chondrocytes but smaller than low-grade chondrosarcoma.[3] Cytologically, chondromyxoid fibroma shows a polymorphic population of ovoid (chondrocytes) and stellate or spindle-shaped cells (fibroblasts) in a chondromyxoid background.[3] The chondrocytes in chondromyxoid fibroma lack the nuclear hyperchromasia and irregularity as seen in chondrosarcoma.[4] Chordoma, which shows characteristic physaliferous cells in a myxoid background, is another cytological differential diagnosis.

It is of paramount importance to distinguish these tumors on crush smears as treatment and prognosis differ. Schwannoma is managed by excision and rarely recurs if excised completely. Treatment of chondromyxoid fibroma includes en bloc or intraslesional curettage and only 25% of recur.[5] Skull base chordoma and chondrosarcoma carry a more favorable outcome than chordoma with regard to survival.[6] Surgical resection remains the key treatment for these tumours. Skull base chondrosarcoma and chordoma requires maximal tumour debulking, however, radiotherapy may not be necessary for patients with chordoma and low-grade chondrosarcoma.[6]

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Conflicts of interest
There are no conflicts of interest.

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REFERENCES
1. Bloch OG, Jian BJ, Yang I, Han SJ, Aranda D, Ahn BJ, et al. Cranial chondrosarcoma and recurrence. Skull Base 2010;20:149‑56.
2. Lerma E, Tani E, Brosjö B, Bauer H, Söderlund V, Skoog L. Diagnosis and grading of chondrosarcomas on FNA biopsy material. Diagn Cytopathol 2003;28:13‑7.
3. Tunc M, Ekinci C. Chondrosarcoma diagnosed by fine needle aspiration cytology. Acta Cytol 1996;40:283‑8.
4. Daswani K, Sudhamani S, Pandit A. Cytological diagnosis of chondrosarcoma: A case report with review of literature. J Sci Soc 2015;42:194‑7.
5. Dey B, Deshpande AH, Brar RK, Ray A. Chondromyxoid fibroma of the metatarsal bone: A diagnosis using fine needle aspiration biopsy. J Cytol 2018;35:67‑8.
6. Almefty K, Pravdenkova S, Colli BO, Almefty O, Gokden M. Chordoma and chondrosarcoma: Similar but quite different skull base tumor. Cancer 2007;110:2457‑67.

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