Case Report

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Pathology

RHABDOMYOSARCOMA OF EXTERNAL AUDITORY CANAL: A DIAGNOSTIC CHALLENGE ON BIOPSY SPECIMEN.

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ABSTRACT Rhabdomyosarcoma is a common soft tissue sarcoma which rarely occurs in ear and temporal bone region. Auditory symptoms often mimic features of chronic infectious diseases, leading to delay in diagnosis. In advanced stage, disease presentation is cranial neuropathies which is unreponsive to usual line of treatment. Therefore histopathology is necessary to establish early and correct diagnosis. We report a case of three year girl child diagnosed as embryonal RMS of external auditory on biopsy. The diagnosis was subsequently confirmed on immunohistochemistry. The correct histological diagnosis forms the basis which helps in planning the management at its earliest.

KEYWORDS : Rhabdomyosarcoma, Embryonal, Immunohistochemistry,External auditory canal.

INTRODUCTION:

Rhabdomyosarcoma (RMS) accounts for about 60% of all sarcoma in the paediatric population and 4-8% of all paediatric cancers. [1, 2] In children, it commonly occurs in head and neck and is seen mostly in parameningeal region. External auditory canal (EAC) is said to be an uncommon site for paediatric RMS.[3] The prognosis of parameningeal RMS was previously thought to be extremely poor, however it has improved due to newer diagnostic modalities.[2] Rhabdomyosarcoma of EAC often clinically mimics inflammatory lesions such as chronic otitis media, which retards early diagnosis. The tissue examination therefore forms the mainstay in arriving at a definitive diagnosis. So that the patient can get benefit of early treatment that prevents further spread of disease. [3] We report a case of embyonal rhabdomyosarcoma in a three year old girl child with advanced disease.

CASE REPORT:

A three year child brought to the outpatient department by parents with complains of purulent to serosanguineous discharge from right ear along with pain and itching since 30 days. They also gave history of sudden onset of incomplete closure of right eye with deviation of angle of mouth on left side since last 7 days. There was no associated history of ear trauma or fever. Otoscopic examination revealed pinkish mass of size 3x2x1.2cm in right external auditory canal obliterating the tympanic membrane. Physical examination confirmed right sided Bell's palsy, however ipsilateral lacrimation was preserved. CT examination revealed abnormal heterogeneous and significantly enhancing soft tissue lesion in right external auditory canal, with lytic bone lesion of tegmen tympani and sigmoid plate with medial extension into right middle ear, posteriorly into right mastoid antrum with permeative destruction on its wall. The lesion was also extending intracranially abutting right sigmoid sinus, suggesting neoplastic lesion. We received a small biopsy specimen from right external auditory canal comprised of multiple, greyish brown, soft tissue pieces aggregating approximately 1.5x1.5x0.5cm in size. Histopathology revealed multiple polypoid tissue bits covered by stratified squamous epithelium. Tumour was seen as subepithelial discohesive round to ovoid to spindled tumour cells separated by loose oedematous to myxoid stroma. Individual cells show round to ovoid nuclei with fine to coarse chromatin and inconspicuous nucleoli. The cytoplasm is predominantly scant and pale eosinophilic. (Figure 1)

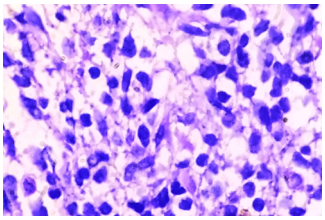

Figure 1 : Round, ovoid to spindled tumour cells in a loose, myxoid background. The cells bear scant cytoplasm and coarse nuclear chromatin (H and E, x20)

Focally the tumour show rhabdomyoblast like strap cells with dense eosinophilic cytoplasm showing striations and eccentrically placed enlarged nuclei.Mitotic figures were evident (Figure 2).

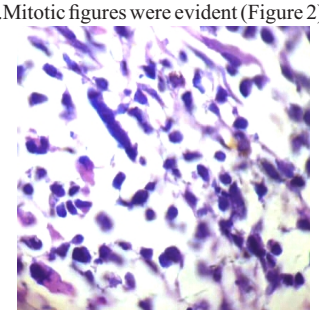

Figure 2 : Rhabdomyoblast, tadpole shaped with eccentrically placed enlarged nuclei and dense eosinophilic cytoplasm showing striations. (H and E, x40)

Considering clinicoradiological findings and histopathological features, the diagnosis of embryonal RMS was offered and immunohistochemistry (IHC) was advised. The tumour showed diffuse cytoplasmic positivity for Desmin and expression of Myogenin as strong nuclear positivity (Figure3), substantiating the diagnosis of Embryonal Rhabdomyosarcoma.

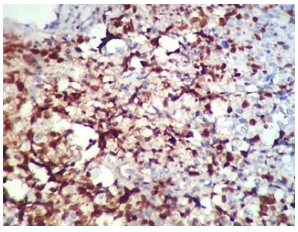

Figure 3 : Strong nuclear positivity of Myogenin shown by tumor cells (IHC-H andE x20)

The patient was referred to higher centre for further management.

DISCUSSION:

Amongst various subclasses of rhabdomyosarcoma, embryonal RMS is a primary malignancy of children and adolescents that arise from embryonic mesenchymal cells which develops into skeletal muscle. Nearly half of these occur in children under the age of 5. Males are affected 1.5 times more than female. [4] RMS in adults is pleomorphic type and involves mainly extremities. In adolescents, it is of alveolar type and has predilection for deep soft tissues of extremities, although tumour may arise in other sites including head and neck, trunk, perineum, pelvis and peritoneum; whereas RMS seen in children are

mostly embryonal and can occur in any anatomical location of body except bone.[5] Approximately 30-40% of these embryonal type occur in head and neck, most common being the base of skull, nasopharynx, nasal cavity and orbit.[1] Involvement of ear and temporal bone are extremely rare.[1,3] Clinically tumours within ear present with symptoms of bloody discharge and persistent otalgia or presented with polypoidal mass mimicking chronic suppurative otitis mdia that is unresponsive to treatment. This is one of the most common reasons for delayed presentation and hindrance in early diagnosis. Intracranial extensions favour a more advanced lesion. [6] Diagnosis is usually confirmed by tissue examination. MRI is used to evaluate the primary lesion and to understand the extent disease. CT scan may be useful to determine bony erosion of skull base and is the best method to assess lung for metastasis. [2] Microscopically, embryonal RMS consists of primitive mesenchymal cells in varying stages of myogenesis with variable content of rhabdomyoblasts. These primitive tumour cells are seen as round, oval to spindled cells with round to ovoid nuclei having coarse chromatin, inconspicuous nucleoli and scant cytoplasm against a loose myxoid background. The diagnostic difficulty arises when these tumours cells together with loose oedematous stroma is mistaken for inflammatory lesion. In our case, histopathological features were identified with great difficulty due to small biopsy sample; however identification of characteristic rhabdomyoblast was the indicator. The rhabdomyoblasts progressively acquire more cytoplasmic eosinophilia and elongation, tadpole or strap cell or spider cells. [4, 7] In difficult situations, clinical features such as long history of non responding otitis media and radiological findings can provide a clue to arrive at a correct diagnosis. The careful search for rhabdomyoblast along with clinicoradiological findings and high index of suspicion can aid the pathologist in arriving at a definitive diagnosis. Immunohistochemical markers of skeletal muscle differentiation typify embryonal RMS. These markers correlate with the degree of tumour cell differentiation. Only vimentin is present in the cytoplasm of most primitive cells. Desmin and Actin are acquired by developing rhabdomyoblasts. Expression of newer markers specific for rhabdomyoblastic differentiation such as MyoD and Myogenin are particularly useful for diagnosis of RMS. [8, 9] The diagnosis of RMS is confirmed if at least one of these markers is positive.[10]

In present case, there was diffuse expression for desmin and strong positivity for myogenin and Myo D1which validated our diagnosis of rhabdomyosarcoma. As RMS a highly malignant tumour, known for its local invasion and early vascular and lymphatic dissemination, therapy totally depend upon clinicopathologic variable such as age, anatomical location of primary tumour, histologic type and extent of disease spread at the time of diagnosis.[11] Management of RMS includes multimodality approach. Surgery followed by combination chemotherapy with or without adjuvant radiotherapy. Radical surgery not possible in advanced disease due to close proximity of tumours to vital structures and also due to possibility of postoperative cosmetic and functional disability as happened in our case.[3,12]Prognosis of RMS is poor, the 5 year survival was about 10% but in last 30yrs, survival is increased to 80%. However with advanced stage disease, the prognosis is still dismal and in patients with meningeal involvement, it is less than 10%.

CONCLUSION:

Even though prognosis of embryonal RMS is relatively good, patients with advanced disease and intracranial spread have poor outcome. Therefore careful and critical search for key morphological features along with immunohistochemistry in a background of clinico radiological findings will assist pathologist in establishing correct histologic diagnosis which eventually leads to prompt treatment and can be life saving to the young child.

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