DESMOPLASTIC SUPRATENTORIAL NEUROEPITHELIAL TUMOR OF INFANCY WITH DIVERGENT DIFFERENTIATION- A RARE CASE REPORT

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ABSTRACT
Desmoplastic infantile gangliogliomas are rare, superficial, supratentorial tumor presenting in early childhood, they occur within first two years of life and represent 1.25% of all intracranial tumors in children. A 3 year old female child with right fronto parietal desmoplastic infantile ganglioglioma who was successfully managed with surgery is presented here.

Key words: Frontoparietal, Desmoplastic, Supratentorial.

Introduction:
Desmoplastic infantile ganglioglioma (DIG) was first described by Vanden Berg et al in 1987. Despite their regular content of poorly differentiated and proliferative small cell elements, desmoplastic infantile astrocytomas and gangliogliomas are currently accorded WHO Grade I status. They are benign CNS tumors that develop virtually exclusively in infants younger than 2 years, often as a remarkably large solid and cystic hemispheric mass that may replace much of the brain on one side. Presents commonly with increasing head circumference and bulging fontanalle. Lymph nodes are common site of metastases for different cancers. Thus clinical recognition and urgent diagnosis of palpable lymphadenopathy is of paramount importance specially to differentiate between inflammatory lesions or metastatic or primary neoplastic tumor.

Case report:
A 3 years old female child presented with hemiparesis and right fronto-parietal space occupying lesion. CT scan revealed solid area and two communicating cystic lesions, one clear and the other with thick internal septations with surrounding edema and midline shift (Fig 1). Gross also showed a well circumscribed solid and cystic mass measuring 6x5x3 cms (Fig 2). Histopathology showed abundant desmoplasia with a neoplasm composed of spindle cells (Fig 3) arranged in a storiform pattern with areas of adipocytic differentiation (Fig 4). PNET (Primitive neuroectodermal tumor) like area is seen focally reflecting neuronal differentiation. Small inconspicuous gemistocyte like cells along with few ganglion cells seen (Fig 5). GFAP was done and is positive (Fig 6).

Fig 1: CT scan showing cystic and solid mass with internal septation and surrounding oedema
Fig 2: Well circumscribed solid and cystic mass measuring 6x5x3 cms

Fig 3: Cellular neoplasm showing spindle cells with areas of desmoplasia

Fig 4: Tumor showing adipocytic differentiation

Discussion:

Vanden Berg et al in 1987 first coined the terms as Desmoplastic infantile ganglioglioma and desmoplastic infantile astrocytoma on the basis of immunohistochemistry and electron microscopy. They have identical clinical and radiological features and have favourable outcome following excision, so they are included as distinctive group in revised WHO classification of brain tumors (1). A frontoparietal localization is most common, many of these sizable lesions spanning more than one cerebral lobe. Desmoplastic infantile astrocytomas and gangliogliomas are characteristically associated with evidences of rapidly evolving supratentorial mass effect as manifested in the very young. Increasing head circumference, bulging fontanelles, and forced downwards deviation of the eyes (the ‘sunset sign’) are commonly observed. CT or MRI show a superficially positioned, multinodular, and brightly contrast-enhancing supratentorial mass attached to dura in plaque-like fashion and associated with a subjacent, uni- or multiloculated cyst with intracranial shifts and other mass effects. Calcification has not been reported in these tumors (2).

The solid components of the desmoplastic cerebral astrocytoma and ganglioglioma are composed of tan to a gray-white tissue that is rubbery in consequence of its collagenisation. Histologically, it resembles a mesenchymal neoplasm such as a fibroma or malignant fibrous histiocytoma because of the dense collagen deposition and prominent spindled elements arranged in fascicular and storiform patterns. Meningeal and dural involvement is also common, thus further contributing to the perception of a mesenchymal derivation. The glial elements are often inconspicuous and
Fig 6: GFAP positivity

range from small gemistocyte-like to spindled cells. Neuronal components are also present in DIG and typically take the form of small ganglion-like cells. These cells are similarly enmeshed in a spindled, reticulin-rich desmoplastic background. Primitive neuroectodermal tumor (PNET)-like foci are common and consist of hypercellular foci with small round cells, increased mitotic activity, or necrosis. Immunostains highlights both GFAP-positive astrocytes and synaptophysin-positive neurons. The MIB-1 labeling index is typically low, except in PNET-like foci, where it may be markedly elevated.

The differential diagnosis includes reticulin rich desmoplastic tumors such as Pleomorphic xanthoastrocytoma (differentiated by age, lipidization and absence of neural component) and Gliofibroma (usually infratentorial and lacks neural component)(2). Because of the desmoplasia, spindled morphology, and frequent dural attachment, the differential diagnosis includes benign and malignant mesenchymal tumors, as well as fibrous meningioma. Careful inspection and Immunohistochemistry reveal the glial, and in the case of DIG, the neuronal elements. The presence of small blue cells with increased proliferative activity raises the differential diagnosis of PNET, although the more differentiated histologic elements and characteristic desmoplasia are also present. Malignant gliomas similarly lack this desmoplasia, a neuronal component, and the characteristic clinic-radiologic features.

The differential diagnosis for large cells with eccentrically located nuclei and abundant unipolar cytoplasm in an aspiration specimen of a cerebral mass in a young person includes DIG, atypical teratoid/rhabdoid tumor (AT/RT), dyssembroplastic neuroepithelial tumor (DNT), ganglioglioma, supratentorial PNET with ganglionic differentiation (ganglioneuroblastoma), anaplastic large cell lymphoma and pleomorphic xanthoastrocytoma(3). Clinical features as well as immunohistochemical analysis will help to differentiate these conditions. AT/RT will have loss of Integrase interactor-1, which can be detected immunohistochemically (INI-1) and the rhabdoid cells in this condition express vimentin. The role of adjuvant therapy is yet very limited. Chemotherapy is given only to those patients having high-grade tumors showing brisk mitosis, aneuploidy and increased MIB labeling. It is also given to patients with tumors involving eloquent regions of the brain not amenable for surgery (4).

Conclusion:

In spite of alarming clinical features with large size and PNET like foci, it is considered a WHO grade 1 neoplasm and has excellent prognosis. Spontaneous regression has been shown to occur in some cases following subtotal resection.

CONFLICT OF INTEREST:

Dr. Chandramouleswari, although a member of Editorial Board, did not participate in the review of this case report which was done by an independent and autonomous panel.
REFERENCES:

