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CASE REPORT

Intradural spinal granular cell tumor

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ABSTRACT

Granular cell tumor is a rare, usually benign tumor with classical histomorphology. Location of tumor varies widely within body, but spine is distinctly a rare location for this tumor. We report a rare case of granular cell tumor involving intradural extramedullary portion of lumbar region of spinal cord. Knowledge of which is important as subsequent prognosis differs from other tumor at same location.

Key words: Granular cell tumor, intradural, spinal cord

Introduction

Granular cell tumor (GCT) is a rare neoplasm that is so-named due to its arrangement of nests of polyhedral cells with abundant granular eosinophilic cytoplasm. GCT, most commonly occurs in the skin, head and neck, with a particular predisposition for the tongue. Other common sites include respiratory tract, breast, and gastrointestinal tract; however, virtually any body site may be affected.[1] GCT of the spine, with intradural extramedullary location, is exceptionally rare but important differential diagnosis. We present a case of GCT involving intradural extramedullary portion of lumbar region of spinal cord.

Case Report

A 13-year-old female was operated outside for spinal tumor. As only subtotal excision was possible, patient was referred to our hospital for further management of tumor which was diagnosed, histopathologically, as low grade glioma. Reviewing clinical details, patient had pain in left lower limb since four years, developed difficulty in walking and swelling in lumbar region of back since one year. She had decreased superficial sensations along with paraesthesia in both the lower limbs below first lumbar dermatome and the motor power was reduced and graded as 4/5. Her bowel-bladder functions were unaffected. Her preoperative magnetic resonance imaging (MRI) of the spine showed well-defined intradural extramedullary lobulated lesion measuring 2.5 cm (AP) × 3.2 cm (RL) × 6.6 cm (SI) which was hypointense on T2-weighted images and isointense on T1-weighted images. [Figure 1] Post contrast study showed homogenous enhancement of lesion. Lesion caused widening of spinal canal, scalloping of adjacent vertebral bodies, splaying of posterior laminae, widening of neural foramina and compressing exiting nerve roots and couda equina nerve roots of L1-3. Radiological consideration included nerve sheath tumor or ependymoma. Her postoperative MRI showed a residual heterogeneously enhancing large intraspinal mass measuring 6.5 × 1.7 cm on sagital plane. Her biopsy slides were reviewed at our department which showed sheets and nests of large polyhedral tumor cells. Cells had round-to-oval, hyperchromatic-to-vesicular nuclei with irregular nuclear membrane, mild pleomorphism, but mitoses were lacking. Many nuclei showed intranuclear pseudoinclusions. Cells had abundant eosinophilic, fine-to-coarse granular cytoplasm with indistinct cellular borders giving a syncytial appearance. The islands of tumor were separated by delicate fibrovascular tissue. [Figure 2] The tumor cells were strongly and diffusely immunoreactive with S-100 protein and neuron-specific enolase (NSE). There was no reaction with epithelial membrane antigen (EMA), glial fibrillary acidic protein (GFAP). Mib-1 index was less than one percent. Overall, these findings were characteristic of a granular cell tumor. [Figure 3] As significant symptomatic/clinical improvement was achieved, patient was managed with regular follow-up only and doing well till date.

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Discussion

GCT, first described by Abrikossoff as granular cell myoblastoma, has been reported in a wide variety of anatomic sites, but only few previous cases has been reported in spinal cord.[2-7] Brain and pituitary are another uncommon site for GCT within

CNS. Average age of patients with intraspinal GCT is 23 years, with a typical age range of 13 to 49 years and striking female preponderance.

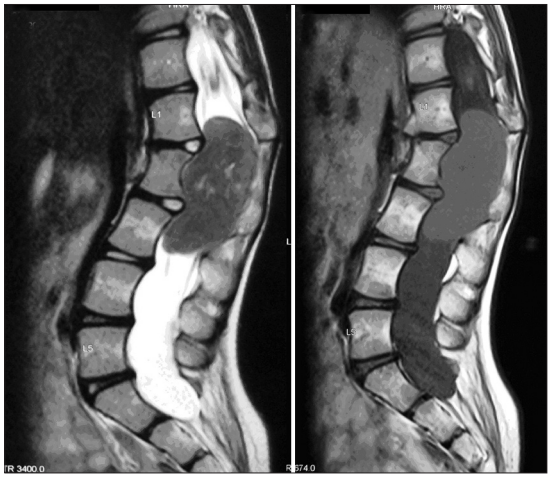

Figure 1: A well-circumscribed mass is seen in the intradural extramedullary space of the spine (sagital plane). The mass is hypointense on this sagital T2-weighted image (left) and hypo/isointense on T1-weighted image (right)

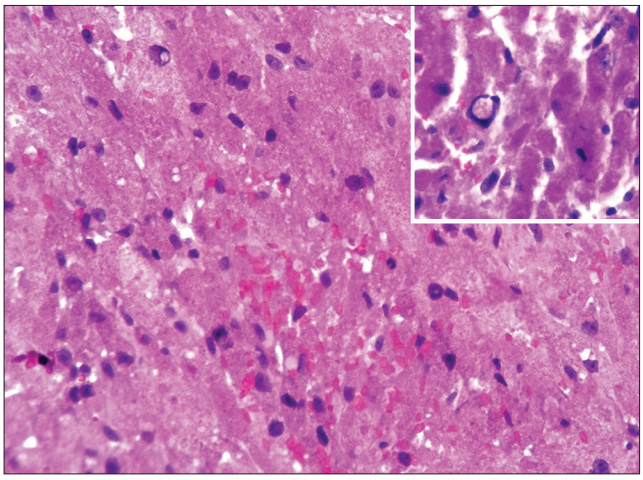

Figure 2: A cellular tumor arranged in sheet-like architecture. The cells are polygonal and elongated. Nuclei are small and round. The neoplastic cells have abundant, granular, eosinophilic cytoplasm surrounding round to oval irregular nuclei with few cells showing intranuclear pseudoinclusion (also show in inset)

The origin and nature of GCT have been debated since its original description. Studies using electron microscopy and immunohistochemistry have proven that these tumors exhibit schwannian differentiation. However, pathogenesis of GCT involving CNS is still debated.[7,8]

Conventional GCTs are benign neoplasms. Malignancy occurs in less than 2% of patients. Malignancy is diagnosed by a combination of histological findings, including cellular pleomorphism, necrosis and elevated mitotic activity, and clinical malignant behavior. Clinical suspicion of malignancy should be higher if the lesion is large or growing rapidly, or if there is evidence of distant spread.[9] Although the intradural tumors described in thus far have been benign histologically, they can cause serious morbidity if not diagnosed and treated correctly due to the anatomic confines of the canal.[3-7]

Clinical symptoms can include back pain, radicular symptoms, and slowly progressive neurological findings, such as weakness, paresthesia, impotence, and problems with gait, bowel and bladder function, upper and lower motor neuron dysfunction etc.[3-7]

Radiologically, they are best evaluated with MRI, being typically intradural extramedullary mass. Typically the tumors are slightly hypointense on T1-weighted sequences, and show homogenous contrast enhancement after intravenous injection of Gadolinium. On T2-weighted sequences, tumors generally show a hypointense signal. The MRI tissue characteristics are not specific as similar type of changes can be seen in other spinal canal tumors, including schwannoma, meningioma, paraganglioma, ependymoma, or metastatic tumor. So histopathologist carries responsibility of definitive diagnosis.[3-6]

The tumors consist of clusters, ribbons, nests and sheets of large, rounded, polygonal or elongated cells with well defined to ill-defined cellular borders, giving a syncytial appearance. The cells have either centrally or eccentrically located small, hyperchromatic to vesicular nuclei, enveloped by abundant, granular, eosinophilic cytoplasm. The granules are small and regular, and focally may aggregate into

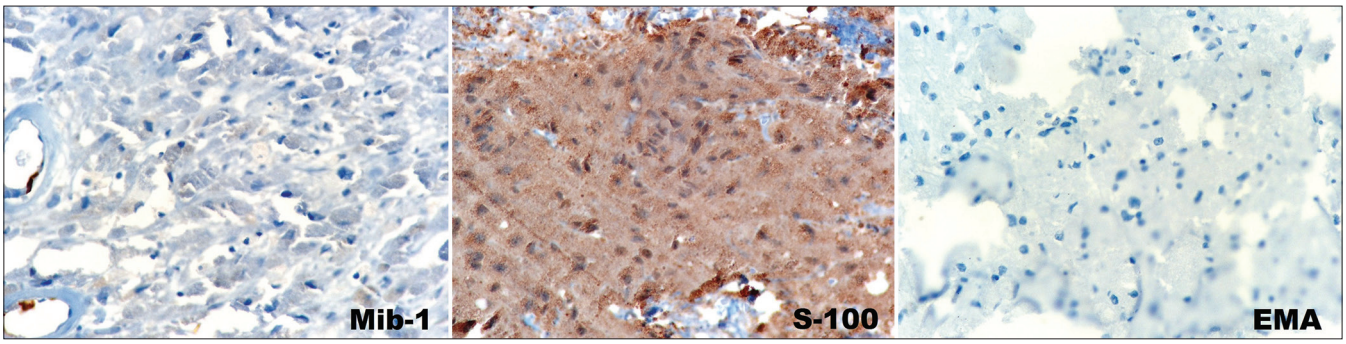

Figure 3: The neoplastic cells are strongly and diffusely immunoreactive with S-100 protein (nuclear and granular cytoplasmic staining). Cells are negative for EMA (right). Mib-1 is less than 1% (left)

larger fragments. In some cells, the granules accumulate at the cell periphery, creating a slightly cleared appearance in the center of the cytoplasm. The spinal GCT does not show atypia, necrosis and significant mitotic activity. The stroma is vascularized with some collagen deposition. The cytoplasmic granules are periodic acid Schiff (PAS) positive, diastase resistant. These cells are also strongly and diffusely immunoreactive with S-100 protein, CD68, NSE, inhibin-α, and vimentin. The cells fail to react with GFAP, neurofilament protein (NFP), HMB-45, keratin, EMA, CK7, chromogranin, and synaptophysin.[8,10]

The treatment advocated for granular cell tumor, at their usual site, is excision with wide margins. Radiotherapy and chemotherapy have been used in the past when malignant disease is present or complete excision of tumor with wide margin is not possible, but effectiveness is yet to be proven. Although there are only few intraspinal GCT reported, radiation treatment has been used successfully to stabilize recurrent/residual disease in one of the case.[5]

Conclusion

Intradural extramedullary tumors of spinal canal encompass a wide variety of tumors, many of which have overlapping clinical and radiographic findings, but histology along with selected immunohistochemistry studies are useful for exact diagnosis. GCT of spine should be taken into consideration with appropriate histological context as it has different biologic behavior.

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