Primary Ewing sarcoma of scalp diagnosed on cytology: A rare case report.

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Abstract

Primary Ewing sarcoma is small round cell tumour of neuroectodermal origin. We report a case of 16 year old female presenting with large cystic, osteolytic mass lesion in temporal region. The cytology showed loose and cohesive cluster of small round epithelial cells that were CD 99 and synaptophysin positive on immunocytochemistry.

Primary scalp Ewing sarcoma is very rare, and can present with cystic lesion. FNAC along with immunocytochemistry provides early opportunity for diagnosis and treatment.

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Introduction
Ewing sarcoma and PNET are round cell sarcoma of childhood that shows varying degree of neuroectodermal differentiation. PNET shows varying degree of neuroectodermal differentiation, while Ewing sarcoma lacks any evidence of it\textsuperscript{1}. The common sites involved are the metaphyseal and diaphyseal portion of long bones. While pelvis and ribs are commonly involved, skull is rarely involved with metastasis being more common than the primary tumour, constituting less than 1% of the total Ewing sarcoma. Males are more commonly affected than females.

Histopathology is considered as gold standard showing characteristic round cells with nuclei containing fine chromatin, scanty eosinophilic cytoplasm with strong PAS positivity and characteristic translocation of EWS-Friend leukemia integration 1 \text{[EWS-FLI1]}.

Case Report
A 16 year old female presented to the cytology department with diffuse mild headache and soft painless slowly progressive swelling in the temporal region for past three months. On local examination the swelling was ill defined, diffuse, cystic and non tender. Physical examination revealed mild weakness in upper and lower limb and localizing signs of raised intracranial tension. No definite clinical impression was made with a provisional diagnosis of cystic lesion scalp was made. On ultrasound a heterogeneous lesion was found in the temporal region with increased internal vascularity and focal calcification with irregular bony margins. MRI revealed a large relatively well defined lobulated heterogeneous mass lesion with its epicenter in the bony skull seen along left temporal bone. Intracranially the lesion was seen to extend in to left temporal lobe and midline shift to right was seen. An impression of primary malignant bone tumour with possibility of Ewing sarcoma was made on radiology.

On FNAC the aspirate was blood mixed, the smear was cellular showed scattered and loose clusters of small round epithelial cells with scant cytoplasm suggestive of a round cell tumour.

Immunocytochemistry for CD 99 and NSE was positive. Based on the cytomorphological feature and immunocytochemistry a diagnosis of primary Ewing sarcoma scalp was rendered. Subsequently done histopathology confirmed the cytological diagnosis. The patient was subsequently administered chemotherapy and she was recovering well till last met.

Discussion
ES is a small round cell tumour of adolescent and young adult of presumably primitive neuroectodermal tissue or pleuripotent migratory neural crest cells arising from bone or soft tissue. The PNET group includes small round cell tumors like Ewing sarcoma, small round cell tumour of thoracopulmonary region (Askin’s tumour), extraskeletal ewing sarcoma, peripheral neuroblastoma and peripheral neuroepithelioma\textsuperscript{2}. The ES/PNET group shares a common signature translocation of EWS-Friend leukemia integration 1 \text{[EWS-FLI1]}. While Ewing sarcoma does not show any neuroectodermal differentiation, PNET shows signs of neuroectodermal differentiation either morphologically, immunohistochemistry or on electron microscopy.

Primary Ewing’s sarcoma (EWS) of the cranium is extremely rare constituting <1% cases. Metastasis of ewing...
sarcoma scalp is more common than primary one. Krishnamani K et al[8] in their study found 7 cases with primary skull involvement while Vohra[4] found 8 cases with primary scalp involvement out of a total of 156 cases. Pritchard and Coley [5,6] reported only 3 cases with skull involvement out of a total of 234 and 149 cases. Frontal, parietal, and temporal bones are the commonly involved skull bones with headache and localized swelling being the commonest clinical features. Marked intracranial involvement increases intracranial pressure and localizing sign and symptoms like lower limb weakness.

With the advent of new advances in ES therapeutic protocols, a significant proportion of patients with ES receive preoperative chemotherapy[7]. FNAC being a primary investigation can provide a rapid and reliable way of diagnosing ES.

On FNAC the smears tend to be cellular with small round epithelial cells arranged in small tight clusters and larger cells with paler nuclei and dispersed chromatin. The cells have often high N: C ratio with scant basophilic cytoplasm that often contains glycogen showing strong PAS positivity. Other cytological findings that may be appreciated are the fibrillary matrix, homer right rosette and mitotic figure. Nuclear molding and crush artifact were occasional finding[1].

While metastasis of Ewing sarcoma to scalp is commonly found, primary skull Ewing sarcoma is rare. The differential diagnosis include other round cell tumors like lymphoblastic lymphoma, desmoplastic small round cell tumor, rhabdomyosarcoma, neuroblastoma and small cell carcinoma. Small cell carcinomas show extensive crush artifact along with prominent necrosis. Lymphoblastic lymphomas are characterized by dyscohesive cell dissociation, scant blue cytoplasm, lymphoglandular bodies, and tingible body macrophages in the background. Neuroblastoma is difficult to distinguish from ES because both tumors share cytologic features like small cell size, high N/C ratio, and rosettes but presence of neurophil in the background of neuroblastomas and in the center of the Homer–Wright rosettes and ganglion cells clinches diagnosis in favor of neuroblastoma. Presence of markedly atypical large bizarre cells and occasional spindle cell leads to diagnosis of rhabdomyosarcoma[8,9,10].

Ewing sarcoma/PNET is characterized by a sensitive and relatively specific antigen, CD99/MIC2, and chromosomal translocation t (11; 22)(q24; q12) which can be used as a fundamental diagnostic marker. Therefore cytomorphology and immunocytochemistry along with newer ancillary techniques like FISH for chromosomal translocation can be accurately used for diagnosis of ES.

**Conclusion**

Cytology with immunocytochemistry can act as primary rapid and reliable diagnostic modality in diagnosis of Ewing sarcoma and aid in initiating early treatment.

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**References**